PCAD Project

Working Resources List on Dementia Care Management and Intellectual Disability

Preparing Community Agencies for Adults Affected by Dementia - "PCAD" Project

v.28b

PCAD Project
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To find resource or content of interest, search by keyword (e.g., assessment, carer, decline, etc.) in PDF reader.

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Abdullah, L,, Zhou, Z., Hall, J., Petersen, M., Zhang, F., & O'Bryant, S.
Association of Alzheimer's disease biomarkers with low premorbid intellectual functioning in a multi-ethnic community-dwelling cohort: A cross-sectional study of HABS-HD.
Journal of Alzheimers Disease, 2025 Apr; 104(4), 1201-1211. doi: 10.1177/13872877251322966. Epub 2025 Mar 21.
Abstract**:** Individuals with intellectual disability (ID) may have a five-fold increased risk for developing Alzheimer's disease (AD). However, studies investigating brain aging among individuals with **ID without Down syndrome** (DS) are lacking. To begin addressing this gap, our study utilized word reading, a widely recognized indicator of an individual's premorbid intellectual ability (pIQ), to examine the effects of ID without DS on plasma AD biomarker outcomes. We investigated the relationship between premorbid intellectual ability (pIQ) and plasma AD biomarkers in individuals with ID without DS, while considering ethnic differences in these associations. Participants were from the Health & Aging Brain Study - Health Disparities (HABS-HD) and were categorized into low ($z \leq -2.00$) or average ($z = 0.00 \pm 1.00$) pIQ groups based on word reading scores. Plasma biomarkers including Aß40, Aß42, Aß42/40, phosphorylated tau 181 (p-Tau181), neurofilament light chain (NfL), and total tau (t-tau) were assayed using Simoa technology.It was found that individuals with low pIQ exhibited significantly higher levels of p-Tau181 ($p < 0.05$), NfL ($p < 0.05$), and t-tau ($p < 0.05$) compared to those with average pIQ. Stratified analysis by ethnicity revealed differential associations, with Hispanic and non-Hispanic White (NHW) participants showing distinct biomarker profiles relative to non-Hispanic Black (NHB) individuals. The findings demonstrate that **low pIQ is a reliable factor associated with plasma AD biomarker outcomes**. Ethnicity appears to modulate these associations, suggesting complex interactions between factors driving AD susceptibility across diverse populations. This study highlights the importance of considering both pIQ and ethnicity in neurodegenerative processes, particularly in individuals with non-DS intellectual developmental disability.

Acton, D., Duncan, C., & Jaydeokar
Co-production of post-diagnostic psychosocial intervention with carers of people with intellectual disability and dementia
Advances in Mental Health and Intellectual Disabilities, 2022, 16(3), 169-178. https://doi.org/10.1108/AMHID-01-2022-0006
Abstract: This paper aims to underline the importance of using a collaborative approach when designing and adapting a post diagnostic psychosocial intervention of cognitive stimulation therapy (CST) for people with intellectual disability and dementia. As part of a service improvement, a manual of CST was adapted, for delivery in clinical practice. A qualitative co-production method allowed participants with a lived experience to provide regular feedback relating to the development of the adapted CST manual and intervention program. This feedback was used to make continual development changes to the CST manual. The study demonstrated co-production with those who provide care is valuable in adapting psychosocial therapies for people with an intellectual disability and dementia. Additional findings identified the need for carer education in ageing, dementia care and the physical health needs for older people with intellectual disability. To the best of the authors' knowledge, this is the first study that has used a co-production approach with families and carers in adapting a group therapy program for people with an intellectual disability. This paper underlines the need for post diagnostic clinical interventions for people with dementia and those who provide care.

Acton, D.J., Jaydeokar, S.,& Jones, S.
Caregivers experiences of caring for people with intellectual disability and dementia: a qualitative evidence synthesis
Advances in Mental Health and Intellectual Disabilities, 2023, 17(1),10-25. https://doi.org/10.1108/AMHID-08-2022-0027
Abstract: A systematic review of the literature was completed to examine the needs of those who provide care to people with intellectual disability and dementia. The purpose of this paper was to develop an understanding of the complexities, challenges and support available to meet the needs of an ageing population. A qualitative evidence synthesis was used to appraise 12 studies. An evidence synthesis approach was used to better understand the challenges caregivers experience in caring for a person with intellectual disability and dementia. Aggregating and integrating findings from multiple studies allowed to identify inconsistencies, quality, relationships and trends to enhance the awareness of gaps in care provision. There were six main domains identified from the available literature which included: gaps in knowledge and skills, early identification of dementia and associated difficulties, managing behavior, coping, burden of care and Impact on confidence.

Acton, D.J., Jaydeokar, S., & Jones, S.
Dementia education and training for caregivers supporting older people with intellectual disability: a scoping review of the literature
Advances in Mental Health and Intellectual Disabilities, 2023, 17(3), 187-201. https://doi.org/10.1108/AMHID-02-2023-0006
Abstract: Education and training is vital in improving age-related care provisions. However, a lack of awareness and understanding of dementia could be a factor in meeting the age-related needs of people. This paper aims to examine the impact dementia education has on caregivers' confidence to provide person-centered care for people with intellectual disability and identifies additional training needs. A systematic scoping review was completed using preferred reporting items for systematic reviews and meta-analyses to guide the process of mapping existing evidence of dementia education and training programs available to caregivers of people with intellectual disability. A search of five electronic databases identified 11 articles that reported on the role of dementia education in improving the knowledge of caregivers in effectively delivering the age-related care. Findings suggest that improved training provision is needed to support early diagnosis and increase caregivers' confidence in meeting the physical and psychological needs of older adults with intellectual disability.

Acton, D.J., Jaydeokar, S., Taylor, R., & Jones, S.
Exploring the lived experiences and care challenges of formal paid caregivers for people with intellectual disability and dementia.
Journal of Intellectual Disabilities, 2024, May 30. doi: 10.1177/17446295241259076
Abstract: A greater number of people with intellectual disability are living into older age and are at increased risk of developing conditions such as dementia. Caring for a person with dementia presents several challenges for formal caregivers due to the progressive nature of the disease. An interpretive phenomenological analysis was used to understand the lived experiences of a purposive sample of formal caregivers in caring for people with intellectual disability and dementia. Discussions from 14 individual interviews generated data were analyzed. Four key super-ordinate themes emerged which were: (1) recognizing early indicators and diagnosis, (2) post diagnostic support, (3) coping with change and (4) need for future development. Themes reflected the experiences, barriers to dementia diagnosis and provide a valuable insight into the challenges faced by formal caregivers in providing aged care services.

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Adams, D., Oliver,C., Kalsy, S., Peters, S., Broquard, M., Basra, T., Konstandinidi, E., & McQuillan, S.
Behavioural characteristics associated with dementia assessment referrals in adults with Down syndrome.
Journal of Intellectual Disability Research. 2008 Apr;52(Pt 4):358-68. Epub 2008 Jan 22.
Abstract: Behavioral changes associated with dementia in Down syndrome are well documented, yet little is known about the effect of such behaviors on carers and referral. By comparing the behavioral and cognitive profiles of individuals referred for a dementia assessment with those of individuals not referred, some insight can be gained into behavioral characteristics that initiate referral for specialist support or interventions. Forty-six adults with Down syndrome were divided into two groups dependent upon method of entry into the study; post-referral to a specialist service for older adults with intellectual disabilities and Down syndrome for a dementia assessment (n = 17) or after receiving information sent out to day centers and residential homes (n = 29). These groups were compared on established measures of dementia alongside two informant measures of behavior. Those referred for a dementia assessment evidenced scores indicative of cognitive decline on both informant and direct Neuropsychological Assessments and showed more behavioral excesses, but not deficits, and lower socialization and coping skills than those in the comparison group. Carers of those referred for a dementia assessment reported a greater impact of behavioral excesses on staff than on the individual showing the behavior in contrast to the comparison group. The behavioral differences between those referred and the comparison group suggest that two factors are involved in the instigation of a referral for a dementia assessment: the nature of the behavioral presentation (excesses rather than deficits) and the effect of that behavioral change upon the care staff.

Adam, E., Sleeman, K.E., Brearley, S., Hunt, K., & Tuffrey-Wijne, I.
The palliative care needs of adults with intellectual disabilities and their access to palliative care services: A systematic review.
Palliative Medicine, 2020 Sep, 34(8), 1006-1018. doi: 10.1177/0269216320932774. Epub 2020 Jun 17.
Abstract: There is evidence that people with intellectual disabilities experience healthcare inequalities, including access to specialist palliative care, but to date, there has not been a systematic review of empirical evidence. To identify the palliative care needs of adults with intellectual disabilities and the barriers and facilitators they face in accessing palliative care. Systematic review using a narrative synthesis approach (International prospective register of systematic reviews (PROSPERO) registration number: CRD42019138974). Five databases were searched in June 2019 (MEDLINE, Embase, PsycINFO, the Cochrane library and CINAHL) along with hand searches and a search of the grey literature. All study designs were included. A total of 52 studies were identified, all of which were conducted in high-income countries, the majority in the United Kingdom (n = 28). From a total of 2970 participants across all studies, only 1% were people with intellectual disabilities and 1.3% were family members; the majority (97%) were health/social care professionals. Identified needs included physical needs, psychosocial and spiritual needs, and information and communication needs. Barriers and facilitators were associated with education (e.g. staff knowledge, training and experience), communication (e.g., staff skill in assessing and addressing needs of people with communication difficulties), collaboration (e.g. importance of sustained multidisciplinary approach) and health and social care delivery (e.g., staffing levels, funding and management support). This review highlights the specific problems in providing equitable palliative care for adults with intellectual disabilities, but there is a lack of research into strategies to improve practice. This should be prioritised using methods that include people with intellectual disabilities and families.

Adams, D., Hastings, R.P., Maidment, I., Shah, C., & Langdon, P.E.
Stakeholder experiences of deprescribing psychotropic medicines for challenging behaviour in people with intellectual disabilities
Tizard Learning Disability Review, 2024, Vol. ahead-of-print No. ahead-of-print. https://doi.org/10.1108/TLDR-09-2023-0020
Abstract: Evidence of overprescribing of psychotropic medicines to manage challenging behaviour in people with intellectual disabilities has led to national programs within the UK to promote deprescribing, such as stopping the overprescribing of medication in people (with intellectual disabilities, autism or both). To successfully implement deprescribing initiatives, we need to understand how to engage stakeholders in the process. In a published systematic review, we reported evidence about the process of deprescribing psychotropic medicines for people of all ages with intellectual disabilities and challenging behavior. As a part of the original review, we searched for evidence about stakeholders' experiences of the psychotropic deprescribing process, which was synthesized and reported within the current study. Six studies were identified. Involving carers and people with intellectual disabilities, providing ongoing support and improving access to non-pharmacological interventions, including positive behavior support, may contribute to successful outcomes, including reducing or stopping psychotropic medicines and improving quality of life. Implementing psychotropic deprescribing requires a multidisciplinary collaborative care approach and education for stakeholders.

Ali, A., Brown, E., Tsang, W., Spector, A., Aguirre, E., Hoare W., & Hassiotis, A.
Individual cognitive stimulation therapy (iCST) for people with intellectual disability and dementia: a feasibility randomised controlled trial
Aging & Mental Health,,2020, 26(4), 698-708, DOI: 10.1080/13607863.2020.1869180
Abstract: Authors examined the feasibility, acceptability and fidelity of individual Cognitive Stimulation Therapy (iCST) in people with intellectual disability (ID) and dementia. We aimed to recruit forty dyads (carer and individual with dementia and ID) who were randomised to iCST or a waiting list control group. Both groups received treatment as usual. Family and paid carers delivered the manualised intervention (40 sessions over 20 weeks). Recruitment and retention of participants, intervention adherence, fidelity and acceptability were assessed. Outcome measures of cognition, adaptive functioning, quality of life (QoL) and carer outcomes were collected at baseline, midpoint (11 weeks) and at 21 weeks. Qualitative interviews were conducted with six carers about their experience of iCST. Forty dyads were recruited over 10 months from 12 National Health Service trusts. One dyad dropped out and 87.5% and 97.5% completed the midpoint and end-point assessments respectively. Assessment of fidelity indicated that the correct session structure was not followed; 70% completed at least 20 sessions and there was a high level of satisfaction with iCST. QoL was significantly higher in the iCST arm at 21 weeks (adjusted mean difference: 3.11; 95% CI: 0.64 to 5.58). There were no differences in the other outcome measures. The intervention was feasible and acceptable. A full-scale trial is warranted but some modifications are needed, including improved training and supervision for carers to improve fidelity.

Ahlström, G., Axmon, A., Sandberg, M., & Flygare Wallén, E.
Health care utilisation among older people with Down syndrome compared to specific medical guidelines for health surveillance: a Swedish national register study
BMC Health Services Research, 2020, Oct 15, 20(1), 949. doi:10.1186/s12913-020-05800-7.
Abstract: Specific medical guidelines for health surveillance exist for people with Down syndrome (DS) since 25 years but knowledge of adherence to the guidelines is lacking. The guidelines were developed to avoid unnecessary suffering from preventable conditions. The aims of the study were to investigate 1) planned health care visits in relation to the co-morbidities described in specific medical guidelines as a measure of adherence, 2) unplanned health care visits as a measure of potentially unmet health care needs and 3) gender differences in health care utilization among older people with DS. This register-based study includes people with DS (n = 472) from a Swedish national cohort of people with intellectual disability (n = 7936), aged 55 years or more, and with at least one support according to the disability law, in 2012. Data on inpatient and outpatient specialist health care utilization were collected from the National Patient Register for 2002-2012. A total of 3854 inpatient and outpatient specialist health care visits were recorded during the 11 years, of which 54.6% (n = 2103) were planned, 44.0% (n = 1695) unplanned and 1.4% (n = 56) lacked information. More than half of the visits, 67.0% (n = 2582) were outpatient health care thus inpatient 33% (n = 1272). Most planned visits (29.4%, n = 618) were to an ophthalmology clinic, and most unplanned visits to an internal medicine clinic (36.6%, n = 621). The most common cause for planned visits was cataract, found at least once for 32.8% in this cohort, followed by arthrosis (8.9%), epilepsy (8.9%) and dementia (6.6%). Pneumonia, pain, fractures and epilepsy each accounted for at least one unplanned visit for approximately one-fourth of the population (27.1, 26.9, 26.3 and 19.7% respectively). Men and women had similar numbers of unplanned visits. However, women were more likely to have visits for epilepsy or fractures, and men more likely for pneumonia. Increased awareness of existing specific medical guidelines for people with DS is vital for preventive measures. The relatively few planned health care visits according to

the medical guidelines together with a high number of unplanned visits caused by conditions which potentially can be prevented suggest a need of improved adherence to medical guidelines.

Ahlström, G., Wallén, E. F., Tideman, M., & Holmgren, M..
Ageing people with intellectual disabilities and the association between frailty factors and social care: A Swedish national register study.
Journal of Intellectual Disabilities, 2021, 26(4), 900-918.
https://doi.org/10.1177/17446295211037170
Abstract: The aim of this study was to describe the social care provided for different age groups of people with intellectual disability, 55 years or above, and to investigate the association between such care and frailty factors for those with diagnosed level of intellectual disabilities. Descriptive and logistic regression analyses were used. Commonest forms of social care among the 7936 people were Residential care, Daily activities and Contact person. Home help and Security alarm increased with age. The frailty factors significantly associated with increased social care were age, polypharmacy and severe levels of intellectual disabilities. Persons most likely to be in residential care were in the age group 65-79 with polypharmacy and severe disability. The results indicate a need for further research of how frailty factors are considered in social care and longstanding medication, especially then severe intellectual disability hinders communication. A national strategic plan for preventive interventions should be developed to ensure the best possible healthy ageing.

Alldred, M..J., Martini, A.C.,, Patterson, D., Hendrix, J., & Granholm, A.C.
Aging with Down syndrome-where are we now and where are we going?
Journal of Clinical Medicine, 2021 Oct 13, 10(20), 4687. doi: 10.3390/jcm10204687.
Abstract: Down syndrome (DS) is a form of accelerated aging, and people with DS are highly prone to aging-related conditions that include vascular and neurological disorders. Due to the overexpression of several genes on Chromosome 21, for example genes encoding amyloid precursor protein (APP), superoxide dismutase (SOD), and some of the interferon receptors, those with DS exhibit significant accumulation of amyloid, phospho-tau, oxidative stress, neuronal loss, and neuroinflammation in the brain as they age. In this review, we will summarize the major strides in this research field that have been made in the last few decades, as well as discuss where we are now, and which research areas are considered essential for the field in the future. Authors examine the scientific history of DS bridging these milestones in research to current efforts in the field. We extrapolate on comorbidities associated with this phenotype and highlight clinical networks in the USA and Europe pursuing clinical research, concluding with funding efforts and recent recommendations to the NIH regarding DS research.

Allen, A.P., McGlinchey, E., Fallon, M., McCallion, P., McCarron, M.
Cognitive reserve and dementia risk management in people with an intellectual disability.
International Journal of Geriatric Psychiatry, 2023 Apr; 38(4), e5906. doi: 10.1002/gps.5906.
Abstract: (not provided). Dementia risk is elevated in people with intellectual disability, particularly for those with Down syndrome. For people with intellectual disability, longitudinal follow-up and reviews of medical databases have confirmed that, as compared to the general population, there are lower levels of hypertension and diabetes, and less smoking and alcohol use, low levels of education and higher levels of traumatic brain injury, physical inactivity and depression. There has been some research interest in associations between these lifestyle factors and dementia in people with intellectual disability. There was no association found between physical activity, diet, weight, or cognitive activity between people with Down syndrome with dementia compared to people with Down syndrome without dementia, although these lifestyle factors were assessed using quite broad ordinal scales. Differences in lifestyle factors have been cited as factors that exacerbate dementia risk in people with intellectual disability. A prevalent belief that there is lower cognitive reserve for people with intellectual disabilities reflects that cognitive reserve in the general population has generally been reported as associated with level of education and employment. Although people with an intellectual disability are increasingly participating in education and employment, they are not doing so to the same extent as the general population. Further and continuing research is required to build a more detailed under standing of both dementia risk in people with an intellectual
disability, and strategies for minimizing such risk, but the existing evidence supports providing more opportunities for people with intellectual disability to engage in education and cognitively stimulating work and pastimes. Lifestyle factors can be targeted to enhance cognitive reserve and reduce dementia risk for people with intellectual disability. The efficacy of lifestyle interventions in general among people with intellectual disabilities deserves further investigation as they have the potential to contribute to cognitive reserve and offer additional beneficial effects as persons age.

Alftberg, Å., Johansson, M., & Ahlström, G.
Ambivalence among staff regarding ageing with intellectual disabilities: Experiences and reflections.
Journal of Intellectual Disabilities, 2021 Jun;25(2):192-209. doi: 10.1177/1744629519874997. Epub 2019 Sep 30.
Abstract: This study explores the experiences and reflections of staff in intellectual disability (ID) services concerning ageing with ID. Qualitative interviews were conducted with 24 staff members in group homes and daily activity centres. The findings showed that the staff were uncertain about the signs of ageing in people with intellectual disabilities; they compared the life conditions of these people with conditions in older people without intellectual disabilities. Their emphasis on an active lifestyle was very strong. The staff members also mentioned uncertainty about how to facilitate assistive devices and whether 'ageing in place' was the best solution. The overall theme was manifested as ambivalence where notions of older people with intellectual disabilities seemed incompatible with notions of old age in general and could be explained by the theoretical concept of age coding. The findings of this study indicate the need to provide education about ageing to staff working in ID services.

Ali, A., Brown, E., Tsang, W., Spector, A., Aguirre, E., Hoare, S., Hassiotis, A.
Individual cognitive stimulation therapy (iCST) for people with intellectual disability and dementia: a feasibility randomised controlled trial.
Aging & Mental Health. 2022 Apr;26(4):698-708. doi: 10.1080/13607863.2020.1869180. Epub 2021 Jan 4. PMID: 33393364.
Abstract: To examine the feasibility, acceptability and fidelity of individual Cognitive Stimulation Therapy (iCST) in people with intellectual disability (ID) and dementia. We aimed to recruit forty dyads (carer and individual with dementia and ID) who were randomised to iCST or a waiting list control group. Both groups received treatment as usual. Family and paid carers delivered the manualised intervention (40 sessions over 20 weeks). Recruitment and retention of participants, intervention adherence, fidelity and acceptability were assessed. Outcome measures of cognition, adaptive functioning, quality of life (QoL) and carer outcomes were collected at baseline, midpoint (11 weeks) and at 21 weeks. Qualitative interviews were conducted with six carers about their experience of iCST. Forty dyads were recruited over 10 months from 12 National Health Service trusts. One dyad dropped out and 87.5% and 97.5% completed the midpoint and end-point assessments respectively. Assessment of fidelity indicated that the correct session structure was not followed; 70% completed at least 20 sessions and there was a high level of satisfaction with iCST. QoL was significantly higher in the iCST arm at 21 weeks (adjusted mean difference: 3.11; 95% CI: 0.64 to 5.58). There were no differences in the other outcome measures. The intervention was feasible and acceptable. A full-scale trial is warranted but some modifications are needed, including improved training and supervision for carers to improve fidelity

Altuna, M., Giménez, S., & Fortea, J.
Epilepsy in Down syndrome: A highly prevalent comorbidity
Journal of Clinical Medicine, 2021 (June 24), 10(13). 2776- [e-print]. doi: 10.3390/jcm10132776.
Abstract: Individuals with Down syndrome (DS) have an increased risk for epilepsy during the whole lifespan, but especially after age 40 years. The increase in the number of individuals with DS living into late middle age due to improved health care is resulting in an increase in epilepsy prevalence in this population. However, these epileptic seizures are probably underdiagnosed and inadequately treated. This late onset epilepsy is linked to the development of symptomatic Alzheimer's disease (AD), which is the main comorbidity in adults with DS with a cumulative incidence of more than 90% of adults by the seventh decade. More than 50% of patients with DS and AD **dementia** will most likely develop epilepsy, which in this context has a specific clinical presentation in the form of generalized myoclonic epilepsy. This epilepsy, named late onset myoclonic epilepsy (LOMEDS) affects the quality of life, might be associated with worse cognitive and functional outcomes in patients with AD dementia and has

an impact on mortality. This review summarizes the current knowledge about the clinical and electrophysiological characteristics, diagnosis and treatment of epileptic seizures in the DS population, with a special emphasis on LOMEDS. Raised awareness and a better understanding of epilepsy in DS from families, caregivers and clinicians could enable earlier diagnoses and better treatments for individuals with DS.

Altuna, M., Estanga, A., Garrido, A., Saldias, J., Cañada, M., Echeverria, M., Larrea, J. A., Ayo, P., Fiz, A., Muñoz, M., Santa-Inés, J., García-Landarte, V., & García-Sebastián, M.
Down Syndrome-Basque Alzheimer Initiative (DS-BAI): Clinic-Biological Cohort.
Journal of Clinical Medicine, 2024, 13(4), 1139.
https://doi.org/10.3390/jcm13041139
Abstract: Down syndrome (DS) is the most common genetically determined intellectual disability. In recent decades, it has experienced an exponential increase in life expectancy, leading to a rise in age-related diseases, including Alzheimer's disease (AD). Specific health plans for the comprehensive care of the DS community are an unmet need, which is crucial for the early and accurate diagnosis of main medical comorbidities. We present the protocol of a newly created clinical and research cohort and its feasibility in real life. Methods: The Down Syndrome-Basque Alzheimer Initiative (DS-BAI) is a population-based, inclusive, multidisciplinary initiative for the clinical-assistance and clinical-biological research approach to aging in DS led by the CITA-Alzheimer Foundation (Donostia, Basque Country). It aims to achieve the following: (1) provide comprehensive care for adults with DS, (2) optimize access to rigorous and quality training for socio-family and healthcare references, and (3) create a valuable multimodal clinical-biological research platform. Results: During the first year, 114 adults with DS joined the initiative, with 36% of them showing symptoms indicative of AD. Furthermore, adherence to training programs for healthcare professionals and families has been high, and the willingness to collaborate in basic and translational research has been encouraging. Conclusion: Specific health plans for DS and conducting clinical and translational research on the challenges of aging, including AD, are necessary and feasible.

Alvarez, N.
Dementia and Alzheimer's disease
In: Rubin, I.L., Merrick, J., Greydanus, D.E., Patel, D.R. (eds), Health Care for People with Intellectual and Developmental Disabilities across the Lifespan. (2016). Springer, Cham. https://doi.org/10.1007/978-3-319-18096-0_85 (pp. 995-1011). doi: 10.1007/978-3-319-18096-0_85
Abstract: Dementia is an acquired syndrome secondary to many causes with a continuous progressive deterioration of intellectual functions and personality changes, associated with a marked decline in activities of daily living. Memory loss, a common early sign, is followed by impairment in other domains like language, judgment, mood changes, psychiatric disorders, like hallucinations, psychosis, and sleep disorders. These symptoms are also associated with gait deterioration, swallowing disorders, loss of activities of daily living and in some cases epileptic seizures. The clinical presentation is variable depending on the cause and the individual affected. This chapter is limited to the presentation of dementia in persons with intellectual and developmental disabilities and because of the high prevalence, special consideration will be given to the development of Alzheimer's Disease in persons with Down Syndrome.

Alzheimer's Association
Guidelines for dignity: Goals of specialized Alzheimer/dementia care in residential settings
47 pp.
Chicago: The Alzheimer's Association [919 North Michigan Avenue, Suite 1000, Chicago, IL 60611-1676] (1992)
Abstract: Standards for care and structure of care settings housing persons affected by Alzheimer's disease. Includes sections on philosophy, pre-admission activities, admission, care planning and implementation, adapting to changes in condition, staffing and training, physical environment and "success indicators."

Alzheimer's Australia
Down syndrome and Alzheimer's disease
12 pp.
[Place of publication not provided] (no date)
Source: http://www.cddh.monash.org/assets/dsad-booklet-final.pdf
Abstract: Informational booklet on dementia and people with Down syndrome jointly issued by Alzheimer's Australia, Down Syndrome Victoria, and Centre for Developmental Disability Health Victoria. Contains three main sections: (1) About Alzheimer's disease and Down syndrome, (2) Diagnosis, and (3) Support, as well as a section on local resources.

Alzheimer's Disease International
Planning and design guide for community based day care centres
21 pp.
London: Alzheimer's Disease International [45/46 Lower Marsh, London SE1 7RG, United Kingdom (www.alz.co.uk)] (1999)
Abstract: An illustrated 21-page booklet highlighting main design issues and suggestions for organizing an effective environment for adults with dementia - with applications for residential environment.

Alzheimer's Disease Society
Safe as houses -- Living alone with dementia (A resource booklet to aid risk management)
London: Alzheimer's Disease Society [Gordon House, 10 Greencoat Place, London SW1P 1PH, United Kingdom] (1994)
30 pp.
Abstract: A 30-page booklet designed for the carer who is concerned about an older person with early to mid-stage dementia who may be living on their own. The booklet examines risks that the older adult may encounter and suggests how they could be minimized. The intent of the booklet is to aid the older person remain functional at home, with as minimal risk, for as long as possible. Covers personal care, finances, wandering, security, medication, utilities, and household safety. Whilst information is generic, resource information is geared toward the UK.

Alzheimer's Society
Learning disabilities and dementia
Alzheimer's Society UK, (2022), Alzheimer's Society, 43-44 Crutched Friars, London, EC3N 2AE
https://www.alzheimers.org.uk/about-dementia/types-dementia/learning-disabilities-dementia
Abstract: Web-based information produced in the UK on the topic of intellectual disabilities and dementia. Contains background information, as well as diagnosis, identificatiion of symptoms and support and care services.

Alzheimer's Society
Supporting a person with dementia who also has a learning disability
Alzheimer's Society UK, (2023), Alzheimer's Society, 43-44 Crutched Friars, London, EC3N 2AE
https://www.alzheimers.org.uk/get-support/publications-and-factsheets/dementia-together/supporting-person-dementia-learning-disability
Abstract: Informational webpage on how to approach helping a relative or friend with an intellectual disability who's diagnosed with dementia.

Anderson, M., Oak, K., Goodey, R., Dodd, K., & Shankar, R.
Do the severity of intellectual disability and /or the presence of neurodevelopmental disorders influence the onset of dementia in people with Down syndrome?,
Journal of Mental Health Research in Intellectual Disabilities, 2020, 13(4), 322-342, DOI: 10.1080/19315864.2020.1822964
Abstract: Having a diagnosis of Down syndrome (DS) is associated with intellectual disability (ID), pervasive developmental disorders and Alzheimer's dementia (AD). The association between these conditions has not been well evaluated. This paper looks to examine the current evidence pertaining to the relationship between dementia in people with DS and severity of ID and the presence of pervasive developmental disorders. A scoping review using PRISMA guidance was undertaken. Medline, Cochrane database, NHS evidence, Trial registers and Open Grey were searched in December 2018 and an updated search was completed in July 2020. Three search strategies were used to retrieve articles relating to DS, dementia, pervasive developmental disorders (including autism, autism spectrum disorder (ASD) and attention-deficit hyperactivity disorder (ADHD)) and severity of ID. Studies were included if they met the pre-defined inclusion criteria of investigating an association between autism/ASD, ADHD, or severity of ID and the development of dementia in people with DS. Studies were excluded if they did not include primary data, if the population included non-Down causes of ID, or if no specific outcome measure

related to comorbid autism/ASD, ADHD, or severity of ID and dementia in people with a diagnosis of DS were reported. There were no exclusions related to study design. Papers were assessed for quality using the Mixed Methods Appraisal Tool (MMAT). The search identified 15 papers, publishing results from 12 studies, relating to severity of ID, DS and dementia. No papers were identified relating to pervasive developmental disorders, DS and dementia. There is limited evidence on how severity of ID impacts on the presentation, diagnosis, management or prognosis of dementia in people with DS. However, no evidence was found on comorbid pervasive developmental disorders, DS, and dementia. This paper has identified multiple areas for future research. There is an urgent need for longitudinal studies into the presentation, development and progression of dementia in people with DS ensuring the severity of ID and comorbid pervasive developmental conditions are captured regularly to understand their influence on the dementia etiology and outcome.

Anderson-Mooney, A.J., Schmitt, F.A., Head, E., Lott, I.T., & Heilman, K.M.
Gait dyspraxia as a clinical marker of cognitive decline in Down syndrome: A review of theory and proposed mechanisms
Brain and Cognition, 2016, Apr, 104, 48-57. doi: 10.1016/j.bandc.2016.02.007. Epub 2016 Feb 27.
Abstract: Down syndrome (DS) is the most common genetic cause of intellectual disability in children. With aging, DS is associated with an increased risk for Alzheimer's disease (AD). The development of AD neuropathology in individuals with DS can result in further disturbances in cognition and behavior and may significantly exacerbate caregiver burden. Early detection may allow for appropriate preparation by caregivers. Recent literature suggests that declines in gait may serve as an early marker of AD-related cognitive disorders; however, this relationship has not been examined in individuals with DS. The theory regarding gait dyspraxia and cognitive decline in the general population is reviewed, and potential applications to the population with individuals with DS are highlighted. Challenges and benefits in the line of inquiry are discussed. In particular, it appears that gait declines in aging individuals with DS may be associated with known declines in frontoparietal gray matter, development of AD-related pathology, and white matter losses in tracts critical to motor control. These changes are also potentially related to the cognitive and functional changes often observed during the same chronological period as gait declines in adults with DS. Gait declines may be an early marker of cognitive change, related to the development of underlying AD-related pathology, in individuals with DS. Future investigations in this area may provide insight into the clinical changes associated with development of AD pathology in both the population with DS and the general population, enhancing efforts for optimal patient and caregiver support and propelling investigations regarding safety/quality of life interventions and disease-modifying interventions.

Andrews, E.J., Martini, A.C., & Head, E.
Exploring the role of sex differences in Alzheimer's disease pathogenesis in Down syndrome.
Frontiers in Neuroscience, 2022 Aug 12;16:954999. doi: 10.3389/fnins.2022.954999.
Abstract: Women are disproportionately affected by Alzheimer's disease (AD), yet little is known about sex-specific effects on the development of AD in the Down syndrome (DS) population. DS is caused by a full or partial triplication of chromosome 21, which harbors the amyloid precursor protein (APP) gene, among others. The majority of people with DS in their early- to mid-40s will accumulate sufficient amyloid-beta (Aß) in their brains along with neurofibrillary tangles (NFT) for a neuropathological diagnosis of AD, and the triplication of the APP gene is regarded as the main cause. Studies addressing sex differences with age and impact on dementia in people with DS are inconsistent. However, women with DS experience earlier age of onset of menopause, marked by a drop in estrogen, than women without DS. This review focuses on key sex differences observed with age and AD in people with DS and a discussion of possible underlying mechanisms that could be driving or protecting from AD development in DS. Understanding how biological sex influences the brain will lead to development of dedicated therapeutics and interventions to improve the quality of life for people with DS and AD.

📖 **Antonangeli, J.M.**
Of two minds: A guide to the care of people with the dual diagnosis of Alzheimer's Disease and mental retardation.
167 pp.
Malden, Mass.: Cooperative for Human Services [110 Pleasant Street, Malden, MA 02148] (1995)
Abstract: Written in training manual format, this text covers a range of topics related to dementia among persons with intellectual disabilities, including the notions behind dementia, structuring physical environments, safety and control issues, communication strategies, assessing and aiding with activities of daily living, behavior management strategies, medical concerns, and aiding carers. Much of the text is drawn from general practice in the Alzheimer's field with reference to application for settings with persons with intellectual disabilities.

Antonangeli. J.M.
The Alzheimer project: formulating a model of care for persons with Alzheimer's disease and mental retardation
American Journal of Alzheimer's Disease, 1995, 10(4), 13-16.
Abstract: Article speaks to a pilot project conducted in Massachusetts to increase staffing, education and Alzheimer case management supports. Special supports were designed and offered to a number of adults with Down syndrome affected by dementia, including specialize assessments, team care planning meetings, home adaptations and behavior loss supports.

Antonarakis, S.E., Skotko, B.G, Rafii, M.S., Strydom, A., Pape, S.E., Bianchi, D.W., Sherman, S.L., & Reeves, R.H.
Down syndrome
Nature Reviews Disease Primers, 2020 Feb 6, 6(1), 9. doi: 10.1038/s41572-019-0143-7.
Abstract: Trisomy 21, the presence of a supernumerary chromosome 21, results in a collection of clinical features commonly known as Down syndrome (DS). DS is among the most genetically complex of the conditions that are compatible with human survival post-term, and the most frequent survivable autosomal aneuploidy. Mouse models of DS, involving trisomy of all or part of human chromosome 21 or orthologous mouse genomic regions, are providing valuable insights into the contribution of triplicated genes or groups of genes to the many clinical manifestations in DS. This endeavor is challenging, as there are >200 protein-coding genes on chromosome 21 and they can have direct and indirect effects on homeostasis in cells, tissues, organs and systems. Although this complexity poses formidable challenges to understanding the underlying molecular basis for each of the many clinical features of DS, it also provides opportunities for improving understanding of genetic mechanisms underlying the development and function of many cell types, tissues, organs and systems. Since the first description of trisomy 21, we have learned much about intellectual disability and genetic risk factors for congenital heart disease. The lower occurrence of solid tumors in individuals with DS supports the identification of chromosome 21 genes that protect against cancer when overexpressed. The universal occurrence of the histopathology of Alzheimer disease and the high prevalence of dementia in DS are providing insights into the pathology and treatment of Alzheimer disease. Clinical trials to ameliorate intellectual disability in DS signal a new era in which therapeutic interventions based on knowledge of the molecular pathophysiology of DS can now be explored; these efforts provide reasonable hope for the future.

Apostolou, A., Kennedy, J.L., Person, M.K., Jackson, E.M.J., Finke, B., McGuire,L.C., Matthews, K.A.
Alzheimer's disease and related dementia diagnoses among American Indian and Alaska Native adults aged ≥45 years,Indian Health Service System, 2016-2020
Journal of the American Geriatrics Society, 2024 Sep, 72(9), C1, 2633-2957. https://agsjournals.onlinelibrary.wiley.com/doi/epdf/10.1111/jgs.19058
Abstract: Alzheimer's disease is the most common type of dementia and isresponsible for up to 80% of dementia diagnoses and is the sixth leading cause ofdeath in the United States. An estimated 38,000 American Indian/Alaska Native(AI/AN) people aged ≥65 years were living with Alzheimer's disease and relateddementias (ADRD) in 2020, a number expected to double by 2030 and quadrupleby 2050. Administrative healthcare data from the Indian Health Service (IHS)were used to estimate ADRD among AI/AN populations.Methods: Administrative IHS healthcare data from federal fiscal years 2016 to2020 from the IHS National Data Warehouse were used to calculate the countand rate per 100,000 AI/AN adults aged ≥45 years with at least one ADRDdiagnosis code on their medical record. This study identified 12,877 AI/AN adults aged ≥45 years with anADRD diagnosis code, with an overall rate of 514 per 100,000. Of those, 1856 peo-ple were aged 45-64. Females were 1.2 times (95% confidence

interval: 1.1-1.2) more likely than males to have a medical visit with an ADRD diagnosis code. [Note unrelated to ID, but useful for Dx dementia in this population.]

Arvio, M., Ajasto, M., Koskinen. J., & Louhiala, L
Middle-aged people with Down syndrome receive good care in home towns (In Finnish).
Finnish Medical Journal, 2013, 15, 1108-1112.
Abstract: Not available

Arvio, M., & Bjelogrlic-Laakso, N.
Down syndrome - Onset age of dementia.
Journal of Alzheimer's Disease & Parkinsonism, 2017, 7(3), 1000329. doi: 10.4172/2161-0460
Abstract: Alzheimer's disease is the most common cause of death in people who have Down syndrome (DS).This prospective, population-based, 15-year follow-up study aimed to define the onset age of dementia. At baseline 98 adults were screened for the first time by using the Present Psychiatric State-Learning Disabilities assessment;19 also participated a second parallel follow up survey. These screenings were repeated twice more during the study. The number of study subjects during the 15 year follow-up period decreased. Another study found that adaptive skills in people with DS start to deteriorate at the age of 35 and our current findings indicate that also the first signs of dementia appear at the same age. The indicative signs for dementia increased rapidly after the age of 35 and appeared most frequently as reduced self-care skills, loss of energy, impaired understanding and forgetfulness. Regular follow-up of people who have Down syndrome from the age of 30 onward enables appropriate interventions to delay the progression of dementia. Early recognition of dementia is of uttermost importance, because any deterioration in daily performance due to some treatable but undiagnosed condition may result in more restrictive residential care. Once memory changes have been observed, they may not reflect the onset of AD (depression, sleep disorders, B12-vitamin deficiency, hypothyroidism, urinary tract infections or polypharmacy may be the underlying causes behind the first dementia signs. When periodic assessments are completed at regular intervals by a carer (who is familiar with the person for a long time) explains why the first dementia signs were recognized at clearly younger age in our study than in other reported surveys (i.e., 35 versus 48-56 years).

Arvio, M., & Bjelogrlic-Laakso. N.
Screening of dementia indicating signs in adults with intellectual disabilities.
Journal of Applied Research in Intellectual Disabilities, 2021 Nov; 34(6), 1463-1467. doi: 10.1111/jar.12888. Epub 2021 May 1.
Abstract: In intellectual disability, the cognitive delay is observed during developmental age, whereas in dementia, cognitive decline occurs during post-developmental period. So far, the risk of dementia in people with intellectual disability, excluding those with Down syndrome, is poorly known. Authors screened dementia signs in a study group of 230 adults (34-80 years of age) with the help of the British Present Psychiatric State-Learning Disabilities assessment. Of the study members, 42% showed two or more signs. The overall frequency of symptoms did not differ between age groups. The number of individuals with a genetic syndrome or disease manifesting with a shortened lifespan was greater in the younger age groups when compared to the older age groups. People with an intellectual disability represent numerous rare syndromes with comorbidities. It seems that dementia signs may affect any age groups of adults with intellectual disability.

Aschenbrenner, A.J,, Baksh, R.A., Benejam, B., Beresford-Webb, J.A., Coppus. A., Fortea, J., Handen, B.L., Hartley, S., Head, E., Jaeger, J., Levin, J., Loos;i. S.V., Rebillat, A-S., Sacco, S., Schmitt, F.A., Thurlow, K.E., Zaman, S., Hassenstab, J., & Strydom, A.
Markers of early changes in cognition across cohorts of adults with Down syndrome at risk of Alzheimer's disease
Alzheimer's and Dementia (Amsterdam, Netherlands), 2021,13(1), e12184. doi: 10.1002/dad2.12184
Abstract: Down syndrome (DS), a genetic variant of early onset Alzheimer's disease (AD), lacks a suitable outcome measure for prevention trials targeting pre-dementia stages. We used cognitive test data collected in several longitudinal aging studies internationally from 312 participants with DS without dementia to identify composites that were sensitive to change over time. We then conducted additional analyses to provide support for the utility of the composites. The composites were presented to an expert panel to determine the most optimal cognitive battery based on predetermined criteria. There were common cognitive domains across site composites, which were sensitive to early decline. The final composite consisted of memory, language/executive functioning, selective attention, orientation, and praxis tests. We have identified a composite that is sensitive to early decline and thus may have utility as an outcome measure in trials to prevent or delay symptoms of AD in DS.

Atri, A., Dickerson, B.C., Clevenger, C., Karlawish, J., Knopman, D., Lin, P.J., Norman, M., Onyike, C., Sano, M., Scanland, S., & Carrillo, M.
Alzheimer's Association clinical practice guideline for the Diagnostic Evaluation, Testing, Counseling, and Disclosure of Suspected Alzheimer's Disease and Related Disorders (DETeCD-ADRD): Executive summary of recommendations for primary care.
Alzheimer's & Dementia, 2025 Jun, 21(6), e14333. doi: 10.1002/alz.14333. Epub 2024 Dec 23.
Abstract: US clinical practice guidelines for the diagnostic evaluation of cognitive impairment due to Alzheimer's disease (AD) or AD and related dementias (ADRD) are decades old and aimed at specialists. This evidence-based guideline was developed to empower all-including primary care-clinicians to implement a structured approach for evaluating a patient with symptoms that may represent clinical AD/ADRD. Through a modified-Delphi approach and guideline-development process (7374 publications were reviewed; 133 met inclusion criteria) an expert workgroup developed recommendations as steps in a patient-centered evaluation process. This summary focuses on recommendations, appropriate for any practice setting, forming core elements of a high-quality, evidence-supported evaluation process aimed at characterizing, diagnosing, and disclosing the patient's cognitive functional status, cognitive-behavioral syndrome, and likely underlying brain disease so that optimal care plans to maximize patient/care partner dyad quality of life can be developed; a companion article summarizes specialist recommendations. If clinicians use this guideline and health-care systems provide adequate resources, outcomes should improve in most patients in most practice settings. Highlights US clinical practice guidelines for the diagnostic evaluation of cognitive impairment due to Alzheimer's disease (AD) or AD and related dementias (ADRD) are decades old and aimed at specialists. This evidence-based guideline was developed to empower all-including primary care-clinicians to implement a structured approach for evaluating a patient with symptoms that may represent clinical AD/ADRD. This summary focuses on recommendations, appropriate for any practice setting, forming core elements of a high-quality, evidence-supported evaluation process aimed at characterizing, diagnosing, and disclosing the patient's cognitive functional status, cognitive-behavioral syndrome, and likely underlying brain disease so that optimal care plans to maximize patient/care partner dyad quality of life can be developed; a companion article summarizes specialist recommendations. If clinicians use this guideline and health-care systems provide adequate resources, outcomes should improve in most patients in most practice settings.

Atri, A., Dickerson, B.C., Clevenger, C., Karlawish, J., Knopman, D., Knopman, D., Lin, P.J., Norman, M., Onyike, C., Sano, M., Scanland, S., & Carrillo, M
The Alzheimer's Association clinical practice guideline for the diagnostic evaluation, testing, counseling, and disclosure of suspected Alzheimer's disease and related disorders (DETeCD-ADRD):Validated clinical assessment instruments
Alzheimer's & Dementia, 2025 Jan, 21(1), e14335. doi: 10.1002/alz.14335. Epub 2024 Dec 23.
Abstract: US clinical practice guidelines for the diagnostic evaluation of cognitive impairment due to Alzheimer's disease (AD) or AD and related dementias (ADRD) are decades old and aimed at specialists. This evidence-based guideline was developed to empower all—including primary care—clinicians to implement a structured approach for evaluating a patient with symptoms that may represent clinical AD/ADRD. Through a modified-Delphi approach and guideline-development process (7374 publications were reviewed; 133 met inclusion criteria) an expert workgroup developed recommendations as steps in a patient-centered evaluation process. This summary focuses on recommendations, appropriate for any practice setting, forming core elements of a high-quality, evidence-supported evaluation process aimed at characterizing, diagnosing,

and disclosing the patient's cognitive functional status, cognitive–behavioral syndrome, and likely underlying brain disease so that optimal care plans to maximize patient/care partner dyad quality of life can be developed; a companion article summarizes specialist recommendations. If clinicians use this guideline and health-care systems provide adequate resources, outcomes should improve in most patients in most practice settings.

Ausserhofer, D., Deschodt, M., De Geest, S., van Achterberg, T., Meyer, G., Verbeek, H., Sjetne, I.S., Malinowska-Lipien, I., Griffiths, P., Schlüter, W., Ellen, M., & Engberg, S.
"There's No Place Like Home": A scoping review on the impact of homelike residential care models on resident-, family-, and staff-related outcomes.
Journal of the American Medical Directors Association, 2016 Aug 1, 17(8), 685-93. doi: 10.1016/j.jamda.2016.03.009. Epub 2016 Apr 26.
Abstract: There is increasing emphasis on promoting "homelike" residential care models enabling care-dependent people to continue living in a self-determined manner. Yet, little is known about the outcomes of homelike residential care models. The authors aimed to (1) identify homelike residential care models for older care-dependent people with and without dementia, and (2) explore the impact of these models on resident-, family-, and staff-related outcomes. They applied a scoping review method and conducted a comprehensive literature search in PubMed, Embase, and CINAHL in May 2015. They included 14 studies, reported in 21 articles. Studies were conducted between 1994 and 2014, most using a quasi-experimental design and comparing the Eden Alternative (n = 5), nondementia-specific small houses (eg Green House homes) (n = 2), and dementia-specific small houses (n = 7) with usual care in traditional nursing homes. The studies revealed evidence of benefit related to physical functioning of residents living in dementia-specific small houses and satisfaction with care of residents living in nondementia-specific small houses compared with those living in traditional nursing homes. We did not find other significant benefits related to physical and psychosocial outcomes of residents, or in family- and staff-related outcomes. The current evidence on homelike residential care models is limited. Comparative-effectiveness research building on a clear theoretical framework and/or logic model and including a standardized set of resident-, family-, and staff-related outcomes, as well as cost evaluation, is needed to provide a stronger evidence base to justify the uptake of more homelike residential care models.

Australian Institute of Health and Welfare
Dementia among people with intellectual disabilities
Web report, 28 Mar 2024, https://www.aihw.gov.au/reports/dementia/dementia-in-aus/contents/dementia-in-priority-groups/dementia-people-intellectual-disabilities
Abstract: (none) People with intellectual disability are at a higher risk of developing dementia than the general Australian population. Down syndrome in particular carries a higher risk of dementia, specifically for Alzheimer's disease. Less research has been undertaken on dementia in Australians with other intellectual and physical disabilities, but the findings show the onset of dementia in people with intellectual disabilities (other than Down syndrome) is on average 10 years younger than the general population. While the strongest risk factor for dementia in people with intellectual disabilities is having Down syndrome, other factors found to increase the risk of dementia include: (a) poor physical and mental health, in particular depression and epilepsy, (b) poor cardiovascular health, which can be common in this group, and (c) a high rate of sensory impairments, including vision and hearing loss, and undiagnosed impairments. There are also many factors known to increase the risk of dementia in the general population that are common in people with intellectual disability, such as poor diet and exercise, poorer social, employment and education engagement and head injury.

Auty, E., & Scior, K.
Psychologists' clinical practices in assessing dementia in individuals with Down syndrome
Journal of Policy and Practice in Intellectual Disabilities, 2008, 5(4), 259-268. DOI:10.1111/j.1741-1130.2008.00187.x
Abstract: There are now ample guidelines for the assessment and diagnosis of possible dementia in individuals with intellectual disabilities (ID) and Down syndrome. However, little is known about their implementation in clinical practice. This study set out to examine the clinical practice of one key professional group, namely clinical psychologists. A national survey of clinical psychologistsin ID services in the United Kingdom was undertaken. Detailed descriptions of clinical practice were obtained from 64 psychologists. Responses were further explored in focus groups. The results suggest marked variability in practice, assessment methods, and explanations clinicians give to service users and carers. Clinicians described struggling with the ethics and practicalities of how to present dementia assessments to individuals with ID and highlighted a need for more research, debate, and guidance. They also noted numerous shortfalls in service provision once aging individuals with ID show signs of dementia. Further research and clearer consensus guidelines are needed to improve assessment and diagnosis for this group.

Axmon, A., Björne, P., Nylander, L., & Ahlström, G.
Psychiatric diagnoses in older people with intellectual disability in comparison with the general population: a register study
Epidemiology and Psychiatric Sciencies, 2018 Oct, 27(5), 479-491. doi: 10.1017/S2045796017000051
Abstract: To describe the occurrence of psychiatric diagnoses in a specialist care setting in older people with intellectual disability (ID) in relation to those found in the same age group in the general population. A cohort of people with ID (n = 7936), aged 55 years or more in 2012, was identified, as was an age and sex-matched cohort from the general population (n = 7936). Information regarding psychiatric diagnoses during 2002–2012 was collected from the National Patient Register, which contains records from all inpatient care episodes and outpatient specialist visits in Sweden. The mean age at the start of data collection (i.e. January 1st, 2002) was 53 years (range 44–85 years).
Seventeen per cent (n = 1382) of the people in the ID cohort had at least one psychiatric diagnosis recorded during the study period. The corresponding number in the general population cohort was 10% (n = 817), which translates to an odds ratio (OR) of 1.84. The diagnoses recorded for the largest number of people in the ID cohort were 'other' (i.e. not included in any of the diagnostic groups) psychiatric diagnoses (10% of the cohort had at least one such diagnosis recorded) and affective disorders (7%). In the general population cohort, the most common diagnoses were affective disorders (4%) and alcohol/substance-abuse-related disorders (4%). An increased odds of having at least one diagnosis was found for all investigated diagnoses except for alcohol/substance-abuse-related disorders (OR = 0.56). The highest odds for the ID cohort was found for diagnosis of psychotic disorder (OR = 10.4) followed by attention deficit/hyperactive disorder (OR = 3.81), **dementia** (OR = 2.71), personality disorder (OR = 2.67), affective disorder (OR = 1.74) and anxiety disorder (OR = 1.36). People with ID also had an increased odds of psychiatric diagnoses not included in any of these groups (OR = 8.02). The percentage of people with ID who had at least one diagnosis recorded during the study period decreased from more than 30% among those aged 55–59 years in 2012 (i.e. born 1953–1957) to approximately 20% among those aged 75+ years in 2012 (i.e. born in or before 1937). Older people with ID seem to be more likely to have psychiatric diagnoses in inpatient or outpatient specialist care than their peers in the general population. If this is an effect of different disorder prevalence, diagnostic difficulties or differences in health care availability remains unknown. More research is needed to understand the diagnostic and treatment challenges of psychiatric disorders in this vulnerable group.

Axmon, A., Karlsson, B., & Ahlström, G.
Health care utilisation among older persons with intellectual disability and dementia: a registry study
Journal of Intellectual Disability Research, 2016, Dec, 60(12), 1165-1177. doi 10.1111/jir.12338. Epub 2016 Oct 11.
Abstract: Both persons with intellectual disability (ID) and persons with dementia have high disease burdens, and consequently also high health care needs. As life expectancy increases for persons with ID, the group of persons with the dual diagnosis of ID and dementia will become larger. Through national registries, we identified 7936 persons who had received support directed to persons with ID during 2012, and an age- and gender-matched sample from the general population. A national registry was also used to collect information on health care utilisation (excluding primary care) for the period 2002-2012. Health care utilization was measured as presence and number of planned and unplanned in-patient and out-patient visits, as well as length of stay. In comparison with persons with ID but without dementia, persons with ID and dementia were more likely to have at least one planned out-patient visit (odds ratio [OR] 8.07), unplanned out-patient visit (OR 2.41), planned in-patient visit (OR 2.76) or unplanned in-patient visit (OR 4.19). However, among those with at least one of

each respective outcome, the average number of visits did not differ between those with and without dementia. Persons with ID and dementia were less likely to have at least one planned out-patient visit than persons with dementia in the general population sample (OR 0.40), but more likely to have at least one unplanned in-patient visit (OR 1.90). No statistically significant differences were found for having at least one unplanned out-patient or planned in-patient visit. Nevertheless, among those with at least one unplanned out-patient visit, the number of visits was higher in the general population sample. Persons with ID and **dementia** are less likely to receive planned health care than persons with dementia in the general population. They have, however, higher levels of unplanned health care utilization. This may be an indication that the current support system is not sufficient to meet the challenges of increased longevity among persons with ID.

Aylward, E., Burt, D., Thorpe, L., Lai. & Dalton, A.J.
Diagnosis of dementia in individuals with intellectual disability:
Journal of Intellectual Disability Research, 1997, 41(Part 2), 152-164.
https://doi.org/10.1111/j.1365-2788.1997.tb00692.x
Abstract: The foremost impediment to progress in the understanding and treatment of dementia in adults with intellectual disability is the lack of standardized criteria and diagnostic procedures. Standardized criteria for the diagnosis of dementia in individuals with intellectual disability are proposed, and their application is discussed. In addition, procedures for determining whether or not criteria are met in individual cases are outlined. It is the intention of the authors, who were participants of an International Colloquium on Alzheimer Disease and Mental Retardation, that these criteria be appropriate for use by both clinicians and researchers. Their use will improve communication among clinicians and researchers, and will allow researchers to test hypotheses concerning discrepancies in findings among research groups (e.g. dementia prevalence ranges and age of onset). {Report of the Task Force for Development of Criteria for the Diagnosis of Dementia in Individuals with [intellectual disability]}.

Baksh, R.A., Pape, S.E., Chan, L.F., Aslam, A.A., Gulliford, M.C., Strydom, A., et al.
Multiple morbidity across the lifespan in people with Down syndrome or intellectual disabilities: a population-based cohort study using electronic health records.
The Lancet Public Health, April 26, 2023.
https://doi.org/10.1016/S2468-2667(23)00057-9.
Abstract: The Down syndrome phenotype is well established, but our understanding of its morbidity patterns is limited. We comprehensively estimated the risk of multiple morbidity across the lifespan in people with Down syndrome compared with the general population and controls with other forms of intellectual disability. In this matched population-based cohort-study design, we used electronic health-record data from the UK Clinical Practice Research Datalink (CRPD) from Jan 1, 1990, to June 29, 2020. We aimed to explore the pattern of morbidities throughout the lifespan of people with Down syndrome compared with people with other intellectual disabilities and the general population, to identify syndrome-specific health conditions and their age-related incidence. We estimated incidence rates per 1000 person-years and incidence rate ratios (IRRs) for 32 common morbidities. Hierarchical clustering was used to identify groups of associated conditions using prevalence data. Between Jan 1, 1990, and June 29, 2020, a total of 10 204 people with Down syndrome, 39 814 controls, and 69 150 people with intellectual disabilities were included. Compared with controls, **people with Down syndrome had increased risk of dementia (IRR 94·7, 95% CI 69·9–128·4),** hypothyroidism (IRR 10·6, 9·6–11·8), epilepsy (IRR 9·7, 8·5–10·9), and haematological malignancy (IRR 4·7, 3·4–6·3), whereas asthma (IRR 0·88, 0·79–0·98), cancer (solid tumour IRR 0·75, 0·62–0·89), ischaemic heart disease (IRR 0·65, 0·51–0·85), and particularly hypertension (IRR 0·26, 0·22–0·32) were less frequent in people with Down syndrome than in controls. Compared to people with intellectual disabilities, risk of dementia (IRR 16·60, 14·23–19·37), hypothyroidism (IRR 7·22, 6·62–7·88), obstructive sleep apnoea (IRR 4·45, 3·72–5·31), and haematological malignancy (IRR 3·44, 2·58–4·59) were higher in people with Down syndrome, with reduced rates for a third of conditions, including new onset of dental inflammation (IRR 0·88, 0·78–0·99), asthma (IRR 0·82, 0·73–0·91), cancer (solid tumour IRR 0·78, 0·65–0·93), sleep disorder (IRR 0·74, 0·68–0·80), hypercholesterolaemia (IRR 0·69, 0·60–0·80), diabetes (IRR 0·59, 0·52–0·66), mood disorder (IRR 0·55, 0·50–0·60), glaucoma (IRR 0·47, 0·29–0·78), and anxiety disorder (IRR 0·43, 0·38–0·48). Morbidities in Down syndrome could be categorized on age-related incidence trajectories, and their prevalence clustered into typical syndromic conditions, cardiovascular diseases, autoimmune disorders, and mental health conditions. Multiple morbidity in Down syndrome shows distinct patterns of age-related incidence trajectories and clustering that differ from those found in the general population and in people with other intellectual disabilities, with implications for provision and timing of health-care screening, prevention, and treatment for people with Down syndrome.

Ball, S.L., Holland, A.J., Hon, J., Huppert, F.A., Treppner, P., & Watson, P.C.
Personality and behaviour changes mark the early stages of Alzheimer's disease in adults with Down's syndrome: findings from a prospective population-based study.
International Journal of Geriatric Psychiatry, 2006, 21(7), 661-673
Abstract: Research based on retrospective reports by carers suggests that the presentation of dementia in people with Down syndrome may differ from that typical of Alzheimer's disease (AD) in the general population, with the earliest changes tending to be in personality or behavior rather than in memory. This is the first long-term prospective study to test the hypothesis that such changes, which are more typical of dementia of frontal type (DFT) in the general population, mark the preclinical stage of AD in DS. A previously identified population sample of older people with DS, first assessed in 1994 and followed-up 18 months later, were reassessed after a further 5 years. This study focuses on the 55 individuals who took part in the second follow-up. Dementia diagnosis was made using the modified CAMDEX informant interview and neuropsychological assessment was undertaken using the CAMCOG. Progression in clinical presentation was examined and degree of cognitive decline over time (on the CAMCOG and derived measures of executive function (EF) and memory) was compared across groups based on diagnosis and age: AD, DFT, personality/behavior changes insufficient for a diagnosis of DFT (PBC), no diagnosis <50 years and no diagnosis 50 + years. Progression was observed from early changes in personality and behavior to an increase in characteristics associated with frontal lobe dysfunction and/or a deterioration in memory, prior to the development of full AD. Individuals who met criteria for DFT were significantly more likely to progress to a diagnosis of AD over the following 5 years than those who did not and those with PBC were significantly more likely to progress to a more severe diagnosis (DFT or AD) than those without. In the 5 years prior to diagnosis, participants with PBC and DFT had shown a degree of global cognitive decline intermediate between those with no dementia and those with AD. Both these groups had shown a significant decline in EF but not in memory, while the AD group had shown significant decline on both measures, with a significantly greater degree of decline in memory. Older participants without informant reported changes showed a more generalized pattern of decline. These findings confirm that the early presentation of AD in DS is characterized by prominent personality and behavior changes, associated with executive dysfunction, providing support for the notion that the functions of the frontal lobes may be compromised early in the course of the disease in this population. This has important implications for the diagnosis, treatment and management of dementia in people with DS.

Ball, S.L., Holland, A.J., Huppert, F.A. Treppner, P., Watson, P.C., & Hon, J.
The modified CAMDEX informant interview is a valid and reliable tool for use in the diagnosis of dementia in adults with Down's syndrome
Journal of Intellectual Disability Research, 2004 Sep;48(Pt 6):611-20.
doi:10.1111/j.1365-2788.2004.00630.x.
Abstract: Dementia because of Alzheimer's disease (AD) commonly affects older adults with Down's syndrome (DS). Methods are needed, with established concurrent and predictive validity, to facilitate the diagnostic assessment of dementia, when it is complicated by pre-existing intellectual disabilities (ID). We report on the reliability and validity of a modified version of the Cambridge Examination for Mental Disorders of the Elderly (CAMDEX) informant interview, for use when assessing people with DS suspected as having dementia. As part of a previous epidemiological study of older people with DS, the CAMDEX informant interview was used to determine the prevalence of dementia. The 74 people with DS included at that time (Time 1) had also completed the Cambridge Cognitive Examination (CAMCOG), the neuropsychological assessment from the CAMDEX schedule. Fifty-six were assessed again 6 years later (Time 2). Based on the CAMDEX informant interview, nine of the 74 at Time 1, and 11 of the 56 at Time 2, were found to meet clinical criteria for AD. Forty-one scored above floor on the CAMCOG at Time 1 and were included in the analysis of cognitive

decline. Concurrent validity was established by comparing diagnosis at Time 2 with independent evidence of objective decline on cognitive tasks since Time 1. Predictive validity was established by examining how accurately diagnosis at Time 1 predicted both cognitive decline and future diagnosis. Inter-rater reliability was determined by comparing the level of agreement between two raters. CAMDEX-based diagnosis of AD was shown to be consistent with objectively observed cognitive decline (good concurrent validity) and to be a good predictor of future diagnosis. Although numbers are small, some support is also provided for the accuracy with which diagnosis predicts cognitive decline. Inter-rater reliability was good with Kappa > 0.8 for 91% of items and > 0.6 for all items. The use of the modified CAMDEX informant interview enables the structured collection of diagnostic information, so that a valid and a reliable diagnosis of dementia can be made in those with pre-existing ID, using established diagnostic criteria.

Ball, S.L., Holland, A.J., Treppner, P., Watson, P.C., & Huppert, F.A.
Executive dysfunction and its association with personality and behaviour changes in the development of Alzheimer's disease in adults with Down syndrome and mild to moderate learning disabilities.
British Journal of Clinical Psychology, 2008, 47(Pt 1), 1-29. doi: 10.1348/014466507X230967.
Abstract: Recent research suggests that preclinical Alzheimer's disease (AD) in people with Down syndrome (DS) is characterized by changes in personality/behavior and executive dysfunction that are more prominent than deterioration in episodic memory. This study examines the relationship between executive dysfunction and the clinical and preclinical features of AD in DS. To determine the specificity of this relationship, performance on executive function (EF) measures is contrasted with performance on memory measures. One hundred and three people with DS (mean age 49 years, range 36-72) with mild to moderate learning disabilities (LD) took part. Dementia diagnosis was based on the CAMDEX informant interview conducted with each participant's main carer. Reported changes in personality/behavior and memory were recorded. Participants completed six EF and six memory measures (two of which also had a strong executive component) and the BPVS (as a measure of general intellectual ability). First, performance was compared between those with and without established dementia of Alzheimer's type (DAT), controlling for age and LD severity using ANCOVA. Next, the degree to which informant-reported changes predicted cognitive test performance was examined within the non-DAT group using multiple regression analyses. The DAT group (N=25) showed a consistent pattern of impaired performance relative to the non-DAT group (N=78), across all measures. Within the non-DAT group, number of informant-reported personality/behavior changes was a significant predictor of performance on two EF and two 'executive memory' tests (but not on episodic memory tests). Informant-reported memory changes, however, were associated with impaired performance on a delayed recall task only. These findings provide further evidence for a specific impairment in frontal-lobe functioning in the preclinical stages of AD in DS.

Ballard, C., Mobley, W., Hardy, J., Williams, G., & Corbett, A.
Dementia in Down's syndrome.
Lancet Neurology, 2016 May;15(6):622-36. doi: 10.1016/S1474-4422(16)00063-6. Epub 2016 Apr 11.
Abstract: Down syndrome is the most common genetic cause of learning difficulties, and individuals with this condition represent the largest group of people with dementia under the age of 50 years. Genetic drivers result in a high frequency of Alzheimer's pathology in these individuals, evident from neuroimaging, biomarker, and neuropathological findings, and a high incidence of cognitive decline and dementia. However, cognitive assessment is challenging, and diagnostic methods have not been fully validated for use in these patients; hence, early diagnosis remains difficult. Evidence regarding the benefits of cholinesterase inhibitors and other therapeutic options to treat or delay progressive cognitive decline or dementia is very scarce. Despite close similarities with late-onset Alzheimer's disease, individuals with Down syndrome respond differently to treatment, and a targeted approach to drug development is thus necessary. Genetic and preclinical studies offer opportunities for treatment development, and potential therapies have been identified using these approaches.

Bauer, A.M., & Shea, T.M.
Alzheimer's disease and Down syndrome: A review and implications for adult services
Education and Training of the Mentally Retarded, 1986, 21, 144-150.
https://eric.ed.gov/?id=EJ341367
Abstract: In this article, the diagnosis of Alzheimer's disease and its progressive behavioral impact on persons with Down syndrome is discussed. Several implications and suggestions for care and service provision for adults with Down syndrome are presented, including that Alzheimer's disease in an adult with Down syndrome has an impact on the carer, adjusting communication strategies to correspond to the stage of dementia, aiding families to seek assistance from social agencies, stressing the remaining abilities and skills, aiding families and carers to develop realistic methods of providing care, and adapting the persons care and environment to help them cope with losses stemming from dementia. The authors also suggest proactive strategies for anticipating decline among adults with Down syndrome associated with dementia.

Baumer NT, Becker ML, Capone GT, Egan K, Fortea J, Handen BL, Head E, Hendrix JE, Litovsky RY, Strydom A, Tapia IE, Rafii MS.
Conducting clinical trials in persons with Down syndrome: summary from the NIH INCLUDE Down syndrome clinical trials readiness working group.
Journal of Neurodevelopmental Disorders, 2022 Mar 23;14(1):22. doi: 10.1186/s11689-022-09435-z.
Abstract: The recent National Institute of Health (NIH) INCLUDE (INvestigation of Co-occurring conditions across the Lifespan to Understand Down syndromE) initiative has bolstered capacity for the current increase in clinical trials involving individuals with Down syndrome (DS). This new NIH funding mechanism offers new opportunities to expand and develop novel approaches in engaging and effectively enrolling a broader representation of clinical trials participants addressing current medical issues faced by individuals with DS. To address this opportunity, the NIH assembled leading clinicians, scientists, and representatives of advocacy groups to review existing methods and to identify those areas where new approaches are needed to engage and prepare DS populations for participation in clinical trial research. This paper summarizes the results of the Clinical Trial Readiness Working Group that was part of the INCLUDE Project Workshop: Planning a Virtual Down Syndrome Cohort Across the Lifespan Workshop held virtually September 23 and 24, 2019.

Bayen, E., Possin, K.L., Chen, Y., Ckeret de Langavant, L., & Yaffe, K.
Prevalence of aging, dementia, and multimorbidity in older adults with Down syndrome
JAMA Neurology, 2018, Nov 1, 75(11), 1399-1406. doi:10.1001/jamaneurol.2018.2210.
Abstract: As the life expectancy of people with Down syndrome (DS) has markedly increased over the past decades, older adults with DS may be experiencing a higher incidence of aging conditions. In addition to longevity, the amyloid precursor protein gene located on chromosome 21 places individuals with DS at a high risk for developing Alzheimer disease. Yet, few studies have determined prevalence of dementia and comorbidities among older people with DS. To determine the prevalence of dementia and aging-related comorbidities in older adult individuals with DS. Cross-sectional analysis of 2015 California Medicare claims data. We examined 1 year of cross-sectional Medicare claims data that included 100% of Californian Medicare beneficiaries enrolled in both Medicare Part A and B in 2015. Of these 3 001 977 Californian Medicare beneficiaries 45 years or older, 878 individuals were identified as having a diagnosis of DS. Data were analyzed between April 2017 and February 2018. The frequency of DS dementia was assessed across different age categories. The number and frequency of 27 comorbidities were compared among individuals with DS with and without dementia and by age and sex groups. A total of 353 DS individuals (40%) were identified as having dementia diagnoses (mean, 58.7 years; 173 women [49%]) and 525 without dementia diagnoses (mean, 55.9 years; 250 women [48%]). The frequency of DS dementia among those 65 years or older rose to 49%. The mean number of comorbidities per individual increased with age in general. Comorbid conditions were more numerous among those with dementia compared with those with DS without dementia (mean, 3.4 vs 2.5, respectively), especially among those younger than 65 years. In particular, 4 treatable conditions, hypothyroidism, epilepsy, anemia, and weight loss, were much more frequent in DS dementia. Older Medicare beneficiaries in California with DS, especially those with dementia, have a high level of multimorbidity including several treatable conditions. While DS follow-up has long been confined to the pediatric sphere, we found that longevity in individuals with DS will necessitate complex adult and geriatric care. More

evidenced-based and standardized follow-up could support better long-term comorbidity management and dementia care among aging adults with DS.

Bayen, E., Yaffe, K., Cleret de Langavant, L., Chen, Y., & Possin, K.L.
The direct health care cost to Medicare of Down syndrome dementia as compared with Alzheimer's disease among 2015 Californian beneficiaries.
Ann Phys Rehabil Med. 2021 Jan;64(1):101430. doi: 10.1016/j.rehab.2020.07.011. Epub 2020 Oct 15.
Abstract: Aging individuals with Down syndrome (DS) are at increased risk of dementia due to trisomy of chromosome 21 on which the amyloid precursor protein gene is located and with increased life expectancy. Yet, little is known about the costs associated with DS dementia and how this compares to Alzheimer's disease (AD). To better understand direct healthcare costs and care consumption in DS dementia, we compared the total cost of care to US Medicare and the drivers of these medical expenditures in individuals with DS with and without dementia, and in those with AD without DS. The effect of dementia in DS on costs and care utilization was estimated with 2015 California Medicare fee-for-service data (parts A and B). Among 3,001,977 Californian Medicare beneficiaries, 353 individuals had DS with dementia (age 45-89 years). We compared their number of chronic comorbidity conditions among 27 and their care and Medicare costs to those of age- and sex-matched individuals with DS without dementia and those with AD without DS. Medicare annual cost per beneficiary was a mean of 43.5% and 82.2% higher with DS dementia (mean $35,011) than DS without dementia (mean $24,401) and AD without dementia (mean $19,212), related to greater utilization of inpatient services. DS dementia was associated with increased level of multimorbidity (mean of 3.4 conditions in addition to dementia vs. 2.7 and 2.2 conditions for DS without dementia and AD, respectively), with more emergency room visits (88% vs. 76.5% and 54.4%) and with more primary care physician visits (91.2% vs. 87.3% and 81.3%). DS adults with dementia have higher health care costs than DS adults without dementia and adults with AD. Understanding costs and complex health care needs in DS dementia could facilitate management of adult and geriatric care resources for these high-need high-cost individuals.

Bejanin, A., Iulita, M.F., Vilaplana, E., Carmona-Iragui, M., Benejam, B,, Videlam, L., Barroeta, I, et al.
Association of apolipoprotein E ε4 allele with clinical and multimodal biomarker changes of Alzheimer disease in adults with Down syndrome.
JAMA Neurology, doi:10.1001/jamaneurol.2021.1893. Published online July 6, 2021.
Abstract: Alzheimer disease (AD) is the leading cause of death in individuals with Down syndrome (DS). Previous studies have suggested that the APOE ε4 allele plays a role in the risk and age at onset of dementia in DS; however, data on in vivo biomarkers remain scarce. To investigate the association of the APOE ε4 allele with clinical and multimodal biomarkers of AD in adults with DS.
his dual-center cohort study recruited adults with DS in Barcelona, Spain, and in Cambridge, UK, between June 1, 2009, and February 28, 2020. Included individuals had been genotyped for APOE and had at least 1 clinical or AD biomarker measurement; 2 individuals were excluded because of the absence of trisomy 21. Participants were either APOE ε4 allele carriers or noncarriers. Participants underwent a neurological and neuropsychological assessment. A subset of participants had biomarker measurements: Aß1-42, Aß1-40, phosphorylated tau 181 (pTau181) and neurofilament light chain (NfL) in cerebrospinal fluid (CSF), pTau181, and NfL in plasma; amyloid positron emission tomography (PET); fluorine 18-labeled-fluorodeoxyglucose PET; and/or magnetic resonance imaging. Age at symptom onset was compared between APOE ε4 allele carriers and noncarriers, and within-group local regression models were used to compare the association of biomarkers with age. Voxelwise analyses were performed to assess topographical differences in gray matter metabolism and volume. Of the 464 adults with DS included in the study, 97 (20.9%) were APOE ε4 allele carriers and 367 (79.1%) were noncarriers. No differences between the 2 groups were found by age (median [interquartile range], 45.9 [36.4-50.2] years vs 43.7 [34.9-50.2] years; P = .56) or sex (51 male carriers [52.6%] vs 199 male noncarriers [54.2%]). APOE ε4 allele carriers compared with noncarriers presented with AD symptoms at a younger age (mean [SD] age, 50.7 [4.4] years vs 52.7 [5.8] years; P = .02) and showed earlier cognitive decline. Locally estimated scatterplot smoothing curves further showed between-group differences in biomarker trajectories with age as reflected by nonoverlapping CIs. Specifically, carriers showed lower levels of the CSF Aß1-42 to Aß1-40 ratio until age 40 years, earlier increases in amyloid PET and plasma pTau181, and earlier loss of cortical metabolism and hippocampal volume. No differences were found in NfL biomarkers or CSF total tau and pTau181. Voxelwise analyses showed lower metabolism in subcortical and parieto-occipital structures and lower medial temporal volume in APOE ε4 allele carriers. In this study, the APOE ε4 allele was associated with earlier clinical and biomarker changes of AD in DS. These results provide insights into the mechanisms by which APOE increases the risk of AD, emphasizing the importance of APOE genotype for future clinical trials in DS.

Benejam, B., Videla, L., Vilaplana, E., Barroeta, I., Carmona-Iragui ,M,. Altuna ,M., Valldeneu, S., Fernandez, S., Giménez, S., Iulita, F., Garzón, D., Bejanin, A., Bartrés-Faz, D., Videla, S., Alcolea, D., Blesa, R., Lleó, A., & Fortea, J.
Diagnosis of prodromal and Alzheimer's disease dementia in adults with Down syndrome using neuropsychological tests.
Alzheimers Dementia (Amst), 2020 Jun 28,12(1), e12047. doi: 10.1002/dad2.12047.
Abstract: We aimed to define prodromal Alzheimer's disease (AD) and AD dementia using normative neuropsychological data in a large population-based cohort of adults with Down syndrome (DS). We employed a cross-sectional study. DS participants were classified into asymptomatic, prodromal AD and AD dementia, based on neurologist's judgment blinded to neuropsychological data (Cambridge Cognitive Examination for Older Adults with Down's syndrome [CAMCOG-DS] and modified Cued Recall Test [mCRT]). We compared the cutoffs derived from the normative data in young adults with DS to those from receiver-operating characteristic curve (ROC) analysis. Diagnostic performance of the CAMCOG-DS and modified Cued Recall Test (mCRT) in subjects with mild and moderate levels of intellectual disability (ID) was high, both for diagnosing prodromal AD and AD dementia (area under the curve [AUC] 0.73-0.83 and 0.90-1, respectively). The cutoffs derived from the normative data were similar to those derived from the ROC analyses. Diagnosing podromal AD and AD dementia in DS with mild and moderate ID using population norms for neuropsychological tests is possible with high diagnostic accuracy.

Beresford-Webb, J.A., Mak, E., Grigorova, M., Daffern, S.J., Holland, A.J., & Zaman, S.H.
Establishing diagnostic thresholds for Alzheimer's disease in adults with Down syndrome: the Cambridge Examination for Mental Disorders of Older People with Down's Syndrome and Others with Intellectual Disabilities (CAMDEX-DS).
BJPsych Open, 2021 Apr 13, 7(3), e79. doi: 10.1192/bjo.2021.36.
Abstract: Diagnosis of prodromal Alzheimer's disease and Alzheimer's disease dementia in people with Down syndrome is a major challenge. The Cambridge Examination for Mental Disorders of Older People with Down's Syndrome and Others with Intellectual Disabilities (CAMDEX-DS) has been validated for diagnosing prodromal Alzheimer's disease and Alzheimer's disease dementia, but the diagnostic process lacks guidance. To derive CAMDEX-DS informant interview threshold scores to enable accurate diagnosis of prodromal Alzheimer's disease and Alzheimer's disease dementia in adults with Down syndrome. Psychiatrists classified participants with Down syndrome into no dementia, prodromal Alzheimer's disease and Alzheimer's disease dementia groups. Receiver operating characteristic analyses assessed the diagnostic accuracy of CAMDEX-DS informant interview-derived scores. Spearman partial correlations investigated associations between CAMDEX-DS scores, regional Aß binding (positron emission tomography) and regional cortical thickness (magnetic resonance imaging). Diagnostic performance of CAMDEX-DS total scores were high for Alzheimer's disease dementia (area under the curve (AUC), 0.998; 95% CI 0.953-0.999) and prodromal Alzheimer's disease (AUC = 0.954; 95% CI 0.887-0.982) when compared with healthy adults with Down syndrome. When compared with those with mental health conditions but no Alzheimer's disease, CAMDEX-DS Section B scores, denoting memory and orientation ability, accurately diagnosed Alzheimer's disease dementia (AUC = 0.958; 95% CI 0.892-0.984), but were unable to diagnose prodromal Alzheimer's disease. CAMDEX-DS total scores exhibited moderate correlations with cortical Aß ($r \sim 0.4$ to 0.6, $P \leq 0.05$) and thickness ($r \sim -0.4$ to -0.44, $P \leq 0.05$) in specific regions. CAMDEX-DS total score accurately diagnoses Alzheimer's disease dementia and prodromal Alzheimer's disease in healthy adults with Down syndrome.

Berry, M.
Alzheimer's and individuals with intellectual and developmental disabilities.

Impact, 2010, 23(1).
https://publications.ici.umn.edu/impact/23-1/alzheimers-and-individuals-with-intellectual-and-developmental-disabilities
Abstract: There are an estimated 5.3 million people in the U.S. with Alzheimer's disease. By the year 2030, that number is expected to increase to 7.7 million. Alzheimer's disease is the most common form of dementia, a group of symptoms characterized by loss in memory and at least one other cognitive skill. Sixty to 80% of people with dementia have Alzheimer's disease (Alzheimer's Association, 2009). The most significant risk factor for Alzheimer's is age. For most people the disease begins with difficulty remembering new or current information, and other cognitive abilities such as the ability to plan, use language, and understand time and spatial relations deteriorate as the disease progresses.

Bevens, S., Dawes, S., Kenshole, A., & Gaussen, K.
Staff views of a music therapy group for people with intellectual disabilities and dementia: A pilot study
Advances in Mental Health and Intellectual Disabilities, 2015, 9(1), 40-48. https://doi.org/10.1108/AMHID-04-2014-0005
Abstract: Despite the longstanding use of music therapy with people with intellectual disabilities and the growing evidence base for using music therapy as a tool to aid behavioural and psychological symptoms of dementia in the general population, there is little work published which details the use of music therapy groups for people with intellectual disabilities who have a diagnosis of dementia. The purpose of this paper is to report a qualitative evaluation of staff views of a music therapy group for people with intellectual disabilities and dementia. Carers of service users attending the group were interviewed either individually or through a focus group in order to ascertain their views about the music therapy group. The interview transcripts were then analysed using thematic analysis.
Two core themes and eight sub themes emerged from the data. These themes show that the group was felt to be pleasurable and enjoyable for the service users and that some tangible benefits of attending the group were observed by staff members. Notwithstanding the positive feedback, the results also suggested that more work is needed to inform carers of the goals and purpose of such groups. Further psycho-education for carers is suggested as a strategy to support future groups to run successfully. There is little published research into the use of music therapy for people with intellectual disabilities who also have dementia. The current paper provides a starting point for future work in the area and further recommendations for future practice and research are considered.

Bigby, C., & Beadle-Brown, J.
Culture in better group homes for people with intellectual disability at severe levels
Intellectual and Developmental Disabilities, 2016 Oct, 54(5), 316-331. doi: 10.1352/1934-9556-54.5.316.
Abstract: Building on cultural dimensions of underperforming group homes this study analyzes culture in better performing services. In depth qualitative case studies were conducted in 3 better group homes using participant observation and interviews. The culture in these homes, reflected in patterns of staff practice and talk, as well as artefacts differed from that found in underperforming services. Formal power holders were undisputed leaders, their values aligned with those of other staff and the organization, responsibility for practice quality was shared enabling teamwork, staff perceived their purpose as "making the life each person wants it to be," working practices were person centered, and new ideas and outsiders were embraced. The culture was characterized as coherent, respectful, "enabling" for residents, and "motivating" for staff. Though it is unclear whether good group homes have a similar culture to better ones the insights from this study provide knowledge to guide service development and evaluation.

Bigby, C., Bowers, B., & Webber, R.
Planning and decision making about the future care of older group home residents and transition to residential aged care
Journal of Intellectual Disability Research, 2011 Aug, 55(8), 777-789. doi: 10.1111/j.1365-2788.2010.01297.x
Abstract: Planning for future care after the death of parental caregivers and adapting disability support systems to achieve the best possible quality of life for people with intellectual disability as they age have been important issues for more than two decades.This study examined perceptions held by family members, group home staff, and organizational managers about the future of older residents and the decisions made that a move to residential aged care was necessary. Grounded Dimensional Analysis was used to guide data collection and analysis by an interdisciplinary research team. Three sets of interviews over a period of 18 months were conducted with a family member, house supervisor and the program manager for each of seventeen older group home residents in Victoria, Australia. For the eight people for whom it was decided a move was necessary and the six who eventually moved focused questions were asked about the decision-making process. While plans for lifelong accommodation in a group home proved unfounded, key person succession plans were effective. However, decisions that a move to a residential aged care facility was necessary were made in haste and seen as a fait accompli to involved family members. Although family members take seriously their mandate to oversee well-being of their older relative, they have little knowledge about their rights or avenues to safeguard untimely or inappropriate decisions being made by professionals.

Bigby, C., Knox, M., Beadle-Brown, J., Clement, T., & Mansell, J.
Uncovering dimensions of culture in underperforming group homes for people with severe intellectual disability
Intellectual and Developmental Disabilities, 2012 Dec, 50(6), 452-467. doi: 10.1352/1934-9556-50.06.452.
Abstract: Culture recurs as an important but under-investigated variable associated with resident outcomes in supported accommodation for people with intellectual disability. This study aimed to conceptualize the potential dimensions of culture in all group homes and describe the culture in underperforming group homes. A secondary analysis, using an inductive interpretative approach, was undertaken of a large qualitative data set from a study that had used ethnographic and action research methods to explore the quality of life outcomes for residents in 5 small group homes. Five categories were developed: misalignment of power-holder values with organizations espoused values, otherness, doing for not with, staff centered, and resistance. Differences from institutional culture are discussed, and the potential of the findings as a starting point to consider culture in high performing group homes and develop a quantitative measure of culture.

Bishop, K.M., Hogan, M., Janicki, M.P., Keller, S.M., Lucchino, R., Mughal, D.T., Perkins, E.A., Singh, B.K., Service, K., Wolfson, S. & Health Planning Work Group of the National Task Group on Intellectual Disabilities and Dementia Practices.
Guidelines for dementia-related health advocacy for adults with intellectual disability and dementia: national task group on intellectual disabilities and dementia practices.
Intellectual and Developmental Disabilities, 2015 Feb, 53(1), 22-29. doi: 10.1352/1934-9556-53.1.2
Abstract: Increasing numbers of adults with intellectual disabilities (ID) are living into old age. Though this indicates the positive effects of improved health care and quality of life, the end result is that more adults with ID are and will be experiencing age-related health problems and also exhibiting symptoms of cognitive impairment and decline, some attributable to dementia. Early symptoms of dementia can be subtle and in adults with ID are often masked by their lifelong cognitive impairment, combined with the benign effects of aging. A challenge for caregivers is to recognize and communicate symptoms, as well as find appropriate practitioners familiar with the medical issues presented by aging adults with lifelong disabilities. Noting changes in behavior and function and raising suspicions with a healthcare practitioner, during routine or ad hoc visits, can help focus the examination and potentially validate that the decline is the result of the onset or progression of dementia. It can also help in ruling out reversible conditions that may have similar presentation of symptoms typical for Alzheimer's disease and related dementias. To enable caregivers, whether family members or staff, to prepare for and advocate during health visits, the National Task Group on Intellectual Disabilities and Dementia Practices has developed guidelines and recommendations for dementia-related health advocacy preparation and assistance that can be undertaken by provider and advocacy organizations.

Bittles, A.H., & Glasson, E.J.
Clinical, social, and ethical implications of changing life expectancy in Down syndrome
Developmental Medicine & Child Neurology, 2004, 46, 282-286.
Abstract: Increased life expectancy generates greater ethical and legal dilemmas

in the treatment of people with Down syndrome. Assumptions that younger cohorts of people with DS will experience healthier lives when compared to previous generations may not be realized as specific health issues associated with DS are genetically encoded and thus contemporary generations may face the same adverse health issues. With respect to dementia, authors note that by age 60 years, dementia involving memory loss, cognitive decline, and changes in adaptive behavior may be present in at least 56% of adults with DS and that some the neuropathological features of Alzheimer disease may be evident as early as age 40.

Bjelogrlic, N.
Insights of dementia in persons with intellectual disability
Medicine & Clinical Science, 2023, 5(7):1-3.
https://www.sciencexcel.com/articles/hmYB13CSGb7zXDU3vBYQdWmxn9EHenriYYGibAaU.pdf
Abstract: Intellectual disability and dementia are age-dependent terms for a cognitive impairment occurring during the developmental age and in adulthood, respectively. Intellectually disabled people like any other people may develop dementia in adulthood. Thus, clinicians should learn to differentiate dementia-indicating signs from intellectual disability related cognitive deficiencies for an early diagnosis and treatment onwards. In intellectual disability, intellectual and adaptive skills of an individual are two standard deviations below the expected age-matched population, and dementia is characterized by a progressive cognitive decline. The cause of both disorders can be genetic, acquired, or multifactorial. An increased risk of Alzheimer's disease in Down syndrome is well known unlike the development of dementia in other intellectual disability syndromes. This commentary discusses 1) how the dementia indicating signs present in intellectually disabled persons, 2) why it is important to distinguish dementia (and its causes) from intellectual disability and 3) why it is important to know the etiology of intellectual disability.

Blesa, R., Trias, C., Fortea J., & Videla. S.
Alzheimer's disease in adults with Down syndrome: a challenge.
T21 Research Science & Society Bulletin, 2015, 2, 4
Abstract: None provided.

Boerwinkle, A. H., Gordon, B. A., Wisch, J., Flores, S., Henson, R. L., Butt, O. H., McKay, N., Chen, C. D., Benzinger, T. L. S., Fagan, A. M., Handen, B. L., Christian, B. T., Head, E., Mapstone, M., Rafii, M. S., O'Bryant, S., Lai, F., Rosas, H. D., Lee, J. H., Silverman, W., . Dominantly Inherited Alzheimer Network
Comparison of amyloid burden in individuals with Down syndrome versus autosomal dominant Alzheimer's disease: a cross-sectional study.
The Lancet Neurology, 2023, 22(1), 55-65.
https://doi.org/10.1016/S1474-4422(22)00408-2
Abstract: Due to the triplication of chromosome 21 and resultant extra copy of the amyloid precursor protein gene, most adults with Down syndrome (DS) develop Alzheimer disease (AD)-like pathology and dementia. Using positron emission tomography (PET) imaging of amyloid, we compared DS with another genetic form of AD, autosomal-dominant AD (ADAD). Participants with DS (n=192), ADAD mutation-carriers (MC) (n=265), and familial controls (n=202) with amyloid PET imaging from the Alzheimer's Biomarker Consortium-Down Syndrome and the Dominantly Inherited Alzheimer Network were included. Global and regional amyloid burdens were compared by cognitive status, APOE ε4 status, sex, age, and estimated years to symptom onset (EYO). Finally, we evaluated the relationship between amyloid PET and CSF Aß42/40 in a subset of participants.
PET and CSF amyloid were inversely correlated in DS and MC participants (? = -0•801 and -0.565 respectively; both p-values < 0•001). There were no significant differences in global amyloid burden between MC and DS when grouped by cognitive status. We also did not observe a significant effect of APOE ε4-positivity or sex in either group. Amyloid accumulation occurred slightly later in participants with DS (EYO -17•5) compared to MC (EYO -23•0) (p-value < 0•001). This difference was mainly driven by PSEN1 MCs. Early amyloid increases were seen in striatal and cortical regions in both MC and DS individuals. While widespread amyloid accumulation was seen in MC, occipital regions were spared in individuals with DS. This study demonstrates that, despite minor differences between DS and ADAD, the overall pathophysiology is similar between these two genetic forms of AD.

Bonne S, Iftimovici A, Mircher C, Conte M, Louveau C, Legrand A, Danset-Alexandre C, Cannarsa C, Debril A, Consoli A, Krebs MO, Ellul P, Chaumette B.
Down syndrome regression disorder, a case series: Clinical characterization and therapeutic approaches.
Frontiers of Neuroscience, 2023 Feb 23, 17, 1126973. doi: 10.3389/fnins.2023.1126973.
Abstract: Down syndrome (DS) is one of the most frequent genetic disorders and represents the first cause of intellectual disability of genetic origin. While the majority of patients with DS follow a harmonious evolution, an unusual neurodevelopmental regression may occur, distinct from that described in the context of autism spectrum disorders, called down syndrome regression disorder (DSRD). Based on four patients, two males and two females, with age range between 20 and 24, treated at the Reference Center for Rare Psychiatric Disorders of the GHU Paris Psychiatry and Neurosciences [Pôle hospitalo-universitaire d'Évaluation Prévention et Innovation Thérapeutique (PEPIT)], we describe this syndrome, discuss its etiologies and propose therapeutic strategies. DSRD often occurs in late adolescence. There is a sudden onset of language disorders, loss of autonomy and daily living skills, as well as behavioral symptoms such as depression, psychosis, or catatonia. These symptoms are non-specific and lead to an overlap with other diagnostic categories, thus complicating diagnosis. The etiologies of the syndrome are not clearly identified but certain predispositions of patients with trisomy 21 have suggested an underlying immune-mediated mechanism. Symptomatic therapeutic approaches (serotonergic antidepressants, atypical antipsychotics, benzodiazepines) were not effective, and generally associated with poor tolerance. Etiological treatments, including anti-inflammatory drugs and corticosteroids, led to partial or good recovery in the four cases. Early recognition of regressive symptoms and rapid implementation of adapted treatments are required to improve the quality of life of patients and their families.

Bodde AE, Brooks JV, Forseth B, Wolfe T, Williams K, Ptomey LT.
Caregiving for adults with Down syndrome: caregiver experiences and support needs.
Journal of Applied Research in Intellectual Disabilities, 2025 Sep;38(5):e70118. doi: 10.1111/jar.70118.
Abstract: Family caregivers of adults with Down syndrome often provide life-long caregiving support for their loved one. Long-term caregiving can impact caregivers' health and well-being, yet their experiences and support needs are underexplored. Semi-structured interviews were conducted with caregivers of adults with Down syndrome to understand their caregiving experiences and perceived caregiver support needs. Transcripts of the recorded interviews were coded and analyzed thematically. Seventeen family caregivers (94.1% female, Mage = 58.8 years) of adults with Down syndrome completed the interviews. We identified four major themes: constancy of caregiving, future planning, significance of social supports, and positive joys and rhythms. Authors' findings demonstrate that caregiving responsibilities can feel constant and unceasing, yet consistent routines and positive appraisal help ease the burden. Family and friends support thriving, but trusted options for transportation services, life transition planning and respite care are needed. Targeting these support needs may improve caregiver well-being.

Bowers, B., Webber, R., & Bigby, C.
Aging and health related changes of people with intellectual disabilities living in group homes in Australia.
Journal of Policy and Practice in Intellectual Disabilities, 2009, 6(2), 98. [SINGLE PAGE]
Abstract: Group homes for people with ID are based on social models, emphasizing inclusion, engagement in community, and quality of life. As age related changes occur, group home staff members are faced with decisions about how to respond, how to support people experiencing health problems, and whether or for how long people can remain in the group homes. This study explored how group home staff members respond to aging and age related health conditions in group home residents and to identify factors that put people at risk of premature or inappropriate relocation. Using a longitudinal design in order to observe, over time, the onset of health problems, the initial responses of housing staff to health, the development of health conditions, the consequences of their initial responses, and the outcomes for both staff and residents were considered. In-depth interviews were conducted—at three 6-month intervals with 18 clusters of the housing manager, family member, the person with the disability, and in

some cases, healthcare providers. A total of 91 interviews were completed, transcribed, and analyzed and in keeping with the theory-generating approach, early interviews were open and exploratory, evolving over time to facilitate comparative analysis across groups, strategies, conditions, and care issues. Staff and family members agreed that aging and the development of associated health conditions was increasingly becoming an issue for them. Significantly, there was wide variation among housing staff in terms of philosophy of care, with some believing that people should be supported to remain at the group homes for as long as possible. This, however, required the acquisition of new resources, a range of organizational changes to support staff and residents, changes to staffing patterns and levels, and a change in recruiting as a strategy to alter skill mix of house workers. Authors concluded that problems identified by most housing staff included: (a) inability of residents to retire despite age and health status; (b) risk of premature moves to aged care; and © disruption to general house activities and routines of other residents. Staff members' experienced altered work routines, concerns about the safety of residents and themselves, and frequent turnover. Availability of resources, such as equipment and home modifications, flexibility of staffing to accommodate changing resident needs, and philosophy of care all had a significant impact on residents' ability to "stay home."

Bowers, B., Webber, R., & Bigby, C.
Health issues of older people with intellectual disability in group homes.
Journal of Intellectual and Developmental Disabilities, 2014, 39(3), 261-269. doi: 10.3109/13668250.2014.936083.
Abstract: This paper explores how group home staff in Victoria, Australia, responded to residents with an intellectual disability (ID) as they developed age-related health conditions. The analysis was based on a longitudinal study that followed 17 ageing group home residents over a 3-year period. Eighty-three interviews were conducted with 30 group home staff in 17 group homes. Dimensional analysis, a variant of grounded theory, guided data collection and analysis. Findings revealed that the organizations all had systems in place to address health issues. However, the results also suggest an inability of staff to differentiate between significant health conditions and normal age-related changes, thus contributing to delays in care for serious medical conditions. Lack of knowledge about normal ageing and an absence of organizational policies influence timeliness of diagnosis and treatment for people with ID. Group home staff could be more effective advocates for older residents, leading to improvements in health outcomes, if they had basic knowledge about normal ageing and symptoms of common age-related illnesses and if group home agencies provided clearer guidance to their staff. The study has implications for staff education and organizational policy development for group homes.

Bowey, L. & McGlaughlin, A.
Adults with a learning disability living with elderly carers talk about planning for the future: Aspirations and concerns.
British Journal of Social Work, 2005, 35(8), 1377-1392. DOI: https://doi.org/10.1093/bjsw/bch241
Abstract: The majority of adults with an intellectual disability live with family carers, many of whom are ageing and have support needs of their own. Planning for the future thus becomes the key to preventing a crisis situation when family care is no longer viable because of death or ill health. Existing knowledge and practice are largely based upon the perspective of professionals and carers. This study explored the views, aspirations and concerns of adults with an intellectual disability, about living at home and planning for the future. Findings show that participants were very aware of the need for alternative housing or support in the future and had clear preferences about their future options. However, they also showed extensive concern for their family carers and this often impacted on their willingness to plan for the future or to move to alternative housing. Their demonstrable awareness of the inevitable death or ill health of family carers, and willingness to engage with the implications, emphasize the importance of involving adults with intellectual disability in planning for their future, as well as providing them with bereavement support.

Bratek, A., Krysta, K., & Kucia, K.
Psychiatric comorbidity in older adults with intellectual disability
Psychiatria Danubina, 2017, 29(Suppl 3), 590-593. PMID: 28953835
Abstact: The population of older adults with intellectual disability (ID) is large and growing due to a significant increase of life expectancy caused by improvements in health and social care. Multimorbidity is highly prevalent in this population and co-morbid psychiatric disorders are especially frequent. This article provides a review of the prevalence and consequences of psychiatric comorbidity in the population of older adults with ID. We therefore performed a literature search of studies relevant to adults with ID, published since January 2006, using the following keywords: intellectual disability and comorbidity, intellectual disability and mental disorders, intellectual disability and polypharmacy. Psychiatric comorbidity is frequent among patients with ID and correlates with older age. Mental disorders are present in up to 40% of older adults with ID and the most prevalent are challenging behaviour, depression, anxiety and dementia. Patients with ID and at least one co-morbid mental disorder are at a high risk of polypharmacy. Importantly, psychiatric comorbidity was found to significantly increase service use and costs of care. Further investigation of the population of older adults with ID is needed, with special attention to development of clear treatment guidelines in order to effectively manage co-morbid mental illnesses and physical health problems.

Brawley, E.C.
Designing for Alzheimer's disease - Strategies for creating better care environments.
313 pp.
New York: Wiley (1997)
Abstract: 20 chapter general text on adapting homes and living environments for persons with dementia; applicable to home and other residential situations for adults with intellectual disabilities and dementia. Chapter sections include Aging and Alzheimer's disease, Sensory environment (light and aging vision, lighting, impact of color, patterns and texture, acoustical changes, and wayfinding guidelines), Special care settings (creating a home feeling, designing spaces, therapeutic gardens and outdoor spaces), Implementing effective interior design (furniture and fabrics, floor-covering, wall and ceiling finishes, windows and window treatments), and the Design process. Contains a directory of resources and a glossary of terms.

British Psychological Society
Dementia and people with intellectual disabilities guidance on the assessment, diagnosis, interventions and support of people with intellectual disabilities who develop dementia.
The British Psychological Society, St Andrews House, 48 Princess Road East, Leicester LE1 7DR, UK (April 2015).
https://cms.bps.org.uk/sites/default/files/2022-09/Dementia%20and%20People%20with%20Intellectual%20Disabilities.pdf
Abstract: This report is a revision to the original joint British Psychological Society and the Royal College of Psychiatrists (2009) guidance on dementia and people with intellectual disabilities. It has been written by a joint working group of the DCP Intellectual Disabilities Faculty of the British Psychological Society and the Royal College of Psychiatrists. The main purpose of the guidance is to enable those working in clinical and social care services to improve the quality of life of people with intellectual disabilities who develop dementia, by providing guidance to inform assessment, diagnosis, interventions and support. The guidance is aimed at clinicians in intellectual disabilities and older peoples' mental health services and services for younger people with dementia. The decision about which services provide which part of the care pathway is a local decision to be taken by commissioners and providers, but ensuring that all elements of this guidance are considered and in place for people with intellectual disabilities and dementia.

Brodaty, H., Seeher,K., & Gibson, L.
Dementia time to death: A systematic literature review on survival time and years of life lost in people with dementia.
International Psychogeriatrics, 2012, 24(7),1034-1045. Published online on 13 February 2012. doi: 10.1017/S1041610211002924.
Abstract: Life expectancy with dementia directly influences rates of prevalence and service needs and is a common question posed by families and patients. As well as years of survival, it is useful to consider years of life lost after a diagnosis of dementia. Authors systematically reviewed the literature on mortality and survival with dementia which were compared to estimated life expectancies in the general population. Both were then compared by age (under 65 years vs. 65+ years), gender, dementia type, severity, and two epochs (prior to and after introduction of cholinesterase inhibitors in 1997). Survival after a diagnosis of dementia varies considerably and depends on numerous factors and their

complex interaction. Relative loss of life expectancy decreases with age at diagnosis across varying sex, dementia subtypes (except for frontotemporal dementia and dementia with Lewy bodies), and severity stages. Numerous study deficiencies precluded a meta-analysis of survival in dementia. Authors concluded that estimates of years of life lost through dementia may be helpful for patients and their families.

Bruinsma, E., van den Hoofdakker, B.J., Groenman, A.P., Hoekstra, P.J., de Kuijper, G.M., Klaver, M., & de Bildt, A.A.
Non-pharmacological interventions for challenging behaviours of adults with intellectual disabilities: A meta-analysis.
Journal of Intellectual Disability Research, 2020 Aug, 64(8), 561-578. doi: 10.1111/jir.12736. Epub 2020 Jun 17
Abstract: Non-pharmacological interventions are recommended for the treatment of challenging behaviors in individuals with intellectual disabilities by clinical guidelines. However, evidence for their effectiveness is ambiguous. The aim of the current meta-analysis is to update the existing evidence, to investigate long-term outcome, and to examine whether intervention type, delivery mode, and study design were associated with differences in effectiveness. An electronic search was conducted using the databases Medline, Eric, PsychINFO and Cinahl. Studies with experimental or quasi-experimental designs were included. We performed an overall random-effect meta-analysis and subgroup analyses. We found a significant moderate overall effect of non-pharmacological interventions on challenging behaviors (d = 0.573, 95% CI [0.352-0.795]), and this effect appears to be longlasting. Interventions combining mindfulness and behavioral techniques showed to be more effective than other interventions. However, this result should be interpreted with care due to possible overestimation of the subgroup analysis. No differences in effectiveness were found across assessment times, delivery modes or study designs. Non-pharmacological interventions appear to be moderately effective on the short and long term in reducing challenging behaviours in adults with intellectual disabilities.

Bunt, C.W.
Medical care for adults with Down syndrome: guidelines from the Global Down Syndrome Foundation
American Family Physician, 2022, 105(4), 436-437.
https://www.aafp.org/pubs/afp/issues/2022/0400/p436.html
Abstract: [None provided]. These guidelines are the first attempt at taking an evidence-based approach to the management of adults with Down syndrome. Although evidence is limited, there are some important recommendations. The screening recommendations for diabetes mellitus, dementia, and hypothyroidism provide an initial template for routine care. Because we all learned about atlantoaxial instability and Down syndrome in medical school, it is important to know that this finding is not clinically significant. Recommendations include assess adults with Down syndrome annually for dementia starting at 40 years of age; screen adults with Down syndrome and obesity for type 2 diabetes mellitus with an A1C level every two to three years starting at 21 years of age and earlier if other diabetes risk factors are present; in adults with Down syndrome at a healthy weight, start screening at 30 years of ages; and avoid routine cervical radiography in adults with Down syndrome unless suggested by neurologic symptoms. Screen adults with Down syndrome for hypothyroidism with a thyroid-stimulating hormone test every one to two years starting at 21 years of age.

Burt, D.B., & Aylward, E.H.
Test battery for the diagnosis of dementia in individuals with intellectual disability. Working Group for the Establishment of Criteria for the Diagnosis of Dementia in Individuals with Intellectual Disability.
Journal of Intellectual Disability Research,. 2000 Apr; 44 (Pt 2), 175-180. doi: 10.1046/j.1365-2788.2000.00264.x
Abstract: A working battery of tests for the diagnosis of dementia, which is applicable to most adults with intellectual disability, is proposed by an international Working Group. The battery, reflecting contemporary research and practice findings, includes scales for informant report of functioning and tests for direct assessment. The Working Group recommends the international use of the battery both as part of ongoing and new longitudinal research, and in clinical practice. The widespread use of a common battery will enhance communication and collaborative opportunities among researchers and clinicians at various sites, and will help to standardize diagnostic protocols and research findings. The collaborative evaluation of such a battery will address one of the greatest challenges in the field, that of differentiating change associated with ageing from that associated with dementia.

Burt, D.B., & Aylward, E.
Assessment methods of diagnosis of dementia
In M.P. Janicki & A.J. Dalton (Eds.), Dementia, Aging, and Intellectual Disabilities.
pp. 141-156
Philadelphia: Brunner-Mazel (1999)
Abstract: Standardized diagnostic criteria and procedures are proposed to further progress in the understanding and treatment of dementia in adults with intellectual disabilities. This book chapter is a revised summary of previous reports prepared by participants of an international working group, which was conducted under the auspices of the International Association on Intellectual Disability and the American Association on Mental Retardation. Similarities in diagnostic issues between adults with intellectual disability and those in the general population are discussed, followed by a summary of issues unique to adults with intellectual disability. A brief overview of the application of ICD-10 diagnostic criteria to adults with intellectual disability is presented, including a description of procedures for determining whether criteria are met in individual cases. Finally, clinical and research recommendations are made.

Burt, D.B., Loveland, K.A., & Lewis, K.R.
Depression and the onset of dementia n adults with mental retardation.
American Journal of Mental Retardation, 1992, 96(5), 502-511. PMID: 1532893.
Abstract: The relation between dementia and depression in 61 adults with Down syndrome or 43 adults with mental retardation due to other causes was examined. Age-matched participants, ranging in age from 20 to 60 years, received a neuropsychological battery to assess declines in functioning and caregiver report measures to assess adaptive behavior and depression. Eight adults with Down syndrome had both depression and declines in functioning. No adults with mental retardation due to other causes had declines. Greater severity of depression was related to lower MA, poorer memory, and lower adaptive functioning in adults with Down syndrome only. Results suggest that dementia and depression are associated in Down syndrome but not in mental retardation due to other causes.

Burt, D.B., Primeaux-Hart, S., Loveland, K.A., Cleveland, L.A., Lewis, K.R., Lesser, J., & Pearson, P.L.
Tests and medical conditions associated with dementia diagnosis
Journal of Policy and Practice in Intellectual Disabilities, 2005, 2(1), 47-56.
https://doi.org/10.1111/j.1741-1130.2005.00007.x
Abstract: Diagnosis of dementia in adults with intellectual disabilities requires documentation of clinically significant declines in memory and other cognitive skills, as well as changes in everyday and emotional functioning. To improve diagnostic accuracy in adults with Down syndrome, the authors examined conditions often associated with dementia, as well as tests useful for documentation of decline. Specific aims were to identify psychiatric disorders or medical conditions that increased the odds of a dementia diagnosis; to evaluate the sensitivity and specificity of widely used dementia scales; and to determine which tests, used singly or in combination, most accurately supported the presence of dementia. Participants were 78 adults with Down syndrome. Two methods based on a large test battery and one method based on clinical judgment were used to diagnose dementia. It was found that combinations of tests lead to increased levels of diagnostic sensitivity compared with single tests. When taken in combination with other investigations, our results suggest that assessment for **psychiatric disorders, delayed memory decline, adaptive behavior decline, and the presence of seizures** would be useful for the diagnosis of dementia and that dementia scales would provide additional useful information. The authors conclude that combinations of tests and scales will be most useful for diagnosing dementia in adults with intellectual disabilities. The authors suggest that further research is needed to promote rapid progress, with studies that focus on common diagnostic methodology, identification of screening instruments, and amounts of decline indicative of dementia.

Burt, D.B., Primeaux-Hart, S., Loveland, K.A, Cleveland,L.A., Lewis, K.R., Lesser, J., & Pearson, P.L
Comparing dementia diagnostic methods used with people with intellectual disabilites

Journal of Policy and Practice in Intellectual Disabilities, 2005, 2(2), 94-115. https://doi.org/10.1111/j.1741-1130.2005.00022.x
Abstract: Accurate detection of dementia in adults with intellectual disabilities is important for clinical care, program planning, andclinical research. This paper reports on a study that examined two major diagnostic methods that varied in the following ways: (1)the extent to which they relied on clinical judgment; (2) the statistical method used to detect declines; and (3) the sensitivity to declines in functioning. Two methods based on testing were compared with one based on clinical judgment. Data were drawn from annual sequential assessments of 168 adults with intellectual disabilities (78 with Down syndrome and 90 with other etiologies).Agreement between testing and clinical judgment methods was 72-75% depending on testing method used. Clinical judgment produced a higher rate of dementia diagnosis for adults with Down syndrome compared with testing methods, suggesting a possible bias. The authors found that diagnostic criteria were useful both for identifying dementia and for describing its characteristics. Our results suggest that clinical judgment could result in a higher number of adults with Down syndrome diagnosed with dementia than methods based on test batteries. Common results across research studies indicate that combinations of sources of information(interviews/direct testing) would be most useful for dementia diagnosis. Future collaboration across research sites is needed to promote rapid progress in this important area, with emphasis on differential diagnosis.

Bush, A., & Beail, N.
Risk factors for dementia in people with Down syndrome: Issues in assessment and diagnosis
American Journal of Mental Retardation, 2004 Mar,109(2), 83-97. doi:10.1352/0895-8017(2004)109<83:RFFDIP>2.0.CO;2.
Abstract: It has been clearly established that there is an increased incidence of early onset dementia of the Alzheimer type (DAT) in people who have Down syndrome. There are variations in the age of onset of the clinical signs of DAT, which may be accounted for by different risk factors. In this review we examined the evidence that different biological and psychological factors may influence the risk for DAT. Limitations in design of early studies, the need for consistent diagnostic criteria for DAT in individuals with Down syndrome, and the lack of adequate psychometric tools to detect cognitive change are highlighted. Implications for research and clinical practice are considered in order to assess potential risk factors.

Cairns, D., Kerr, D., Chapman, A.
Difference realities: a training guide for people with Down's syndrome and Alzheimer's disease
pp. 54
University of Stirling (Dementia Services Development Centre), Stirling, Scotland FK9 4LA
A working guide for staff who are working with people with intellectual disabilities affected by Alzheimer's disease. Topical sections cover the definitions of dementia and deal with diagnostic suggestions, as well as dealing with communication, helping maintenance of skills, dealing with challenging behaviors, structuring activities, and overall management of dementia. Written in an easy style, this guide is a very useful addition to any materials given to staff to help them understand and related to people affected by dementia.

Cairns, V., Lamb, I., & Smith, E.
Reflections upon the development of a dementia screening service for individuals with Down's syndrome across the Hyndburn and Ribble Valley area.
British Journal of Learning Disabilities, 2011, 39(3), 198-208. https://doi.org/10.1111/j.1468-3156.2010.00651.x
Abstract: The high prevalence of dementia in individuals with Down's syndrome has led intellectual disability services in the Hyndburn and Ribble Valley (HRV) area of England to develop a screening service to address this need. The authors offer reflections upon this process by its members after the first 12 months of operation. A multidisciplinary team, comprising professionals from intellectual disability psychology, intellectual disability speech and language therapy, intellectual disability community nursing and older adults psychiatry, has developed, and begun to implement, screening care pathways. The service conducts routine screening assessments, provides intervention for individuals where concerns arise and delivers training to carers. At the point of writing, 27 service users have received screening assessments and six have been identified as at moderate–high risk of developing dementia. Reflection and feedback has highlighted issues for consideration throughout the service development process, and an evaluation of the training provided by the service has found this to be effective in increasing carers understanding about dementia and intellectual disabilities.

Callahan, B.L., Wong, C., Ngang, P.N., Wolf, U., Freedman, M.
Interventions targeting reversible dementia in Down syndrome.
Journal of the American Geriatrics Society, 2016 Apr;64(4):917-9. doi: 10.1111/jgs.14068.
Abstract: A 49-year-old man with high-functioning Down syndrome was admitted to the Behavioural Neurology Unit, Baycrest Centre for Geriatric Care (Toronto, Ontario, Canada), with a 1-year history of cognitive and behavioral decline. Behavioral problems included physical and verbal aggression (kicking, hitting, swearing, screaming), voiding and defecating on the floor, and refusing medication and assistance with hygiene. Hoarding (toilet paper, incontinence briefs) and disrobing were long-standing but had worsened. Although Alzheimer's disease was strongly suspected, he made a remarkable recovery to his premorbid level after a combination of pharmacological and behavioral interventions. Iindividuals with Down syndrome who present with dementia-like symptoms require thorough assessment and treatment for reversible causes of decline, regardless of how impaired they may seem, before being diagnosed with Alzheimer's disease or other dementia. Additional studies should examine these interventions separately to determine their individual effectiveness in individuals with Down syndrome who have progressive behavioral and cognitive decline.

Caoimh, R.O., Clune, Y., & Molloy, D.W.
Screening for Alzheimer's disease in Downs syndrome.
Journal of Alzheimers Disease & Parkinsonism, 2013, S7:001. doi:10.4172/2161-0460.S7-001
Abstract: Downs syndrome (DS) is associated with an increased incidence of Alzheimer's disease (AD). Although pathological changes are ubiquitous by 60 years of age, prevalence rates are lower. The diagnosis of AD in persons with DS is challenging, complicated by atypical presentations, baseline intellectual disability and normal age associated cognitive decline. Effective screening is limited by a paucity of diagnostic criteria, cognitive screening instruments and screening programs. Both observer-rated questionnaires and direct neuropsychological testing are suggested to screen for cognitive impairment, each with different strengths and weaknesses. This paper reviews commonly used screening instruments and explores the unique challenges of screening for AD in persons with DS. It concludes that single, one-dimensional screening tools and opportunistic evaluations are insufficient for detecting dementia in this population. These should be replaced by batteries of tests, incorporating informant questionnaires, direct neuropsychological testing, assessment of activities of daily living and behaviors, measured at baseline and reassessed at intervals. Developing these strategies into organized screening programs should improve diagnostic efficiency and management.

Carter, J., Spector, A., Ali, A., McFeeters, A., Butt, S., & Charlesworth, G.
Views and experiences of dementia in people with intellectual disabilities: a systematic review of qualitative research.
Journal of Intellectual Disability Research, 2025 Jul, 69(7), 533-545. doi: 10.1111/jir.13227. Epub 2025 Mar 16.
Abstract: It is important to hear the perspectives of people with intellectual disabilities on dementia. This review aimed to explore views and experiences of dementia from the perspective of people with intellectual disabilities and methodologies enabling people with intellectual disabilities and dementia to participate in qualitative research. Studies were identified in database searches, along with reference and citation searches. Qualitative data were reviewed using thematic synthesis and risk of bias assessed using the Critical Appraisal Skills Programme (2018). Methodologies used to include participants with intellectual disabilities and dementia were reviewed. Findings from 11 studies, with a total of 47 participants, highlighted loss of ability, relationships and connection associated with dementia, counteracted by support from others, and maintenance of a sense of self through choice, relational connection and competence. A range of methodologies were identified to enable participants with intellectual disabilities and dementia to participate in research. This review highlights emerging, albeit demographically limited, qualitative research in this field. It suggests ways to build on this including methodologies to facilitate inclusion of people with intellectual disabilities and dementia in further research.

Carfi, A., Antociccio, M., Brandi, V., Cipriani, C., Fiore, F., Mascia, D., Vetrano, D.L., Onder, G..
Down syndrome in adulthood: A disease for geriatricians
European Geriatric Medicine, 2014, 5(Supp 1), 549.
Abstract: Authors evaluated 89 adults with Down syndrome at a clinic in Rome, Italy, using a range of physiological and neurological methods, including nutritional and sensory assessments. The S's mean age was 42 years (range 18 to 72); 51% were females. Authors found behavioral disorders (53%), mood disorders (43%), seizures (22%), osteoporosis (40%), hypothyroidism (53%), diastolic dysfunction (80%), OSAS (90%), and hearing impairment (82%). Authors noted severe cognitive impairments in 67%, BMI greater than 25 in 66%, and low scores on physical performance measures (50%). Authors conclude that the pattern of diseases and conditions noted resemble those of other older adults and recommend that a mandatory geriatric evaluation be undertaken in older adults with Down.

Carling-Jenkins, R., Torr, J., Iacono, T., & Bigby, C.
Experiences of supporting people with Down syndrome and Alzheimer's disease in aged care and family environments.
Journal of Intellectual and Developmental Disability, 2012, 37(1), 54-60. doi: 10.3109/13668250.2011.645473. Epub 2012 Jan 3
Abstract: Australian research addressing the experiences of families of adults with Down syndrome and Alzheimer's disease in seeking diagnosis and gaining support is limited. The aim of this study was to gain a greater understanding of these processes by exploring the experiences of families and carers in supporting people with Down syndrome and Alzheimer's disease who had lived most or all of their lives with family. Three detailed case studies were created from multiple data sources, and then analyzed thematically. Families of adults with Down syndrome experienced stress and confusion as they negotiated a service system poorly equipped to meet their needs and professionals more focused on longstanding disability than the recent diagnosis of Alzheimer's disease. Such overshadowing led to mismanagement by services. Authors conclude that this research advances understandings of the support needs of people with Down syndrome and Alzheimer's disease and their families and exposes gaps in the service system.

Carmeli, E., Ariav, C., Bar-Yossef, T., Levy, R., & Imam, B.
Movement skills of younger versus older adults with and without Down syndrome.
Research in Developmental Disabilities, 2012 Jan-Feb;33(1):165-71. doi: 10.1016/j.ridd.2011.09.008. Epub 2011 Oct 4. Erratum in: Res Dev Disabil. 2012 Mar-Apr;33(2):781. PMID: 22093661.
Abstract: Adults with Down syndrome (DS) are often physically inactive, which may accelerate the onset of disease and aging symptoms. Eight older persons with DS (aged 54-61), and 10 younger persons with DS (aged 26-35) living in a residential care center were examined. Eighteen age- and gender-matched individuals without DS served as control groups. Sensory-motor tasks and Posture Scale Analyzer (PSA) were used to examine coordination and standing stability. The isokinetic muscle strength test was used for muscle strength investigation. The functional performance, coordination, and leg muscle strength of the older adults with DS were more impaired than both the younger DS and the control groups. The older DS group showed lower sway rate and more symmetrical weight-bearing distribution during quiet standing than both the younger DS and the control groups. Our observations may have significant implications for understanding movement dysfunction in older adults with DS.

Carmeli, E., Ariav, C., Bar-Yossef, T., & Levy, R.
Movement skills in persons with Down syndrome decrease with aging
International Journal on Disability and Human Development, 2010, 9(1), 29–34. https://www.degruyter.com/document/doi/10.1515/IJDHD.2010.005/html
Abstract: Persons with Down syndrome (DS) are comparatively physically inactive, which could accelerate the onset of disease, resulting in symptoms associated with aging that are detrimental to health. The aim was to evaluate movement abilities across the life span in persons with DS. Eleven persons with DS (>50 years, mean age 58 years), and 10 younger persons with DS (<49 years, mean age 28 years) who resided in a residential living center were included in the study. Age- and gender-matched people without DS (n=22) served as control group. Five sensory-motor tasks that involved the integration of hand movements with visual information were used, as well as the posture scale analyzer system to examine postural stability. Results showed that the older persons with DS had more medical problems than the young persons with and without DS. The hand coordination and postural stability of the older adults with DS were more impaired in comparison with the young group and both control groups. It is postulated that their poor motor function and slower responses might be explained by a less active lifestyle, that could accelerate the onset of disease, resulting in symptoms associated with aging that are detrimental to health. Our observations could have significant implications for understanding the mechanisms underlying movement dysfunction in older adults with DS and might offer new approaches for possible prevention.

Carr, J.
Stability and change in cognitive ability over the life span: a comparison of populations with and without Down's syndrome
Journal of Intellectual Disability Research, 2005 Dec;49(Pt 12):915-28. doi: 10.1111/j.1365-2788.2005.00735.x.
Abstract: Longitudinal studies show that in the general population IQ declines with age: early and rapidly in the case of performance IQ, later and more slowly in the case of verbal IQ. These populations have not apparently included people with intellectual disabilities (ID). A literature search identified 11 studies, some cross-sectional and others longitudinal, which provided data on a variety of verbal and performance tests, over periods from 3 to 19 years, on older people with ID. Following statistical advice the results from the different tests were converted into the equivalent of, for verbal scores, British Picture Vocabulary Scale, and for performance scores, Leiter International Performance Scale, raw scores. Percentage change between earlier and later scores was then calculated. With one exception the studies considered tend to show verbal ability declining relatively more, and performance ability declining relatively less, than has been shown to occur in the general population. Potential confounding factors, such as population attrition, cohort effects, etc., are thought not to have affected these results. The pattern of change with age in verbal and performance ability appears different in people with ID from that seen in the general population. Some possible reasons for this difference are discussed.

Carr, J., & Collins, S.
Ageing and dementia in a longitudinal study of a cohort with Down syndrome.
Journal of Applied Research in Intellectual Disabilities, 2014, 27(6), 555-563. doi: 10.1111/jar.12093. Epub 2014 Mar 29.
Abstract: A population sample of people with Down syndrome has been studied from infancy and has now been followed up again at age 47 years. Intelligence and language skills were tested and daily living skills assessed. Memory/cognitive deterioration was examined using two test instruments. Scores on verbal tests of intelligence changed little. Those on a non-verbal test, on self-help skills and on both memory tests showed some decline, even when the scores of those already suffering from dementia were discounted. At age 47, scores on most tests of even the majority of the cohort (i.e. those not definitely diagnosed with dementia) showed some decline. While this includes the scores of people who may subsequently develop dementia, it may also reflect the normal ageing process in this population.

Carr, J., & Collins, S.
50 years with Down syndrome: A longitudinal study.
Journal of Applied Research in Intellectual Disabilities, 2018 Sep, 31(5), 743-750. doi: 10.1111/jar.12438. ://doi.org/10.1111/jar.12438
Abstract: A population sample of people with Down syndrome, studied from infancy, has now been followed up at the age of 50 years. From the original sample of 54, there were 27 still in the study at the age of 50, all but four of the losses resulting from deaths. Intelligence and language skills were tested and daily living skills assessed. Memory/cognitive deterioration was examined using two test instruments. Other aspects of the people's lives were examined via carers' reports. Scores on verbal tests showed little change. Those on a non-verbal test, on self-help skills and on both memory tests showed some decline, even when the scores of those already suffering from dementia were discounted. **At the age of 50, those not already diagnosed with dementia showed some decline on most tests**. While this may include scores of people who subsequently develop dementia, it may also reflect the normal ageing process in this population.

Castro, P., Zaman, S., & Holland, A.
Alzheimer's disease in people with Down's syndrome: the prospects for and the challenges of developing preventative treatments.

Journal of Neurology, 2017 Apr, 264(4), 804-813.
doi:10.1007/s00415-016-8308-8. Epub 2016 Oct 24.
Abstract: People with Down's syndrome (DS) are at high risk for developing Alzheimer's disease (AD) at a relatively young age. This increased risk is not observed in people with intellectual disabilities for reasons other than DS and for this reason it is unlikely to be due to non-specific effects of having a neurodevelopmental disorder but, instead, a direct consequence of the genetics of DS (trisomy 21). Given the location of the amyloid precursor protein (APP) gene on chromosome 21, the amyloid cascade hypothesis is the dominant theory accounting for this risk, with other genetic and environmental factors modifying the age of onset and the course of the disease. Several potential therapies targeting the amyloid pathway and aiming to modify the course of AD are currently being investigated, which may also be useful for treating AD in DS. However, given that the neuropathology associated with AD starts many years before dementia manifests, any preventative treatment must start well before the onset of symptoms. To enable trials of such interventions, plasma, CSF, brain, and retinal biomarkers are being studied as proxy early diagnostic and outcome measures for AD. In this systematic review, we consider the prospects for the development of potential preventative treatments of AD in the DS population and their evaluation.

Centre for Developmental Disability Health Victoria
Dementia and Intellectual Disability – A guide to supporting people with intellectual disabilities through their journey with dementia: Online Learning for Disability Support Workers
http://www.cddh.monash.org/online-learning/
Abstract: These are on-line learning modules for disability staff supporting people who were at risk of developing, or had already been identified as having, dementia. There are four 16 minute nodules in the series addressing key questions you may have when supporting someone with dementia. They cover helpful information related to dementia and ID. Module 1: Understanding dementia and intellectual disability; Module 2: Taking action – The role of the support worker in assessment; Module 3: Supporting someone with intellectual disability and dementia; and Module 4: Supporting people through environment and activity. There are also a series self-taking test questions.

Chang, Z., Yao, H., Sun, S., Zhang, L., Liu, S., Brikell, I., D'Onofrio, B. M., Larsson, H., Lichtenstein, P., Kuja-Halkola, R., Hägg, S., Happé, F., & Taylor, M. J.
Association between autism and dementia across generations: evidence from a family study of the Swedish population.
Molecular Psychiatry, 2025, 30, 4605-4612.
https://doi.org/10.1038/s41380-025-03045-6
Abstract: There is emerging evidence to suggest that autistic individuals are at an increased risk for cognitive decline or dementia. It is unknown whether this association is due to shared familial influences between autism and dementia. The main purpose of this study was, thus, to investigate the risk of dementia in relatives of autistic individuals. We conducted a family study based on linking Swedish registers. We identified all individuals born in Sweden from 1980-2013, followed until 2020, and clinical diagnoses of autism among these individuals. We linked these index individuals with their parents, grandparents, and aunts/uncles. The risk of dementia (including any type of dementia, Alzheimer's disease, and other types of dementia) in relatives of autistic individuals was estimated using Cox proportional hazards models. Analyses were then stratified by sex of the relatives and intellectual disability in autistic individuals. Relatives of autistic individuals were at an increased risk of dementia. The risk was strongest in parents (hazards ratio [HR] = 1.36, 95% confidence intervals = 1.25-1.49), and weaker in grandparents (HR = 1.08, 1.06-1.10) and aunts/uncles (HR = 1.15, 0.96-1.38). Furthermore, there were indications of a stronger association between autism in index individuals and dementia in mothers (HR = 1.51, 1.29-1.77) compared to dementia in fathers (HR = 1.30, 1.16-1.45). There was only a small difference in relatives of autistic individuals with and without intellectual disability. Our results provide evidence of familial co-aggregation between autism and different types of dementia, and a potential genetic link. Future research now needs to clarify the risk of dementia in autistic individuals.

Chao, S.F., McCallion, P., & Nickle, T.
Factorial validity and consistency of the Maslach Burnout Inventory among staff working with persons with intellectual disability and dementia
*Journal of Intellectual Disability Research***,** 2011, 55(5), 529-536.
https://doi.org/10.1111/j.1365-2788.2011.01413.x
Abstract: Burnout has been considered important to understanding the well-being of workers in the intellectual disabilities (ID) field and the quality of services delivered to clients/consumers. However, little research has examined the psychometric properties and applicability to staff in ID services of one of the most widely used burnout measurements – the Human Services Survey version of the Maslach Burnout Inventory (MBI-HSS). Data were gathered using a mailed questionnaire comprising the MBI-HSS and demographic information. The sample consisted of 435 staff delivering direct care and working in out-of-home community placements for persons with ID in New York state. The factorial structure of the scale was examined using confirmatory and exploratory factor analysis. Internal consistency estimates of reliability of the MBI-HSS were determined using Cronbach's alpha. Confirmatory factor analysis supported the MBI-HSS as an acceptable measure to evaluate burnout in ID services staff. However, the reliability statistics obtained for the Depersonalization (DP) sub-scale was much lower than what has been reported in studies with other staff populations. An exploratory factor analysis suggested that a four-factor solution, dividing the DP sub-scale into two factors, provided a somewhat better fit for the sample. The use of the MBI-HHS as an instrument for measuring burnout among ID workers has its attraction but also its limitations. In particular, the DP sub-scale should be used with caution because there appear to be wording issues for staff in ID settings that may lead to inconsistent responses.

Chapman, M., Lacey, H., & Jervis, N.
Improving services for people with learning disabilities and dementia: Findings from a service evaluation exploring the perspectives of health and social care professionals.
British Journal of Learning Disabilities, 2017, 45(1), 33-44. Doi: 10.1111/bld.12210.
Abstract: Dementia prevalence rates are higher amongst people with intellectual [learning] disabilities than the general population. People with Down's syndrome are at even greater risk of developing dementia and of developing dementia at an earlier age. This study, conducted as part of a wider service evaluation, explored community learning disability team perspectives on screening, pathways, training, information and supports developed to improve services for people with learning disabilities and dementia. A focus group was held with health and social care professionals working in community learning disability services. Thematic analysis was used to analyze the data. The dementia screening, pathways and processes had become embedded in practice, leading to a common framework, an efficient, multidisciplinary, proactive approach, earlier detection and diagnosis of dementia and identification of other health needs and issues. This avoided crisis situations supporting people to remain at home longer. Training and information were felt to improve care quality and reduce caregiver anxiety. People with intellectual disabilities and caregivers were involved to varying extents. External influences impacting on support included the availability, appropriateness, cost and effectiveness of different models of service provision. Service developments have been made as a result of the findings which suggest that dementia pathways and supports improve service provision and outcomes for people with intellectual disabilities. It is important to develop the evidence base on the effectiveness of different service models for people with intellectual disabilities and dementia.

Chaput, J.L.
Housing people with Alzheimer disease as a result of Down syndrome: a quality of life comparison between group homes and special care units in long term care facilities.
Master's thesis, Department of City Planning, University of Manitoba (1998).
https://www.collectionscanada.gc.ca/obj/s4/f2/dsk2/tape15/PQDD_0014/MQ32072.pdf
Abstract: Report of study to determine which form of housing, group homes or special care units (SCUs), provided an enhanced quality of life for individuals with Down syndrome (DS) and Alzheimer disease (AD). Ten long term care (LTC) facilities with SCUs for people with AD in the Winnipeg, Canada area and ten group homes for people with DS and AD across Canada participated in the study. Results indicated that the group homes seemed to provide an enhanced quality of life for adults with DS and AD because they provided a home-like environment and they operated according to a therapeutic philosophy of care. In addition, costs for caregiving seemed to be more economical in group homes than in SCUs because group homes utilized lower staff wages and medical costs. Report provides information on practices and costs.

Chaput, J.L., & Udell, L.
Housing people with Alzheimer disease as a result of Down syndrome: a quality of life comparison between group homes and special care units in long term care facilities.
Journal of Intellectual Disability Research, 2000, 44, 236 (abstract No. 186) [Paper presented at the 11th World Congress of the International Association for the Scientific Study of Intellectual Disabilities, Seattle, Washington (USA), August 1-6, 2000]
Abstract: The purpose of the study was to determine which form of housing, i.e., group homes or special care units (SCUs), provided a better quality of life for individuals with Alzheimer disease (AD) as a result of Down syndrome (DS). The study also provided Winnserv Inc. (a non-profit housing organization that houses people with mental disabilities) with important information. Using the study results, Winnserv Inc. was able to determine that their group homes were suitable to maintain individuals with DS and AD and that their group homes were more cost-effective than SCUs in terms of caregiving. Twenty caregivers from both group homes and SCUs were selected to participate in this study. Ten long term care (LTC) facilities with SCUs for people with AD were selected in the Winnipeg area and ten group homes for people with Down syndrome and AD were chosen in Winnipeg and across Canada. The results indicated that the group homes seemed to provide the best quality of life for people with AD as a result of Down syndrome because they provided a home-like environment and they operated according to a therapeutic philosophy of care. In addition, costs for caregiving seemed to be more economical in group homes than in SCUs because group homes utilized lower staff wages and medical costs. Based on the results, it was recommended that Winnserv Inc. continue to house people with DS and AD.

Chaput, J.L.
Adults with Down syndrome and Alzheimer's disease: Comparisons of services received in group homes and in special care units
Journal of Gerontological Social Work, 2002, 38, 197-211
https://doi.org/10.1300/J083v38n01_05
Abstract: An increasing number of people with Down syndrome are at risk of dementia resulting from Alzheimer's disease. Many reside in community group homes. When they are affected by dementia, the challenge to agencies providing group homes is how to best provide continued housing and provide effective dementia-related care management. In the general population, long term care is typically provided in nursing facilities, often in special care units (SCUs). This study evaluated select factors found in group homes and SCUs to determine which is able to provide a better quality of life for people with Down syndrome affected by dementia. Interviews, using quality of life indicators, were conducted at 20 sites, equally selected from group homes and SCUs, on the basis of their experience with people with dementia. Results indicate that group homes can provide conditions associated with better quality of life and, additionally, operate with lower staffing costs due to the non-utilization of medical staff.

Chaudhury, H., Zhang, Z & Salmon, A.
Pre-relocation evaluation of the physical environment of a traditional care home before transitioning to a dementia village,
The Gerontologist, 2025, 65(10), gnaf167.
https://doi.org/10.1093/geront/gnaf167
Abstract: The physical environment in long-term care homes is a critical component for providing quality care and support to residents with dementia. The first publicly funded "dementia village" model-inspired care home in Canada will offer a neighborhood environment for the residents. This study provides a baseline assessment of the physical environment's influence in a traditional dementia care unit before residents relocate to the new care village, as part of a pre and post transfer evaluation project. Multi-method evaluation approaches were used, including standardized physical environmental assessments, behavioral observations of residents and staff, and staff focus groups to generate findings for a comprehensive and in-depth understanding of the role of the physical environment on residents' functioning and engagement in the selected care setting. The pre-relocation baseline evaluation in the existing care unit revealed challenges arising from the unit's physical design, space use, and care practices. Three themes emerged from observations and focus group findings: "Main Common Area: Managing Environmental Stimulation," "Corridor: Wayfinding and Mobility Autonomy," and "Lack of Engagement Opportunities." The study provided evidence on how a large, institutional care setting can pose challenges to residents' well-being and staff practices. It also highlighted the interrelations among the physical environment, residents, and staff in both positive and negative aspects.

Chicoine, B.A. & McGuire, D.
Longevity of a woman with Down syndrome: A case study
Mental Retardation, 1998, 35(6), 477-479
DOI:10.1352/0047-6765(1997)035<0477:LOAWWD>2.0.CO;2
Abstract: [Adapted] A case of a woman at age 83 who is among the longest surviving people with Down syndrome was described. The life expectancy of persons with Down syndrome has increased more than six-fold to 56 years since the turn of the century. The literature regarding life expectancy for persons with Down syndrome was reviewed, and the implications regarding Down syndrome and Alzheimer's disease were discussed. Of particular interest about the patient reported in this article is that she was reported to have no decline in mental function and performance of activities of daily living. The longevity of our 83 year-old patient can probably be attributed to the absence of congenital heart disease and the fact that she was not institutionalized at a time many years ago when conditions in institutions tended to be poor.

Chicoine, B., Rivelli, A., Fitzpatrick, V., Chicoine, L., Jia, G., Rzhetsky, A.
Prevalence of common disease conditions in a large cohort of individuals with Down syndrome in the United States.
Journal of Patient-Centered Research and Reviews, 2021, 8(2), 86-97. doi: 10.17294/2330-0698.1824
Abstract: Given the current life expectancy and number of individuals living with Down syndrome (DS), it is important to learn common occurrences of disease conditions across the developmental lifespan. This study analyzed data from a large cohort of individuals with DS in an effort to better understand these disease conditions, inform future screening practices, tailor medical care guidelines, and improve utilization of health care resources. This retrospective, descriptive study incorporated up to 28 years of data, compiled from 6078 individuals with DS and 30,326 controls matched on age and sex. Data were abstracted from electronic medical records within a large Midwestern health system. In general, individuals with DS experienced higher prevalence of testicular cancer, leukemias, moyamoya disease, mental health conditions, bronchitis and pneumonia, gastrointestinal conditions, thyroid disorder, neurological conditions, atlantoaxial subluxation, osteoporosis, dysphagia, diseases of the eyes/adnexa and of the ears/mastoid process, and sleep apnea, relative to matched controls. Individuals with DS experienced lower prevalence of solid tumors, heart disease conditions, sexually transmitted diseases, HIV, influenza, sinusitis, urinary tract infections, and diabetes. Similar rates of prevalence were seen for lymphomas, skin melanomas, stroke, acute myocardial infarction, hepatitis, cellulitis, and osteoarthritis. While it is challenging to draw a widespread conclusion about comorbidities in individuals with Down syndrome, it is safe to conclude that care for individuals with DS should not automatically mirror screening, prevention, or treatment guidelines for the general U.S. population. Rather, care for those with DS should reflect the unique needs and common comorbidities of this population.

Cho, I.Y., Koo, H.Y., Um, Y.J., Park, Y-M. M., Kim, K.M., Lee, C.E., & Han, K.
Intellectual disabilities and risk of cardiovascular diseases: A population-based cohort study.

Disability and Health Journal, 2025, 18(2),101754.
https://doi.org/10.1016/j.dhjo.2024.101754.
Abstract: While intellectual disability is associated with higher mortality rates due to circulatory diseases, it is unclear whether intellectual disability is associated with higher risk of myocardial infarction (MI) and stroke than the general population. We aimed to analyze the risk of cardiovascular diseases (CVD), specifically myocardial infarction (MI) and ischemic stroke, and death due to circulatory diseases in individuals with disability. This retrospective cohort study used data from the National Disability Registration System linked to the Korean National Health Insurance Service database. Individuals who underwent national health examinations in 2009 were followed until 2020. Cox-proportional hazard analyses were performed to estimate the risk of CVD, MI, ischemic stroke, and circulatory disease deaths with adjustment for covariates. A total of 3642 individuals with intellectual disability (mean [SD] age 39.1 [12.6], 28.8 % female) and 3,889,794 individuals without intellectual disability (mean [SD] age 47.1 [13.9], 45.6 % female) were included. Compared to those without intellectual

disability, those with intellectual disability had higher risk of CVD (adjusted hazard ratio [aHR] 1.71, 95 % confidence interval [CI] 1.45-2.02), ischemic stroke (aHR 2.21, 95 % CI 1.81-2.69), and death due to circulatory diseases (aHR 4.20, 95 % CI 3.24-5.45), and a non-significant risk for MI (aHR 1.24, 95 % CI 0.95-1.63) after full adjustment for covariates. Individuals with intellectual disability were at increased risk of CVD, in particular ischemic stroke, and death due to circulatory diseases. Healthcare professionals should be aware of increased CVD risk in individuals with intellectual disability.

Choi, P., Motl, R.W., & Agiovlasitis, S.
Feasibility of social cognitive theory-based fall prevention intervention for people with intellectual disabilities living in group-home.
Journal of Intellectual Disability Research, 2022 Dec 18. doi: 10.1111/jir.13001.
Abstract: Adults with intellectual disability (ID) have a higher rate of fall events than the general population. Consequently, interventions for reducing fall events and improving health are highly required for individuals with ID. One essential step towards effectively delivering fall prevention interventions among adults with ID involves evaluating their feasibility. This study examined the feasibility of a home-based exercise intervention, supplemented with behavioural change strategies, among individuals with ID living in residential settings. This study provided an 8-week intervention, consisting of a workshop for support workers and sessions for participants with ID, focusing on behavioural reward/s, education regarding fall prevention/exercise and exercise training. One week prior to and 1 week following such an intervention, such participants underwent measurements for (1) physical performance, (2) fall efficacy, (3) self-efficacy for activity and (4) social support. Participants having ID (n = 33), support workers (n = 11) and one administrator participated in this study. There were no adverse events during the intervention, and the mean adherence rate was 70.8 ± 19.5%. Two participants with ID dropped out of the programme due to a lack of interest. The participants with ID significantly improved individual physical performance, self-efficacy for activity, fall efficacy and support from friends and support workers. Fall prevention interventions for adults with ID living in group-homes were highly promising for eventual large-scale implementation within such communities.

Cipriani, G., Danti, S., Carlesi, C., & DiFiorino, M.
Aging with Down syndrome: the dual diagnosis - Alzheimer's disease and Down syndrome
American Journal of Alzheimer's Disease & Other Dementias, 2018, 33(4), 253-262. doi: 10.1177/1533317518761093. Epub 2018 Mar 5.
Abstact: People with Down syndrome (DS) enjoy a longer life expectancy now than they ever have before and are therefore at greater risk of developing conditions associated with aging, including dementia. Authors undertook at review to explore the phenomenon of dementia in DS. Medline and Google Scholar searches were conducted for relevant articles, chapters, and books published until 2017. Search terms included Alzheimer's disease, cognitive impairment, dementia, DS, and trisomy 21. Publications found through this indexed search were reviewed for further references. Authors concluded that virtually, all subject aged 35 to 40 show key neuropathologic changes characteristic of Alzheimer's disease, but only a part of them show clinical signs of dementia, usually around the age of 50 years. Early signs of dementia in people with DS may be different from those experienced by the general population. Failure to recognize this can delay diagnosis and subsequent interventions.

Cipriani, G., Picchi, L., Dolciotti, C., & Bonucecelli, U.
Alzheimer's disease and Down's syndrome: An unhappy union [La malattia di Alzheimer e la sindrome di Down: un infelice connubio]
Quaderni Italiani di Psichiatria, 2011 March, 30(1), 26-32.
https://www.researchgate.net/publication/235977633_La_malattia_di_Alzheimer_e_la_sindrome_di_Down_un_infelice_connubio_Alzheimer's_disease_and_Down's_syndrome_an_unhappy_union
Abstract: People with Down's syndrome (DS) have an increased risk of developing Alzheimer's disease (AD) during middle age. Both disorders can present with a decline in cognitive skills and behavioral symptoms. Therefore, dementia, particularly in its early stages, can be difficult to diagnose in this population. We conducted a search of electronic databases for literature on the relationship between AD and DS. The key words used were: ''Down syndrome'', ''Alzheimer's disease'', ''dementia'', and ''mental retardation''. AD onset has been reported as early as age 30 in individuals with DS, and there is a dramatic increase in prevalence rates in older age groups. This trend reflects increased survival of persons with DS probably as a result of advances in medical treatment and improved living conditions. Even with careful clinical assessment, it can be very difficult to identify early symptoms of dementia when it is superimposed on a background of intellectual disability. The reasons include the wide intra-individual variability in cognitive functioning and difficulties involved in establishing baseline levels of the premorbid condition. Many frontal lobe-related symptoms usually associated with later stages of dementia in the general population are commonly seen in the early stage of the dementia that develops in adults with DS. After onset, the clinical symptoms of dementia progress rapidly in all subjects with DS. Research suggests that the presentation of dementia in people with DS may differ from that typical of AD in the general population. Early changes tend to involve personality and behavior rather than memory. DS can be best understood as a complex syndrome of genetic origin that has protean neurobiological consequences and numerous clinical characteristics. [Note: article text in Italian]

Cleary, J., & Doody, O.
Professional carers' experiences of caring for individuals with intellectual disability and dementia: A review of the literature.
Journal of Intellectual Disabilities, 2017 Mar, 21(1), 68-86. doi:10.1177/1744629516638245.
Abstract: The number of people with intellectual disability living into old age and developing dementia continues to increase. Dementia presents a wide range of challenges for staff due to progressive deterioration. This article presents the findings from a narrative literature review of professional caregivers' experiences of caring for individuals with intellectual disability and dementia. Seven electronic databases were searched using Boolean operators and truncation to identify relevant literature. Search results were combined and narrowed to articles relevant to staff working with individuals with intellectual disability and dementia, and 14 articles met the criteria for review. Themes outlined in the review include staff knowledge of dementia, staff training in dementia, caregiving, challenging behavior, pain management, mealtime support and coping strategies. Overall carers must review and adjust their care delivery and support to people with intellectual disability and dementia, not only in terms of identifying and responding to their health needs but also through collaborative team working within and across services.

Cleary J, & Doody O.
Nurses' experience of caring for people with intellectual disability and dementia.
Journal of Clinical Nursing, 2017, 26(5-6), 620-631. doi: 10.1111/jocn.13431. Epub 2016 Nov 14.
Abstract: The authors endeavored to explore nurses' experiences of caring for older people with intellectual disability and dementia. Ageing and dementia prevalence is increasing along with the life expectancy of people with intellectual disability. As a population group, people with intellectual disability have a high prevalence of dementia, which is higher within the subpopulation of Down syndrome. People with intellectual disability live in residential care, community or residential settings, and nurses are required to adapt their practices to meet the changed needs of the individual. A qualitative Husserlian descriptive phenomenological methodology was undertaken by the researchers so as to be able to become absorbed in the quintessence of meaning and explore nurses' experience of working with older people with intellectual disability and dementia. Ethical approval was obtained, and data were collected utilizing semistructured interviews (n = 11). Interviews were transcribed and analyzed using Colaizzi's framework for data analysis. The authors extracted three key themes were identified: 'knowledge of dementia', 'person-centred care' and 'transitioning within the service'. The study highlights the need for proactive planning, life story books of the patient, and funding to support client and staff. The authors concluded that overall, the study highlights the importance of knowing the person, supporting the individual and recognizing presenting behaviors as outside the control of the individual. The article presents the experiences of nurses caring for the older person with intellectual disability and dementia. Transitions are often very difficult for both the person and their peers, and they experience benefit from the efforts of a multidisciplinary team facilitating a person-centered approach.

Clifford, C., & Lauer, E.
Evaluating dementia capability of service systems for people with intellectual and developmental disorders and dementia
Journal of Applied Research in Intellectual Disabilities, 2021, 34(5), 1215-1216.
https://onlinelibrary.wiley.com/toc/14683148/2021/34/5

Abstract: People with an intellectual or developmental disability (IDD) experience complex age-related issues, including dementia-related disorders, at higher rates and earlier ages than the general population. Increased support needs of this subpopulation can strain caregivers and existing community supports. Patterns of resource awareness and utilization and unmet needs are not well understood for this subpopulation. A collaboration of the Massachusetts Council on Aging (MCOA), the Massachusetts Department of Developmental Services (DDS), and the Center for Developmental Disability Evaluation and Research (CDDER) at University of Massachusetts Medical School conducted a needs assessment with caregivers of people with intellectual and developmental disabilities to assess awareness and utilization of community-based resources, and unmet needs including for caregiver supports. Home visits, including an environmental assessment, and interviews with caregivers of 95 adults with dementia-related diagnoses were conducted. Interviews asked about changes in the adult's condition since diagnosis including a needs assessment about the dementia-related knowledge and training, care confidence levels, perceived barriers and/or concerns to care provision and resource use. About half of the caregivers reported significant changes in the person's skills, function, and memory since diagnosis, as well as worsening of the person's gait, continence, and swallowing. 78% of caregivers reported feeling confident providing care currently and 68% were confident about providing future care. 100% of the respondents found outreach provided by a nurse practitioner helpful. Caregiver concerns included lack of suitable day programming, future planning resources, and caregiver burnout/stress. Most caregivers were currently aware of some local resources but with scattered use. Environmental assessments indicated most homes, while accessible, warranted additional lighting and clutter removal. Caregivers requested additional training in addressing the behavioral and mental health needs. Findings suggest a need for increased collaboration across the intellectual and developmental disability and aging/human services systems, and additional training and resource navigation guides for caregivers.

Clifford, A., Omokanye, M. and Bagalkote, D.
"Commentary on "Stakeholder experiences of deprescribing psychotropic medicines for challenging behaviour in people with intellectual disabilities"",
Tizard Learning Disability Review, 2024, Vol. ahead-of-print No. ahead-of-print. https://doi.org/10.1108/TLDR-05-2024-0022.
Abstract: This paper provides a commentary response to "Stakeholder experiences of deprescribing psychotropic medicines for challenging behaviour in people with intellectual disabilities". The commentary provides some evidence-based perspectives on the risks and challenges of psychiatric drug-withdrawal, emphasizing the importance of the patient experience in deprescribing decisions. It also makes the case for clinical clarity and carer-engagement in optimising deprescribing outcomes for individuals. Some clinical reflections are provided. Whilst efforts to reduce the overmedication of people with intellectual disabilities are welcomed, there is lacking evidence around this population's experience of adverse effects from psychiatric deprescribing decisions, including differentiating between withdrawal and relapse effects. Clinical clarity and carer engagement are key to safe and effective prescribing, although in reality, this can be challenging.

 **Cohen, U., & Wiesman, G.D.**
Holding on to home: Designing environments for people with dementia.
181 pp.
Baltimore: Johns Hopkins University Press (1991)
Abstract: General text on adapting homes and living environments for persons with dementia; applicable to home and other residential situations for adults with intellectual disabilities and dementia.

Collacott R.A.
Epilepsy, dementia and adaptive behaviour in Down's syndrome.
Journal of Intellectual Disability Research, 1993, 37(2), 153-60. doi: 10.1111/j.1365-2788.1993.tb00582.x.
Abstract: Widespread inquiry identified 378 adults with Down syndrome resident in Leicestershire, England. The immediate carer of 351 of these (92.8%) was interviewed for the purpose of establishing a past history of seizures, including the age at which the seizures began. The immediate carer was also invited to provide information to enable the completion of an Adaptive Behaviour Scale (A.B.S.) rating. Individuals with a history of seizures were divided into two groups on the basis of whether or not seizures commenced prior to or after age 35 years. Two control groups of individuals with Down syndrome, but without a history of seizures were selected. Adaptive Behavior Scale scores for those in whom seizures commenced at a younger age were similar to those who had no recorded history of seizures. However, in those in whom seizures began in later life, scores on all domains of the A.B.S. were significantly reduced compared to both young epileptic patients and their controls. Adaptive Behavior Scale scores for the older control group held an intermediate position, suggesting that late-onset epilepsy may be a late manifestation of a dementing process. A clinical diagnosis of dementia recorded in the case records was significantly associated with the presence of late-onset epilepsy. This is supportive of the hypothesis that late-onset epilepsy in individuals with Down's syndrome is associated with Alzheimer's disease.

Conceiçao, A.S.G.G., Sant'Ana, L.F.G., Mattar, G.P., de Fátima R. Silva, M., Ramos, A.R., Oliveira, A.M., Carvalho, C.L., Gonçalves, O.R., Varotto, B.L.R, Martinez, L.D., Leduc, V., Fonseca, L.M., & Forlenza, O.V.
Balance and gait: Associations with cognitive impairment and dementia in individuals with Down syndrome.
Alzheimer Disease & Associated Disorders, 2023 (Oct-Dec), 37(4), 349-356. doi: 10.1097/WAD.0000000000000580
Abstract: Atypical aging in Down syndrome (DS) is associated with neuropathological characteristics consistent with Alzheimer disease. Gait abnormalities have been shown to be associated with an increased risk of dementia for the general population. The aim of this study was to determine whether gait disorders are associated with worse cognitive performance and dementia in adults with DS. We evaluated 66 individuals with DS (≥20 y of age), divided into 3 groups: stable cognition, prodromal dementia, and dementia (presumed Alzheimer disease). Each individual was evaluated with the Performance-Oriented Mobility Assessment (POMA), Timed Up and Go test, and Cambridge Examination for Mental Disorders of Older People with Down's Syndrome and Others with Intellectual Disabilities (CAMDEX-DS), in addition to a comprehensive clinical protocol to ascertain the occurrence of medical or psychiatric comorbidities. The score on the POMA-Gait subscale score and body mass index were found to be independent predictors of prodromal dementia and dementia (P<0.001 for both). With the exception of perception, all cognitive domains correlated with the POMA-Total score (P<0.05). A lower POMA-Gait score increases the chance of prodromal dementia and dementia in adults with DS. Unlike other research, in this study higher body mass index was also found to increase the chance of prodromal dementia and dementia. In those individuals, applying the POMA could facilitate the early diagnosis of dementia, help identify fall risks, and promote the adoption of geriatric interventions focused on improving functional mobility.

Coppus, A.M.W
People wtih intellectual disabiltiy: What do we know about adulthood and life expectancy?
Developmental Disabilities Research Review, 2013, 18(1), 6-16. doi: 10.1002/ddrr.1123.
Abstract: Increases in the life expectancy of people with Intellectual Disability have followed similar trends to those found in the general population. With the exception of people with severe and multiple disabilities or Down syndrome, the life expectancy of this group now closely approximates with that of the general population. Middle and old age, which until 30 years ago were not recognized in this population, are now important parts of the life course of these individuals. Older adults with intellectual disabilities form a small, but significant and growing proportion of older people in the community. How these persons grow older and how symptoms and complications of the underlying cause of the intellectual disability will influence their life expectancy is of the utmost importance.

Coppus, A. , Evenhuis, H., Verberne, G.J., Visser, F., van Gool, P., Eikelenboom, P., & van Duijin, C.
Dementia and mortality in persons with Down's syndrome.
Journal of Intellectual Disability Research, 2006, 50(10), 768-77. doi:10.1111/j.1365-2788.2006.00842.x.
Abstract: Numerous studies have documented that persons with Down's syndrome (DS) are at an increased risk of Alzheimer's disease (AD). However, at present it is still not clear whether or not all persons with DS will develop dementia as they reach old age. We studied 506 people with DS, aged 45 years and above. A standardized assessment of cognitive, functional and physical status was repeated annually. If deterioration occurred, the patients were examined and the differential diagnosis of dementia was made according to the

revised Dutch consensus protocol and according to the ICD-10 Symptom Checklist for Mental Disorders. We compared our findings with those reported in the literature. The overall prevalence of dementia was 16.8%. Up to the age of 60, the prevalence of dementia doubled with each 5-year interval. Up to the age of 49, the prevalence is 8.9%, from 50 to 54, it is 17.7%, and from 55 to 59, it is 32.1%. In the age category of 60 and above, there is a small decrease in prevalence of dementia to 25.6%. The lack of increase after the age of 60 may be explained by the increased mortality among elderly demented DS patients (44.4%) in comparison with non-demented patients (10.7%) who we observed during a 3.3-year follow-up. There was no decrease in incidence of dementia in the age group of 60 and above. Our findings are very similar to those published in the literature. Patients with dementia were more frequently treated with antiepileptic, antipsychotic and antidepressant drugs. The history of depression was strongly associated with dementia. Our study is one of the largest population-based studies to date. We found that despite the exponential increase in prevalence with age, the prevalence of dementia in the oldest persons with DS was not higher than 25.6%.

Coppus, A.M.W , Evenhuis, H.M, Verberne, G-J., Visser, F.E., Oostra, B.A. Eikelenboom, P., van Gool, W.A., Cecile, A., Janssens, J.W. , van Duijn, C.M.
Survival in elderly persons with Down syndrome.
Journal of the American Geriatrics Society, 2008, 56(12), 2311-2316. doi: 10.1111/j.1532-5415.2008.01999.x.
Abstract: The longer life expectancy now experienced by persons with Down syndrome (DS) makes it necessary to know the factors influencing survival in older persons with this syndrome. In a prospective longitudinal cohort study of dementia and mortality, 506 persons with DS aged 45 and older were followed for a mean of 4.5 years (range 0.0–7.6 years). Cognitive and social functioning were tested at baseline and annual follow-up. The diagnosis of dementia was determined according to a standardized protocol. Cox proportional hazards modeling was used for survival analysis. Relative preservation of cognitive and functional ability is associated with better survival in this study population. Clinically, the most important disorders in persons with DS that are related to mortality are dementia, mobility restrictions, visual impairment, and epilepsy -- but not cardiovascular diseases. Also, level of intellectual disability and institutionalization were associated with mortality.

Coppus, A., Evenhuis, H., Verberne, G.J., Visser, F., van Gool, P., Eikelenboom, P., & van Duijin, C.
Dementia and mortality in persons with Down's syndrome.
Journal of Intellectual Disability Research, 2006, Oct;50(Pt 10):768-77. doi: 10.1111/j.1365-2788.2006.00842.x.
Abstract: Numerous studies have documented that persons with Down syndrome (DS) are at an increased risk of Alzheimer's disease (AD). However, at present it is still not clear whether or not all persons with DS will develop dementia as they reach old age. The authors studied 506 people with DS, aged 45 years and above. A standardized assessment of cognitive, functional and physical status was repeated annually. If deterioration occurred, the patients were examined and the differential diagnosis of dementia was made according to the revised Dutch consensus protocol and according to the ICD-10 Symptom Checklist for Mental Disorders. We compared our findings with those reported in the literature. The overall prevalence of dementia was 16.8%. Up to the age of 60, the prevalence of dementia doubled with each 5-year interval. Up to the age of 49, the prevalence is 8.9%, from 50 to 54, it is 17.7%, and from 55 to 59, it is 32.1%. In the age category of 60 and above, there is a small decrease in prevalence of dementia to 25.6%. The lack of increase after the age of 60 may be explained by the increased mortality among elderly demented DS patients (44.4%) in comparison with non-demented patients (10.7%) who we observed during a 3.3-year follow-up. There was no decrease in incidence of dementia in the age group of 60 and above. Our findings are very similar to those published in the literature. Patients with dementia were more frequently treated with antiepileptic, antipsychotic and antidepressant drugs. The history of depression was strongly associated with dementia. The authors concluded that their study is one of the largest population-based studies to date. We found that despite the exponential increase in prevalence with age, the prevalence of dementia in the oldest persons with DS was not higher than 25.6%.

Coppus, A.M., Evenhuis, H.M., Verberne, G.J., Visser, F.E., Eikelenboom, P., van Gool, W.A., Janssens, A.C., & van Duijn, C.M.
Early age at menopause is associated with increased risk of dementia and mortality in women with Down syndrome.
Journa of Alzheimer's Disease, 2010;19(2):545-50. doi: 10.3233/JAD-2010-1247.
Abstract: In a prospective longitudinal cohort study of dementia and mortality in persons with Down syndrome aged 45 years and older, 85 postmenopausal women were followed for a mean follow-up time of 4.3 years (range 0.0 to 7.4 years). The effect of age at menopause on age at diagnosis of dementia and survival was estimated using correlation analysis and Cox Proportional Hazard Model. We found a significant correlation between age at menopause and age at diagnosis of dementia (rho=0.52; p< 0.001), and between age at menopause and age at death (rho=0.49; p=0.01). Early age at menopause is associated with a 1.8 fold increased risk of dementia: Hazard Ratio (HR): 1.82 (95%Confidence Interval (CI): 1.31-2.52) and with risk of death: HR: 2.05 (95%CI: 1.33-3.16). Our study suggests that age at menopause in women with Down syndrome is a determinant of age at onset of dementia and mortality.

Cooper, S-A.
High prevalence of dementia among people with leaming disabilities not attributable to Down's syndrome.
Psychological Medicine, 1997, 27(3), 609-616. doi: 10.1017/s0033291796004655.
Abstract: For many years, it has been known that dementia can occur in people with learning disabilities, but there have been few research studies. Studies that do quote rates for dementia show these to be high, but this important fact has received remarkably little attention. Comprehensive psychiatric and medical assessments were undertaken on the whole population (ascertained as far as is possible) of people with learning disabilities aged 65 years and over living in Leicestershire, UK (N=134), in order to ascertain rates of DCR defined dementia, and associated factors. Dementia was diagnosed in 21·6%, against an expected prevalence of 5·7%, for a group with this age structure. The rate of dementia increased in successive age cohorts: 15·6% aged 65-74 years; 23·5% aged 65-84 years; and 70·0% aged 85-94 years. People with dementia tended to be older, female, with more poorly controlled epilepsy, a larger number of additional physical disorders, less likely to be smokers and had lower adaptive behavior scores than did the elderly people without dementia. They were more likely to live in health service accommodation. Dementia occurs at a much higher rate among elderly people with learning disabilities than it does among the general population; this is independent of the association between dementia and Down's syndrome. Whether this relates etiologically to genetics, lack of brain 'reserve' or history of brain damage is yet to be determined.

Cooper, S.
Psychiatric symptoms of dementia among elderly people with learning disabilities.
International Journal of Geriatric Psychiatry, 1997, 12(6), 622-666. PMID: 9215950.
Abstract: Author proceeded to determine the rate of psychiatric symptoms among elderly people with learning disabilities who have dementia via a survey of the general community of a county in the UK. Participants included the whole population of people with [intellectual] disabilities who were 65 years or over. The total population was 143, of whom 134 participated (93.7%). From the total population, those with dementia were determined (N = 29). Description of psychopathologies were captured using a semi-structured psychiatric rating scale. Author determined that psychotic symptoms occurred in 27.6%, with the most common types being delusions of thefts, other persecutory delusions and visual hallucinations of strangers in the house. The onset of other psychiatric symptoms as part of the dementia was also common, in particular changed sleep pattern, loss of concentration, worry, reduced quantity of speech, change in appetite and onset of or increase in aggression. It was found that people with [intellectual] disabilities are living longer, and so the number with dementia is rising. Psychiatric symptoms occur commonly in dementia, can cause significant distress and require recognition, understanding and the development of effective managements.

Cooper, S-A., & Prasher, V.
Maladaptive behaviours and symptoms of dementia in adults with Down's syndrome compared with adults with intellectual disability of other aetiologies
Journal of Intellectual Disability Research, 1998, 42(4), 293-300. https://doi.org/10.1046/j.1365-2788.1998.00135.x
Abstract: Dementia commonly occurs in elderly people with intellectual disability,

especially those with Down's syndrome. The non-cognitive symptoms of dementia can be of greater significance to individuals and carers than the cognitive changes caused by this condition. It is not known whether there are differences between people with Down's syndrome and those with intellectual disability of other causes with regard to the prevalence of such symptoms. The present study was undertaken to draw a comparison between a group with Down's syndrome and dementia (n= 19), and a group with intellectual disability of other causes and dementia (n= 26). Maladaptive behaviours and psychiatric symptomatology were assessed in both groups. The group with Down's syndrome had a higher prevalence of low mood, restlessness/excessive overactivity, disturbed sleep, being excessively uncooperative and auditory hallucinations. Aggression occurred with greater frequency in those subjects with intellectual disability of other causes. These findings are of epidemiological importance in terms of service planning and understanding psychiatric presentation.

Cooper, S-A., & van der Speck
Epidemiology of mental ill health in adults with intellectual disabilities
Current Opinion in Psychiatry, 2009, Sep, 22(5), 431-436.
doi:10.1097/YCO.0b013e32832e2a1e.
Abstract: Adults with intellectual disabilities experience higher rates of mental ill health than the general population. Despite this, the epidemiological knowledge base remains limited. The purpose of this article is to review mental health epidemiological studies relevant to adults with intellectual disabilities, published since January 2008. Several studies have aimed to build the epidemiological evidence base, particularly with regards to problem behaviours, which appear to be remitting-relapsing conditions rather than necessarily being chronic. Most of such work confirms prevalence and incidence rates, and conducts exploratory analyses to determine factors independently related to mental ill health. Down syndrome protects against problem behaviours and mental ill health (except dementia that occurs at a higher rate), whereas epilepsy does not appear to affect risk for mental ill health. Dementia is four times more common in older persons with intellectual disabilities without Down syndrome than in the general population. Persons with borderline intellectual disabilities also experience higher rates of mental ill health than the general population, but receive fewer treatments.

Cordell, C.B., Borson, S., Boustani, M., Chodosh, J., Reuben, D., Verghese, J., Thies,W., Fried, L.B.,& Medicare Detection of Cognitive Impairment Workgroup
Alzheimer's Association recommendations for operationalizing the detection of cognitive impairment during the Medicare Annual Wellness Visit in a primary care setting
Alzheimer's & Dementia, 2013 Mar, 9(2),141-150. doi:
10.1016/j.jalz.2012.09.011. Epub 2012 Dec 20.
Abstract: The Patient Protection and Affordable Care Act added a new Medicare benefit, the Annual Wellness Visit (AWV), effective January 1, 2011. The AWV requires an assessment to detect cognitive impairment. The Centers for Medicare and Medicaid Services (CMS) elected not to recommend a specific assessment tool because there is no single, universally accepted screen that satisfies all needs in the detection of cognitive impairment. To provide primary care physicians with guidance on cognitive assessment during the AWV, and when referral or further testing is needed, the Alzheimer's Association convened a group of experts to develop recommendations. The resulting Alzheimer's Association Medicare Annual Wellness Visit Algorithm for Assessment of Cognition includes review of patient Health Risk Assessment (HRA) information, patient observation, unstructured queries during the AWV, and use of structured cognitive assessment tools for both patients and informants. Widespread implementation of this algorithm could be the first step in reducing the prevalence of missed or delayed dementia diagnosis, thus allowing for better healthcare management and more favorable outcomes for affected patients and their families and caregivers.

Cosgrave, M., Anderson, M., Tyrrell, J., Gill, Michael., Lawlor, B.
Cognitive decline in Down syndrome: A validity/reliability study of the Test for Severe Impairment.
American Journal of Mental Retardation,1998,103(2), 193-197.
doi: 10.1352/0895-8017(1998)103<0193:CDIDSA>2.0.CO;2.
Abstract: The utility of the Test for Severe Impairment was studied with 60 older persons who had Down syndrome. Construct validity, test-retest reliability, and interrater reliability were established for the full study group and for subgroups based on degree of mental retardation and dementia status. There was no difference in scores by sex. There were some interesting findings for the group with moderate intellectual disability and dementia and the group with severe intellectual disability without dementia that may signal specific applications and limitations of the test for use with individuals who have Down syndrome. The internal consistency of the instrument was satisfactory. Results suggest that this test is a useful performance-based task for persons with Down syndrome.

Cosgrave, M.P., Tyrrell, J., McCarron, M., Gill, M., & Lawlor, B.A.
Determinants of aggression, and adaptive and maladaptive behavior in older people with Down's syndrome with and without dementia.
Journal of Intellectual Disability Research, 1999, 43(5), 393-399.
DOI:10.1046/j.1365-2788.1999.043005393.x
Abstract: In a cross-sectional study of aggression, and adaptive and maladaptive behavior in 128 subjects with Down syndrome (DS), 29 of whom had dementia, the current authors found that the presence of dementia was not predictive of aggression or maladaptive behavior. However, the level of adaptive behavior was shown to be lower in subjects with dementia, and in those with lower levels of cognitive functioning, as measured on a rating instrument, the Test for Severe Impairment. Although the presence of aggressive behaviors is not higher in subjects with dementia and DS on cross-sectional review, it remains to be seen whether aggression will increase in individual cases with the onset or progression of dementia. The decline in adaptive behavior shown in the present study confirms the findings of previous studies and indicates a direction for service development for persons with the dual diagnosis of dementia and DS.

Cosgrove, M.P., Tyrrell, J., McCarron, M., Gill, M., & Lawlor, B.A.
Age at onset of dementia and age of menopause in women with Down's syndrome.
Journal of Intellectual Disability Research, 1999, 43(6), 461-465. doi:
10.1046/j.1365-2788.1999.00192.x.
Abstract: Menstrual status and the age of menopause were investigated in 143 Irish females with Down syndrome (DS). The average age of menopause in 42 subjects (44.7 years) was younger than in the general population (about age 51). The age at onset of dementia correlated with the age of menopause. This finding may be a manifestation of accelerated ageing in DS or point to oestrogen deficiency being an independent risk factor for the development of Alzheimer's.

Cosgrave, M., Tyrrell, J., Mccarron, M., Gill, M., & Lawlor, B.
Determinants of aggression, and adaptive and maladaptive behaviour in older people with Down's syndrome with and without dementia
*Journal of Intellectual Disability Research,*1999, 43(5), 393-399. doi:
10.1046/j.1365-2788.1999.043005393.x
Abstract: In a cross-sectional study of aggression, and adaptive and maladaptive behavior in 128 subjects with Down syndrome (DS), 29 of whom had dementia, the current authors found that the presence of dementia was not predictive of aggression or maladaptive behavior. However, the level of adaptive behaviour was shown to be lower in subjects with dementia, and in those with lower levels of cognitive functioning, as measured on a rating instrument, the Test for Severe Impairment. Although the presence of aggressive behaviours is not higher in subjects with dementia and DS on cross-sectional review, it remains to be seen whether aggression will increase in individual cases with the onset or progression of dementia. The decline in adaptive behaviour shown in the present study confirms the findings of previous studies and indicates a direction for service development for persons with the dual diagnosis of dementia and DS.

Cosgrove, M., Tyrrell, J., Mccarron, M., Gill, M., & Lawlor, B.
A five year follow-up study of dementia in persons with Down's syndrome: Early symptoms and patterns of deterioration
Indian Journal of Psychological Medicine, 2000, 17(1), 5-11. doi:
10.1017/S0790966700003943
Abstract: To investigate the development of dementia over a five year follow up period in a population of females with Down syndrome; to examine age at onset and duration of dementia in the population; to document the clinical features of dementia and to highlight scores on functional and cognitive rating scales at diagnosis of dementia and at the onset of complete dependency. Authors undertook a five year follow-up study of 80 female subjects on prevalence of dementia, early clinical features of dementia and patterns of scoring on rating scales at diagnosis and end-stage dementia was completed. Over the five year

study period the number of subjects diagnosed with dementia rose from seven (8.75%) to 35 (43.75%). Age related prevalence figres showed that dementia was more common with increasing age. The earliest recognizable symptoms of dementia were memory loss, spatial disorientation, and loss of independence especially in the area of personal hygiene. These findings were confirmed by the rating scales used in the study. Authors concluded that the earliest recognizable clinical features of dementia include memory loss and increased dependency. The results of this study should facilitate earlier diagnosis of dementia in DS.

Courtenay, K., Jokinen, N.S., & Strydom, A.
Caregiving and adults with intellectual disabilities affected by dementia
Journal of Policy and Practice in Intellectual Disabilities, 2010, 7(1), 26-33.
https://doi.org/10.1111/j.1741-1130.2010.00244.x
Abstract: Authors conducted a systematic review of the available Dutch, English, and German language literature for the period 1997–2008 on the current knowledge on social-psychological and pharmacological caregiving with respect to older adults with intellectual disabilities (ID) affected by dementia. Authors note that caregiving occurs on a personal level between the person and their carer and organizational and interorganizational supports have an impact on the quality of care provided. However, the lack of robust evidence to meet the needs of adults with ID affected by dementia means that service organizations often have to extrapolate from the evidence base of dementia care practices in the general population. The review showed that concerns over staff burden, behavioral interventions, and staff training, and applications of models of care were emerging, but were not systematically studied. Authors noted that pharmacological agents and nonpharmacological, psychosocial techniques were being used to assist carers manage behavior, but the evidence base of both nonpharmacological and pharmacological interventions that can help people with ID and dementia and their carers is insufficient because of the absence of systematic and robust studies. The authors note a need for an international research agenda that begins to address gaps in knowledge. With more adults projected to be affected by dementia, a robust evidence-based body of literature on dementia care in people with ID can help with planning for and providing quality dementia-capable services.

Cox, S.
Home solutions: Housing & support for people with dementia
London: The Housing Associations Charitable Trust [78 Quaker Street, London, England E1 6SW; e/m: hact@hact.org.uk] (1998)
112 pp.
Abstract: Publication details some 10 case studies of housing options and accommodations for persons affected by dementia (and applicable to adults with intellectual disabilities). Models covered include: support in a person's own home, support in a shared home, specialist dementia support with communal facilities, and different types and levels of support on one site. Sections also deal with housing and support solutions for people with dementia from ethnic minority communities and the repair, remodeling, adaptation and renovation of ordinary housing. Case models contain full descriptions of settings and accommodations.

Coyle, C.E., Kramer, J., & Mutchler, J.E.
Aging together: Sibling carers of adults with intellectual and developmental disabiliities
Journal of Policy and Practice in Intellectual Disabilities, 2014, 11(4), 302-312; doi: 10.1111/jppi.12094
Abstract: Family care provision is the norm for adults with intellectual and developmental disabilities (I/DD), even as they and their support networks grow older. As families age together, the role of primary carer frequently transitions from the parent to a sibling, as aging parents die or become too frail to provide continued support. This paper explores the transition in care from the perspective of a sibling who has replaced parents as the primary carer for an individual aging with I/DD. Data are drawn from semi-structured, in-depth interviews with a sample of adults over age 40, living in the United States, and caring for a sibling with I/DD(n = 15). Data were analyzed using a constant comparative qualitative approach. Results reveal themes impacting the adjustment to the role of primary carer, the extent to which aging transformed the content of care needs, the importance of planning, and the availability of supplementary support. First, we found that the aging process permeated the careproviding role, requiring ongoing modi?cation of that role as a result of the aging of the adult with I/DD and their family support system. A second key theme is that planning shapes the adjustment to the carer role.A third key theme is that the support systems surrounding the sibling dyad contribute to successful adjustment to care providing.Findings from this study underscore the need to develop long-term services and supports as well as educational resources that accommodate this population of carers as they age together with their sibling with I/DD.

Crook, N., Adams, M., Shorten, N., & Langdon, P. E.
Does the well-being of individuals with Down syndrome and dementia improve when using life story books and rummage boxes? A randomized single case series experiment.
Journal of Applied Research in Intellectual Disabilities, 2016, 29(1), 1-10.
https://doi.org/10.1111/jar.12151
Abstract: This study investigated whether a personalized life story book and rummage box enhanced well-being and led to changes in behavior for people with Down syndrome (DS) who have dementia. A randomized single case series design was used with five participants who had DS and a diagnosis of dementia. Participants were invited to take part in three conditions at random (i) life story book, (ii) rummage box and (iii) no-intervention condition. The two reminiscence conditions were significantly associated with enhanced well-being as compared to the no-intervention condition. However, for one participant, the life story book was associated with significantly higher well-being, while for another participant, the rummage box was associated with significantly higher well-being, suggesting some participants may prefer one method over another.
Personalized life story books and rummage boxes are associated with higher levels of well-being for people with DS and dementia.

Cutler, N.R., Heston, L.L., Davies, P., Haxby, J.V., & Schapiro, M.B.
NIH Conference. Alzheimer's disease and Down's syndrome: new insights.
Annuals of Internal Medicine, 1985,103(4), 566-578.
doi:10.7326/0003-4819-103-4-566.
Abstract: Neuropathologic and neurochemical studies of older adults with Down's syndrome and those with Alzheimer's disease reveal striking similarities. Genetic studies indicate that near relatives of patients with Alzheimer's disease are at increased risk of developing Alzheimer's disease, and the risk appears to be age specific. These families with familial Alzheimer's disease have also been found to have a high incidence of Down's syndrome. Neurochemical data suggest that a cholinergic deficiency must be present for dementia to develop, and serial assessments of brain metabolic function with positron emission tomography in Alzheimer's disease have shown that the parietal lobe has reductions in metabolic function before the onset of neuropsychologic deficits in this brain region. Neuropsychologic testing indicates that patients with Down's syndrome over 35 years old have poorer cognitive skills than do younger patients. Brain metabolic function is excessively reduced in the demented adults with Down's syndrome.

Dalton, A.J., Fedor, B.L., Patti, P.J., Tsiouris, J.A., & Mehta, P.D.
The Multidimensional Observation Scale for Elderly Subjects (MOSES): studies in adults with intellectual disability
Journal of Intellectual & Developmental Disability, 2002, 27(4), 310-324.
https://doi.org/10.1080/1366825021000029348
Abstract: This report describes the results of five studies aimed at evaluating the usefulness, reliability, and validity of the Multidimensional Observation Scale for Elderly Subjects (MOSES) in the assessment of change in ageing persons with intellectual disability. Three hundred and thirty-six individuals with an average age of 49.8 years, including an equal number of men and women, were participants in one or more of the five studies. There were 220 participants with Down syndrome, 81 persons without Down syndrome with intellectual disability, and 35 persons from the general ageing population who were clinically diagnosed with Alzheimer's disease using NINCD/ADRDA criteria. Persons with Down syndrome 40 years of age and older could be distinguished from their younger peers with Down syndrome by statistically significant poorer scores on the MOSES, with those 50 years of age and older showing the worst scores. Comparisons of adults with intellectual disability diagnosed with dementia of the Alzheimer type (DAT), using DSM-IV criteria, with or without Down syndrome, as well as comparisons of patients with clinically diagnosed depression, provided evidence that subtests of the MOSES were sensitive to DAT but less so to depression. These clinical groups also showed significantly poorer scores on the MOSES when compared with those of the normative sample. It was concluded that the MOSES is a behavioural observation scale that can provide useful

information in clinical settings as well as in research.

Dalton, A.J., Mehta, P.D., Fedor, B.L. & Patti, P.J.
Cognitive changes in memory precede those in praxis in aging persons with Down syndrome.
*Journal of Intellectual & Developmental Disability,*1999, *24*(2), 169-187. https://doi.org/10.1080/13668259900033961
Abstract: Experimental tests of cognitive functions were developed and standardised to detect the onset and progression of the early stage of Alzheimer disease in persons with Down syndrome. The aim was to determine whether or not there was a specific sequence of cognitive changes over a 3-year period for the test measures. When compared with a young group (17-39years of age at the start), an old group of persons with Down syndrome (40-58years of age at the start) showed small but statistically significant changes over time suggestive of "pre-clinical signs" of dementia. When the data were sorted into 4 subgroups on the basis of age, a more detailed analysis revealed that the subgroup that was 50 years of age and older at the start showed changes in scores which were of a magnitude more clearly indicative of early dementia on the test measures. Deterioration in learning/ memory functions began at a mean age of 54.2 years, followed later by deterioration in movement-related functions (praxis) at a mean age of 56.9 years. Deterioration in scores on an informant-based behavior rating scale (MOSES) occurred at an intermediate age of 55.0 years. The results provide preliminary support for the hypothesis that persons with Down syndrome who are 50 years of age and older may develop a specific sequence of functional changes during the early stage of dementia. They also illustrate ways in which small sample norms can be effectively used to increase the practical usefulness of tests intended to evaluate dementia in persons with intellectual disabilities.

Das J.P., Davis B., Alexander J., Rauno K.P. & Naglieri J.A.
Cognitive decline due to aging among persons with Down's syndrome.
Research in Developmental Disabilities, 1995, Nov-Dec, 16(6): 461-4788 doi: 10.1016/0891-4222(95)00030-5.
Abstract: This study examined decline in cognitive functions in individuals with Down syndrome (DS) over the age of 40 in comparison to participants of the same age and comparable mental handicap without Down syndrome (NonDS). Both DS (n = 32) and NonDS (n = 31) samples were divided into "younger" (40-49 years) and "older" (50-62) groups. Cognitive processes were examined by tests of general intellectual functioning (Dementia Rating Scale, Peabody Picture Vocabulary Test-Revised, and the Matrix Analogies Test-Expanded form), as well as planning, attention, simultaneous, and successive processing tests taken from Das-Naglieri Cognitive Assessment System. The older individuals with Down syndrome performed more poorly than those in the other three groups. The differences were particularly evident in tasks requiring planning and attention. Authors note the possibility of using these tests as indicators of the early signs of Alzheimer's disease.

Davis, D.R.
A parent's perspective
In M.P. Janicki & A.J. Dalton (Eds.), Dementia, aging, and intellectual disabilities.
pp. 42-50
Philadelphia: Brunner-Mazel (1999)
Abstract: Book chapter that provides an account of the experiences of a family with an adult son with Down syndrome who eventually succumbs to dementia of the Alzheimer's type. Includes a discussion of the difficult early years of the son's life and the challenges the family faced as he aged. It also examines the family's problems in recognizing that their son was experiencing the onset of dementia and his gradual decline until his death at age 46.

Day, K., Carreon, D., & Stump, C.
The therapeutic design of environments for people with dementia: A review of the empirical research
Gerontologist, 2000, 40, 397-416. https://doi.org/10.1093/geront/40.4.397
Abstract: Design of the physical environment is increasingly recognized as an important aid in caring for people with dementia. This article reviews the empirical research on design and dementia, including research concerning facility planning (relocation, respite and day care, special care units, group size), research on environmental attributes (noninstitutional character, sensory stimulation, lighting, safety), studies concerning building organization (orientation, outdoor space), and research on specific rooms and activity spaces (bathrooms, toilet rooms, dining rooms, kitchens, and resident rooms). The analysis reveals major themes in research and characterizes strengths and shortcomings in methodology, theoretical conceptualization, and application of findings.

Davies, M., McGllade, A., & Bickerstaff, D.
A needs assessment of people in the Eastern Health and Social Services Board (Northern Ireland) with intellectual disability and dementia
Journal of Learning Disabilities, 2002, 6, 23-33.
https://doi.org/10.1177/146900470200600
Abstract: Article details a study undertaken by the Eastern Health and Social Services Board (Northern Ireland) which aimed to identify the number of people with intellectual disability within this area who were diagnosed with or were thought to have dementia. The objectives of the study were to collate demographic details and to profile the needs of this group. Key workers were asked to provide this information and were invited to comment on gaps in existing service provision and on future needs. A number findings emerged: diagnostic services were patchy; people with dementia were living in a range of residential settings; carers wished to care for their clients for as long as practically possible, but they required extra resources and training to do so; and some individuals with an intellectual disability were excluded from elderly services. A report was compiled incorporating 12 recommendations.

Davys, D., & Haigh, C.
Older parents of people who have a learning disability: Perceptions of future accommodation needs*British Journal of Learning Disabilities,* 2007, 36, 66–72. doi:10.1111/j.1468-3156.2007.00447.x
Abstract: The aim of this qualitative study was to provide an insight into the perceptions of older parents of adults with intellectual diability on the future accommodation needs of their adult children. Semi-structured interviews were used to seek parental awareness of residential options available, concerns in relation to future accommodation and the preferred accommodation options for their offspring. Four couples who shared the family home with an adult who has an intellectual disabilty took part in the study and data were analyzed using a step-by-step form of content analysis as described by Burnard *[Nurse Educ Today 11 (1991), 461*]. Emergent themes from transcripts were then organized into main categories. The results of this study suggest that older parents are dissatisfied with both statutory and private services, that they have concerns for their other children, and their own aging. Being a parent to a person who has an intellectual disability is seen to be a difficult task, were worried about what the adult's brothers and sisters may have to do in the future, but also wanted to provide support at home for as long a possible. Of the parents who participated in this study, three couples wanted to maintain their adult child at home for as long as possible and the parents who were actively seeking accommodation outside the family home expected to be involved in all aspects of their daughter's care for the long-term future.

de Franca Bram, J.M., Talib, L.L., Giroud Joaquim, H.P., Carvalho, C.L., Gattaz, W.G., & Forlenza, O.V.
Alzheimer's disease-related biomarkers in aging adults with Down syndrome: systematic review
Current Psychiatry Research and Reviews, 2019, 15(1), 49-57. doi: 10.2174/1573400515666190122152855
Abstract: Down syndrome (DS) is associated with a high prevalence of cognitive impairment and dementia in middle age and older adults. Given the presence of common neuropathological findings and similar pathogenic mechanisms, dementia in DS is regarded as a form of genetically determined, early-onset AD. The clinical characterization of cognitive decline in persons with DS is a difficult task, due to the presence intellectual disability and pre-existing cognitive impairment. Subtle changes that occur at early stages of the dementing process may not be perceived clinically, given that most cognitive screening tests are not sensitive enough to detect them. Therefore, biological markers will provide support to the diagnosis of DS-related cognitive impairment and dementia, particularly at early stages of this process. To perform a systematic review of the literature on AD-related biomarkers in DS. We searched PubMed, Web of Science and Cochrane Library for scientific papers published between 2008 and 2018 using as primary mesh terms 'Down', 'Alzheimer', 'biomarker'. 79 studies were retrieved, and 39 were considered eligible for inclusion in the systematic review: 14 post-mortem studies, 10 neuroimaging, 4 addressing cerebrospinal

fluid biomarkers, and 11 on peripheral markers. There is consistent growth in the number of publication in this field over the past years. Studies in DS-related dementia tend to incorporate many of the diagnostic technologies that have been more extensively studied and validated in AD. In many instances, the study of CNS and peripheral biomarkers reinforces the presence of AD pathology in DS.

de Sola, S., de la Torre, R., Sánchez-Benavides, G., Benejam, B. , Cuenca-Royo, A., del Hoyo, L., Rodríguez, J., Catuara-Solarz, S., Sanchez-Gutierrez, J., Dueñas-Espin, I., Hernandez, G., Peña-Casanova, J., Langohr, K., Videla, S., Blehaut, H., Farre,M,. Dierssen, M. & The TESDADStudy Group
A new cognitive evaluation battery for Down syndrome and its relevance for clinical trials.
Frontiers in Psychology, 2015, 6, 708. doi:10.3389/fpsyg.2015.00708.
Abstract: The recent prospect of pharmaceutical interventions for cognitive impairment of Down syndrome (DS) has boosted a number of clinical trials in this population. However, running the trials has raised some methodological challenges and questioned the prevailing methodology used to evaluate cognitive functioning of DS individuals. This is usually achieved by comparing DS individuals to matched healthy controls of the same mental age. We propose a new tool, the TESDAD Battery that uses comparison with age-matched typically developed adults. This is an advantageous method for probing the clinical efficacy of DS therapies, allowing the interpretation and prediction of functional outcomes in clinical trials. In our DS population the TESDAD battery permitted a quantitative assessment of cognitive defects, which indicated language dysfunction and deficits in executive function, as the most important contributors toother cognitive and adaptive behavior outcomes as predictors of functional change in DS. Concretely, auditory comprehension and functional academics showed the highest potential as end-point measures of therapeutic intervention for clinical trials: the former as a cognitive key target for therapeutic intervention, and the latter as a primary functional outcome measure of clinical efficacy. Our results also emphasize the need to explore the modulating effects of IQ, sex, and age on cognitive enhancing treatments. Noticeably, women performed significantly better than men of the same age and IQ in most cognitive tests, with the most consistent differences occurring in memory and executive functioning and negative trends rarely emerged on quality of life linked to the effect of age after adjusting for IQ and sex. In sum, the TESDAD battery is a useful neurocognitive tool for probing the clinical efficacy of experimental therapies in interventional studies in the DS population suggesting that age-matched controls are advantageous for determining normalization of DS.

De La Garza, E., Scott, A., Hillerstrom , H, Hendrix, J., Rubenstein, E.
Caregivers' concerns and supports needed to care for adults with Down syndrome.
Am J Med Genet C Semin Med Genet. 2024 Mar;196(1):e32041. doi: 10.1002/ajmg.c.32041. Epub 2023 Apr 18.
Abstract: Research regarding caregivers for individuals with Down syndrome mainly focuses on outcomes for the pediatric population and not on the experience of caregivers themselves. Our objective was to understand caregiver-reported experiences and concerns for themselves and the individual they care for through a survey of caregivers of adults with Down syndrome. We conducted a survey of N = 438 caregivers of adults with Down syndrome and asked about the perspectives of the respondents surrounding caregiving and demographics. The most common concerns among caregivers were planning for future needs (72.1%) and what happens when they (the caregiver) are gone (68.3%). Concerns they had for the individual they cared for were employment (63.2%) and friendships/relationships (63.2%). We found no significant difference in responses based on caregiver education level. Our survey identified six themes for the feedback about what clinical and research professionals should know to better serve individuals with Down syndrome, their families, and those who support them. Many caregivers discussed topics including healthcare, coordination, competence, and ability. More efforts for research into the caregiver experience for adults with Down syndrome are needed.

De Vreese, L.P., Gomiero, T., Uberti M., De Bastiani, E., Weger, E., Mantesso, U., & Marangoni, A.
Functional abilities and cognitive decline in adult and aging intellectual disabilities. Psychometric validation of an Italian version of the Alzheimer's Functional Assessment Tool (AFAST): analysis of its clinical significance with linear statistics and artificial neural networks.
Journal of Intellectual Disability Research, 2015, Apr; 59(4), 370-84. doi: 10.1111/jir.12113.
Abstract: A psychometric validation of an Italian version of the Alzheimer's Functional Assessment Tool scale (AFAST-I), designed for informant-based assessment of the degree of impairment and of assistance required in seven basic daily activities in adult/elderly people with intellectual disabilities (ID) and (suspected) dementia; and a pilot analysis of its clinical significance with traditional statistical procedures and with an artificial neural network. AFAST-I was administered to the professional caregivers of 61 adults/seniors with ID with a mean age (± SD) of 53.4 (± 7.7) years (36% with Down syndrome). Internal consistency (Cronbach's a coefficient), inter/intra-rater reliabilities (intra-class coefficients, ICC) and concurrent, convergent and discriminant validity (Pearson's r coefficients) were computed. Clinical significance was probed by analysing the relationships among AFAST-I scores and the Sum of Cognitive Scores (SCS) and the Sum of Social Scores (SOS) of the Dementia Questionnaire for Persons with Intellectual Disabilities (DMR-I) after standardization of their raw scores in equivalent scores (ES). An adaptive artificial system (AutoContractive Maps, AutoCM) was applied to all the variables recorded in the study sample, aimed at uncovering which variable occupies a central position and supports the entire network made up of the remaining variables interconnected among themselves with different weights. AFAST-I shows a high level of internal homogeneity with a Cronbach's a coefficient of 0.92. Inter-rater and intra-rater reliabilities were also excellent with ICC correlations of 0.96 and 0.93, respectively. The results of the analyses of the different AFAST-I validities all go in the expected direction: concurrent validity (r=-0.87 with ADL); convergent validity (r=0.63 with SCS; r=0.61 with SOS); discriminant validity (r=0.21 with the frequency of occurrence of dementia-related Behavioral Excesses of the Assessment for Adults with Developmental Disabilities, AADS-I). In our sample age and gender do not correlate with the scale and comparing the distribution of the AFAST-I and DMR-SCS and DMR-SOS expressed as ES, it appears that memory disorders and temporal and spatial disorientation (SCS) precede the loss of functional abilities, whereas changes in social behaviour (SOS) are less specific in detecting cognitive deterioration sufficient to provoke functional disability and vice versa. The results of AutoCM analysis reveal that the hub (core) of the entire network is represented by the functional domain 'personal/oral hygiene' in the entire study sample and 'use of toilet' in a subgroup of subjects who obtained an ES equal to 0 at DMR-SCS. These results confirm the reliability and validity of AFAST-I and emphasise the complexity of the relationship among functional status, cognitive functioning and behaviour also in adults/seniors with ID.

De Vreese, L. P., Mantesso, U., De Bastiani, E., Weger, E., Marangoni, A.C., & Gomiero, T.
Impact of dementia-derived nonpharmacological intervention procedures on cognition and behavior in older adults with intellectual disabilities: a 3-year follow-up study
Journal of Policy and Practice in Intellectual Disabilities, 2012, 9(2), 92-102. https://doi.org/10.1111/j.1741-1130.2012.00344.x
Abstract: Dementia appears at a higher rate among some adults with intellectual disabilities (ID) and this potentially poses a greater risk of nursing home admission. Yet, to date, there is no evidence on the efficacy of general dementia-derived environment-, personnel-, and patient-oriented intervention strategies in delaying onset of dementia or in slowing down its rate of progression in this population. To investigate the feasibility and efficacy of a multicomponent nonpharmacological approach, the authors studied a sample of 14 adults with worsening cognition and everyday functioning who were no longer manageable by their family or staff in day centers or, and who were relocated in a model special care unit (SCU) designed to proactively accommodate the needs of people with ID and dementia. Baseline level and rate of decline across a 3-year period were assessed by means of the Dementia Questionnaire for Persons with Intellectual Disabilities and compared to two control groups not in dementia-capable programs matched for age, sex, and severity of ID. After 3 years, the authors found some improvement in cognition and stabilization in everyday functioning and behaviors in the SCU residents and a worsening in the control groups. The authors noted that enrollment in a dementia-capable program facilitated daily practice of residents' residual skills and abilities, enhancing their memory and verbal communication, that the prosthetic environment contributed to activity maintenance and appropriate intellectual challenges, and that the greater participation on an individual level added to the skill maintenance. Although the interpretation of these positive findings is not straightforward, they

confirm the validity of this "in-place progression" model and provide a platform for continuing progress in person-centered services and care for aging persons with ID.

De Vreese, L.P. Mantesso, U., de Bastiani, E., Marangoni, A., & Gomiero, T.
Psychometric evaluation of the Italian version of the AADS questionnaire: A caregiver-rated tool for the assessment of behavioral deficits and excesses in persons with intellectual disabilities and dementia
International Psychogeriatrics, 2011, 23, 1124-1132. doi: 10.1017/S1041610211000342.
Abstract: The aim of this study was to verify the reliability and validity of the Italian version of the Assessment for Adults with Developmental Disabilities (AADS-I), the only available measure specifically designed to assess the frequency, management difficulties and impact on the quality of life (QoL) of positive and negative non-cognitive symptoms in persons with intellectual disabilities (ID) and dementia. AADS-I was administered to professional carers of 63 aging ID individuals. We computed the internal consistency separately of the frequency, management difficulty and effect on the QoL subscales of Behavioral Excesses and Behavioral Deficits and their inter-rater and test-retest reliabilities. Homogeneity of AADS-I was found to range from good to excellent: Cronbach's a coefficients were 0.77, 0.83 and 0.82, respectively for frequency, management difficulty and effect on the QoL of Behavioral Excesses, and 0.82, 0.76 and 0.79 of Behavioral Deficits. Intraclass correlation coefficients (ICC) between two independent carers were 0.67, 0.79 and 0.73 and 0.67, 0.67 and 0.67 for frequency, management difficulty and effect on the QoL of Behavioral Excesses and Deficits, respectively. Corresponding ICC for test-retest reliability were 0.80, 0.75, 0.78 and 0.70, 0.81, 0.81. Age, sex and typology of ID did not correlate with the AADS-I subscale scores, whereas the severity of ID related only with the frequency subscale of Behavioral Deficits. This subscale also correlated with the Dementia Questionnaire for Persons with Intellectual Disabilities. Behavioral deficits are more frequent in subjects with dementia. These results confirm the reliability and validity of the Italian version of AADS.

De Vreese, L.P., Uberti, M., Mantesso, U., De Bastiani, E., Weger, E., Marangoni, A.C., Weiner, M.F. & Gomiero, T.
Measuring quality of life in intellectually disabled persons with dementia with the Italian version of the quality of life in late-stage dementia (QUALID) scale.
Journal of Alzheimer's Disease and Parkinsonism, 2012, 2:104e. doi:10.4172/2161-0460.1000104
Abstract: The aim of this study is to verify a cross-cultural adaptation of an Italian version of the Quality of Life in Late-Stage Dementia (QUALID) scale in a sample of aging people with intellectual disabilities (ID). The QUALID was translated according to standardized procedures. Internal consistency was analyzed using Cronbach's alpha. A Principal Component Analysis verified its multidimensionality. Inter-rater and test-retest reliabilities were also assessed using the Intraclass Correlation Coefficient (ICC). Convergent validity was probed by Spearman's correlations among the QUALID score and the six sub-scores of the Assessment for Adults with Development Disabilities (AADS), a proxy-based questionnaire rating behavioral excesses and deficits commonly found in people with intellectual disabilities and dementia. Clinical validity was assessed by comparing QUALID scores obtained by subjects with and without dementia using the Mann-Whitney U test. A total of 40 adults/older people with ID at five ID-specific centers in the province of Trento and Cremona participated in the study. Findings show optimal levels of internal consistency (a = 0.80) and confirm the factors identified in the Spanish validation study (symptoms of discomfort, positive social interaction and depression). The scale has high inter-rater (ICC = 0.95) and good test-retest reliabilities (ICC = 0.89). The total QUALID score correlates significantly with the AADS sub-scores for behavioral excesses, but does not differ between individuals with and without dementia, though two out of the three identified factor scores are significantly higher in the dementia subgroup. The authors conclude that the Italian version of the QUALID is a reliable and valid instrument for estimating quality of life in aging adults with ID and dementia.

Deb, S., & Braganza. J.
Comparison of rating scales for the diagnosis of dementia in adults with Down's syndrome.
Journal of Intellectual Disability Research,1999 Oct, 43(Pt 5), 400-407. doi: 10.1046/j.1365-2788.1999.043005400.x.
Abstract: Making a diagnosis of dementia, particularly in its early stages, in a person with intellectual disability can be difficult Some neuropsychological tests which were originally devised for the diagnosis of dementia in the non-intellectually disabled population have been modified for use in people with intellectual disability. Observer-rated scales have also been used for making a diagnosis of dementia in people with intellectual disability. Within the context of a genetic study, the rates of diagnosis of dementia according to different criteria, namely the clinician's diagnosis (ICD-10), the Dementia Questionnaire for Persons with Mental Retardation (DMR), the Dementia Scale for Down Syndrome (DSDS) and the Mini Mental State Examination (MMSE), were compared among 62 adults with Down's syndrome (26 demented and 36 non-demented adults according to the clinician's diagnosis). A comparison between the clinician's diagnosis and the diagnosis according to DMR criteria showed specificity and sensitivity at the 0.92 level for both categories. Similarly, a comparison between the clinician's diagnosis and the diagnosis according to the DSDS criteria showed a specificity of 0.89 and a sensitivity of 0.85. A good positive correlation was also shown between the scores of the DSDS and the DMR (Pearson's r = +0.868, P < 0.001). A similar positive correlation was found between the overall DSDS score and the scores in the main subcategories of the DMR. An MMSE could be performed in only 34 (55%) out of the 62 subjects with Down's syndrome. Out of the 30 subjects who had an MMSE score of less than 24 (the usual cut-off for the diagnosis of possible dementia), 23 (77%) did not have a diagnosis of dementia according to any criteria. It seems that the observer-rated scales, rather than the direct neuropsychological tests, are more useful for the diagnosis of dementia in people with an intellectual disability.

Deb, S., Hare, M.A., Prior, L., & Bhaumik, S.
Dementia screening questionnaire for individuals with intellectual disabilities.
British Journal of Psychiatry, 2007, 190, 440-444. doi:10.1192/bjp.bp.106.024984.
Abstract: Many adults with Down syndrome develop Alzheimer's dementia relatively early in their lives, but accurate clinical diagnosis remains difficult. The authors set out too develop a user-friendly observer-rated dementia screening questionnaire with strong psychometric properties for adults with intellectual disabilities. They used qualitative methods to gather information from carers of people with Down syndrome about the symptoms of dementia. This provided the items for the Dementia Screening Questionnaire for Individuals with Intellectual Disabilities (DSQIID) which was then tested for its psychometric properties. The DSQIID was administered to carers of 193 adults with Down syndrome, 117 of whom were examined by clinicians who confirmed a diagnosis of dementia for 49 according to modified ICD-10 criteria. They established that a total score of 20 provides maximum sensitivity (0.92) and optimum specificity (0.97) for screening. The DSQIID has sound internal consistency ($\propto$=0.91) for all its 53 items, and good test-retest and interrater reliability. The authors established a good construct validity by dividing the questionnaire items into four factors. The authors conclude that the DSQIID is valid, reliable and user-friendly observer-rated questionnaire for screening for dementia among adults with Down syndrome.

Deb, S., Hare, M. & Prior, L.
Symptoms of dementia among adults with Down's syndrome: a qualitative study.
Journal of Intellectual Disability Research, 2007, 51, 726-739. DOI:10.1111/j.1365-2788.2007.00956.x
Abstract: Dementia is common among adults with Down's syndrome (DS); yet the diagnosis of dementia, particularly in its early stage, can be difficult in this population. One possible reason for this may be the different clinical manifestation of dementia among people with intellectual disabilities. The aim of this study was to map out the carers' perspective of symptoms of dementia among adults with DS in order to inform the development of an informant-rated screening questionnaire. Unconstrained information from carers of people with DS and dementia regarding the symptoms, particularly the early symptoms of dementia, was gathered using a qualitative methodology. Carers of 24 adults with DS and dementia were interviewed. The interviews were recorded and fully transcribed. The transcripts were then analyzed using qualitative software. There appeared to be many similarities in the clinical presentation of dementia in adults with DS and the non-intellectually disabled general population. Like in the non-intellectually disabled general population, forgetfulness especially, impairment of recent memory combined with a relatively intact distant memory and confusion were common, and presented early in dementia among adults with DS. However, many 'frontal lobe'-related symptoms that are usually manifested later in the process of dementia among the general population were common at

an early stage of dementia among adults with DS. A general slowness including slowness in activities and speech, other language problems, loss of interest in activities, social withdrawal, balance problems, sleep problems, loss of pre-existing skills along with the emergence of emotional and behavior problems were common among adults with DS in our study. This study highlighted the similarities in the clinical presentation of dementia among the general population and people with DS with a particular emphasis on the earlier appearance of symptoms associated with the frontal lobe dysfunction among adults with DS.

Deb, S.S. , Strydom, A., Hithersay, T., Gomiero, T., De Vreese,. L.P., Janicki, M.P., Jokinen, N.S., Service, K.
Dementia in people with intellectual disabilities.
In: Bertelli, M.O., Deb, S., Munir, K., Hassiotis, A., Salvador-Carulla, L. (eds) Textbook of Psychiatry for Intellectual Disability and Autism Spectrum Disorder, 2022. Springer, Cham. https://doi.org/10.1007/978-3-319-95720-3_28
Abstract: With the increasing life expectancy in the last five decades, people with intellectual disabilities (ID) are exposed to the risk of age-related neurodegenerative disorders including dementia. The prevalence of dementia is increased in people with ID compared with that in the general population. People with Down syndrome (DS) are at even a higher risk of developing Alzheimer's dementia (AD) compared with non-DS people with ID. However, there are difficulties in making an early and accurate diagnosis of dementia in individuals with ID and the screening instruments such as the Mini Mental Status Examination that are used in the general population often show floor effects when used for individuals with ID because of their pre-existing cognitive impairment, the level of which varies depending on the severity of ID. Both informant-rated and direct neuropsychological tests have been used for the case detection of dementia for individuals with ID. However, direct neuropsychological tests cannot be used for those who have severe ID and their validity could still be questionable in a number of cases of mild to moderate ID. There are many similarities and some differences in the clinical manifestation of dementia in individuals with ID and the non-ID general population. Impaired recent memory and confusion in the context of relatively intact distant memory are likely to be the early symptoms in individuals with ID who have a mild ID, whereas loss of skills and change in behavior are likely to be the early features for those with more severe ID. Many symptoms, including features of `frontal lobe dysfunction' that tend to appear late in dementia in the general population, may appear early in individuals with ID and DS. Ideally, individuals with ID should be screened for signs of dementia from before the age of 30/35. A multi-disciplinary approach should be taken for diagnosis of dementia in individuals with ID using a combination of informant-rated scales and neuropsychological tests in a longitudinal fashion over time. Important differential diagnoses include hypothyroidism, depression, and sensory impairment. Assessment should include physical, psychological, and social aspects including appropriate examinations and investigations. The evidence-base for the pharmacological management of dementia in ID and DS is poor, which does not allow to draw any definitive conclusion about their efficacy in this population. Therefore, the same pharmacological regime that is used in the non-ID population along with non-pharmacological interventions should be considered for people with ID.

Dekker, A.D., Coppus, A.M.W., Strydom, A., Vermeiren, Y., Grefelman, S., Eleveld, J., Beugelsdijkm G., ... De Deyn, P.P.
Behavioural and psychological symptoms of dementia in Down syndrome: European development of the novel BPSD-DS evaluation scale
European Neuropsychopharmacology, 2016 (Oct.), 26, S641 [Presentation at ECNP Congress 2016, Vienna, Austria] DOI: 10.1016/S0924-977X(16)31739-4
Abstract: Core to the dementia are behavioral and psychological symptoms of dementia (BPSD), an umbrella term for various behavioral changes associated with the presence of dementia, such as apathy, anxiety, disinhibition, irritability and psychosis. BPSD are associated with increased suffering, a reduced quality of life, earlier institutionalization and accelerated cognitive decline for patients, and increased caregiver burden. Although accurate recognition of BPSD may increase acceptance and understanding, enable adaptive caregiving and, importantly, allow for therapeutic interventions, BPSD have not been comprehensively evaluated in the Down syndrome (DS) population. BPSD are extensively assessed in AD patients in the general population using focussed behavioural scales, such as the Neuropsychiatric Inventory. However, not a single behavioural assessment scale has been adapted for the DS population, thus not considering the DS-specific circumstances. The authors sought to develop an adapted evaluation scale for comprehensive assessment of BPSD in DS, taking pre-existing behavior and limitations associated with intellectual disability into account. A multidisciplinary collaboration of major DS expertise centers in Europe has been established to develop the novel BPSD-DS evaluation scale. Development consisted of a series of phases, starting with a thorough evaluation of the rather limited knowledge on BPSD in DS and extensive expert feedback rounds to identify relevant behavioral items. A structured, informant interview has been adopted, since people with DS often have difficulty to verbalize their emotions and feelings. Using defined scoring definitions for frequency, severity and caregiver burden at two periods of time, the scales enables identification of specific changes over time, i.e. disentangling behavioral alterations from characteristic behavior that has always been typical for an individual. Multiple optimization rounds and a pilot study yielded the final version that is tested until summer 2016 in a multi-center European validation phase including approx. 250 DS individuals with and without dementia. The BPSD-DS evaluation scale exclusively focuses on behaviour, including 83 items in twelve clusters: anxiety & nervousness, sleep disturbances, irritability, obstinacy, agitation & stereotypical behaviour, aggression, apathy & aspontaneity, depression, delusions, hallucinations, disinhibition & sexual behaviour, and eating and drinking behavior. Scale development, the final version, as well as the first results from the validation phase will be presented. Current clinical experiences and preliminary results for interrater reliability (>.90) are promising. BPSD have been largely neglected in DS. The novel BPSD-DS evaluation scale is the first comprehensive tool for its systematic assessment in the DS population, and serves a three-fold purpose in the future: (1) to monitor behavioural changes in daily care, (2) to identify early BPSD in DS in longitudinal research studies, and (3) to assess behavioural outcome measures in clinical trials for dementia in DS.

Dekker, A.D., Sacco, S., Carfi, A., Benejam, B., Vermeiren, Y., Beugelsdijk, G., Schippers, M., Hassefras, L,, Eleveld, J,, Grefelman, S,, Fopma, R,, Bomer-Veenboer, M,, Boti, M,, Oosterling, G.D.E, Scholten, E., Tollenaere, M., Checkley, L., Strydom, A., Van Goethem, G., Onder, G., Blesa, R., Zu Eulenburg, C., Coppus, A.M.W., Rebillat, A.S., Fortea, J., De Deyn, P.P.
The Behavioral and Psychological Symptoms of Dementia in Down Syndrome (BPSD-DS) Scale: Comprehensive assessment of psychopathology in Down syndrome
Journal of Alzheimers Disease, 2018, 63(2), 797-819. doi: 10.3233/JAD-170920.
Abstract: People with Down syndrome (DS) are prone to develop Alzheimer's disease (AD). Behavioral and psychological symptoms of dementia (BPSD) are core features, but have not been comprehensively evaluated in DS. In a European multidisciplinary study, the novel Behavioral and Psychological Symptoms of Dementia in Down Syndrome (BPSD-DS) scale was developed to identify frequency and severity of behavioral changes taking account of life-long characteristic behavior. 83 behavioral items in 12 clinically defined sections were evaluated. The central aim was to identify items that change in relation to the dementia status, and thus may differentiate between diagnostic groups. Structured interviews were conducted with informants of persons with DS without dementia (DS, n = 149), with questionable dementia (DS+Q, n = 65), and with diagnosed dementia (DS+AD, n = 67). First exploratory data suggest promising interrater, test-retest, and internal consistency reliability measures. Concerning item relevance, group comparisons revealed pronounced increases in frequency and severity in items of anxiety, sleep disturbances, agitation & stereotypical behavior, aggression, apathy, depressive symptoms, and eating/drinking behavior. The proportion of individuals presenting an increase was highest in DS+AD, intermediate in DS+Q, and lowest in DS. Interestingly, among DS+Q individuals, a substantial proportion already presented increased anxiety, sleep disturbances, apathy, and depressive symptoms, suggesting that these changes occur early in the course of AD. Future efforts should optimize the scale based on current results and clinical experiences, and further study applicability, reliability, and validity. Future application of the scale in daily care may aid caregivers to understand changes, and contribute to timely interventions and adaptation of caregiving.

Dekker, A.D., Strydom, A., Coppus, A,M,W., Nizetic, D., Vermeiren,, Y., Naude, P.J.W., Van Dam, D., Potier, M-C., Fortea, J., De Deyn, P.P.
Behavioural and psychological symptoms of dementia in Down syndrome: Early indicators of clinical Alzheimer's disease?
Cortex, 2015, 75, 36-61. doi: 10.1016/j.cortex.2015.07.032. Epub 2015 Aug 13.
Abstract: Behavioral and psychological symptoms of dementia (BPSD) are a core symptom of dementia and are associated with earlier institutionalization and accelerated cognitive decline for adults with Down syndrome (DS) and increased

caregiver burden. Despite the extremely high risk for DS individuals to develop dementia due to Alzheimer's disease (AD), BPSD have not been comprehensively assessed in the DS population. Due to the great variety of DS cohorts, diagnostic methodologies, sub-optimal scales, covariates and outcome measures, it is questionable whether BPSD have always been accurately assessed. However, accurate recognition of BPSD may increase awareness and understanding of these behavioral aberrations, thus enabling adaptive caregiving and, importantly, allowing for therapeutic interventions. Particular BPSD can be observed (long) before the clinical dementia diagnosis and could therefore serve as early indicators of those at risk, and provide a new, non-invasive way to monitor, or at least give an indication of, the complex progression to dementia in DS. This review found that various BPSD appear to be altered in demented DS individuals, but study results have not always been consistent. From childhood to adulthood, externalizing behavior likely decreases and internalizing behavior increases. Frontal lobe symptoms have been suggested as early signs of AD in DS. Disinhibition and apathy, as well as executive dysfunction, seem to be omnipresent in the prodromal phase, whereas reports are inconsistent for depression. Regarding activity disturbances, studies indicated decreasing hyperactivity levels towards adulthood. Excessive activity in demented DS individuals should be a fairly easy observable sign, however, general slowness has been reported and apathy itself might cause reduced activity. Agitation appears to be more prevalent in demented than in non-demented DS individuals, but reports on aggression are inconsistent, though aggression seems to be reduced in the overall DS population. Sleep disturbances are markedly present in both demented and non-demented DS individuals. Although sleep disorders may not yet differentiate between those with and without AD, they are important to consider as such sleep disorders may aggravate cognitive decline and BPSD.

Dekker, A.D., Wissing, M.B.G., Ulgiati, A.M., Bijl, B., van Gool, G., Groen, M.R., Grootendorst, E.S., van der Wal, I.A., Hobbelen, J.S.M., De Deyn, P.P., & Waninge, A.
Dementia in people with severe or profound intellectual (and multiple) disabilities: Focus group research into relevance, symptoms and training needs.
Journal of Applied Research in Intellectual Disability, 2021, Jul 2. doi: 10.1111/jar.12912. Online ahead of print.
Abstract: Differentiating dementia from baseline level of functioning is difficult among people with severe/profound intellectual (and multiple) disabilities. Moreover, studies on observable dementia symptoms are scarce. This study examined (a) the relevance of dementia diagnosis, (b) observable symptoms and (c) training/information needs. Four explorative focus groups were held with care professionals and family members who have experience with people with severe/profound intellectual (and multiple) disabilities (≥40 years) and decline/dementia. Thematic analysis showed that participants wanted to know about a dementia diagnosis for a better understanding and to be able to make informed choices (question 1). Using a categorization matrix, cognitive and behavioural changes were shown to be most prominent (question 2). Participants indicated that they needed enhanced training, more knowledge development and translation, and supportive organizational choices/policies (question 3). Timely identifying/diagnosing dementia allows for a timely response to changing needs. This requires a better understanding of symptoms.

Dekker, A.D., Sacco, S., Carfi, A., Benejam, B., Vermeiren, Y., Beugelsdijk, G., Schippers, M., Hassefras, L., Eleveld, J., Grefelman, S., Fopma, R., Bomer-Veenboer, M., Boti, M., Oosterling, G.D.E., Scholten, E., Tollenaere, M., Checkley, L., Strydom, A., Van Goethem, G., Onder, G., Blesa, R., Eulenburg, Z.C., Coppus, A.M.W., Rebillat, A.S., Fortea, J., & De Deyn, P.P.
The Behavioral and Psychological Symptoms of Dementia in Down Syndrome (BPSD-DS) Scale: Comprehensive Assessment of Psychopathology in Down Syndrome.
Journal of Alzheimer's Disease, 2018, 63(2), 797-819. doi: 10.3233/JAD-170920.
Abstracat: People with Down syndrome (DS) are prone to develop Alzheimer's disease (AD). Behavioral and psychological symptoms of dementia (BPSD) are core features, but have not been comprehensively evaluated in DS. In a European multidisciplinary study, the novel Behavioral and Psychological Symptoms of Dementia in Down Syndrome (BPSD-DS) scale was developed to identify frequency and severity of behavioral changes taking account of life-long characteristic behavior. 83 behavioral items in 12 clinically defined sections were evaluated. The central aim was to identify items that change in relation to the dementia status, and thus may differentiate between diagnostic groups. Structured interviews were conducted with informants of persons with DS without dementia (DS, n = 149), with questionable dementia (DS+Q, n = 65), and with diagnosed dementia (DS+AD, n = 67). First exploratory data suggest promising interrater, test-retest, and internal consistency reliability measures. Concerning item relevance, group comparisons revealed pronounced increases in frequency and severity in items of anxiety, sleep disturbances, agitation & stereotypical behavior, aggression, apathy, depressive symptoms, and eating/drinking behavior. The proportion of individuals presenting an increase was highest in DS+AD, intermediate in DS+Q, and lowest in DS. Interestingly, among DS+Q individuals, a substantial proportion already presented increased anxiety, sleep disturbances, apathy, and depressive symptoms, suggesting that these changes occur early in the course of AD. Future efforts should optimize the scale based on current results and clinical experiences, and further study applicability, reliability, and validity. Future application of the scale in daily care may aid caregivers to understand changes, and contribute to timely interventions and adaptation of caregiving.

Dekker, A.D., Ulgiati, A.M., Groen, H., Boxelaar, V.A., Sacco, S., Falquero, S., Carfi, A.., di Paola, A., Benejam, B., Valldeneu, S., Fopma, R., Oosterik, M,. Hermelink, M., Beugelsdijk, G., Schippers, M., Henstra, H., Scholten-Kuiper, M., Willink-Vos, J., de Ruiter, L., Willems, L., Loonstra-de Jong, A., Coppus, A.M.W., Tollenaere, M., Fortea, J., Onder, G.,Rebillat, A-S., Van Dam, D., De Deyn, P.P.
The Behavioral and Psychological Symptoms of Dementia in Down Syndrome Scale (BPSD-DS II): Optimization and Further Validation.
Journal of Alzheimer's Disease, 2021, 82(3), 1371-1371, 2021
Abstract: [Corrections: In Figure 8, p. 1519, the second graph (item 10.3) reads: "10.3 making inappropriate comments". `Making inappropriate comments' is the description belonging to item 10.2 of the BPSD-DS II. Instead, the second graph (item 10.3) should read: "10.3 loss of decorum".]

Dekker, A.D., Ulgiati, A.M., Groen, H., Boxelaar, V.A., Sacco, S., Falquero, S., Carfi, A.., di Paola, A., Benejam, B., Valldeneu, S., Fopma, R., Oosterik, M,. Hermelink, M., Beugelsdijk, G., Schippers, M., Henstra, H., Scholten-Kuiper, M., Willink-Vos, J., de Ruiter, L., Willems, L., Loonstra-de Jong, A., Coppus, A.M.W., Tollenaere, M., Fortea, J., Onder, G.,Rebillat, A-S., Van Dam, D., De Deyn, P.P.
'The Behavioral and Psychological Symptoms of Dementia in Down Syndrome Scale (BPSD-DS II): Optimization and further validation'.
Journal of Alzheimer's Disease, 2021, 81(4), 1505-1527. DOI 10.3233/JAD-201427
Abstract: People with Down syndrome (DS) are at high risk to develop Alzheimer's disease dementia (AD). Behavioral and psychological symptoms of dementia (BPSD) are common and may also serve as early signals for dementia. However, comprehensive evaluation scales for BPSD, adapted to DS, are lacking. Therefore, we previously developed the BPSD-DS scale to identify behavioral changes between the last six months and pre-existing life-long characteristic behavior. To optimize and further study the scale (discriminative ability and reliability) in a large representative DS study population. Optimization was based on item irrelevance and clinical experiences obtained in the initial study. Using the shortened and refined BPSD-DS II, informant interviews were conducted to evaluate 524 individuals with DS grouped according to dementia status: no dementia (DS, N = 292), questionable dementia (DS + Q, N = 119), and clinically diagnosed dementia (DS + AD, N = 113). Comparing item change scores between groups revealed prominent changes in frequency and severity for anxious, sleep-related, irritable, restless/stereotypic, apathetic, depressive, and eating/drinking behavior. For most items, the proportion of individuals displaying an increased frequency was highest in DS + AD, intermediate in DS + Q, and lowest in DS. For various items within sections about anxious, sleep-related, irritable, apathetic, and depressive behaviors, the proportion of individuals showing an increased frequency was already substantial in DS + Q, suggesting that these changes may serve as early signals of AD in DS. Reliability data were promising. The optimized scale yields largely similar results as obtained with the initial version. Systematically evaluating BPSD in DS may increase understanding of changes among caregivers and (timely) adaptation of care/treatment.

Dennehy, H., Allen AP, McGlinchey E, Buttery N, García-Domínguez L, Chansler R, Corr C, Dunne P, Kennelly S, Daly L, McCallion P, McCarron M.
A scoping review of post-diagnostic dementia supports for people with intellectual disability.

Aging & Mental Health, 2023, 27(8), 1456-1465, doi: 10.1080/13607863.2022.2130171
Abstract: People with intellectual disability, particularly people with Down syndrome, are at an increased risk for early-onset dementia, in comparison to people without an intellectual disability. The aim of this review was to scope the current landscape of post-diagnostic dementia supports for people with intellectual disability.Method: A systematic search of five electronic databases (CINAHL, Medline, PsycArticles, PsycInfo and Web of Science) was conducted for this scoping review. Results were screened independently by two reviewers, with a third reviewer for arbitration where necessary. Forty-two studies met the inclusion criteria, and relevant information was extracted. The articles included focused on the experiences of people with intellectual disability and dementia, as well as the role of carers, family members and staff. Key themes included ageing in place, environmental supports for people with intellectual disability and dementia, dementia-specific interventions and therapies, as well as the feasibility of these interventions. Besides the studies that focussed on these themes, other studies focused on staff training and family supports. This review highlights the importance of implementing timely and appropriate post-diagnostic supports for people living with intellectual disability and dementia. More controlled trials are required on post-diagnostic dementia supports for people with intellectual disability.

Devenny, D.A., Krinsky-McHale, S.J., Sersen, G., & Silverman, W.P.
Sequence of cognitive decline in dementia in adults with Down's syndrome.
Journal of Intellectual Disability Research 2000, 44(6), 654-665. doi:10.1046/j.1365-2788.2000.00305.x
Abstract: Because of lifelong intellectual deficits, it is difficult to determine the earliest signs and characteristics of age-associated decline and dementia among adults with Down syndrome. In a longitudinal study in which all participants were healthy at the time of their entry into the study, the present authors compared the amount of decline on the subtests of the WISC-R to determine the sequence of cognitive decline associated with varying stages of dementia. Twenty-two individuals with varying degrees of cognitive decline were compared to 44 adults with DS who have remained healthy. All participants functioned in the mild or moderate range of intellectual disability at initial testing. On each subtest of the WISC-R, the amount of change experienced by the healthy participants over the study period was compared to the amount of change found for each of the groups with decline. Out of the individuals who showed declines, 10 adults with DS were classified as having 'questionable' decline based on the presence of memory impairment, and five and seven adults with DS were classified as in the 'early stage' and 'middle stage' of DAT, respectively, based on the presence of memory impairment, score on the Dementia Scale for Down Syndrome and a physician's diagnosis. It was found that participants who were identified as 'questionable', in addition to the memory loss that determined their classification, also showed significant declines on the Block Design and Coding subtests. The five adults in the early stage of dementia showed declines on these subtests, and in addition, on the Object Assembly, Picture Completion, Arithmetic and Comprehension subtests. The seven adults in the middle stage of dementia showed declines on these subtests, plus declines on Information, Vocabulary and Digit Span subtests. The Picture Arrangement and Similarities subtests were not useful in distinguishing between the groups because of baseline floor effects for a substantial proportion of participants. The present longitudinal study showed a sequence of cognitive decline associated with DAT, beginning with a possible 'pre-clinical' stage, and progressing through the early and middle stages. This approach begins to define the sequence of declining cognitive capacities that contributes to the observed functional deterioration caused by Alzheimer's disease and that is likely to reflect the involvement of cortical areas as the disease progresses.

Devenny, D.A., Wegiel, J., Schupf, N., Jenkins, E., Zigman, W., Krinsky-McHale, S.J., & Silverman, W.P.
Dementia of the Alzheimer's type and accelerated aging in Down syndrome.
Science of Aging Knowledge Environment, 2005 Apr 6, 2005(14), dn1. doi: 10.1126/sageke.2005.14.dn1
Abstract: This case study, of a woman with Down syndrome and dementia of the Alzheimer's type (DAT), follows the course of her decline over an 11-year period until death at age 57. Detailed neuropathological findings are also presented. This case illustrates features of premature aging that are typically associated with Down syndrome, and the progressive changes in memory and cognition that are usually associated with DAT. Although the subject's cardiovascular condition and thyroid disorder were treated, they may have contributed to the decline of her memory. This case shows the difficulty in diagnosing dementia in an individual with [intellectual disability] who suffered comorbid episodes of depression and psychosis.

Dick, M.B., Doran, E., & Phalen, M., & Lott,I.
Cognitive profiles on the Severe Impairment Battery are similar in Alzheimer disease and Down syndrome with dementia
Alzheimer Disease and Associated Disorders, 30(3), 251-257. https://doi.org/10.1097/WAD.0000000000000132]
Abstract: Previous research has revealed similarities in the neuropathology, clinical presentation, and risk factors between persons with Alzheimer disease from the general population (GP-AD) and those with Down syndrome (DS-AD). Less is known, however, about the extent of similarities and differences in the cognitive profiles of these 2 populations. Fifty-one moderate to severely demented GP-AD and 59 DS-AD individuals participated in this study which compared the cognitive profiles of these 2 populations on the Severe Impairment Battery (SIB), controlling for sex as well as level of functional ability using a modified version of the Bristol Activities of Daily Living Scale. Overall, the neuropsychological profiles of the higher-functioning individuals within the DS-AD and advanced GP-AD groups, as represented by mean difference scores on the SIB as a whole and across the 9 separate cognitive domains, were very similar to one another after adjusting for sex and functional impairment. To our knowledge, this is the first study to directly compare the cognitive profiles of these 2 populations on the SIB. Findings suggest that the underlying dementia in GP-AD and DS-AD may have corresponding and parallel effects on cognition.

Dillane, I., & Dood, O.
Nursing people with intellectual disability and dementia experiencing pain: An integrative review.
Journal of Clinical Nursing, 2019, doi: 10.1111/jocn.14834, 28(13-14), 2472-2485
Abstract: To explore the current evidence of nurses caring for people with intellectual disability and dementia who experience pain. People with intellectual disability are aging and are experiencing age-related health conditions including dementia and conditions associated with pain, but at an earlier age. Addressing the needs of people with intellectual disability who develop dementia is a new challenge for nurses. The authors undertook an integrative literature review and a systematic search of databases: CINAHL, MEDLINE, PsycINFO, Cochrane, EMBASE, Academic Search Complete, Scopus and Web of Science between 27 October 2017-7 November 2017. Hand searching and review of secondary references were also undertaken. Quality appraisal (Crowe Critical Appraisal Tool), thematic data analysis (Braun and Clarke, Qualitative Research in Psychology, 3, 2006, 77) and reporting using the PRISMA guidelines. Seven papers met the inclusion criteria, and three themes emerged from this review: nurses knowledge of ageing, dementia and pain; recognizing pain in people with intellectual disability and dementia; and the role of nurse education. People with intellectual disability and dementia have difficulty communicating their pain experience compounded by pre-existing communication difficulties. A pain experience can present similar to behavioral and psychological symptoms of dementia, and diagnostic overshadowing often occurs whereby a pain need is misinterpreted as behavioral and psychological symptoms resulting in inappropriate treatment. Nurses need greater knowledge about the presence of pain and potential causes in people with intellectual disability and dementia, and education can be effective in addressing this knowledge deficit. Pain assessment tools for people with intellectual disability and dementia need to include behavioural elements, and baseline assessments are required to identify changes in presentation. Nurses need to recognize and respond to pain based on the evidence in order to deliver quality care.

Dillenburger, K., & McKerr,L.
'How long are we able to go on?' Issues faced by older family caregivers of adults with disabilities
British Journal of Learning Disabilities, 2011 March, 39(1), 29-38. https://doi.org/10.1111/j.1468-3156.2010.00613.x
Abstract: Research-informed policy and practice is needed for older caregivers of adult sons/daughters with disabilities. These caregivers are often under tremendous stress because of failing health, financial pressures, bereavement and worry about the future of their sons/daughters. Twenty-nine older parents/caregivers of 27 adults with intellectual and/or developmental disabilities were interviewed to explore their views and experiences regarding long-term care

and service arrangements, health and psychological needs and 'future planning'. Findings show a severe lack of support, respite care and future planning which causes high stress levels for caregivers. Policy makers and researchers working in this field need to take into consideration the needs of older caregivers when making future plans for adults with disabilities.

Dodd, K.
Commentary on "Estimating the number of people with Down's syndrome in Scotland and the cohort at elevated risk of early onset dementia"
Tizard Learning Disability Review, 2017, 22(3), 172-176.
10.1108/TLDR-04-2017-0019
Abstract: The purpose of this paper is to consider the implications for people with Down's syndrome and their families of identifying those people who are at risk of developing dementia from the research study "Estimating the number of people with Down's syndrome in Scotland and the cohort at elevated risk of early onset dementia". The commentary is based on a review of the associated literature. Estimating the numbers is important but has serious implications for people who have an elevated risk and their families. Preparation and ongoing support and planning are vital to ensure that quality of life is maintained as dementia is identified and progresses.

Dodd, K.
Supporting people with Down's syndrome and dementia
Tizard Learning Disability Review, 2003, 8(4), 14-18.
https://www.emerald.com/insight/publication/issn/1359-5474/vol/8/iss/4
Abstract: Brief review of literature and concepts dealing with the prevalence of dementia among people with Down syndrome in England, ethical issues in assessment and diagnosis, the value of early diagnosis, and an explication of service options and management strategies. Review concludes with a prognosis for services in the future.

Dodd, K., Bush, A., & Livesey, A.
Developing and piloting the QOMID - quality outcome measure for individuals with intellectual disabilities and dementia
Advances in Mental Health and Intellectual Disabilities, 2015, 9(6), 298-311.
https://doi.org/10.1108/AMHID-12-2014-0041
There are no papers on specific overall quality outcome measures for people with intellectual disabilities who have dementia. The purpose of this paper is to describe the development and piloting of a new measure. A process was developed to measure quality outcomes across all stages of dementia. The reliability of the tool was measured using Cronbach's α coefficients, along with data about its clinical utility. It was found that the QOMID has good reliability, face validity and internal reliability suggesting that all domains contribute equally towards the construct of quality outcome. An exploratory factor analysis revealed that there may be four or five sub-factors within the QOMID, The clinical utility of the assessment tool was explored and it can be concluded that the QOMID is simple, fairly quick and effective. The scale has good psychometric properties and the initial parameters for the QOMID were met. Further exploration of factors needs to be considered with a larger sample of participants. The scale was liked by assessors and gives a practical tool that can both measure the quality outcome for people at each stage of their dementia, and help to develop more effective dementia care plans.

Dodd, K., Watchman, K., Janicki, M.P., Coppus, A., Gaertner, C., Fortea, J., Santos, F. H., Keller, S.M., & Strydom. A.
Consensus Statement of the International Summit on Intellectual Disability and Dementia related to post-diagnostic support
Aging & Mental Health, 2018, 22(11), 1406-1415. doi:
10.1080/13607863.2017.1373065
Abstract: Post diagnostic support (PDS) has varied definitions within mainstream dementia services and different health and social care organizations, encompassing a range of supports that are offered to adults once diagnosed with dementia until death. An international summit on intellectual disability and dementia held in Glasgow, Scotland in 2016 identified how PDS applies to adults with an intellectual disability and dementia. The Summit proposed a model that encompassed seven focal areas: post-diagnostic counseling; psychological and medical surveillance; periodic reviews and adjustments to the dementia care plan; early identification of behavior and psychological symptoms; reviews of care practices and supports for advanced dementia and end of life; supports to carers/ support staff; and evaluation of quality of life. It also explored current practices in providing PDS in intellectual disability services. The Summit concluded that although there is limited research evidence for pharmacological or non-pharmacological interventions for people with intellectual disability and dementia, viable resources and guidelines describe practical approaches drawn from clinical practice. Post diagnostic support is essential, and the model components in place for the general population, and proposed here for use within the intellectual disability field, need to be individualized and adapted to the person's needs as dementia progresses. Recommendations for future research include examining the prevalence and nature of behavioral and psychological symptoms (BPSD) in adults with an intellectual disability who develop dementia, the effectiveness of different non-pharmacological interventions, the interaction between pharmacological and non-pharmacological interventions, and the utility of different models of support.

Donaldson S.
Work stress and people with Down syndrome and dementia.
Down's Syndrome, Research and Practice, 2002, 8(2), 74-78. doi:
10.3104/reports.133.
Abstract: Author assessed how staff ratings of challenging behavior for people with Down syndrome and dementia affected the self-reported well-being of care staff. Data were collected from 60 care staff in 5 day centers in a large city in England. The data were collected by use of a questionnaire. There was no significant difference between those who cared for individuals with Down syndrome and dementia and those caring for service users with other non-specified learning disabilities without dementia, regarding their self-reported well-being. Self-reported well-being did correlate with staff rating of challenging behavior in both those who cared for people with Down syndrome and dementia and those who did not care for such service users, with well-being declining as perceived challenging behavior increased. The findings indicate that challenging behavior prevention and reduction may be of benefit to both service users and care staff well-being.

Donaway, A.R.
Dementia of the Alzheimer's type in adults with Down syndrome: knowledge among graduate students in physical therapy, occupational therapy, and speech-language pathology.
Electronic Theses and Dissertations. Paper 4290. (2024).
https://doi.org/10.18297/etd/4290
Abstract: This non-experimental study utilized a convenience sample (N = 322) to investigate graduate students in physical therapy (PT), occupational therapy (OT), and speech-language pathology' s (SLP) knowledge of the connection between Dwn syndrome and dementia of the Alzheimer's type (DAT). Ninety-two-point-five percent of participants were female, and 79.80% were white. All participants completed a survey comprised of items relating to information about general knowledge (i.e. facts) of the association between DS and DAT, the signs/symptoms that occur in each stage of dementia of the Alzheimer's type for people with Down syndrome, and familiarity regarding the diagnosis and care. Descriptive and summary statistics indicated the persistence of misconceptions regarding the connection between DS and DAT among graduate students in PT, OT, and SLP training programs. Group comparisons were conducted to identify differences according to discipline (PT, OT, or SLP) and the relationship of knowing individuals diagnosed with DAT, DS, neither, or both. ANOVA analyses revealed knowledge of the stages of DAT as they relate to DS appeared stronger for OT students when compared to their PT peers; no statistically significant difference was noted per SLP students. Kruskall-Wallis and ANOVA analyses revealed no statistically significant difference in knowledge across disciplines regarding general knowledge about DS and DAT and familiarity of diagnosis and care parameters, respectively. Spearman's rank correlation revealed a statistically significant result among the respondents' connection to an individual with both DS and DAT and increased scores per accuracy of facts and familiarity (diagnosis and care) when compared to knowing someone with only DS or DAT.

Down, J.L.H
Observations on an ethnic classification of idiots
London Hospital Reports, 1866, 3, 1866, 259-262.
https://www.romolocapuano.com/wp-content/uploads/2013/07/Langdon-Down-1866.pdf
Abstract: [None provided; text extract]. A very large number of congenital idiots are typical Mongols. So marked is this, that when placed side by side, it

is difficult to believe that the specimens compared are not children of the same parents.The number of idiots who arrange themselves around the Mongolian type is so great,and they present such a close resemblance to one another in mental power, that I shall describe an idiot member of this racial division, selected from the large number that have fallen under my observation. The hair is not black, as in the real Mongol, but of a brownish colour, straight and scanty. The face is flat and broad, and destitute of prominence. The cheeks are roundish, and extended laterally. The eyes are obliquely placed, and the internal canthi more than normally distant from one another. The palpebral fissure is very narrow. The forehead is wrinkled transversely from the constant assistance which the levatores palpebrarum derive from the occipito-frontalis muscle in the opening of the eyes. The lips are large and thick with transverse fissures. The tongue is long, thick, and is much roughened. The nose is small. The skin has a slight dirty yellowish tinge, and is deficient in elasticity, giving the appearance of being too large for the body. The improvement which training effects in them is greatly in excess of what would be predicted if one did not know the characteristics of the type. The life expectancy, however, is far below the average

Duncan, C., Wilkinson, E., Jaydeokar, S., & Acton, D.J.
Assessing adherence to National Institute for Health and Care Excellence dementia assessment and diagnosis guidelines in adults with intellectual disability: a retrospective cohort study.
Advances in Mental Health and Intellectual Disabilities, 2023, 1 November 2023, Vol. No. ahead-of-print. https://doi.org/10.1108/AMHID-07-2023-0022.
Abstract: This study evaluated the dementia assessment and diagnosis care provided to adults with intellectual disability. The authors selected recommendations from the National Institute for Health and Care Excellence (NICE) standards which could be evidenced in clinical notes and aimed to identify characteristics which may be associated with improved adherence to these recommendations. The study population was adults with an intellectual disability who were diagnosed with dementia between January 2019 and December 2022 by a UK-based intellectual disability service. Data to demonstrate adherence to selected recommendations and demographic and clinical characteristics were extracted from electronic patient records. The authors identified 41 individuals. A mean of six of the eight recommendations were adhered to. There was low adherence with structural imaging to support dementia subtype diagnosis (9 individuals, 22%). This may be linked with the low percentage of people diagnosed with vascular dementia (1 individual, 2%) despite a national figure of 20%. No demographic or clinical characteristics were associated with level of adherence recorded. The authors found incomplete recording of diagnostic clinical coding in electronic patient records. This may disadvantage this population, as they cannot be readily identified for post diagnostic support or resource allocation. To the best of the authors' knowledge, this is the first study to examine adherence to these NICE guidelines in this population.

Dunne, P., Reilly, E., Judge, R., Lowe, F. & McCarron, M.
Giving meaning to life - the role of digital life stories in supporting people with intellectual disability and dementia.. World Congress of the International Association for the Scientific Study of Intellectual and Developmental Disabilities, Glasgow, Scotland, 6th - 9th August, 2019
Journal of Intellectual Disability Research, 2019, 63(8), 643.
Abstract: Over a four-year period individuals with an intellectual disability (ID) in a large service provider in the Republic of Ireland were supported to create their personalized digital life story using selected multi media apps. An easy read survey was distributed to 380 people with ID to gauge their readiness and interest in engaging in digital life story. A bespoke training course was developed to support the introduction of digital life story activities. The use of the personalized digital life stories by the individuals and their support staff, family and social network was captured through recorded observations and individual use was tracked through a developed audit tool. Over the four-year period eighty people with an ID commenced their digital life story. Case studies within this cohort showed a variety of use which varied depending on degree of ID and stage of dementia. In all cases life stories were instrumental to enhanced communication and social interactions. Key factors in uptake and sustainability were staff training, iPad clubs and champions across the organization. Digital life stories were key in supporting meaningful interactions across the continuum of dementia both in day to day interactions with family, staff, volunteers, and peers and as a tool for social engagement through digital life story clubs.

Dykens, E.
Psychiatric and behavioral disorders in persons with Down syndrome
Mental Retardation and Developmental Disabilities Research Reviews, 2007, 13(3), 272-278. https://onlinelibrary.wiley.com/doi/10.1002/mrdd.20159
Abstract: Similar to the state of the broader intellectual disabilities field, many gaps exist in the research and treatment of mental health concerns in people with Down syndrome. This review summarizes key findings on the type and prevalence of behavior and emotional problems in children, adolescents, and adults with Down syndrome. Such findings include relatively low rates of severe problems in children, and well-documented risks of depression and Alzheimer's disease in older adults. The review also considers emerging data on autism, and the paucity of studies on adolescents. Three next steps for research are highlighted, including a need to: (1) connect research on psychiatric status and diagnoses across developmental periods, including adolescence, and to examine such associated processes as sociability, anxiety and attention; (2) unravel complicated biopsycho-social risk and protective factors that serve to increase or diminish psychopathology; and (3) identify evidence-based treatments that both reduce distressful symptoms and enhance well-being in individuals with Down syndrome.

Eady, N., Sheehan, R., Rantell, K., Sinai, A., Bernal, J., Bohnen, I., Courtney, K., Dodd, K., Gazizova, D., Hassiotis, A., ... Strydom, A.
Impact of cholinesterase inhibitors or memantine on survival in adults with Down syndrome and demenita: Clincal cohort study.
British Journal of Psychiatry, 2018 Mar, 212(3), 155-160. doi:10.1192/bjp.2017.21
Abstract: There is little evidence to guide pharmacological treatment in adults with Down syndrome and Alzheimer's disease. Authors investigated the effect of cholinesterase inhibitors or memantine on survival and function in adults with Down syndrome and Alzheimer's disease. This was a naturalistic longitudinal follow-up of a clinical cohort of 310 people with Down syndrome diagnosed with Alzheimer's disease collected from specialist community services in England. Median survival time (5.59 years, 95% CI 4.67-6.67) for those on medication (n = 145, mainly cholinesterase inhibitors) was significantly greater than for those not prescribed medication (n = 165) (3.45 years, 95% CI 2.91-4.13, log-rank test P<0.001). Sequential assessments demonstrated an early effect in maintaining cognitive function. Cholinesterase inhibitors appear to offer benefit for people with Down syndrome and Alzheimer's disease that is comparable with sporadic Alzheimer's disease; a trial to test the effect of earlier treatment (prodromal Alzheimer's disease) in Down syndrome may be indicated.

Ebbing, K., von Gunten, A., Guinchat, V., Georgescu, D., Bersier, T., Moad, D., & Verloo, H.
Barriers facing direct support professionals when supporting older adults presenting with intellectual disabilities and unusual dementia-related behavior: A multi-site, multi-methods study.
Disabilities, 2022, 2(4), 662-680. https://doi.org/10.3390/disabilities2040047
Abstract: Increased life expectancy among people with intellectual disabilities (ID) raises the risk of their diagnosis being superimposed by behavioral and psychological symptoms of dementia (BPSD). The difficulties facing direct support professionals dealing with this is an emerging, under-investigated issue. The study investigates direct support professionals' perceptions and experiences of their daily support for aging people with ID presenting with superimposed BPSD. Twenty-four direct support professionals from long-term care facilities responded to clinical vignettes and attended focus groups conducted to investigate perceptions and lived experiences of the barriers and struggles they faced. Direct support professionals' reactions to vignettes revealed their difficulties recognizing BPSD superimposed on the known challenging behaviors of people with ID. Focus groups highlighted daily struggles with BPSD, the lack of knowledge about detecting and dealing with them, and associated somatic and psychopathological diseases of aging. Improved knowledge transfer about good practices for person-centered support to aging people with ID presenting with BPSD is strongly recommended.

Egan, C., & Dalton, C.T.
An exploration of care-burden experienced by older caregivers of adults with intellectual disabilities in Ireland
British Journal of Learning Disabilities, 2019, 47(3),188-194. https://doi.org/10.1111/bld.12273C
Abstract: People with intellectual disabilities are experiencing increased longevity, and in parallel, their family caregivers are also ageing. The literature identifies that these caregivers are at risk of burden. The aim of this study was to

measure the level of caregiver burden among older carers of adults with intellectual disabilities in an Irish sample and to analyze the effect of socio-demographic factors upon experiences of caregiver burden. Methods Thirty caregivers completed a survey questionnaire. Data were collected based upon participants' self-reports of burden using the Zarit Burden Interview (ZBI) and a socio-demographic questionnaire. Data were analyzed using SPSS version 24. Over 57% of carers indicated a mild-to-moderate level of burden. Analysis indicated that younger caregivers experience significantly higher levels of burden, when compared to older caregivers. This study contributes to our understanding of burden among an Irish population of older caregivers supporting an adult with an intellectual disability. It identified that carers do experience burden. The importance of proactive assessments and supports for these caregivers was revealed. This study highlights a lack of Irish research in this area and may pave the way for future research which could build upon its findings.

Eisner, D.A.
Down's syndrome and aging: Is senile dementia inevitable?
Psychological Reports 1983, 52(1), 119-124.
https://doi.org/10.2466/pr0.1983.52.1.119
Abstract: Numerous studies have reported that in elderly Down syndrome individuals there is a high preponderance of senile dementia. An examination of these investigations shows that, while there is accelerated neurological aging, there is not a high incidence of behavioral or overt senile dementia. Changes in cognitive functioning for Down syndrome persons are similar to those found in non-Down "retarded" populations.

Elliott-King, J., Shaw, S., Bandelow, S., Devshi, R., Kassam, S., & Hogervorst, E.
A critical literature review of the effectiveness of various instruments in the diagnosis of dementia in adults with intellectual disabilities
Alzheimers and Dementia (Amst), 2016,Jun30, 4(1), 126-148. doi: 10.1016/j.dadm.2016.06.002. eCollection 2016.
Abstract: Currently, there is no consensus on dementia diagnostics in adults with intellectual disabilities (ID). There are three types of assessments available: direct cognitive tests, test batteries, and informant reports. A systematic literature search was conducted in four databases yielding 9840 records. Relevant studies were identified and selected using predefined inclusion and exclusion criteria and then coded and classified according to assessment type. This was completed by two independent researchers, with a third consulted when discrepancies arose. The review collates diagnostic instruments and presents strengths and weaknesses. Overall 47 studies met the search criteria, and 43 instruments were extracted from the selected studies. Of which, 10 instruments were classified as test batteries, 23 were classified as direct cognitive tests, and the remaining 10 were informant reports. This review can recommend that cognitive test batteries can offer the most practical and efficient method for dementia diagnosis in individuals with ID.

Engelborghs S.
Adults with cerebral palsy and Alzheimer disease: a missing link?
Dev Med Child Neurol. 2022 Mar;64(3):284. doi: 10.1111/dmcn.15080.
Abstract: Although the brain injury causing CP is non-progressive, prevalence of comorbidities that are usually age-related is already elevated early in adulthood among individuals with CP, with a more pronounced increase by the age of 50 years.2 Examples of such comorbidities are arterial hypertension, diabetes, frailty, sarcopenia, osteoporosis, osteoarthritis, cardiorespiratory diseases, and cancer. Core symptoms of dementia are cognitive deficits, behavioural and personality changes, and functional disabilities. Due to a loss of autonomy, people with dementia often must rely on carers for their activities of daily living. Since people with CP have a normal life expectancy, dementia should be as prevalent as in age-matched non-CP individuals. However, as CP seems to predispose to age-related disorders at a younger age, individuals with CP might also have a higher risk of dementia, which remained an unanswered question so far. Therefore, the study by Mahmoudi et al. is interesting and relevant. In a large population of 5176 adults with CP (vs 1 119 131 adults without CP as a control group), the authors have demonstrated that adults with CP have a significantly higher risk of both early- and late-onset dementia as compared to controls. This study triggers many questions for future research, which makes it a valuable addition to existing literature. In the absence of biomarker-based diagnoses, we can only speculate whether CP selectively increases the likelihood to develop one specific type of dementia like Alzheimer disease. If this is the case, an interaction between the pathophysiology of CP and Alzheimer disease would be suspected, and neuroinflammation might be a possible link between both conditions. Comorbidities among individuals with CP might also attenuate the risk for dementia, as was recently suggested. On the other hand, CP might affect an individual's cognitive reserve, making them more vulnerable to develop symptoms of dementia at a younger age as compared to controls.

Engdahl, J.M.K.
Alzheimer's disease & Down syndrome: A practical guide for caregivers.
36 pp.
Bozeman, Montana: Author [723 South 13th Street, Bozeman, MT 59715] (1995)
Abstract: Training manual developed to provide primary information about care practices for parents and other primary carers of adults with Down syndrome affected by Alzheimer's disease. Covers, in brief format, recognizing signs and symptoms, diagnostic advice, care management practice (communication, dealing with problem behaviors, helping with activities of daily living, promoting alternative activities) and help for carers.

Englund, A., Jonsson, B., Zander, C.S., Gustafsson, J,. & Annerén, G.
Changes in mortality and causes of death in the Swedish Down syndrome population
American Journal of Medical Genetics [Part A], 2013 Apr, 161A(4), 642-649, doi: 10.1002/ajmg.a.35706.
Abstract: During the past few decades age at death for individuals with Down syndrome (DS) has increased dramatically. The birth frequency of infants with DS has long been constant in Sweden. Thus, the prevalence of DS in the population is increasing. The aim of the present study was to analyze mortality and causes of death in individuals with DS during the period 1969-2003. All individuals with DS that died between 1969 and 2003 in Sweden, and all individuals born with DS in Sweden between 1974 and 2003 were included. Data were obtained from the Swedish Medical Birth Register, the Swedish Birth Defects Register, and the National Cause of Death Register. Median age at death has increased by 1.8 years per year. The main cause of death was pneumonia. Death from congenital heart defects decreased. Death from atherosclerosis was rare but more frequent than reported previously. Dementia was not reported in any subjects with DS before 40 years of age, but was a main or contributing cause of death in 30% of the older subjects. Except for childhood leukemia, cancer as a cause of death was rare in all age groups. Mortality in DS, particularly infant mortality, has decreased markedly during the past decades. Median age at death is increasing and is now almost 60 years. Death from cancer is rare in DS, but death from dementia is common.

ENIDA
Face to face: Respectful coping with dementia in older people with intellectual disability
52 minutes
Working Group on Coping with Dementia in Older People with Intellectual Disability, European Network on Intellectual Disability and Ageing [ENIDA - c/o Patricia Noonan Walsh, Ph.D., Director, Centre for the Study of Developmental Disabilities, University College Dublin, Belfield, Dublin 4, IRELAND – e-mail: patricia.walsh@ucd.ie] (2000)
Abstract: A 52-minute video with an accompanying information booklet, which uses a number of case vignettes from France, Belgium and the Netherlands to illustrate the various symptoms and stages of dementia among older people with intellectual disability. Examples of practices to promote "respectful coping" with dementia, death and dying on the part of direct support professionals and clinicians are presented. Devised for staff training and development, Face to Face may be viewed in short segments. A version with English subtitles and English booklet is available in formats suitable for Europe and for North America. Developed with funding and support from: ENIDA, Fondation de France, the European Union, and University College Dublin, Ireland.

Ericksson, M., & Sundin, M.
Developing early detection of dementia with people with intellectual and developmental disabilities
Poster presented at the 27th Annual Conference of Alzheimer Europe, Berlin, Germany, October 3, 2017. (PO3.26).
https://www.alzheimer-europe.org/conferences/past-conferences/2017-berlin?language_content_entity=en

Abstract: In Finland there is a lack of a unified approach to the early detection of dementia for people with intellectual and developmental disabilities (IDD). The aim of this presentation is to describe a currently underway collaboration project, which responds to this challenge. In our opinion, there are two essential elements in early detection of dementia with people with IDD: 1) an overall description of the psychosocial functioning of the person and 2) a screening method for dementia, which developed for persons with IDD. These two components are shortly outlined. Psychosocial functioning: Early detection of neurocognitive disorders in people with IDD is multidisciplinary teamwork. The Finnish Association of Intellectual and Developmental Disabilities (FAIDD) has published two methods for this purpose (the Toimi and the Psyto), and we recommend using these methods in the assessment of dementia in people with IDD as well. For example, it is important to distinguish the symptoms of dementia from other possible psychological disorders (like mood disorders and psychotic symptoms). Translation of the National Task Group Early Detection Screen for Dementia (NTG-EDSD): Commonly used assessment methods (like the Cerad and the Mini-Mental) may not be applicable for people with IDD. In our project, we decided to translate the NTG-EDSD into Finnish. The EDSD is already available online in many other languages (www.the-ntg.org/screening). As the authors write, the NTG-EDSD is not an assessment or diagnostic instrument, but an administrative screen that can be used by people who know the client well. The Finnish version of the NTG-EDSD is being introduced into practice in 2017. FAIDD trains staff and other people working with people with IDD in using the screen within the perspective of psychosocial functioning.

Esbensen, A.J.
Health conditions associated with aging and end of life of adults with Down syndrome.
International Review of Research in Mental Retardation, 2010, 39c, 107-126. https://www.ncbi.nlm.nih.gov/pmc/articles/PMC3010180/
Abstract: Expectations for the life course of individuals with Down syndrome (DS) have changed, with life expectancy estimates increasing from 12 in 1949 to nearly 60 years of age today. Along with this longer life expectancy comes a larger population of adults with DS who display premature age-related changes in their health. There is thus a need to provide specialized health care to this aging population of adults with DS who are at high risk for some conditions and at lower risk for others. This review focuses on the rates and contributing factors to medical conditions that are common in adults with DS or that show changes with age. The review of medical conditions includes the increased risk for skin and hair changes, early onset menopause, visual and hearing impairments, adult onset seizure disorder, thyroid dysfunction, diabetes, obesity, sleep apnea and musculoskeletal problems. The different pattern of conditions associated with the mortality of adults with DS is also reviewed.

Esbensen, A.J., Boshkoff Johnson, E., Amaral, J.L., Tan, C.M., & Macks, R.
Differentiating aging among adults with Down syndrome and comorbid psychopathology.
American Journal on Intellectual and Developmental Disabilities, 2016, 121 (1), 13-24. doi: 10.1352/1944-7558-121.1.13.
Abstract: Differences were examined between three groups of adults with Down syndrome in their behavioral presentation, social life/activities, health, and support needs. We compared those with comorbid dementia, with comorbid psychopathology, and with no comorbid conditions. Adults with comorbid dementia were more likely to be older, have lower functional abilities, have worse health and more health conditions, and need more support in self-care. Adults with comorbid psychopathology were more likely to exhibit more behavior problems and to be living at home with their families. Adults with no comorbidities were most likely to be involved in community employment. Differences in behavioral presentation can help facilitate clinical diagnoses in aging in Down syndrome, and implications for differential diagnosis and service supports are discussed.

Esbensen, A.J., Mailick, M.R., & Silverman, W.
Long-term Impact of parental well-being on adult outcomes and dementia status in individuals with Down syndrome.
American Journal on Intellectual and Developmental Disabilities, 2013, 118(4), 294-309. doi: 10.1352/1944-7558-118.4.294.
Abstract: Parental characteristics were significant predictors of health, functional abilities, and behavior problems in adults with Down syndrome (n = 75) over a 22-year time span, controlling for initial levels and earlier changes in these outcomes. Lower levels of behavior problems were predicted by improvements in maternal depressive symptoms. Higher levels of functional abilities were predicted by prior measures of and improvements in maternal depressive symptoms. Better health was predicted by prior measures of maternal depressive symptoms, paternal positive psychological well-being, relationship quality between fathers and their adult children, and improvements in maternal positive psychological well-being. Dementia status was also predicted by parental characteristics. The study suggests the importance of the family context for healthy aging in adults with Down syndrome.

Esralew, L., Janicki, M.P., & Keller, S.M.
National Task Group Early Detection Screen for Dementia (NTG-EDSD) Chapter in V. Prasher (ed.), *Neuropsychological Assessments of Dementia in Down Syndrome and Intellectual Disabilities*. pp. 197-213. 2018. doi:10.1007/978-3-319-61720-6-11
Abstract: Early identification of signs and symptoms of cognitive and functional decline associated with dementia is an important first step in managing the course of dementia and providing quality care. Studies show that persons with intellectual disabilities are as susceptible to the causes of dementia as are other adults, with the relative distribution of etiologies mirroring those of other adults. The challenge, as noted by the World Health Organization, is to identity early those adults susceptible and affected so that assessment and diagnostic work-ups can be undertaken. A screening tool can be used to substantiate changes in adaptive skills, behavior and cognition. With early detection, assessment and diagnosis can be carried out to determine whether cognitive changes are the result of a neuropathological process related to disease or trauma to the brain, or attributable to other causes, which may be treatable and reversible. The National Task Group Early Detection Screen for Dementia (NTG-EDSD) is an informant-based rating tool for use with adults with intellectual and developmental disabilities who are suspected of experiencing changes in thinking, behavior, and adaptive skills suggestive of mild cognitive impairment or dementia. This chapter offers background on the NTG-EDSD and describes benefits that may accrue from using a dementia screening instrument.

Esteba-Castillo S, Dalmau-Bueno A, Ribas-Vidal N, Vilà-Alsina M, Novell-Alsina R, García-Alba J.
Adaptacion y validacion del Cambridge Examination for Mental Disorders of Older People with Down's Syndrome and Others with Intellectual Disabilities (CAMDEX-DS) en poblacion española con discapacidad intelectual [Adaptation and validation of CAMDEX-DS (Cambridge Examination for Mental Disorders of Older People with Down's Syndrome and Others with Intellectual Disabilities) in Spanish population with intellectual disabilities].
Revista de Neurologia, 2013 Oct 16, 57(8), 337-346. [In Spanish]. PMID: 24081888.
Abstract: Dementia caused by Alzheimer's disease commonly affects the adult population with Down's syndrome. This population presents two characteristic clinical features: a semiologic pattern that differs from the typical Alzheimer's disease, and previous intellectual deficits that may confound the clinical diagnosis. There is a clear need to validate specific instruments adapted to Spanish population. To adapt and to validate CAMDEX-DS (Cambridge Examination for Mental Disorders of Older People with Down's Syndrome and Others with Intellectual Disabilities) in Spanish population, 146 adults with intellectual disability (mild to moderate) were recruited and assessed with CAMDEX-DS, K-BIT I and DMR tests. Test-retest reliability, inter-rater concordance and validity statistic were performed between CAMDEX-DS and clinical diagnosis. This is an observational, multicenter, cross-sectional and validation study. Test-retest and inter-rater reliability achieved kappa coefficient values of 0.92 and 0.91, respectively. Agreement (kappa index) for CAMDEX-DS on clinical diagnosis compared to other clinical criteria was high: CAMDEX-DS vs DSM-IV (kappa = 0.95; p < 0,001); CAMDEX-DS vs ICD-10 (kappa = 0.97; p < 0.001). All item-test correlations ranged between 0,31 and 0,69. Internal reliability-calculated using Chronbach's alpha scored 0.93. The Spanish version of CAMDEX-DS is a valid instrument with high applicability for people with intellectual disability. It shows good psychometric properties. The Cambridge Cognitive Examination for Older Adults with Down's Syndrome (CAMCOG-DS) can set two key points by the level of intellectual disability on the suspicion of cognitive impairment in people with Down's syndrome.

Evans, E., & Trollor, J.N.
Dementia in people with Intellectual Disability: Guidelines for Australian GPs.

University of New South Wales, Department of Developmental Disability Neuropsychiatry, 2018 (*webpage)*
https://www.3dn.unsw.edu.au/resources/professionals/health-mental-health-professionals/dementia-people-intellectual-disability-guidelines-australian-gps
Abstract: (none). Summary of key recommendations: (1) GPs need to be aware that people with intellectual disability (ID) are at increased risk of dementia; (2) Alzheimer's disease in particular is very common in Down syndrome, is often of early onset, and typically begins with changes in personality and executive function. People with Down syndrome should receive a comprehensive baseline `healthy' assessment at around 30 years of age, and again at 40. From 30 onwards, questions regarding signs of decline should be incorporated into annual health checks; (3) In people with other forms of intellectual disability (ID), average dementia onset is approximately 10 years prior to that experienced by the general population. A practical approach would be to screen for evidence of decline at around the age of 40, by asking questions about decline and using a carer-report checklist. This checklist should be repeated at the age of 50 and each year thereafter. Those with signs of potential decline should receive a comprehensive assessment; (4) After performing a standard dementia work-up, refer a person with suspected cognitive declines to an experienced psychologist or psychiatrist for a full cognitive assessment; (5) Diagnosing dementia in people with ID requires establishing longitudinal declines in function across at least 3 sequential assessments. Standard tests used with the general population are unsuitable for this group; (6) A number of screening checklists are available. The US National Task Group on Intellectual Disabilities and Dementia Practices Early Detection Screen for Dementia (NTG-EDSD) is free to download and can be used qualitatively to examine declines' (7) Important principles for managing dementia in patients with ID include: (a) Be equipped to manage mental disorders in people with ID. Recommended adjustments to practice can be found here https://3dn.unsw.edu.au/the-guide (b) Screen and examine for other potential causes of cognitive decline; (c) Use principles of dementia care applicable to people without ID, including communicating the diagnosis to the patient as early as possible in a manner they understand; seeking their preferences for care; coordinating services across relevant sectors (which may include the disability sector); and, where applicable, encouraging family carers to access emotional support and to make use of respite services.

Evans, E., Bhardwaj, A., Brodaty, H., Sachdev, P., Draper, B., & Trollor, J.N.
Dementia in people with intellectual disability: insights and challenges in epidemiological research with an at-risk population
International Review of Psychiatry, 2013, 25(6), 755-763. doi: 10.3109/09540261.2013.866938.
Abstract: The population with intellectual disability (ID) is aging, but age-related health concerns such as dementia have received little research attention thus far. We review evidence regarding the prevalence and incidence of dementia in people with ID, and discuss some possible explanations for an increased risk, such as shared genetic risk factors, co-morbid physical and mental disorders, lifestyle factors, trauma, and lowered brain reserve. We discuss practical and theoretical challenges facing researchers in this field, before highlighting the implications of findings to date for future research and clinical care. Research on dementia in this at-risk population has the potential to help us understand dementia in general and to improve services for this group of vulnerable individuals.

Evenhuis, H.M.
The natural history of dementia in Down's syndrome.
Archives of Neurology, 1990, Mar, 47(3), 263-267. doi:10.1001/archneur.1990.00530030029011.
Abstract: In a prospective longitudinal study with death as the end point in 17 middle-aged patients with Down's syndrome, dementia was clinically diagnosed in 15 patients, by means of careful observations in daily circumstances. Autopsies were performed in 10 cases: 8 demented patients and 2 nondemented patients. Neuropathologically, Alzheimer-type abnormalities were demonstrated in 9 patients, both demented and nondemented, and combined Alzheimer-type abnormalities with infarctions were demonstrated in 1 patient. In the 14 demented patients who did not show evidence of cerebrovascular or systemic vascular disease, dementia had an early onset and was rapidly progressive (mean age at onset, 51.3 years in the moderately retarded patients and 52.6 years in the severely retarded patients; mean duration of symptoms, respectively, 4.9 and 5.2 years). Cognitive and behavioral decline corresponded to symptoms of dementia of the Alzheimer's type in patients without Down's syndrome, but often were not recognized early. In the present group of patients, there was a remarkably high incidence of gait and speech deterioration. Also, the incidence of epileptic seizures and myoclonus was about eightfold, as compared with dementia of the Alzheimer's type in patients without Down's syndrome.

Evenhuis, H.M.
The Dementia Questionnaire for People with Learning Disabilities.
Chapter in V. Prasher (ed.), *Neuropsychological Assessments of Dementia in Down Syndrome and Intellectual Disabilities*. pp. 43-56. 2018. doi: 10.1007/978-3-319-61720-6
Abstract: To facilitate the diagnosis of dementia in persons with intellectual disabilities (ID), based on observations of caregivers, since 1980 the Dementie Vragenlijst voor Zwakzinnigen (DVZ) has been developed by Heleen Evenhuis, ID physician, and Margeen Kengen and Harry Eurlings, behavioral therapists, all working in De Bruggen center for people with ID, Zwammerdam, the Netherlands. The Dementia Questionnaire for People with Learning Disabilities (DLD) is an English translation of this instrument. Formally known as the Dementia Questionnaire for Mentally Retarded Persons (DMR). After many years of distribution through De Bruggen, its publication has now been taken over by Harcourt Test Publishers]. In this chapter, the author reviews the development of the DMR (DLD) along with its clinical applications.

Evenhuis, H.M., Hermans, H., Hilgenkamp, T.I.M., Bastiaanse, L.P., & Echteld, M.A.
Frailty and disability in older adults with intellectual disabilities: results from the healthy ageing and intellectual disability study
Journal of the American Geriatrics Society, 2012, May, 60(5), 934-938. doi: 10.1111/j.1532-5415.2012.03925.x.
Abstract: To obtain first insight into prevalence and correlates of frailty in older people with intellectual disability (ID). This was a population-based cross-sectional study in persons using formal ID services. Three Dutch care provider services, with 848 individuals with borderline to profound ID aged 50 and older participated in the Healthy Ageing and Intellectual Disability (HA-ID) Study. All participants underwent an extensive health examination. Frailty was diagnosed according to Cardiovascular Health Study criteria. Associations between frailty and participant characteristics were investigated using multivariate logistic regression analysis. Prevalence of frailty was 11% at age 50 to 64 and 18% at age 65 and older. Age, Down syndrome, dementia, motor disability, and severe ID were significantly associated with frailty, but only motor disability had a unique association with frailty. In a regression model with these variables, 25% of the variance of frailty was explained. At age 50 to 64, prevalence of frailty is as high as in the general population aged 65 and older (7-9%), with a further increase after the age of 65. Motor disability only partially explains frailty. Future studies should address health outcomes, causes, and prevention of frailty in this population.

Fahey-McCarthy, E., McCarron, M., Connaire, K., & McCallion, P.
Developing an education intervention for staff supporting persons with an intellectual disability and advanced dementia.
Journal of Policy and Practice in Intellectual Disabilities, 2009, 6(4), 267-275. https://doi.org/10.1111/j.1741-1130.2009.00231.x
Abstract: Generally, staff working in settings that provide care for adults with intellectual disabilities (ID) have not received specific education with respect to extended care for terminal illnesses or late-stage dementia. Equally, staff working in specialist palliative care often are not familiar with the unique issues of supporting persons with intellectual disabilities affected by dementia. To understand care concerns with respect to supporting persons with ID and advanced dementia, and to develop, deliver, and evaluate an educational intervention with staff in ID settings and specialist palliative care services, 14 focus group interviews were conducted with staff across six ID services and one specialist palliative care provider in the greater Dublin (Ireland) area. Qualitative descriptive analysis resulted in the emergence of key themes and formed the development of an educational intervention. Pre- and posttest questionnaires assessed responses to a pilot delivery of the educational intervention. Formal feedback from staff indicated that the educational intervention was highly valued and addressed key training concerns. They agreed that the training supported "aging in place," and the preparation for a "good death" including support for staff, peers, and family in their grief and bereavement. An educational intervention in the form of a trainer manual was produced to support cross-service system in-service training on issues of addressing advanced dementia in persons with

ID.

Fazio, S., Pace, D., Kallmyer, B., & Pike, J.
Alzheimer's Association towards Guidelines for Dementia Care Practice: Recommendations with emphasis on high-quality, person-centered care in long-term and community-based care settings.
Alzheimer's & Dementia, 2018, 14(4), 520-521.
https://doi.org/10.1016/j.jalz.2018.03.001
Abstract: Speaks to the Dementia Care Practice Recommendations that were developed to better define quality care across all care settings and throughout the disease course. Notes that they are intended for professional care providers who work with individuals living with dementia and their families in residential and community based care settings.

Fazio, S., Pace, D., Kallmyer, B., & Maslow, K., & Zimmerman, S.
Alzheimer's Association dementia care practice recommendations.
Gerontologist, 2018, Jan 18, 58(suppl1), S1-S9.
doi: 10.1093/geront/gnx182.
Abstract: The Alzheimer's Association 'Dementia Care Practice Recommendations' outline guidance on quality care practices based on a comprehensive review of current evidence, best practice, and expert opinion. The 'Dementia Care Practice Recommendations' were developed to better define quality care across all care settings, and throughout the disease course. They are intended for professional care providers who work with individuals living with dementia and their families in residential and community-based care settings. With the fundamentals of person-centered care as the foundation, the 'Dementia Care Practice Recommendations' posit goals for quality dementia care in the following areas: (a) person-centered care, (b) detection and diagnosis, (c) assessment and care planning, (d) medical management, (e) Information, education, and support, (f) ongoing care for behavioral and psychological symptoms of dementia, and support for activities of daily living, (g) staffing, (h) supportive and therapeutic environments, and (I) transitions and coordination of services.

Firth, N.C., Startin, C.M., Hithersay, R., Hamburg, S., Wijeratne, P.A., Mok, K.Y., Hardy, J.. Alexander, D.C., LonDownS Consortium, & Strydom, A.
Aging related cognitive changes associated with Alzheimer's disease in Down syndrome.
Annals of Clinical and Translational Neurology, 2018, May 20, 5(6), 741-751.
doi: 10.1002/acn3.571. eCollection 2018 Jun.
Abstract: Individuals with Down syndrome (DS) have an extremely high genetic risk for Alzheimer's disease (AD), however, the course of cognitive decline associated with progression to dementia is ill-defined. Data-driven methods can estimate long-term trends from cross-sectional data while adjusting for variability in baseline ability, which complicates dementia assessment in those with DS. We applied an event-based model to cognitive test data and informant-rated questionnaire data from 283 adults with DS (the largest study of cognitive functioning in DS to date) to estimate the sequence of cognitive decline and individuals' disease stage. Decline in tests of memory, sustained attention/motor coordination, and verbal fluency occurred early, demonstrating that AD in DS follows a similar pattern of change to other forms of AD. Later decline was found for informant measures. Using the resulting staging model, we showed that adults with a clinical diagnosis of dementia and those with APOE 3:4 or 4:4 genotype were significantly more likely to be staged later, suggesting that the model is valid. Our results identify tests of memory and sustained attention may be particularly useful measures to track decline in the preclinical/prodromal stages of AD in DS whereas informant-measures may be useful in later stages (i.e. during conversion into dementia, or postdiagnosis). These results have implications for the selection of outcome measures of treatment trials to delay or prevent cognitive decline due to AD in DS. As clinical diagnoses are generally made late into AD progression, early assessment is essential.

Fleming, V., & Litzelman, K.
Caregiver resource utilization: Intellectual and development disability and dementia.
Journal of Applied Research in Intellectual Disabilities, 2021, 34(6), 1468-1476.
doi: 10.1111/jar.12889
Abstract: Adults with intellectual and developmental disabilities and their families have high need for support services. This study assessed resource utilization among caregivers of intellectual and developmental disabilities and other conditions. We assessed 366 caregivers of adults with intellectual and developmental disabilities, dementia or other conditions Regressions assessed group differences in number of agency contacts and frequency of service use. A secondary analysis assessed reasons for underutilization of services. Caregivers of individuals with dementia contacted twice as many agencies as other caregivers and were more likely to report using suggested services. Agency contact and service utilization were similar among caregivers of adults with intellectual and developmental disabilities compared to other caregivers. Caregivers of adults with intellectual and developmental indicated that suggested services were unavailable to them. The findings of this study shed light on challenges with access to and utilization of support services.

Fleming, V., Helsel, B.C., Ptomey, L.T., Rosas, H.D., Handen, B., Laymon, C., Christian, B.T., Head, E., Mapstone, M., Lai, F., Krinsky-McHale, S., Zaman, S., Ances, B.M., Lee, J.H., Hartley, S.L. & Alzheimer's Biomarker Consortium -Down Syndrome (ABC-DS) Consortium.
Weight loss and Alzheimer's disease in Down syndrome.
Journal of Alzheimers Disease, 2023, 91(3), 1215-1227. doi: 10.3233/JAD-220865.
Abstract: Virtually all adults with Down syndrome (DS) develop Alzheimer's disease (AD) pathology, but research gaps remain in understanding early signs of AD in DS. The goal of the present study was to determine if unintentional weight loss is part of AD in DS. The specific aims were to: 1) examine relation between chronological age, weight, AD pathology, and AD-related cognitive decline were assessed in a large cohort of adults with DS, and 2) determine if baseline PET amyloid-ß (Aß) and tau PET status (-versus+) and/or decline in memory and mental status were associated with weight loss prior to AD progression. Analyses included 261 adults with DS. PET data were acquired using [11C] PiB for Aß and [18F] AV-1451 for tau. Body mass index (BMI) was calculated from weight and height. Direct measures assessed dementia and memory. Clinical AD status was determined using a case consensus process. Percent weight decline across 16-20 months was assessed in a subset of participants (n = 77). Polynomial regressions indicated an 0.23 kg/m2 decrease in BMI per year beginning at age 36.5 years, which occurs alongside the period during which Aß and tau increase and memory and mental status decline. At a within-person level, elevated Aß, decline in memory and mental status were associated with higher percent weight loss across 16-20 months. Unintentional weight loss occurs alongside Aß deposition and prior to onset of AD dementia, and thus may be a useful sign of AD in DS.

Fleming, V., Hartley, S., Handen, B., Tudorascu, D., Lee, L., Mchale, S., Hom, C., Cohen, A., Schworer, E., Peven, J., Zammit, M., Klunk, W., Laymon, C., Zaman, S., Ances, B., & Christian, B.
Does the timing of Alzheimer's disease vary by severity of intellectual disability in Down syndrome?
Alzheimer's & Dementia, 2013, 19(S15), doi:10.1002/alz.079095
Abstract: Trisomy 21 causes Down syndrome (DS) and is a recognized cause of Alzheimer's disease (AD). The triplication of the amyloid precursor protein gene leads to an overexpression of amyloid-beta (Aß), which accumulates into extracellular plaques, followed by intracellular neurofibrillary tau tangles (Lott & Head, 2019). However, variability in the age of AD onset in DS spans 25+ years (Iulita et al., 2022). As the DS field moves to AD clinical trials, it is important to identify factors related to this variability, such as premorbid severity of intellectual disability (ID). We compared the age-trajectory of the AD biomarkers Aß and tau and cognitive decline across premorbid ID levels (mild, moderate, and severe/profound), in models controlling trisomy type (full, mosaic, translocation) and APOE status. Analyses involved 361 adults with DS (M = 45.22 years, SD = 9.92) from the Alzheimer's Biomarkers Consortium-Down Syndrome. Participants completed measures of memory, mental status, and visuospatial ability. Premorbid ID level was recorded. PET was acquired using [11C] PiB for Aß, and [18F] AV-1451 for tau in 162 participants. Aß was quantified using the centiloid method and tau was quantified as SUVR (reference: cerebellar cortex) in a composite region determined as the volume-weighted average of select FreeSurfer 5.3, T1 MR-based components. General linear models controlling for site, age, trisomy type and APOE status indicated no significant effect of premorbid ID level by age on the cognitive outcomes (Figure 1a-e). There was not a significant effect of premorbid ID by age on PET Aß or on tau PET (Figure 2a-b). There was not a significant difference in age of those with mild cognitive impairment-DS (MCI-DS) or dementia by premorbid ID level (Figure 3). This study adds to the characterization of the time course of AD pathology and clinical symptomology in DS. Findings provide robust evidence of a similar time course in

AD trajectory across severity of ID, laying the groundwork for the inclusion of individuals with DS with a variety of IQ levels in clinical AD trials. Premorbid IQ may have little effect on AD biomarkers in DS in contrast to findings in neurotypical populations.

Fleming, V., Hom, C.L., Clare, I.C.H., Hurd-Thomas, S.L., Krinsky-McHale, S., Handen, B., & Hartley, S.L.
Cognitive outcome measures for tracking Alzheimer's disease in Down syndrome.
International Review of Research in Developmental Disabilities, 2022, 62, 227-263. doi: 10.1016/bs.irrdd.2022.05.006. Epub 2022 Jul 21
Abstract: Down syndrome (DS) is now viewed as a genetic type of Alzheimer's disease (AD), given the near-universal presence of AD pathology in middle adulthood and the elevated risk for developing clinical AD in DS. As the field of DS prepares for AD clinical intervention trials, there is a strong need to **identify cognitive measures** that are specific and sensitive to the transition from being cognitively stable to the prodromal (e.g., Mild Cognitive Impairment-Down syndrome) and clinical AD (e.g., Dementia) stages of the disease in DS. It is also important to determine cognitive measures that map onto biomarkers of early AD pathology during the transition from the preclinical to the prodromal stage of the disease, as this transition period is likely to be targeted and tracked in AD clinical trials. This article covers the current state of research on cognitive measures that could be used to screen/select study participants and as potential outcome measures in future AD clinical trials with adults with DS. This article also identifies key challenges that need to be overcome and questions that need to be addressed by the DS field as it prepares for AD clinical trials in the coming years.

Flygare Wallén, E., Ljunggren, G., Wahlström, L., Pettersson, D., Carlsson, A. C., & Wändell, P.
The prevalence of self-harm and mental disorders among individuals with intellectual disabilities.
Nordic Journal of Psychiatry, 2023, 77(7), 712-720. https://doi.org/10.1080/08039488.2023.2228292
Abstract: Mental health disorders are prevalent among individuals with intellectual disabilities (ID). However, there is a lack of research on the impact of concomitant autism spectrum disorders (ASD) or attention deficit hyperactivity disorder (ADHD) on the mental health within this population. We aimed to investigate the prevalence of mental health disorders and registered healthcare visits due to self-harm among individuals with ID. We used administrative data for all healthcare with at least one recorded diagnosis of mental health disorder or self-harm during 2007-2017 among people with a diagnosis of Down syndrome (DS; n = 1298) and with ID without DS (IDnonDS; n = 10,671) using the rest of the population in Stockholm Region (n = 2,048,488) for comparison.The highest odds ratios for a mental health disorder were present in females with IDnonDS (9.01) followed by males with IDnonDS (8.50), compared to the general population. The ORs for self-harm among individuals with IDnonDS were high (8.00 for females and 6.60 for males). There were no registered cases of self-harm among individuals with DS. The prevalence of an anxiety or affective disorder was higher among individuals with ID including DS with concomitant ASD or ADHD. There was a strong increase in the prevalence of psychotic disorders from childhood to adulthood and even more so after the age of 50. In this older group, **dementia** could be the underlying cause especially in the population with DS. Neighborhood socio-economic status was associated with a lower occurrence of mental health disorders and self-harm in wealthier areas for all outcomes and for all groups. Self-harm and psychiatric comorbidities were common among individuals with ID without DS with an attenuated difference among those with concomitant ASD or ADHD, which calls for attention.

Folin, M., Baiguera, S., Conconi, M.T., Pati, T., Grandi, C., Parnigotto, P.P., & Nussdorfer, G.G.
The impact of risk factors of Alzheimer's disease in the Down syndrome
International Journal of Molecular Medicine, 2003, Feb,11(2), 267-270. https://pubmed.ncbi.nlm.nih.gov/12525890/
Abstract: Down syndrome (DS) patients, after the fourth decade of life, display some neuropathological features of the Alzheimer's disease (AD). Several hypotheses suggested that apoE4 protein, an AD risk factor, might promote amyloid formation by stabilizing an aggregated conformation of the beta amyloid protein (Abeta). This peptide is the major proteinaceous component of the senile plaques either in AD or DS, and it is a proteolytic product of the amyloid precursor protein (APP). Both brain and platelets express three APP transcripts of the apparent molecular weight of 106, 110 and 130 kDa. In DS the Abeta deposits may ensue, at least in part, from the overexpression of the Abeta precursor gene located on chromosome 21. Aims of the present study were to evaluate the frequency of apoE4 isoform in DS population, and to ascertain whether the ratio between the 130 and the 106-110 kDa platelet APP isoforms is lower in DS, as seems to occur in AD patients. ApoE4 frequency was significantly lower in DS when compared to AD patients. E4 allele frequency of older DS patients was about half that of younger ones. The 130 to 106-110 kDa APP isoform ratio was similar in young DS and control subjects, and markedly lower in AD patients. Our results indicate that: i) in DS patients the early, selective accumulation of Abeta peptides is independent of the ApoE genotype, but the allele epsilon4 predisposes to various causes of premature death; and ii) platelet APP isoform abnormalities, which can be observed in AD patients, do not occur in young DS patients, suggesting a different processing of APP platelets in DS with respect to AD.

Forster-Gibson, C,J..
Behaviour changes in an adult with Down syndrome.
Canadian Family Physician, 2019 Apr, 65(Suppl 1), S25-S26. PMID: 31023775; PMCID: PMC6501721
Abstract: A case review of a 43-year-old man with Down syndrome who had a 2-year history of behavior changes, paranoia, and auditory hallucinations, increased fatigue, and changes in gait and speech, as well as losses in his social, work, and daily living skills formed the basis of this article. Author notes that adults with Down syndrome are at increased risk of depression, hypothyroidism, and Alzheimer dementia (AD) as they age, all of which can cause behavior changes. Challenges in the primary care of aging adults with developmental disability can include limited information on differences in the aging process; complex health needs; causes of developmental delay; behaviors that challenge; and communication problems. In persons with Down syndrome, pathology studies of the brain have shown the presence of neurofibrillary tangles and plaques typical of AD in 7.5% of teens, 80% of adults in their 30s, and almost all adults older than 40 years of age, although the condition might not be symptomatic. The prevalence of diagnosed AD in adults with Down syndrome has been estimated to be 9.4% for those aged 40 to 49, increasing to 54.5% for those aged 60 to 69. Differentiating between depression and early-stage AD can be very difficult, and depression can be a sign of early AD. Progressive changes that might suggest early-stage AD include loss of language, behavior changes (including social withdrawal), loss of daily living skills, gait disorder, and, in some, psychosis (including hallucinations and delusions), seizures, and dysphagia. Assessment of behavioral changes in adults with Down syndrome should begin with assessment of medical concerns common to Down syndrome. The diagnostic assessment of adults with Down syndrome for Alzheimer dementia is similar to accepted practice in the general population. Caveats include limitations on the usefulness of neuropsychological assessment tools and neuroimaging, and the common occurrence of hypothyroidism and depression.

Fonseca, L. M., Prado Mattar, G., Guerra Haddad, G., Burduli, E., McPherson, S.M., Maria de Figueiredo Ferreira Guilhoto, L., Busatto Filho, G., Sanches Yassuda, M., Bottino, C.M.C., Queiroz Hoexter, M. & Chaytor, N.S.
Prevalence of neuropsychiatric symptoms related to dementia in individuals with Down syndrome
AAIC2020, Poster presentation, July 29, 2020. *Alzheimer's & Dementia,* 16(S6), First published: 07 December 2020. https://doi.org/10.1002/alz.047603
Abstract: Neuropsychiatric symptoms (NPS) are significant manifestations of dementia, with important consequences for patients and caregivers. Despite the established genetic link between Down syndrome (DS) and Alzheimer's disease (AD), studies investigating NPS in individuals with DS and dementia are scarce. The Neuropsychiatric Inventory (NPI) was developed to identify symptoms of dementia and while it is widely used for the assessment of individuals with dementia in the general population, we have found no studies using the NPI in those with DS. The aim of this study is to characterize NPS in a sample with DS with heterogeneous cognitive profiles using the NPI Participants (N=92) with DS, =30 years of age were assessed with the Cambridge Examination for Mental Disorders of Older People with Down's Syndrome and Others with Intellectual Disabilities (CAMDEX-DS). They were classified by a psychiatrist into three categories: AD, prodromal dementia, and stable cognition. Another psychiatrist

blinded to the CAMDEX-DS dementia diagnosis, evaluated the participants using the NPI. Chi-square tests were used to check for significant differences in frequency of symptoms. Thirteen participants (14.1%) had AD, 17 (18.4%) were classified as prodromal dementia and 62 (67.5%) were in the stable cognition group. Prevalence of delusion, depression, anxiety, disinhibition, irritability, appetite abnormalities and total NPI did not differ between groups. Anxiety and irritability were common across all groups (~50% of the total), while euphoria was not present in any participant. Hallucination, agitation, apathy, aberrant motor behaviour and night-time behaviour disturbance showed significant difference among the groups ($p<0.05$), with higher prevalence in the group with AD. Authors note that the results indicate that individuals with DS and AD have some of the symptoms that are characteristically present among the general population with AD. Future studies are needed to understand if AD in DS is associated with a similar pattern of NPS observed among people with AD in the general population or may follow a specific NPS pattern. The high frequency of some NPS in individuals with DS and stable cognition should be considered in the diagnostic process in order to reduce the odds of generating a false positive.

Fonseca, L.M., Haddad, G.G., Mattar, G.P., Oliveira, M.C., Simon, S.S., Guilhoto, L.M., Busatto, G.F., Zaman, S., Holland, A.J., Hoexter, M.Q., & Bottino, C.M.
The validity and reliability of the CAMDEX-DS for assessing dementia in adults with Down syndrome in Brazil.
Brazilian Journal of Psychiatry. 2019 May-Jun;41(3):225-233. doi: 10.1590/1516-4446-2018-0033. Epub 2018 Oct 22.
Abstract: Alzheimer's disease occurs at a higher prevalence and an earlier age in individuals with Down syndrome (DS) than typically developing individuals. However, diagnosing dementia in individuals with intellectual disability remains a challenge due to pre-existing cognitive deficits. The aim of this study was to investigate the validity and reliability of the Brazilian version of the Cambridge Examination for Mental Disorders of Older People with Down's syndrome and Others with Intellectual Disabilities (CAMDEX-DS) for individuals with DS. Two psychiatrists, working independently, evaluated 92 adults with DS ≥ 30 years of age. The concurrent validity of the CAMDEX-DS was analyzed in relation to the gold standard of established international criteria. In a subgroup of 20 subjects, the concurrent validity of the CAMDEX-DS was analyzed in relation to an independent objective assessment of cognitive decline over three years. We analyzed the inter-rater reliability of cognitive assessment. The diagnostic accuracy of the CAMDEX-DS compared to the gold standard was 96.7%. CAMDEX-DS-based diagnosis was considered consistent with cognitive decline. The probability of a participant with dementia having cognitive decline was 83%. Inter-rater reliability for the participant assessment was good, with a kappa of > 0.8 for 93% of the CAMDEX-DS items. The CAMDEX-DS can be considered the first valid and reliable instrument for evaluating dementia in adults with DS in Brazil. Its use in such individuals could improve clinical practice and research.

Fonseca, L.M., Mattar, G.P., Haddad, G.G., Burduli, E., McPherson, S.M., Guilhoto, L.M.F.F., Yassuda, M.S., Busatto, G.F., Bottino, C.M.C., Hoexter, M.Q., Chaytor, N.S..
Neuropsychiatric symptoms of Alzheimer's disease in Down syndrome and its impact on caregiver distress.
Journal of Alzheimer's Disease, 2021, 81(1), 137-154. doi: 10.3233/JAD-201009. ://doi.org/10.3233/JAD-201009
Abstract: Neuropsychiatric symptoms (NPS) are non-cognitive manifestations common to dementia and other medical conditions, with important consequences for the patient, caregivers, and society. Studies investigating NPS in individuals with Down syndrome (DS) and dementia are scarce. Authors sought to characterize NPS and caregiver distress among adults with DS using the Neuropsychiatric Inventory (NPI). We evaluated 92 individuals with DS (=30 years of age), divided by clinical diagnosis: stable cognition, prodromal dementia, and AD. Diagnosis was determined by a psychiatrist using the Cambridge Examination for Mental Disorders of Older People with Down's Syndrome and Others with Intellectual Disabilities (CAMDEX-DS). NPS and caregiver distress were evaluated by an independent psychiatrist using the NPI, and participants underwent a neuropsychological assessment with Cambridge Cognitive Examination (CAMCOG-DS). Symptom severity differed between-groups for delusion, agitation, apathy, aberrant motor behavior, nighttime behavior disturbance, and total NPI scores, with NPS total score being found to be a predictor of AD in comparison to stable cognition (OR for one-point increase in the NPI = 1.342, p = 0.012). Agitation, apathy, nighttime behavior disturbances, and total NPI were associated with CAMCOG-DS, and 62% of caregivers of individuals with AD reported severe distress related to NPS. Caregiver distress was most impacted by symptoms of apathy followed by nighttime behavior, appetite/eating abnormalities, anxiety, irritability, disinhibition, and depression ($R2 = 0.627$, $F(15,76) = 8.510$, $p < 0.001$). NPS are frequent and severe in individuals with DS and AD, contributing to caregiver distress. NPS in DS must be considered of critical relevance demanding management and treatment. Further studies are warranted to understand the biological underpinnings of such symptoms.

Fonseca, L.M.,, Padilla, C., Jones, E., Neale, N., Haddad, G.G., Mattar, G.P., Barros, E., Clare, I.C.H., Busatto, G.F., Bottino, C.M.C., Hoexter, M.Q., Holland, A.J., & Zaman, S.
Amnestic and non-amnestic symptoms of dementia: An international study of Alzheimer's disease in people with Down's syndrome
International Journal of Geriatric Psychiatry, 2020 Jun, 35(6), 650-661. doi: 10.1002/gps.5283. Epub 2020 Mar 15.
Abstract: The presence of age-related neuropathology characteristic of Alzheimer's disease (AD) in people with Down syndrome (DS) is well-established. However, the early symptoms of dementia may be atypical and appear related to dysfunction of prefrontal circuitry. The authors sought to characterize the initial informant reported age-related neuropsychiatric symptoms of dementia in people with DS, and their relationship to AD and frontal lobe function. Non-amnestic informant reported symptoms (disinhibition, apathy, and executive dysfunction) and amnestic symptoms from the CAMDEX-DS informant interview were analyzed in a cross-sectional cohort of 162 participants with DS over 30 years of age, divided into three groups: stable cognition, prodromal dementia, and AD. To investigate age-related symptoms prior to evidence of prodromal dementia we stratified the stable cognition group by age. Amnestic and non-amnestic symptoms were present before evidence of informant-reported cognitive decline. In those who received the diagnosis of AD, symptoms tended to be more marked. Memory impairments were more marked in the prodromal dementia than the stable cognition group (OR = 35.07; P < .001), as was executive dysfunction (OR = 7.16; P < .001). Disinhibition was greater in the AD than in the prodromal dementia group (OR = 3.54; P = .04). Apathy was more pronounced in the AD than in the stable cognition group (OR = 34.18; P < .001). Premorbid amnestic and non-amnestic symptoms as reported by informants increase with the progression to AD. For the formal diagnosis of AD in DS this progression of symptoms needs to be taken into account. An understanding of the unique clinical presentation of DS in AD should inform treatment options.

Fonseca, L M.., Rufino Navatta, A.C., Bottino, C.M.C., & Miotto, E.C.
Cognitive rehabilitation of dementia in adults with Down syndrome: A review of non-pharmacological interventions
Dementia and Geriatric Cognitive Disorders Extra, 2015, Sep-Dec, 5(3), 330–340.
Published online 2015 Sep 18. doi: 10.1159/000438858
Abstract: There is a close genetic relationship between Alzheimer's disease (AD) and Down syndrome (DS), AD being the most severe mental disorder affecting aging individuals with DS. The objective of the present study was to evaluate the efficacy of cognitive rehabilitation interventions in DS patients with AD by means of a critical literature review. Because AD is progressive and irreversible, treatment is aimed at delaying and reducing the cognitive and functional decline in order to preserve or improve quality of life. The effects that pharmacological treatments and cognitive interventions have on elderly individuals with AD are well documented. Recent clinical trials have investigated the use of pharmacological treatment in DS patients with AD, generating preliminary results that have been unfavorable. There is a clear lack of studies addressing the efficacy of cognitive rehabilitation interventions in DS patients with AD, and there is an urgent need for studies providing evidence to inform decisions regarding the appropriate choice of treatment strategies.

Fonseca, L.M., Prado Mattar, G., Guerra Haddad, G., Burduli, E., McPherson, S.M., de Figueiredo Ferreira Guilhoto, L.M., Sanches Yassuda, M. Filho Busatto, G., Machado de Campos Bottino, C., Queiroz Hoexter, M., & Sage Chaytor, N.
Neuropsychiatric symptoms of Alzheimer's disease in Down Syndrome and Its impact on caregiver distress
Journal of Alzheimer's Disease, 2021, 81(1),137-154.doi: 10.3233/JAD-201009.
Abstract: Neuropsychiatric symptoms (NPS) are non-cognitive manifestations common to dementia and other medical conditions, with important consequences for the patient, caregivers, and society. Studies investigating NPS in individuals

with Down syndrome (DS) and dementia are scarce. Characterize NPS and caregiver distress among adults with DS using the Neuropsychiatric Inventory (NPI). We evaluated 92 individuals with DS (≥30 years of age), divided by clinical diagnosis: stable cognition, prodromal dementia, and AD. Diagnosis was determined by a psychiatrist using the Cambridge Examination for Mental Disorders of Older People with Down's Syndrome and Others with Intellectual Disabilities (CAMDEX-DS). NPS and caregiver distress were evaluated by an independent psychiatrist using the NPI, and participants underwent a neuropsychological assessment with Cambridge Cognitive Examination (CAMCOG-DS). Symptom severity differed between-groups for delusion, agitation, apathy, aberrant motor behavior, nighttime behavior disturbance, and total NPI scores, with NPS total score being found to be a predictor of AD in comparison to stable cognition (OR for one-point increase in the NPI = 1.342, p = 0.012). Agitation, apathy, nighttime behavior disturbances, and total NPI were associated with CAMCOG-DS, and 62% of caregivers of individuals with AD reported severe distress related to NPS. Caregiver distress was most impacted by symptoms of apathy followed by nighttime behavior, appetite/eating abnormalities, anxiety, irritability, disinhibition, and depression (R2 = 0.627, F(15,76) = 8.510, p < 0.001). NPS are frequent and severe in individuals with DS and AD, contributing to caregiver distress. NPS in DS must be considered of critical relevance demanding management and treatment. Further studies are warranted to understand the biological underpinnings of such symptoms.

Fonseca, L.M., Yokomizo, J.E., Bottino, C.M., & Fuentes, D.
Frontal lobe degeneration in adults with Down syndrome and Alzheimer's disease: A review.
Dementia and Geriatric Cognitive Disorders. 2016;41(3-4):123-36. doi: 10.1159/000442941.
Abstract: here is a proven link between Down syndrome and the early development of the neuropathological features of Alzheimer's disease (AD). Changes in the personality and behavior of adults with Down syndrome might indicate the early stages of dementia or of frontotemporal lobar degeneration. The objective of this study was to investigate the executive functions and changes in behavior associated with frontal lobe degeneration in individuals with Down syndrome who develop AD. We conducted a systematic review selecting studies employing cognitive assessments. We identified few studies using objective measurements to determine whether cognitive aspects associated with the frontal lobe correlate with dementia in this population. We observed a tendency toward such correlations. There is a need for further studies in which objective measures of cognitive and behavioral factors are evaluated together with data related to brain function and morphology.

Forbat, L., & Service, K.P.
Who cares? Contextual layers in end-of-life care for people with intellectual disability and dementia.
Dementia, 2005, 4(3), 413-431.
https://go.gale.com/ps/i.do?id=GALE%7CA137361004&sid=googleScholar&v=2.1&it=r&linkaccess=abs&issn=14713012&p=AONE&sw=w&userGroupName=maine_oweb&isGeoAuthType=true&aty=geo
Abstract: The complexity of the relationship between intellectual disability (ID) and dementia is increasingly acknowledged. In order to operationalize a route towards person-centered care, we introduce the hierarchy model (Pearce, 1999) as a tool to focus the attention of policy and practice on all aspects of caregiving. This tool, which is taken from the family therapy literature, enables practitioners to examine the broad systems that impact on the delivery and receipt of care. In this article, we focus on its utility in scrutinizing end-of-life and later stages of dementia by illustrating its use with three key areas in dementia care. These three areas provide some of the most challenging situations at the end stages, because of the possible treatment options, they are: nutrition, medical interventions, and the location of care provision. This model enables a focused approach to understanding how meaning is created within social interaction. The article draws out implications for practice and policy and has applications for practice internationally.

Fortea, J., Zaman,S,H., Hartley, S., Rafii, M.S., & Head, H., & Maria Carmona-Iragui, M.
Alzheimer's disease associated with Down syndrome: a genetic form of dementia
The Lancet Neurology, 2021 (Nov. 01), 20(11), 930-942. doi: https://doi.org/10.1016/S1474-4422(21)00245-3
Abstract: Adults with Down syndrome develop the neuropathological hallmarks of Alzheimer's disease and are at very high risk of developing early-onset dementia, which is now the leading cause of death in this population. Diagnosis of dementia remains a clinical challenge because of the lack of validated diagnostic criteria in this population, and because symptoms are overshadowed by the intellectual disability associated with Down syndrome. In people with Down syndrome, fluid and imaging biomarkers have shown good diagnostic performances and a strikingly similar temporality of changes with respect to sporadic and autosomal dominant Alzheimer's disease. Most importantly, there are no treatments to prevent Alzheimer's disease, even though adults with Down syndrome could be an optimal population in whom to conduct Alzheimer's disease prevention trials. Unprecedented research activity in Down syndrome is rapidly changing this bleak scenario that will translate into disease-modifying therapies that could benefit other populations.

Fortea J, Vilaplana E, Carmona-Iragui M, Benejam B, Videla L, Barroeta I, Fernández S, Altuna M, Pegueroles J, Montal V, Valldeneu S, Giménez S, González-Ortiz S, Muñoz L, Estellés T, Illán-Gala I, Belbin O, Camacho V, Wilson LR, Annus T, Osorio RS, Videla S, Lehmann S, Holland AJ, Alcolea D, Clarimón J, Zaman SH, Blesa R, Lleó A.
Clinical and biomarker changes of Alzheimer's disease in adults with Down syndrome: a cross-sectional study.
Lancet. 2020 Jun 27;395(10242):1988-1997. doi: 10.1016/S0140-6736(20)30689-9
Abstract: Alzheimer's disease and its complications are the leading cause of death in adults with Down syndrome. Studies have assessed Alzheimer's disease in individuals with Down syndrome, but the natural history of biomarker changes in Down syndrome has not been established. We characterised the order and timing of changes in biomarkers of Alzheimer's disease in a population of adults with Down syndrome. We did a dual-centre cross-sectional study of adults with Down syndrome recruited through a population-based health plan in Barcelona (Spain) and through services for people with intellectual disabilities in Cambridge (UK). Cognitive impairment in participants with Down syndrome was classified with the Cambridge Cognitive Examination for Older Adults with Down Syndrome (CAMCOG-DS). Only participants with mild or moderate disability were included who had at least one of the following Alzheimer's disease measures: apolipoprotein E allele carrier status; plasma concentrations of amyloid ß peptides 1-42 and 1-40 and their ratio (Aß1-42/1-40), total tau protein, and neurofilament light chain (NFL); tau phosphorylated at threonine 181 (p-tau), and NFL in cerebrospinal fluid (CSF); and one or more of PET with 18F-fluorodeoxyglucose, PET with amyloid tracers, and MRI. Cognitively healthy euploid controls aged up to 75 years who had no biomarker abnormalities were recruited from the Sant Pau Initiative on Neurodegeneration. We used a first-order locally estimated scatterplot smoothing curve to determine the order and age at onset of the biomarker changes, and the lowest ages at the divergence with 95% CIs are also reported where appropriate. Between Feb 1, 2013, and June 28, 2019 (Barcelona), and between June 1, 2009, and Dec 31, 2014 (Cambridge), we included 388 participants with Down syndrome (257 [66%] asymptomatic, 48 [12%] with prodromal Alzheimer's disease, and 83 [21%] with Alzheimer's disease dementia) and 242 euploid controls. CSF Aß1-42/1-40 and plasma NFL values changed in individuals with Down syndrome as early as the third decade of life, and amyloid PET uptake changed in the fourth decade. 18F-fluorodeoxyglucose PET and CSF p-tau changes occurred later in the fourth decade of life, followed by hippocampal atrophy and changes in cognition in the fifth decade of life. Prodromal Alzheimer's disease was diagnosed at a median age of 50·2 years (IQR 47·5-54·1), and Alzheimer's disease dementia at 53·7 years (49·5-57·2). **Symptomatic Alzheimer's disease prevalence increased with age in individuals with Down syndrome, reaching 90-100% in the seventh decade of life.** Alzheimer's disease in individuals with Down syndrome has a long preclinical phase in which biomarkers follow a predictable order of changes over more than two decades. The similarities with sporadic and autosomal dominant Alzheimer's disease and the prevalence of Down syndrome make this population a suitable target for Alzheimer's disease preventive treatments.

Foundation for People with Learning Disabilities
Down's syndrome and dementia - Briefing for Commissioners
London: The Foundation for People with Learning Disabilities [c/o Mental Health Foundation, 20/21 Cornwall Terrace, London, England NW1 4QL; e/m mhf@mhf.org.uk; www.learningdisabilities.org.uk] (February 2001)

8 pp.
Abstract: Backgrounder document, written for funders of services in the United Kingdom, outlines the epidemiology of dementia and Down's syndrome and identifies key support services necessary as part of a package of local services to be established for persons affected by dementia and intellectual disabilities (ID). While titled for dementia and Down's syndrome applicable for all persons with ID. Written in brief style, covers main issues and funding considerations and serves as an excellent planning tool for establishing services. Also covers basic clinical diagnostic information and basis for care management decision making. Routes the reader to associated organizations for further information

Fray, M.T.
Caring for Kathleen: A sister's story about Down's syndrome and dementia. Kidderminster, United Kingdom: British Institute of Learning Disabilities [BILD, Wolverhampton Road, Kidderminster, Worcestershire, UK DY10 3PP – www.bild.demon.co.uk] (2000)
44 pp.
Abstract: Biographical monograph on the aging and eventual decline and death of a woman with Down syndrome as told by her sister. Provides many insights in service barriers and successes, while also providing a vivid case example of how Alzheimer's disease affects a family carer of a person with an intellectual disability.

Fredericksen, J. & Fabbre, V.
Down syndrome and Alzheimer's disease: Issues and implications for social work.
Journal of Gerontological Social Work, 2018, 61(1), 4-10. DOI: 10.1080/01634372.2017.1393480.
Owing to recent medical advancements, people with Down Syndrome (DS) are now able to live considerably longer lives and thus experience a variety of complex issues as they age. Alzheimer's Disease (AD) frequently occurs in older adults who have DS, but few practice guidelines exist to inform social work practice with older adults who have this dual diagnosis. This commentary will highlight the connection between these two conditions within a neurobiological framework and discuss implications for practice based on the available literature on this intersection of ability status, cognitive status, and age.

Frederiksen, K..S., Nielsen, T.R., Winblad, B., Schmidt, R., Kramberger, M.G., Jones, R.W., Hort, J., Grimmer, T., Georges, J., Frölichm L., et al.
European Academy of Neurology/European Alzheimer's Disease Consortium position statement on diagnostic disclosure, biomarker counseling, and management of patients with mild cognitive impairment
European Journal of Neurology, 2021, 28(7), 2147-2155. doi: 10.1111/ene.14668.
Abstract: Careful counseling through the diagnostic process and adequate postdiagnostic support in patients with mild cognitive impairment (MCI) is important. Previous studies have indicated heterogeneity in practice and the need for guidance for clinicians. A joint European Academy of Neurology/European Alzheimer's Disease Consortium panel of dementia specialists was appointed. Through online meetings and emails, positions were developed regarding disclosing a syndrome diagnosis of MCI, pre- and postbiomarker sampling counseling, and postdiagnostic support. Prior to diagnostic evaluation, motives and wishes of the patient should be sought. Diagnostic disclosure should be carried out by a dementia specialist taking the ethical principles of "the right to know" versus "the wish not to know" into account. Disclosure should be accompanied by written information and a follow-up plan. It should be made clear that MCI is not dementia. Prebiomarker counseling should always be carried out if biomarker sampling is considered and postbiomarker counseling if sampling is carried out. A dementia specialist knowledgeable about biomarkers should inform about pros and cons, including alternatives, to enable an autonomous and informed decision. Postbiomarker counseling will depend in part on the results of biomarkers. Follow-up should be considered for all patients with MCI and include brain-healthy advice and possibly treatment for specific underlying causes. Advice on advance directives may be relevant. Guidance to clinicians on various aspects of the diagnostic process in patients with MCI is presented here as position statements. Further studies are needed to enable more evidence-based and standardized recommendations in the future.

Fromage, B., & Anglade, P.
[Le vieillissement des sujets trisomiques] [The aging of adults with Down syndrome] *Article in French*
L'Encéphale, 2002, May-June, 28(3 Pt 1), 212-216.
https://pubmed.ncbi.nlm.nih.gov/12091781/
Abstract: The normal aging of adults with Down syndrome is comparable to that observed in individuals who have an equivalent cognitive deficit. However it is earlier and is related to the former intellectual level and life story of the person. Before 50 years, there is no significant reduction of memory. After this age short-term memory, the speed of information processing and selective attention weaken. These changes are similar to those in older adults with intellectual disability, giving the impression of early ageing in adults with Down syndrome. In terms of autonomy in everyday life, it is possible to establish an average evolutionary profile. From 60 years old, deterioration is estimated at 45% of the score obtained at 40 years, affecting in particular the skills necessary for the carrying out daily tasks (washing, dressing, feeding without assistance.). We have little knowledge of the psychiatric evolution of this people because older handicapped people are a new group in society. In the three fields of cognition, autonomy and mental health, the ageing of adults with Down syndrome is very sensitive to their environment. Pathological aging of adults with Down syndrome is associated with the dementia syndrome that, with varying degrees, combines disorders of the cognitive functions and behavior, modifying the personality. The clinical diagnosis of dementia is difficult to establish in adults with Down syndrome and opinions diverge, also it is important to comply with three rules: 1) to establish an individual base line and to observe, with the help of regular evaluations, a clear change in performance; this must be confirmed by similar modifications in daily conducts; 2) the decline depends not only on the resources of the subject, but also on the demands made by environment; 3) lasting deterioration of capacities must be clearly greater than that observable in normal ageing to signify dementia. As a function of actual age, adults with Down syndrome show early signs of ageing compared to the general population. One notes the presence of pathological anatomic lesions from 36 years old, which are similar to those observable among adults diagnosed with Alzheimer disease. However it seems that about 20% of adults with Down syndrome do not show clinical signs of dementia 20 years later. The diagnosis, which is delicate to establish, requires an ecological process consistent over time, underlining the influence of the context and the human environment on aging amond adiults with Down syndrome.

Fromage, B., & Anglade, P.
Avancée en âge du sujet atteint d'une trisomie 21 [The aging of Down's syndrome subjects].
Encephale, 2002, May-Jun, 28(3 Pt 1), 212-216.
https://www.lissa.fr/rep/articles/12091781
Abstract: The normal ageing of adults with Down syndrome is comparable to that observed in individuals who have an equivalent cognitive deficit. However it is earlier and is related to the former intellectual level and life story of the person. Before 50 years, there is no significant reduction of memory. After this age short-term memory, the speed of information processing and selective attention weaken. These changes are similar to those in older adults with an intellectual disabiliy, giving the impression of early ageing in Down syndrome subjects. In terms of autonomy in everyday life, it is possible to establish an average evolutionary profile. From 60 years old, deterioration is estimated at 45% of the score obtained at 40 years, affecting in particular the skills necessary for the carrying out daily tasks (washing, dressing, feeding without assistance.). We have little knowledge of the psychiatric evolution of this people because older handicapped people are a new group in society. In the three fields of cognition, autonomy and mental health, the ageing of Down syndrome subjects is very sensitive to their environment. Pathological aging of adults ith Down syndrome is associated with the dementia syndrome that, with varying degrees, combines disorders of the cognitive functions and behavior, modifying their personality. The clinical diagnosis of dementia is difficult to establish in subjects with Down syndrome and opinions diverge, also it is important to comply with three rules: 1) to establish an individual base line and to observe, with the help of regular evaluations, a clear change in performance; this must be confirmed by similar modifications in daily conducts; 2) the decline depends not only on the resources of the subject, but also on the demands made by environment; 3) lasting deterioration of capacities must be clearly greater than that observable in normal ageing to signify dementia. As a function of actual age, adults with Down syndrome show early signs of aging compared to the general population. One notes the presence of pathological anatomic lesions from 36 years old, which are

similar to those observable among patients with Alzheimer's disease. However it seems that about 20% of adults with Down syndrome do not show clinical signs of dementia 20 years later. The diagnosis, which is delicate to establish, requires an ecological process consistent over time, underlining the influence of the context and the human environment on the ageing of adults with Down syndrome.

Fujino, H., & Moritsugu, A.
Dohsa-hou for unexplained regression in Down syndrome in a 19-year-old man: A case report.
Clinical Case Reports, 2022, 10(5), e05827. https://doi.org/10.1002/ccr3.5827
Abstract: Unexplained regression in Down syndrome (URDS) has become a significant issue in clinical practice and research. This report illustrates the case of a patient with URDS treated with psychological treatment using Dohsa-hou, in addition to medication. Dohsa-hou, a form of psychological treatment, used for people with mental disorders in Asian countries. In Dohsa-hou treatment communication using body movement and physical interactions between the patient and therapist is emphasized. The intervention included relaxation procedures and empathic and communicative physical interaction with the therapist in treatment sessions, intended to alleviate the patient's psychological distress and agitation in interactions with others. After seven months, from Dohsa-hou initiation, the patient showed partial-to-moderate improvement in the NPI total symptoms (37%) and large improvement in caregiver's distress (59%), although some persistent symptoms remained (agitation and apathy). Although psychological treatment may be helpful, monitoring potential risks of acute aggression is necessary.

Funaki, Y., Kaneko, F., & Okamura, H.
Study of factors associated with changes in quality of life of demented elderly persons in group homes
Scandinavian Journal of Occupational Therapy, 2005, 12(1), 4-9. doi:10.1080/11038120510031725.
Abstract: The purpose of the present study was to attempt to identify changes in quality of life (QOL) and factors associated with them shortly after demented elderly residents entered a group home. The subjects were 25 demented elderly persons who had entered a group home within the previous 3 months. Their QOL and factors associated with it were evaluated on two occasions, at baseline and 3 months later. An objective scale for dementia, the Quality of Life Questionnaire for Dementia (QOL-D), was used to evaluate their QOL. The results showed a significant change between the QOL-D scores at baseline and 3 months later, and changes in housekeeping item scores were extracted as factors associated with changes in QOL-D. These findings suggest that the QOL score rises soon after entering a group home, and that the acquisition of roles within the group home may influence the increase in QOL.

Furness, K.A., Loverseed, A., Lippold, T., & Dodd, K.
The views of people who care for adults with Down's syndrome and dementia: A service evaluation.
British Journal of Learning Disabilities, 40(4), 318-327.
https://doi.org/10.1111/j.1468-3156.2011.00714.x
Abstract: It is well established that people with Down's syndrome are more likely to develop dementia than other people and that onset of dementia is likely to occur earlier at an earlier age. The article reports on a specialist service for people with Down's syndrome and dementia. The service has offered dementia screening and assessment to people with Down's syndrome for over 10 years and has also developed to offer support and training for carers. Semi-structured interviews were conducted with family carers, relatives and staff about the impact on them of caring for someone with Down's syndrome and how the dementia service supports them in this role. The resulting data were analyzed using interpretative phenomenological analysis. The responses provide rich insights into the areas of 'knowledge and information', 'coping and support' and 'concerns about the future'. Interviewees also identified services they wanted for the future. As a result of this evaluation, a number of changes have been proposed and begun to be implemented within the service. The results have important implications for other health, social care, and voluntary organizations.

Gangadharan, S.K. & Jovia Maria Jesu. A.
Dementia in people with intellectual disability
In Sabyasachi Bhaumik, David Branford, Mary Barrett,Satheesh Kumar Gangadharan (Eds), The Frith Prescribing Guidelines for People with Intellectual Disability (Chapter 5). 08 September 2015. Oxford, UK: Wiley. https://doi.org/10.1002/9781118897164.ch5.
Abstract: People with intellectual disabilities (ID) have a higher risk of developing dementia when compared to the general population. The four most common causes of dementia are Alzheimer's disease, Lewy body dementia, vascular dementia and fronto-temporal dementia. The common presentation of dementia in people with ID includes memory loss and deterioration in speech, personality and behavioral changes, disorientation and functional deterioration. The diagnosis of dementia requires evidence of a definitive change in cognitive functioning such as impairment in memory, language ability (aphasia), ability to perform complex tasks (apraxia) and orientation in time and place. A number of medications are used in delaying the progress of dementia as in general population. Evidence base for the use of medication in people with ID is currently limited. Psychotropic medications have only a limited role in the management of neuropsychiatric symptoms in people with ID and dementia.

Garcia-Alba, J., Ramírez Toraño, F., Esteba-Castillo, S., Bruña, R., Moldenhauer, F., Novell, R., Romero-Medina, V., & Maestú, F.
Neuropsychological and neurophysiological characterization of mild cognitive impairment and Alzheimer's disease in Down Syndrome
Neurobiology of Aging, 2019 Dec, 84, 70-79. doi: 10.1016/j.neurobiolaging.2019.07.017. Epub 2019 Aug 3.
Abstract: Down syndrome (DS) has been considered a unique model for the investigation of Alzheimer's disease (AD) but intermediate stages in the continuum are poorly defined. Considering this, we investigated the neurophysiological (i.e., magnetoencephalography [MEG]) and neuropsychological patterns of mild cognitive impairment (MCI) and AD in middle-aged adults with DS. The sample was composed of four groups: Control-DS (n = 14, mean age 44.64 ± 3.30 years), MCI-DS (n = 14, 51.64 ± 3.95 years), AD-DS (n = 13, 53.54 ± 6.58 years), and Control-no-DS (healthy controls, n = 14, 45.21 ± 4.39 years). DS individuals were studied with neuropsychological tests and MEG, whereas the Control-no-DS group completed only the MEG session. Our results showed that the AD-DS group exhibited a significantly poorer performance as compared with the Control-DS group in all tests. Furthermore, this effect was crucially evident in AD-DS individuals when compared with the MCI-DS group in verbal and working memory abilities. In the neurophysiological domain, the Control-DS group showed a widespread increase of theta activity when compared with the Control-no-DS group. With disease progression, this increased theta was substituted by an augmented delta, accompanied with a reduction of alpha activity. Such spectral pattern-specifically observed in occipital, posterior temporal, cuneus, and precuneus regions-correlated with the performance in cognitive tests. This is the first MEG study in the field incorporating both neuropsychological and neurophysiological information, and demonstrating that this combination of markers is sensitive enough to characterize different stages along the AD continuum in DS.

Ghazirad, M., Hewitt, O. & Walden, S.
What outcome measures are most useful in measuring the effectiveness of anti-dementia medication in people with intellectual disabilities and dementia?
Advances in Mental Health and Intellectual Disabilities, 2022, 16(2), 87-101.
https://doi.org/10.1108/AMHID-10-2021-0038
Abstract: The use of anti-dementia medication in people with intellectual disabilities has been controversial and requires additional research to assess the efficacy of such medications. An essential part of this treatment (both in terms of research and clinical practice) is having robust outcome measures to assess the efficacy of these medications for individuals. Currently there is no consensus in the UK regarding which outcome measures, in conjunction with clinical judgement, are effective in informing clinicians' decision-making regarding anti-dementia medication management and this paper aims to present useful outcome measures. A comprehensive literature search was conducted to identify relevant outcome measures. Outcome measures focused on aspects of patients' presentation such as cognition, activities of daily living, neuropsychiatric presentation or the impact of their presentation (either on themselves, or on others). These outcome measures were critically appraised to ascertain their suitability in informing clinician's decisions regarding management of anti-dementia medication. The focus of this appraisal was on good quality measures that are practical and accessible and can be easily used within clinical NHS services. This paper provides advice for clinicians on using appropriate outcome measures, depending on patients' presentations and the symptoms of dementia being targeted, that can be used alongside their clinical assessment to

enhance their anti-dementia medication management. Two case studies are presented to illustrate the use of such outcome measures. The case for using a range of assessments that are both broad in focus, and those specifically selected to measure the areas of functioning targeted by the anti-dementia medication, is presented.

Ghezzo, A., Salvioli, S., Solimando, M.C., Palmieri, A., Chiostergi, C., Scurti,M., Lomartire,L., Bedetti, F., Cocchi, G., Follo, D., Pipitone, E,, Rovatti,P., Zamberletti, J., Gomiero, T., Castellani, G., & Franceschi, C.
Age-related changes of adaptive and neuropsychological features in persons with Down syndrome.
PLoSONE. 2014, 9(11): e113111. doi:10.1371/journal.pone.0113111
Abstract: Down syndrome (DS) is characterised by premature aging and an acceleratedecline of cognitive functions in the vast majority of cases. As the life expectancy of DS persons is rapidly increasing, this decline is becoming a dramatic healthproblem. The aim of this study was to thoroughly evaluate a group of 67 non-demented persons with DS of different ages (11 to 66 years), from neuropsychological, neuropsychiatric and psychomotor point of view in order to evaluate in a cross-sectional study the age-related adaptive and neuropsychological features, and to possibly identify early signs predictive of cognitive decline. The main finding of this study is that both neuropsychological functions and adaptive skills are lower in adult DS persons over 40 years old, compared to younger ones. In particular, language and short memory skills, frontal lobe functions, visuo-spatial abilities and adaptive behavior appear to be the more affected domains. A growing deficit in verbal comprehension, along with social isolation, loss of interest and greater fatigue in daily tasks, are the main features found in older, non demented DS persons evaluated in our study. It is proposed that these signs can be alarm bells for incipient dementia, and that neuro-cognitive rehabilitation and psycho-pharmacological interventions must start as soon as the fourth decade (or even earlier) in DS persons, i.e. at an age where interventions can have the greatest efficacy.

Gholipour., T, Mitchell, S., Sarkis, R.A., & Chemali, Z.
The clinical and neurobehavioral course of Down syndrome and dementia with or without new-onset epilepsy.
Epilepsy & Behavior, 2017 Mar; 68, 11-16. doi: 10.1016/j.yebeh.2016.12.014. Epub 2017 Feb 10.
Abstract: Adult patients with Down syndrome (DS) are at higher risk of developing Alzheimer-type dementia and epilepsy. The relationship between developing dementia and the risk of developing seizures in DS is poorly characterized to date. In addition, treatment response and medication tolerability have not been rigorously studied. We identified 220 patients with a diagnosis of DS and dementia. Those without a history of developing seizures (DD) were compared to patients with new-onset seizures (DD+S) after the age of 35. Electronic records were reviewed for demographics, seizure characteristics, cognitive status, and psychiatric comorbidities. Of the patients included for analysis, twenty-six out of 60 patients had new-onset seizures or developed seizures during the follow-up period (the DD+S group) with a median onset of 2.0years after the dementia diagnosis. Generalized tonic-clonic seizures were the most common seizure type (61.5% of DD+S). Sixteen (61.5%) patients were reported to have myoclonus. Levetiracetam was the most commonly used initial medication, with the majority (73%) of patients treated achieving partial or complete seizure control. The DD+S patients tended to have a similar burden of new-onset neuropsychiatric symptoms compared to the DD group. New-onset epilepsy seems to occur early in the course of dementia in DS patients. Patients generally respond to treatment. A great burden of neuropsychiatric symptoms is seen. Future studies need to explore the relationship between ß-amyloid accumulation and epileptiform activity and attend to the care and needs of DS patients with dementia and seizures.

Giménez, S., Tapia, I.E., Fortea, J., Levedowski, D., Osorio, R., Hendrix, J., & Hillerstrom, H.
Caregiver knowledge of obstructive sleep apnoea in Down syndrome.
Journal of Intellectual Disability Research, 2023 Jan;67(1), 77-88. doi: 10.1111/jir.12990. Epub 2022 Nov 22. PMID: 36416001.
Abstract: Down syndrome (DS) population has a very high prevalence of obstructive sleep apnoea (OSA), but this remains underdiagnosed. Hence, we aimed to evaluate caregiver's knowledge of OSA and related sociodemographic factors that could contribute to OSA screening patterns in this population. An online survey though the LuMind IDSC Foundation focused on OSA diagnosis, treatments and the number of sleep studies performed. Data were compared between subjects born before and after the American Academy of Pediatrics (AAP) recommendations for OSA screening. Of the caregivers, 724 (parents 96.3%), responded to the survey. The median [interquartile (IQR)] age of the subjects with DS was 12 [20;7] years. The majority (84.3%) had sleep apnoea diagnosis, and half of them were initially referred for a sleep study due to disturbed sleep symptoms. Only 58.7% of the responders were aware of the AAP recommendations. This was linked to higher socioeconomic and/or educational level and to an earlier OSA diagnosis. The median (IQR) age of OSA diagnosis was lowered after the AAP guidelines publication compared with before its publication (3 [4;2] years vs. 10 [18;5] years, P < 0.000). Adenotonsillectomy (81.9%) and continuous positive airway pressure (61.5%) were the most commonly prescribed treatments. Few had discussed other new therapies such as hypoglossal nerve stimulation (16.0%). Only 16.0% of the subjects repeated the sleep study to monitor OSA with ageing, and 30.2% had to wait more than 4 years between studies. This study reinforces the need to improve OSA knowledge of caregivers and clinicians of individuals with DS to promote an earlier diagnosis and optimal treatment of OSA in this population.

Gitlin, L.N., & Corcoran, M.
Making homes safer: environmental adaptations for people with dementia
Alzheimer's Care Quarterly, 2000, 1(1), 50-58.
https://journals.lww.com/actjournalonline/abstract/2000/01010/making_homes_safer__environmental__adaptations_for.9.aspx
Abstract: Evaluating the safety of the home environment is an important component of clinical care for persons with dementia. This article discusses safety concerns for persons with dementia living at home alone or with family members, specific modifications to the physical environment to address these issues, and guiding principles for implementing environmental changes. A wide range of environmental strategies can be introduced to maximize home safety. Different adaptations may need to be implemented with progressive memory loss thus necessitating periodic reevaluation of the home.

Glasson, E.J., Dye. D.E., & Bittles, A.H.
The triple challenges associated with age-related comorbidities in Down syndrome.
Journal of Intellectual Disability Research, 2014, Apr; 58(4), 393-398. doi:10.1111/jir.12026. Epub 2013 Mar 19. PMID: 23510031
Abstract: Major increases in the survival of people with Down syndrome during the last two generations have resulted in extended periods of adulthood requiring specialist care, which in turn necessitates greater understanding of the nature, timing and impact of comorbidities associated with the disorder. The prevalence of five comorbidities reported as common in adults with Down syndrome, visual impairment, hearing impairment, epilepsy, thyroid disorders and dementia was assessed by decade of life. From early adulthood, people with Down syndrome are at enhanced risk of developing new comorbidities and they may present with multiple conditions. Three specific challenges are identified and discussed: are comorbidities detected in a timely manner, is the clinical progress of the disorder adequately understood, and who is responsible for the provision of care? Further detailed investigations into the development and treatment of comorbidities across the lifespan are needed for a successful longitudinal approach to healthcare in people with Down syndrome. Authors note that iImplementation of this approach will better inform healthcare providers to ensure continuity of care with advancing age.

Golaz, J., Charnay, Y., Vallet, P., & Bouras, C.
Maladie d'Alzheimer et syndrome de Down. Quelques données étiopathogéniques récentes [Alzheimer's disease and Down's syndrome. Some recent etiopathogenic data].
Encephale,1991 Jan-Feb, 17(1), 29-31. French. PMID: 1669030.
Abstract: The present status of research clearly demonstrates the occurrence of lesions characteristic of Alzheimer's disease in patients suffering from a Down's syndrome or trisomy 21. The senile plaques appear very early in trisomy 21 (from the age of 20) and are constant after 40 or 45 years. In these two illnesses, the beta-amyloid protein or A4 protein (4.2 kD) leads to deposits in preferential regions of the central nervous system within two compartments: 1) intracellular, contributing to the formation of neurofibrillary tangles and 2) extracellular, making up the amyloid center of senile plaques as well as around the wall of some blood vessels, then corresponding to the amyloid congophilic angiopathies. Unexpectedly, larger proteins including the A4 sequence have been shown to be normally present in several tissues of normal as well as sick individuals and

represent possible precursors of the A4 protein. Alzheimer's disease may happen either sporadically or following a familial incidence associated with an autosomic dominant mode of transmission. Moreover, the risk of incidence of trisomy 21 seems to be enhanced for collaterals of Alzheimer's disease patients. Since 1987, the use of molecular biology tools has revealed particularly fruitful. A linkage analysis has been undertaken that showed an association of the putative gene for the familial form of Alzheimer's disease (FAD) with the gene coding for amyloid precursor proteins

Gomiero, T., Bertelli, M., Deb, S., Weger, E., Marangoni, A., De Bastiani, E., Mantesso, U., & De Vreese, L.P.
A multicentre Italian validation study in aging adults with Down syndrome and other forms of intellectual disabilities: Dementia Screening Questionnaire for Individuals with Intellectual Disabilities.
Current Alzheimer Research, 2017, 14(7), 709-721.
doi:10.2174/1567205014666170117094757.
Abstract: The USA National Task Group (NTG) guidelines advocate the use of an adapted version of Dementia Screening Questionnaire for Individuals with Intellectual Disabilities (DSQIID) for dementia screening of individuals with Down syndrome (DS) and with other forms of ID (non-DS). In order to meet these guidelines, this study verifies the psychometric properties of an Italian version of the original DSQIID in a population composed of adults aged 40 years and over with DS and non-DS ID. Internal consistency, inter-rater and intra-rater reliabilities, structural validity, convergent validity and known group differences of DSQIID-I were assessed with 200 individuals with ID (mean of 55.2 years; range: 40-80 years) recruited from 15 different centers in Italy. Diagnosis of dementia was done according to IASSID diagnostic criteria and its degree of clinical certainty was defined according to Silverman et al.'s classification (2004). Cronbach's alpha for the DSQIID-I was 0.94. The ICCs for inter-rater and test-retest reliability were both 0.89. A Principal Component analysis revealed three domains, namely memory and confusion- related items, motor and functional disabilities, depression and apathy, which explained almost 40% of the overall variance. The total DSQIID-I score correlated significantly with DMR and differed significantly among those individuals (n = 34) with cognitive decline from those without (n = 166). Age, gender and severity of ID were unrelated to the DSQIID-I. The present study confirms the cross-cultural value of DSQIID which was proved to be a psychometrically valid and user-friendly observer-rated scale for dementia screening in adults with both DS and non-DS ID.

Gonçalves, A.S., Prado Mattar, G., Fidalgo Sant'Ana, L.V., Rebouças da Silva, M.F., Dongue Martinez, L., De-Paula, V.VJ., Lopes Carvalho, C., Martine, A., Mascarenhas Fonseca, L., & Vicente Forlenza, O.
Associations of gait and cognitive impairment and dementia in individuals with Down syndrome.
Alzheimer's & Dementia, 2023 Dec, 19(S23), Supplement: Public Health - Part 2,
e071602. https://alz-journals.onlinelibrary.wiley.com/doi/abs/10.1002/alz.071602
Abstract: Life expectancy of individuals with Down syndrome (DS) has increased considerably over the years. Atypical aging in Down syndrome (DS) is associated with neuropathological characteristics consistent with Alzheimer disease (AD). Significant gait alterations are associated with an increased risk of dementia for the general population. The aim of this study was to determine whether gait performance is associated with cognitive performance and dementia in adults with DS. This study included 66 individuals with DS over 20 years of age, divided into three diagnostic groups: stable cognition, prodromal dementia, and dementia. They were evaluated with the Performance-Oriented Mobility Assessment (POMA), Timed Up and Go test, and Cambridge Examination for Mental Disorders of Older People with Down's Syndrome and Others with Intellectual Disabilities (CAMDEX-DS), which includes the Cambridge Cognitive Examination (CAMCOG). Logistic regression model was used to assess the effects of gait and balance on the diagnosis (prodromal dementia and dementia versus stable cognition) adjusting for age, sex, body mass index (BMI), degree of intellectual disability and physical inactivity. The score on the POMA-Gait subscale score and body mass index (BMI) were found to be independent predictors of prodromal dementia and dementia (Poma-Gait OR = 0.67, 0.53-0.85 CI, p<0.001; and BMI OR = 1.12, 1.021.24 CI, p<0.001). A lower POMA-Total score indicates a higher fall risk. With the exception of perception, all cognitive domains correlated with the POMA total score (P<0.05). These data indicate that poor gait performance, as assessed by the POMA, as well as a high BMI, were associated with a diagnosis of prodromal dementia or dementia. In those individuals, applying the POMA could facilitate the early diagnosis of dementia, help identify fall risks, and promote the adoption of geriatric interventions focused on improving functional mobility. There is a need for longitudinal studies to characterize the true nature of the associations found, to validate gait instruments specifics to this population, and for future analyses in order to clarify the role that gait and balance play in cognitive decline and the development of dementia.

Gonçalves Gomes da Conceiçao, A.S., Assunçao Cecilio, A.P., Fidalgo Garcêz Sant'Ana, L.C., Fonseca, L., & Forlenza, O.V.
Associations of gait, cognitive imparment, dementia and obstructive sleep apnea in individuals with Down syndrome: preliminary findings
Paper presented at the 2024 AAIC Conference, Philadelphia, PA USA, Wednesday, July 31, 2024.
https://alz.confex.com/alz/2024/meetingapp.cgi/Paper/94955
Abstract: It is known that atipical aging in DS is related to a high risk of early dementia, with neuropathological charactheristics compatible with Alzheimer's disease (AD). Changes in functional mobility are expected throughout the aging process, with impairments in motor performance, involving balance and gait. Growing evidence suggests that sleep disruption may also accelerate the progression to symptomatic Alzheimer's disease (AD) in this population. Therefore, it is mandatory to assess sleep quality and diagnose sleep-disordered breathing, especially obstructive sleep apnea (OSA), associated with DS, to improve cognition and quality of life, preventing related comorbidities.
Method: We evaluated 66 individuals with DS (≥20 years), divided into 3 groups: stable cognition, prodromal dementia, and Alzheimer's disease. Each individual was evaluated with the Performance-Oriented Mobility Assessment (POMA) protocol, Polysomnography, and Cambridge Examination for Mental Disorders of Older People with Down's Syndrome and Others with Intellectual Disabilities interview adapted for adults with DS (CAMDEX-DS). The logistic regression model with the stepwise backward technique, with the initial model including age, gender, body mass index (BMI), degree of intellectual disability, and apnea-hypopnea index (AHI), showed that gait performance in POMA are independent factors associated with the diagnosis of control and dementia (POMA $p < 0.005$). All cognitive domains were correlated with OSA, but only orientation had a significant correlation (p-value 0.268). We did not find a difference in OSA scores among POMA. The POMA results indicate that those with AD have worse gait performance and a higher risk of falls relative to those with dementia and stable cognition. There was no evidence of correlation between gait speed and OSA, which may have been influenced by the sample size (n:17). Further longitudinal studies with larger samples are needed to confirm our results.

Goodman, C., Evans, C., Wilcock, J., Froggatt, K., Drennan, V., Sampson, E., Blanchard, M, Bissett, M., & Iliffe, S.
End of life care for community dwelling older people with dementia: an integrated review.
International Journal of Geriatric Psychiatry, 2010, Apr, 25(4), 329-337.
doi:10.1002/gps.2343.
Abstract: [Non-ID population study] Authors reviewed the evidence for end-of-life care for community dwelling older people with dementia (including those resident in care homes). An integrated review synthesised the qualitative and quantitative evidence on end-of-life care for community dwelling older people with dementia. English language studies that focused on prognostic indicators for end-of-life care, assessment, support/relief, respite and educational interventions for community dwelling older people with dementia were included. A user representative group informed decisions on the breadth of literature used. Each study selected was screened independently by two reviewers using a standardised check list. Sixty eight papers were included. Only 17% (12) exclusively concerned living and dying with dementia at home. Six studies included direct evidence from people with dementia. The studies grouped into four broad categories: Dementia care towards the end of life, palliative symptom management for people with dementia, predicting the approach of death for people with dementia and decision-making. The majority of studies were descriptive. The few studies that developed dementia specific tools to guide end of life care and outcome measures specific to improve comfort and communication, demonstrated what could be achieved, and how much more needs to be done. Research on end-of-life care for people with dementia has yet to develop interventions that address the particular challenges that dying with dementia poses. There is a need for investigation of interventions and outcome measures for providing end-of-life care in the settings where the majority of this

population live and die.

Greenwood, N., Pound, C., & Brearley, S.
'What happens when I can no longer care?' Informal carers' concerns about facing their own illness or death: a qualitative focus group study
BMJ Open, 2019 Sep 3;9(8):e030590. doi: 10.1136/bmjopen-2019-030590.
Abstract: Older informal carers play an increasingly important role in supporting others with long-term health conditions. This study aimed to explore in depth the perspectives of older carers (70+ years) supporting others with a variety of conditions and disabilities focusing on their thoughts and experiences about when they are unable to continue caring. The design was qualitative with four focus groups and the setting was Greater London in England. Participants were 28 older carers (70+ years) recruited from the voluntary sector participated in this study. Most were women and many were spouses caring for partners with age-related conditions such as dementia, arthritis and visual impairment. Nearly a third were parents of adult children with severe physical or cognitive disabilities. Findings were that the thematic analysis identified two main aspects for carers when contemplating the future-when they are unable to care in the short term or long term if they die or can no longer manage. Themes included the following: the impact of age, health conditions and relationships on future planning; anxiety about future care; carers' ambivalence and challenges in broaching the subject; interventions that might help older carers talk about and plan for the future of those they care for. Authors note that services need to be open to talking about this difficult topic. Our findings suggest that frank discussions about when older carers cannot care and having plans in place, whether these are financial or address other practical issues, makes it easier for all concerned. However, this issue is not easily broached and its timing and ways to access this support must be carefully and individually gauged. Future research with more diverse demographic groups is needed to improve understanding of these carers' perspectives. Research is also needed to develop interventions to support older carers to talk about and plan for the future.

Grohmann, D., Wellsted, D., & Mengoni, S.E.
Definition, assessment and management of frailty for people with intellectual disabilities: A scoping review.
Journal of Applied Research in Intellectual Disabilities, 2024 May, 37(3), e13219. doi: 10.1111/jar.13219.
Abstract: People with intellectual disabilities may experience frailty earlier than the general population. This scoping review aimed to investigate how frailty is defined, assessed, and managed in adults with an intellectual disability; factors associated with frailty; and the potential impact of COVID-19 on frailty identification and management. Databases were searched from January 2016 to July 2023 for studies that investigated frailty in individuals with intellectual disabilities. Twenty studies met the inclusion criteria. Frailty prevalence varied between 9% and 84%. Greater severity of intellectual disability, presence of **Down syndrome**, older age, polypharmacy, and group home living were associated with frailty. Multiagency working, trusted relationships and provision of evidence-based information may all be beneficial in frailty management. Frailty is common for people with intellectual disabilities and is best identified with measures specifically designed for this population. Future research should evaluate interventions to manage frailty and improve lives.

Gustavson, K.-H., Umn-Carlsson, Õ., & Sonnader, K.
A follow-up study of mortality, health conditions and associated disabilities of people with intellectual disabilities in a Swedish county.
Journal of Intellectual Disability Research, 2005 Dec., 49(12), 905-914, https://doi.org/10.1111/j.1365-2788.2005.00728.x
Abstract: In the planning of services and health care for individuals with intellectual disability (ID), information is needed on the special requirements for habilitation and medical service and associated disabilities. An unselected consecutive series of 82 adult persons with ID was studied. The medical examination consisted of the individual's health condition, associated impairments and disabilities. Medical and habilitation services and support were studied. The results indicated that 71% of the persons in the series had severe and 29% mild ID. Forty-seven per cent of the persons with severe ID and 35% of those with mild ID had one or more additional central nervous system (CNS) disabilities. Of the persons with ID, 99% had access to a family doctor and 84% attended regular health visits. Notably, half of persons were referred to a specialist examination as a consequence of their present medical examination. Half of the persons with mental health problems were previously undiagnosed and only a few of these had access to a psychiatrist. Our study clearly demonstrates the magnitude and importance of neurological and psychiatric impairments in ID. The findings suggest a strong need for multidisciplinary health service.

Hahn, H.E., & Cadogan, M.P.
Development and Evaluation of a staff training program on palliative care for persons with intellectual and developmental disabilities
Journal of Policy and Practice in Intellectual Disabilities, 2011, Mar, 8(1), 42–52
Abstract: Persons with intellectual and developmental disabilities (I/DD) face barriers and disparities at end of life. Among thesebarriers are limited educational opportunities and a paucity of targeted training materials on palliative care for staff who providetheir day-to-day care. This paper reports on a three-phase project undertaken to develop, implement, and evaluate a palliative carecurriculum and educational program that is responsive to the unique learning needs of staff providing services and supports forindividuals with I/DD living in long-term care settings. Participants' ratings of their levels of preparation and con?dence to providepalliative care improved from pretraining to posttraining. Posttraining use of materials and practice changes in palliative careoccurred.When training is developed in partnership with the staff who will use these training resources,it has the potential to sustainits use and to alter the care practices to address the palliative care needs of persons with I/DD.

Hamadelseed O., Elkhidir, I.H., & Skutella, T.
Psychosocial risk factors for Alzheimer's disease in patients with Down syndrome and their association with brain changes: A Narrative Review.
Neurol ogy and Therapy, 2022 Sep.,11(3), 931-953. doi: 10.1007/s40120-022-00361-9. Epub 2022 May 21.
Abstract: Several recent epidemiological studies attempted to identify risk factors for Alzheimer's disease. Age, family history, genetic factors (APOE genotype, trisomy 21), physical activity, and a low level of schooling are significant risk factors. In this review, we summarize the known psychosocial risk factors for the development of Alzheimer's disease in patients with Down syndrome and their association with neuroanatomical changes in the brains of people with Down syndrome. We completed a comprehensive review of the literature on PubMed, Google Scholar, and Web of Science about psychosocial risk factors for Alzheimer's disease, for Alzheimer's disease in Down syndrome, and Alzheimer's disease in Down syndrome and their association with neuroanatomical changes in the brains of people with Down syndrome. Alzheimer's disease causes early pathological changes in individuals with Down syndrome, especially in the hippocampus and corpus callosum. People with Down syndrome living with dementia showed reduced volumes of brain areas affected by Alzheimer's disease as the hippocampus and corpus callosum in association with cognitive decline. These changes occur with increasing age, and the presence or absence of psychosocial risk factors impacts the degree of cognitive function. Correlating Alzheimer's disease biomarkers in Down syndrome and cognitive function scores while considering the effect of psychosocial risk factors helps us identify the mechanisms leading to Alzheimer's disease at an early age. Also, this approach enables us to create more sensitive and relevant clinical, memory, and reasoning assessments for people with Down syndrome.

📖 **Hammond, B., & Beneditti, P.**
Perspectives of a care provider
In M.P. Janicki & A.J. Dalton (Eds.), Dementia, Aging, and Intellectual Disabilities, pp. 32-41
Philadelphia: Brunner-Mazel (1999)
Abstract: Book chapter that provides a descriptive chronology of a middle-aged woman with Down syndrome who, once diagnosed with Alzheimer disease, follows a classic course of decline and eventual debilitation and death. Staff of her residence chronicled the progression of her dementia and provide some insights into the care management practices used in providing for her care. The authors place the course of her disease in perspective and offer comments on the stresses and strains on agency resources. Suggestions are offered for agencies facing similar challenge in providing day to day care for adults with dementia.

Handen, B.L.
The search for biomarkers of Alzheimer's disease in Down syndrome
American Journal of Intellectual and Developmental Disabilities, 2020,125(2),

97-99. doi: 10.1352/1944-7558-125.2.97.
Abstract: Adults with Down syndrome are at high risk for Alzheimer's disease (AD), with most individuals developing clinical dementia by their late 60s. This increased risk for AD has been attributed, at least in part, to triplication and overexpression of the gene for amyloid precursor protein (APP) on chromosome 21, leading to elevated levels of amyloid ß peptides. This article offers a brief overview of our current knowledge of AD in the DS population. In addition, the NIA/NICHD-funded, multicenter longitudinal study of biomarkers of AD in adults with DS is explored.The Alzheimer's Biomarkers Consortium–Down Syndrome (ABC-DS) is a longitudinal study of Alzheimer Disease biomarkers in adults with Down syndrome supported by federal grants from the National Institute on Aging (NIA) and the Eunice Kennedy Shriver National Institute of Child Health and Human Development (NICHD). The primary goal of ABC-DS is to understand the factors that moderate the relationship between Aß, neurodegeneration and dementia in DS and biomarkers for those factors that could be critically important in the design of effective therapeutic trials for AD, not only in DS, but in the general population as well.

Handen, B., Clare, I., Laymon, C., Petersen, M.., Zaman, S.., O'Bryant, S., Minhas, D., Tudorascu, D., Brown, S., Christian, B., on behalf of the Alzheimer's Biomarker Consortium-Down Syndrome (ABC-DS).
Acute regression in Down syndrome.
Brain Sciences, 2021, 11, 1109. https://doi.org/10.3390/brainsci11081109
Abstract: Acute regression has been reported in some individuals with Down syndrome (DS), typically occurring between the teenage years and mid to late 20s. Characterized by sudden, and often unexplained, reductions in language skills, functional living skills and reduced psychomotor activity, some individuals have been incorrectly diagnosed with Alzheimer's disease (AD). This paper compares five individuals with DS who previously experienced acute regression with a matched group of 15 unaffected individuals with DS using a set of AD biomarkers. Results: While the sample was too small to conduct statistical analyses, findings suggest there are possible meaningful differences between the groups on proteomics biomarkers (e.g., NfL, total tau). Hippocampal, caudate and putamen volumes were slightly larger in the regression group, the opposite of what was hypothesized. A slightly lower amyloid load was found on the PET scans for the regression group, but no differences were noted on tau PET. Some proteomics biomarker findings suggest that individuals with DS who experience acute regression may be at increased risk for AD at an earlier age in comparison to unaffected adults with DS. However, due to the age of the group (mean 38 years), it may be too early to observe meaningful group differences on image-based biomarkers.

Handen, B.L., Lott, I.T., Christian, B.T., Schupf, N., OBryant, S., Mapstone, M., Fagan, A.M., Lee, J.H., Tudorascu, D., Wang, M.C., Head, E., Klunk, W., Ances, B., Lai, F., Zaman, S., Krinsky-McHale, S., Brickman, A.M., Rosas, H.D., Cohen, A., Andrews, H., Hartley, S., Silverman, W. & Alzheimer's Biomarker Consortium-Down Syndrome (ABC-DS).
The Alzheimer's Biomarker Consortium-Down Syndrome: Rationale and methodology.
Alzheimer's & Dementia (Amst). 2020 Aug 3;12(1):e12065. doi: 10.1002/dad2.12065.
Abstract: Adults with Down syndrome (DS) are at exceptionally high risk for Alzheimer's disease (AD), with virtually all individuals developing key neuropathological features by age 40. Identifying biomarkers of AD progression in DS can provide valuable insights into pathogenesis and suggest targets for disease modifying treatments. This article describes the development of a multi-center, longitudinal study of biomarkers of AD in DS. The protocol includes longitudinal examination of clinical, cognitive, blood and cerebrospinal fluid-based biomarkers, magnetic resonance imaging and positron emission tomography measures (at 16-month intervals), as well as genetic modifiers of AD risk and progression. Approximately 400 individuals will be enrolled in the study (more than 370 to date). The methodological approach from the administrative, clinical, neuroimaging, omics, neuropathology, and statistical cores is provided. This represents the largest U.S.-based, multi-site, biomarker initiative of AD in DS. Findings can inform other multidisciplinary networks studying AD in the general population.

Handen, B.L., Mapstone, M., Hartley, S., Andrews, H., Christian, B., Lee, J.H., Tudorascu, D., Hom, C., Ances, B.M., Zaman, S., Krinsky-McHale, S., Brickman, A.M., Rosas, H.D., Cohen, A., Petersen, M., O'Bryant, S., Harp, J.P., Schmitt, F., Ptomey, L., Burns, J., Lott, I.T., Lai, F., Silverman, W., Laymon, C., Head, E., & Alzheimer's Biomarker Consortium - Down Syndrome (ABC-DS).
The Alzheimer's Biomarker Consortium-Down Syndrome (ABC-DS): A 10-year report.
Alzheimer's & Dementia, 2025 May, 21(5), e70294. doi: 10.1002/alz.70294.
Abstract: Virtually all adults with Down syndrome (DS) will accumulate the neuropathologies associated with Alzheimer's disease (AD) by age 40, with the majority having a clinical dementia diagnosis by their middle 50s.This paper complements a 2020 publication describing the Alzheimer's Biomarker Consortium-Down Syndrome (ABC-DS) methodology by highlighting protocol changes since initial funding in 2015. It describes available clinical, neuropsychological, neuroimaging, and biofluid data and bio-specimen repository. Ten years of accomplishments are summarized. Over 500 adults with DS and 59 sibling controls have been enrolled since 2015 with nearly 800 follow-up visits. More than 900 magnetic resonance imaging (MRI), 800 amyloid positron emission tomography (PET), and 600 tau PET scans have been conducted; multiple omics data have been generated using over 1100 blood and 100 cerebrospinal fluid (CSF) samples. ABC-DS is the largest U.S.-based, multi-site (including the United Kingdom and Puerto Rico), longitudinal biomarker initiative to target adults with DS at risk for AD.

Hanney, M., Prasher, V., Williams, N., Jones, E.L., Aarsland, D., Corbett, A., Lawrence, D., Yu, L.M., Tyrer, S., Francis, P.T., Johnson, T., Bullock, R., Ballard, C & MEADOWS trial researchers.
Memantine for dementia in adults older than 40 years with Down's syndrome (MEADOWS): a randomised, double-blind, placebo-controlled trial.
Lancet, 2012 Feb 11, 379(9815), 528-36. doi: 10.1016/S0140-6736(11)61676-0.
Abstract: Prevalence of Alzheimer's disease in people with Down's syndrome is very high, and many such individuals who are older than 40 years have pathological changes characteristic of Alzheimer's disease. Evidence to support treatment with Alzheimer's drugs is inadequate, although memantine is beneficial in transgenic mice. We aimed to assess safety and efficacy of memantine on cognition and function in individuals with Down's syndrome. In our prospective randomised double-blind trial, we enrolled adults (>40 years) with karyotypic or clinically diagnosed Down's syndrome, with and without dementia, at four learning disability centres in the UK and Norway. We randomly allocated participants (1:1) to receive memantine or placebo for 52 weeks by use of a computer-generated sequence and a minimisation algorithm to ensure balanced allocation for five prognostic factors (sex, dementia, age group, total Down's syndrome attention, memory, and executive function scales [DAMES] score, and centre). The primary outcome was change in cognition and function, measured with DAMES scores and the adaptive behaviour scale (ABS) parts I and II. We analysed differences in DAMES and ABS scores between groups with analyses of covariance or quantile regression in all patients who completed the 52 week assessment and had available follow-up data. We randomly allocated 88 patients to receive memantine (72 [82%] had DAMES data and 75 [85%] had ABS data at 52 weeks) and 85 to receive placebo (74 [87%] and 73 [86%]). Both groups declined in cognition and function but rates did not differ between groups for any outcomes. After adjustment for baseline score, there were non-significant differences between groups of -4·1 (95% CI -13·1 to 4·8) in DAMES scores, -8·5 (-20·1 to 3·1) in ABS I scores, and 2·0 (-7·2 to 11·3) in ABS II scores, all in favour of controls. 10 (11%) of 88 participants in the memantine group and six (7%) of 85 controls had serious adverse events (p=0·33). Five participants in the memantine group and four controls died from serious adverse events (p=0·77). There is a striking absence of evidence about pharmacological treatment of cognitive impairment and dementia in people older than 40 years with Down's syndrome. Despite promising indications, memantine is not an effective treatment. Therapies that are effective for Alzheimer's disease are not necessarily effective in this group of patients.

Harisinghani, A., Cottrell, C., Donelan, K., Lam, A.D., Pulsifer, M., & Santoro, S.L.
Practicalities (and real-life experiences) of dementia in adults with Down syndrome.
American Journal of Medical Genetics Part C: Seminars in Medical Genetics, 2024, Jul 5, e32098. doi: 10.1002/ajmg.c.32098. Epub ahead of print. PMID: 38967370.
Abstract: Adults with down syndrome (DS) have a lifetime dementia risk in excess of 95%, with a median age of onset of 55 years, due to trisomy 21. Co-occurring Alzheimer's disease (AD) has increased morbidity and mortality, and it is now recommended to screen for AD in all adults with DS beginning at 40

years of age. In this manuscript, we present two clinical cases of adults with DS who developed AD summarizing their medical histories, presenting symptoms, path to diagnosis and psychosocial aspects of care collected from retrospective chart review with caregiver consent. These two cases were chosen due to their complexity and interwoven nature of the medical and psychosocial aspects, and highlight the complexity and nuance of caring for patients with DS and AD.

Harp, J.P., Koehl, L.M., Van Pelt, K.I., Hom, C.I., Doran, E., Hea\d, E., Lott, I.T., & Schmitt, F.A.
Cognitive and behavioral domains that reliably differentiate normal aging and dementia in Down syndrome
Brain Science, 2021 Aug 25, 11(9), 1128.
https://doi.org/10.3390/brainsci11091128
Abstract: : Primary care integration of Down syndrome (DS)-specific dementia screening is strongly advised. The current study employed principal components analysis (PCA) and classification and regression tree (CART) analyses to identify an abbreviated battery for dementia classification. Scale- and subscale-level scores from 141 participants (no dementia n = 68; probable Alzheimer's disease n = 73), for the Severe Impairment Battery (SIB), Dementia Scale for People with Learning Disabilities (DLD), and Vineland Adaptive Behavior Scales—Second Edition (Vineland-II) were analyzed. Two principle components (PC1, PC2) were identified with the odds of a probable dementia diagnosis increasing 2.54 times per PC1 unit increase and by 3.73 times per PC2 unit increase. CART analysis identified that the DLD sum of cognitive scores (SCS < 35 raw) and Vineland-II community subdomain (< 36 raw) scores best classified dementia. No significant difference in the PCA versus CART area under the curve (AUC) was noted (D(65.196) = -0.57683; p = 0.57; PCA AUC = 0.87; CART AUC = 0.91). The PCA sensitivity was 80% and specificity was 70%; CART was 100% and specificity was 81%. These results support an abbreviated dementia screening battery to identify at-risk individuals with DS in primary care settings to guide specialized diagnostic referral.

Harper, D.C., & Wadsworth, J.S.
Dementia and depression in elders with mental retardation: a pilot study.
Research in Developmental Disabilities, 1990, 11(2), 177-98. doi: 10.1016/0891-4222(90)90034-6.
Abstract: A preliminary investigation of cognitive decline and depressive symptomatology is presented with older adults who have mental retardation. A series of different assessment instruments are reviewed and tested in a pilot study. A review of dementia and depression with respect to elders with mental retardation is presented to place the study in perspective. Findings reveal decreasing cognitive ability is associated with higher rates of observed depression and reported behavioral problems. Trends suggested those more elderly displayed more depressive behaviors, psychotropic medication was a common treatment, and cognitive decline was associated with lower initial intellectual levels. Dementia and depression is a complicated symptom complex to identify in aging adults with mental retardation.

Harrison, S.L., Dyer, S.M., Laver, K.E., Milte, R.K., Fleming, R., Crotty, M.
Physical environmental designs in residential care to improve quality of life of older people.
Cochrane Database Systemic Reviews, 2022 Mar 7, 3(3), CD012892. doi: 10.1002/14651858.CD012892.pub2.
Abstract: The demand for residential aged care is increasing due to the ageing population. Optimizing the design or adapting the physical environment of residential aged care facilities has the potential to influence quality of life, mood and function. Authors assessed the effects of changes to the physical environment, which include alternative models of residential aged care such as a 'home-like' model of care (where residents live in small living units) on quality of life, behavior, mood and depression and function in older people living in residential aged care. Search methods included CENTRAL, MEDLINE, Embase, six other databases and two trial registries were searched on 11 February 2021. Reference lists and grey literature sources were also searched. Non-randomized trials, repeated measures or interrupted time series studies and controlled before-after studies with a comparison group were included. Interventions which had modified the physical design of a care home or built a care home with an alternative model of residential aged care (including design alterations) in order to enhance the environment to promote independence and well-being were included. Studies which examined quality of life or outcomes related to quality of life were included. Two reviewers independently assessed the abstracts identified in the search and the full texts of all retrieved studies. Two reviewers independently extracted data, assessed the risk of bias in each included study and evaluated the certainty of evidence according to GRADE criteria. Where possible, data were represented in forest plots and pooled. Twenty studies were included with 77,265 participants, although one large study included the majority of participants (n = 74,449). The main comparison was home-like models of care incorporating changes to the scale of the building which limit the capacity of the living units to smaller numbers of residents and encourage the participation of residents with domestic activities and a person-centered care approach, compared to traditional designs which may include larger-scale buildings with a larger number of residents, hospital-like features such as nurses' stations, traditional hierarchical organizational structures and design which prioritizes safety. Six controlled before-after studies compared the home-like model and the traditional environment (75,074 participants), but one controlled before-after study included 74,449 of the participants (estimated on weighting). It is uncertain whether home-like models improve health-related quality of life, behavior, mood and depression, function or serious adverse effects compared to traditional designs because the certainty of the evidence is very low. The certainty of the evidence was downgraded from low-certainty to very low-certainty for all outcomes due to very serious concerns due to risk of bias, and also serious concerns due to imprecision for outcomes with more than 400 participants. One controlled before-after study examined the effect of home-like models on quality of life. The author stated "No statistically significant differences were observed between the intervention and control groups." Three studies reported on global behavior (N = 257). One study found little or no difference in global behavior change at six months using the Neuropsychiatric Inventory where lower scores indicate fewer behavioral symptoms (mean difference (MD) -0.04 (95% confidence interval (CI) -0.13 to 0.04, n = 164)), and two additional studies (N = 93) examined global behavior, but these were unsuitable for determining a summary effect estimate. Two controlled before-after studies examined the effect of home-like models of care compared to traditional design on depression. After 18 months, one study (n = 242) reported an increase in the rate of depressive symptoms (rate ratio 1.15 (95% CI 1.02 to 1.29)), but the effect of home-like models of care on the probability of no depressive symptoms was uncertain (odds ratio 0.36 (95% CI 0.12 to 1.07)). One study (n = 164) reported little or no difference in depressive symptoms at six months using the Revised Memory and Behavior Problems Checklist where lower scores indicate fewer depressive symptoms (MD 0.01 (95% CI -0.12 to 0.14)). Four controlled before-after studies examined function. One study (n = 242) reported little or no difference in function over 18 months using the Activities of Daily Living long-form scale where lower scores indicate better function (MD -0.09 (95% CI -0.46 to 0.28)), and one study (n = 164) reported better function scores at six months using the Interview for the Deterioration of Daily Living activities in Dementia where lower scores indicate better function (MD -4.37 (95% CI -7.06 to -1.69)). Two additional studies measured function but could not be included in the quantitative analysis. One study examined serious adverse effects (physical restraints), and reported a slight reduction in the important outcome of physical restraint use in a home-like model of care compared to a traditional design (MD between the home-like model of care and traditional design -0.3% (95% CI -0.5% to -0.1%), estimate weighted n = 74,449 participants at enrolment). The remaining studies examined smaller design interventions including refurbishment without changes to the scale of the building, special care units for people with dementia, group living corridors compared to a non-corridor design, lighting interventions, dining area redesign and a garden vignette. Authors noted there is currently insufficient evidence on which to draw conclusions about the impact of physical environment design changes for older people living in residential aged care. Outcomes directly associated with the design of the built environment in a supported setting are difficult to isolate from other influences such as health changes of the residents, changes to care practices over time or different staff providing care across shifts. Cluster-randomized trials may be feasible for studies of refurbishment or specific design components within residential aged care. Studies which use a non-randomize design or cluster-randomized trials should consider approaches to reduce risk of bias to improve the certainty of evidence.

Hassiotis, A., Strydom, A., Allen, K., & Walker, Z.
A memory clinic for older people with intellectual disabilities
Aging & Mental Health, 2003, 7(6), 418-423. doi: 10.1080/13607860310001594664.
Abstract: Cognitive decline in older people with intellectual disabilities (ID) is often under-recognized. Following the publication of the National Service

Framework for Older People and the white paper Valuing People, older people with intellectual disabilities of all aetiologies should have access to a systematic assessment of their cognitive function in order to detect decline in cognition and adaptive skills and implement appropriate treatments as early as possible. The development of a memory clinic for older people with ID is described, including instruments used and characteristics of attendees. Such projects are in line with current UK government policies and can contribute to the improvement of standards of care and support research in this vulnerable group of people.

Harp, J.P., Koehl, L.M., Pelt, K.L.V., Hom, C.L., Doran, E., Head, E., Lott, I.T., & Schmitt, F.A.
Cognitive and behavioral domains that reliably differentiate normal aging and dementia in Down syndrome.
Brain Science, 2021 Aug 25, 11(9), 1128. doi: 10.3390/brainsci11091128.
Abstract: Primary care integration of Down syndrome (DS)-specific dementia screening is strongly advised. The current study employed principal components analysis (PCA) and classification and regression tree (CART) analyses to identify an abbreviated battery for dementia classification. Scale- and subscale-level scores from 141 participants (no dementia n = 68; probable Alzheimer's disease n = 73), for the Severe Impairment Battery (SIB), Dementia Scale for People with Learning Disabilities (DLD), and Vineland Adaptive Behavior Scales-Second Edition (Vineland-II) were analyzed. Two principle components (PC1, PC2) were identified with the odds of a probable dementia diagnosis increasing 2.54 times per PC1 unit increase and by 3.73 times per PC2 unit increase. CART analysis identified that the DLD sum of cognitive scores (SCS < 35 raw) and Vineland-II community subdomain (<36 raw) scores best classified dementia. No significant difference in the PCA versus CART area under the curve (AUC) was noted (D(65.196) = -0.57683; p = 0.57; PCA AUC = 0.87; CART AUC = 0.91). The PCA sensitivity was 80% and specificity was 70%; CART was 100% and specificity was 81%. **These results support an abbreviated dementia screening battery to identify at-risk individuals with DS** in primary care settings to guide specialized diagnostic referral.

Hartley, D., Blumenthal, T., Carrillo, M., DiPaolo, G., Esralew, L., Gardiner, K., Granholm, A.C., Iqbal, K., Krams, M., Lemere, C., Lott, I., Mobley, W., Ness, S., Nixon, R., Potter, H., Reeves, R., Sabbagh, M., Silverman, W., Tycko, B., Whitten, M., & Wisniewski, T.
Down syndrome and Alzheimer's disease: Common pathways, common goals.
Alzheimer's & Dementia - the Journal of the Alzheimer's Association, 2015 Jun, 11(6), 700-709. doi: 10.1016/j.jalz.2014.10.007. Epub 2014 Dec 12.
Abstract: In the United States, estimates indicate there are between 250,000 and 400,000 individuals with Down syndrome (DS), and nearly all will develop Alzheimer's disease (AD) pathology starting in their 30s. With the current lifespan being 55 to 60 years, approximately 70% will develop dementia, and if their life expectancy continues to increase, the number of individuals developing AD will concomitantly increase. Pathogenic and mechanistic links between DS and Alzheimer's prompted the Alzheimer's Association to partner with the Linda Crnic Institute for Down Syndrome and the Global Down Syndrome Foundation at a workshop of AD and DS experts to discuss similarities and differences, challenges, and future directions for this field. The workshop articulated a set of research priorities: (1) target identification and drug development, (2) clinical and pathological staging, (3) cognitive assessment and clinical trials, and (4) partnerships and collaborations with the ultimate goal to deliver effective disease-modifying treatments.

Hartley, S.L., Fleming, V., Schworer, E.K., Peven, J., Handen, B.L., Krinsky-McHale, S., Hom, C., Lee, L., Tudorascu, D.L., Laymon, C., Minhas, D., Luo, W., Cohen, A., Zaman, S., Ances, B.M., Mapstone, M., Head, E., Lai, F., Rosas, H.D., Klunk, W., Christian, B. & Alzheimer Biomarker Consortium-Down Syndrome.
Timing of Alzheimer's disease by intellectual disability level in Down syndrome.
Journal of Alzheimers Disease, 2023, 95(1),213-225. doi: 10.3233/JAD-230200.
Abstract: Trisomy 21 causes Down syndrome (DS) and is a recognized cause of early-onset Alzheimer's disease (AD). The current study sought to determine if premorbid intellectual disability level (ID) was associated with variability in age-trajectories of AD biomarkers and cognitive impairments. General linear mixed models compared the age-trajectory of the AD biomarkers PET Aß and tau and cognitive decline across premorbid ID levels (mild, moderate, and severe/profound), in models controlling trisomy type, APOE status, biological sex, and site. Analyses involved adults with DS from the Alzheimer's Biomarkers Consortium-Down Syndrome. Participants completed measures of memory, mental status, and visuospatial ability. Premorbid ID level was based on IQ or mental age scores prior to dementia concerns. PET was acquired using [11C] PiB for Aß, and [18F] AV-1451 for tau. Cognitive data was available for 361 participants with a mean age of 45.2 (SD = 9.92) and PET biomarker data was available for 154 participants. There was not a significant effect of premorbid ID level by age on cognitive outcomes. There was not a significant effect of premorbid ID by age on PET Aß or on tau PET. There was not a significant difference in age at time of study visit of those with mild cognitive impairment-DS or dementia by premorbid ID level. Findings provide robust evidence of a similar time course in AD trajectory across premorbid ID levels, laying the groundwork for the inclusion of individuals with DS with a variety of IQ levels in clinical AD trials.

Hartley, S.L., Handen, B.L., Devenny, D., Tudorascu, D. Piro-Gambetti, B., Zammit, M.B., Laymon, C.V., Klunk, W.E., Zaman,S., Cohen, A., & Christian, B.T.
Cognitive indicators of transition to preclinical and prodromal stages of Alzheimer's disease in Down syndrome.
Alzheimer's & Dementia: Diagnosis, Assessment & Disease Monitoring, 2020, 12, 1-10 e12096. https://doi.org/10.1002/dad2.12096
Abstract: There is a critical need to identify measures of cognitive functioning sensitive to early Alzheimer's disease (AD) pathophysiology in Down syndrome to advance clinical trial research in this at-risk population. The objective of the study was to longitudinally track performance on cognitive measures in relation to neocortical and striatal amyloid beta (Aß) in non-demented Down syndrome. The study included 118 non-demented adults with Down syndrome who participated in two to five points of data collection, spanning 1.5 to 8 years. Episodic memory, visual attention and executive functioning, and motor planning and coordination were assessed. Aß was measured via [C-11] Pittsburgh Compound-B (PiB) PET. PiB was associated with level and rate of decline in cognitive performance in episodic memory, visual attention, executive functioning, and visuospatial ability in models controlling for chronological age. The Cued Recall Test emerged as a promising indicator of transition from preclinical to prodromal AD.

Hartley, S.L., Fleming, V., Schworer, E.K., Peven, J., Handen, B.L., Krinsky-McHale, S., Hom, C., Lee, L., Tudorascu, D.L., Laymon, C., Minhas, D., Luo, W., Cohen, A., Zaman, S., Ances, B.M., Mapstone, M., Head, E., Lai, F., Rosas, H.D., Klunk, W., Christian, B., & Alzheimer Biomarker Consortium-Down Syndrome.
Timing of Alzheimer's disease by intellectual disability level in Down syndrome.
J*ournal of Alzheimer's Disease,* 2023, 95(1), 213-225. doi: 10.3233/JAD-230200.
https://content.iospress.com/articles/journal-of-alzheimers-disease/jad230200
Abstract: Trisomy 21 causes Down syndrome (DS) and is a recognized cause of early-onset Alzheimer's disease (AD). The current study sought to determine if premorbid intellectual disability level (ID) was associated with variability in age-trajectories of AD biomarkers and cognitive impairments. General linear mixed models compared the age-trajectory of the AD biomarkers PET Aß and tau and cognitive decline across premorbid ID levels (mild, moderate, and severe/profound), in models controlling trisomy type, APOE status, biological sex, and site. Analyses involved adults with DS from the Alzheimer's Biomarkers Consortium-Down Syndrome. Participants completed measures of memory, mental status, and visuospatial ability. Premorbid ID level was based on IQ or mental age scores prior to dementia concerns. PET was acquired using [11C] PiB for Aß, and [18F] AV-1451 for tau. Cognitive data was available for 361 participants with a mean age of 45.22 (SD = 9.92) and PET biomarker data was available for 154 participants. There was not a significant effect of premorbid ID level by age on cognitive outcomes. There was not a significant effect of premorbid ID by age on PET Aß or on tau PET. There was not a significant difference in age at time of study visit of those with mild cognitive impairment-DS or dementia by premorbid ID level. Findings provide robust evidence of a similar time course in AD trajectory across premorbid ID levels, laying the groundwork for the inclusion of individuals with DS with a variety of IQ levels in clinical AD trials.

Hatzidimitriadou, E., & Alisoun Milne, A.
Planning ahead: Meeting the needs of older people with intellectual disabilities in the United Kingdom
Dementia, 2005, 4(3), 341-359. https://doi.org/10.1177/1471301205055027

Abstract: Despite the acknowledged increase in the number of older people with intellectual disabilities (ID) in the UK, the age-related health and social care needs of this population have yet to be fully understood and addressed. Although there is some evidence of positive development, the current picture of service provision is characterized by fragmentation and limited choice of resources and specialist care. Policy aims are variably met and inconsistently applied. Research suggests that service planning is often incoherent, that many older people with ID and their carers receive poor quality non-specialist care and that staff are inadequately trained to manage the often multiple and complex needs of this user group. There is a considerable co-joined service development and research challenge in this emerging field. If older people with ID and their carers are to receive quality provision, a coherent and well-funded service planning system is required which is underpinned by articulated agency partnerships, informed by good practice developments in the fields of ID, gerontology and dementia care, and linked to evidence about effective models of care and services. The incorporation of the perspectives of users and carers in the planning process is an essential pre-requisite as is a commitment to the development of effective support across the life course of all individuals with ID.

Hawkes, T., Luff, T., & Gee, S. B.
Supporting person-centred dementia care for people with intellectual disabilities using the person, interaction, environment programme.
New Zealand Journal of Occupational Therapy, 2019, 66(1), 12-18.
https://go.gale.com/ps/i.do?p=AONE&u=googlescholar&id=GALE|A619219947&v=2.1&it=r&sid=AONE&asid=6fba816b
Abstract: This small scale study explored person-centered care for people with a dual diagnosis of intellectual disability and dementia. The goal was to identify areas of improvement in relation to personalized care, interactions with staff, and the environment. The Person, Interaction, Environment program was used in staff training. Data was gathered using a cyclical process of observation, feedback and goal setting. The findings revealed ongoing improvement in the quality of residents' communication and care. The study outcomes suggest that the Person, Interaction and Environment program is a practical tool that can make a positive difference to the experience of intellectually disabled people living with dementia.

He, P., Chen, G., Wang, Z., Guo., C., Li, N., Yun, C., & Zheng, X.
Adults with intellectual disabilities in China: comorbid psychiatric disorder and its association with health service utilisation
Journal of Intellectual Disability Research, 2018 Feb, 62(2), 16-114. doi: 10.1111/jir.12451. Epub 2017 Nov 26.
Abstract: Adults with intellectual disabilities (ID) often have multiple comorbidities. Psychiatric disorders in this population have been poorly studied in developing countries. We aimed to investigate the prevalence of psychiatric disorders in adults with ID and whether comorbid psychiatric disorders were associated with health service utilization. We obtained data from the Second National Sample Survey on Disability, conducted in 31 provinces of China and selected a subsample of 13 631 adults aged 18 years and above with ID. ID were defined by intelligence quotient score under 70, deficits in two or more adaptive behaviours and age of onset under 18 years. Psychiatric disorders were identified according to the International Statistical Classification of Diseases, Tenth Revision. Logistic regressions were used for data analyses. The prevalence of psychiatric disorders in adults with ID was 16.7%. The most prevalent type of psychiatric disorder was **dementia.** Older adults, females, being minorities, urban residents, being literate, low-income groups and having severe ID, were associated with elevated risk of psychiatric disorder among adults with ID. Compared with individuals without psychiatric disorders, those with comorbid psychiatric disorders were more likely to use medical service and less likely to use rehabilitation service. The prevalence of psychiatric disorder in adults with ID was strikingly higher than that in the general population. Health service utilisation among Chinese adults with ID remained a big challenge. There is a possibility of diagnostic overshadowing by local clinicians, which may have resulted in overdiagnosis of dementia and underdiagnosis of common mental disorders. This study informs further investigations regarding common mental disorders among people with ID and has implications for public health strategies and health policies to meet health service need for this population.

Head, E., Lott, I.T., Wilcock, D.M., & Lemere, C.A.
Aging in Down syndrome and the development of Alzheimer's disease neuropathology
Current Alzheimer Research, 2016, 13(1),18-29.
doi:10.2174/1567205012666151020114607.
Abstract: Chromosome 21, triplicated in Down Syndrome, contains several genes that are thought to play a critical role in the development of AD neuropathology. The overexpression of the gene for the amyloid precursor protein (APP), on chromosome 21, leads to early onset beta-amyloid (Aß) plaques in DS. In addition to Aß accumulation, middle-aged people with DS develop neurofibrillary tangles, cerebrovascular pathology, white matter pathology, oxidative damage, neuroinflammation and neuron loss. There is also evidence of potential compensatory responses in DS that benefit the brain and delay the onset of dementia after there is sufficient neuropathology for a diagnosis of AD. This review describes some of the existing literature and also highlights gaps in our knowledge regarding AD neuropathology in DS. It will be critical in the future to develop networked brain banks with standardized collection procedures to fully characterize the regional and temporal pathological events associated with aging in DS. As more information is acquired regarding AD evolution in DS, there will be opportunities to develop interventions that are age-appropriate to delay AD in DS

Head, E., Powell, D., Gold, B.T., Schmitt, F.A..
Alzheimer's disease in Down syndrome
European Journal of Neurodegenerative Diseases, 2012, 1(3): 353–364.
https://www.ncbi.nlm.nih.gov/pmc/articles/PMC4184282/
Abstract: A key challenge to adults with Down syndrome (DS) as they age is an increased risk for cognitive decline, dementia, and Alzheimer disease (AD). In DS persons ranging from 40-49 years of age, 5.7-55% may be clinically demented and between 50-59 years, dementia prevalence ranges from 4-55% (reviewed in [1]). Despite the wide ranges reported for dementia prevalence, a consistent feature of aging in DS is the progressive accumulation of AD brain pathologies. By the age of 40 years, virtually all have sufficient senile plaques and neurofibrillary tangles for a neuropathological diagnosis of AD [2]. Thus, there is dissociation between the age of onset of AD neuropathology (40 years) and increasing signs of clinical dementia. We discuss the hypothesis that frontal impairments are a critical factor affecting cognitive function and are associated with white matter (WM) and AD neuropathology. While these may be an early sign of conversion to dementia, we also review several other clinical comorbidities that may also contribute to dementia onset.

Hellen, C.R.
Alzheimer's disease - activity-focused care (2nd Ed.)
Boston: Butterworth-Heinemann (1998)
436 pp.
Abstract: A 13-chapter text that provide voluminous information on developing and provision of activities for persons affected by Alzheimer's disease and related dementias - with application to persons with intellectual disabilities. Written from a practitioner viewpoint, it is designed to promote an individual's cognitive, physical and psychosocial well-being. It includes forms and profiles for use by program personnel, presents a holistic intervention program, features content on refocusing activities for physically combative or violent situations. Contains chapters on communication, daily living care activities, aiding at mealtimes, facilitating physical wellness (mobility and exercise), addressing dementia induced behaviors, creating meaningful activities for daily life, and aiding in terminal care, among others.

Heller, T., Scott, H.M., & Janicki, M.P.
Caregiving, intellectual disability, and dementia: Report of the Summit Workgroup on Caregiving and Intellectual and Developmental Disabilities
Alzheimers & Dementia - Translational Research & Clinical Interventions,. 2018, 4, 272-282.
Published online 2018 Jul 10. doi: 10.1016/j.trci.2018.06.002
Abstract: A specially commissioned working group produced a report on caregiving, intellectual and developmental disabilities (IDDs), and dementia for the National Institutes of Health-located National Research Summit on Care, Services, and Supports for Persons with Dementia and Their Caregivers. Experts in caregiving, dementia, and IDDs examined the current state of research, policy, and practice related to caregiving and supports; identified the similarities and dissimilarities between IDD-related care and services and the general population affected by dementia; and considered how these findings might contribute to the conversation on developing a dementia care research and services development agenda. Five major areas related to programs and caregiving were assessed: (1)

challenges of dementia; (2) family caregiving interventions; (3) supportive care settings; (4) effects of diversity; and (5) bridging service networks of aging and disability. Recommendations included increasing supports for caregivers of adults with IDDs and dementia; increasing research on community living settings and including caregivers of persons with IDDs in dementia research; acknowledging cultural values and practice diversity in caregiving; increasing screening for dementia and raising awareness; and leveraging integration of aging and disability networks.

Herron, D.L., & Priest, H.M.
Support workers' knowledge about dementia: A vignette study.
Advances in Mental Health and Intellectual Disabilities, 2013, 7(1), 27-39.
https://doi.org/10.1108/20441281311294675
Abstract: It is widely acknowledged that people with intellectual disabilities are highly likely to experience mental health problems, but that support workers' knowledge and skill in this area is sometimes lacking. There is little research explicitly exploring knowledge about the mental health of older people with intellectual disabilities and the purpose of this paper is to attempt to fill this gap. In total, 14 support workers completed a questionnaire in which three vignettes presented progressively worsening indicators of dementia in an older person with intellectual disabilities. Participants explained what they thought was happening and what action they would take. Data were analysed using Braun and Clarke's framework. Few participants had undertaken any mental health training, and only one in relation to older people. They were generally poor at judging early and intermediate indicators of dementia, but were able to identify more overt later signs. However, they believed these advanced indicators to be the onset of dementia. Nonetheless, they would generally take appropriate action, such as observation and referral. Abuse was often considered as a causal factor. The most significant implication is the need for training in the mental health needs of older people and in particular, the general and specific indicators and expected trajectory of dementia in this population.

Herron, D.L., Priest, H.M., & Read, S.
Supporting people with an intellectual disability and dementia: A constructivist grounded theory study exploring care providers' views and experiences in the UK.
Journal of Applied Research in Intellectual Disabilities, 2020, 33(6), 1405-1417.
https://doi: 10.1111/jar.12768
Abstract: There is a need to better understand the experiences and support needs of paid and family carers of people with an intellectual disability and dementia, and the role of Intellectual Disability Dementia Care Pathways (IDDCPs). This study explored the experiences of carers, and IDDCPs and other support structures within those experiences. A constructivist grounded theory methodology was implemented. Data were obtained through 23 semi-structured interviews with two family carers, eight paid carers and eight healthcare professionals. The study's theory produced five interrelated categories: Impact of Dementia, Challenging the Diagnosis Process, Continuum of Support, Continuity and Continuum of Understanding. Findings have demonstrated the importance of planning and supporting carers' holistic needs; the role of an IDDCP in the post-diagnostic support (or lack of it) for carers; and the importance of a timely diagnosis of dementia. An important implication of these findings is the need for local services to develop inclusive specialized IDDCPs. Having IDDCP healthcare professionals, who have expertise in both intellectual disability and dementia, allows support, advice and information to be tailored to both the person's intellectual disability and dementia. This may also help to address some of the challenges people with an intellectual disability and dementia, and their carers, experience when encountering generic services without the appropriate expertise in intellectual disability care. Knowledge and understanding of both intellectual disability and dementia are essential to initiating the diagnosis process and providing person-centred support. Carers need both a theoretical understanding of dementia and how to address dementia-related changes through dementia training courses, and hands on experience where they could apply this training. Based on their work, the authors recommend the (1) need for local health services to develop inclusive specialized IDDCPs, (2) development of a comprehensive, accessible training package, informed by these findings and the concept of person-centred care, (3) need for organizations and services to address the reactive culture sometimes seen, and implement procedures for effective dementia care planning, and (4) need to ensure a reliable, timely diagnosis and early dementia care planning, through reactive assessments, proactive baselining and screening, and associated guidance.

Heston, L.L.
Down's syndrome and Alzheimer's dementia: defining an association
Psychiatric Developments,1984, Winter, 2(4), 287-294.
https://pubmed.ncbi.nlm.nih.gov/6241313/
Abstract: The typical neuropathological features of Alzheimer's disease, plaques and tangles, appear in virtually all patients with Down's Syndrome after the age of 40. Clinically, changes in cognitive performance and behavior appear to correlate with these neuropathological changes, although a satisfactory operational definition of dementia in a context of mental retardation is not available. It is unknown whether the cholinergic losses in the nucleus basalis, which are a feature of early onset Alzheimer's disease, also occur late in Down's syndrome. Two family studies have supported a greater than expected incidence of Down's cases among relatives of probands dying with Alzheimer-type dementia, but the association is not strong. It is noteworthy that in both studies, phenotypically normal carriers of the rare 15/21 translocation had severe early onset dementia, although this translocation is responsible for less than 0.4 per cent of Down's cases. An increased incidence of dementia among carriers of the more common 14/21 translocation has not been reported. In any case, it is proposed that a gene product originating from the long arm of chromosome 21 (21q) is necessary for Alzheimer-type pathology, since a segregating gene could not be responsible for the 100 per cent incidence of these changes among 21q trisomics.

Higgins, L., & Mansell, J.
Quality of life in group homes and older persons' homes.
British Journal of Learning Disabilities, 2009, 37(3), 207–212.
https://doi.org/10.1111/j.1468-3156.2009.00550.x
Abstract: Older people with intellectual disabilities sometimes live in older people's homes rather than homes for people with intellectual disabilities. Little is known about their quality of life in these homes. A non-equivalent comparison group design was used to compare the quality of life of 59 people in three groups; older people without an intellectual disability living in older people's homes (n = 20), older people with an intellectual disability living in older people's homes (n = 19) and older people with an intellectual disability living in intellectual disability homes (n = 20). Data were collected on participant characteristics, adaptive behavior and three aspects of quality of life; community involvement, participation in domestic living and choice making. The three groups were comparable in terms of gender, ethnicity and additional impairments but the older people without an intellectual disability were older and had more adaptive skills than the other groups. Older people with an intellectual disability experienced better quality of life outcomes in terms of participation in meaningful activity and community access when they lived in intellectual disability homes compared with older people's homes. It was not possible to achieve reliability on the measure of choice-making. This study provides some evidence to suggest that older people with an intellectual disability may be best served in intellectual disability homes rather than older people homes and that it is an area of research which needs further exploration.

Hill-Smith, A.J., & Hollins, S.C.
Mortality of parents of people with intellectual disabilities.
Journal of Applied Research in Intellectual Disabilities, 2002 Mar., 15(1), 18-27.
https://doi.org/10.1046/j.1360-2322.2002.00100.x
Abstract: The experience of caring for a son or daughter with an intellectual disability has long been recognized as stressful. The long-term health costs for parents of people with intellectual disability have attracted some recent research attention, but mortality has not been studied. The present authors examined mortality as measured by the standardized mortality ratio (SMR), and cause of death for parents of people with intellectual disability, identified through an intellectual disability register in Merton, South London. Although there was a trend for lower SMRs particularly for mothers, SMRs were not significantly different from unity. Subgroups of parents whose child was cared for predominantly in an institution, or in the family home were analysed and similarly showed no significant difference from unity. The same applied to cause of death analyses. These findings offer some reassurance to parents of people with intellectual disability. There is an urgent need for further research in this area.

Hillerstrom, H., Fisher, R., Janicki, M.P., Chicoine, B., Christian, B.T., Esbensen, A., Esralew, L., Fortea, J., Hartley, S., Hassenstab, J., Keller, S..M., Krinsky-McHale, S., Lai, F., Levin, J., McCarron, M., McDade, E.,

Rebillat, A.S., Rosas, H.D., Silverman, W., Strydom, A., Zaman, S.H., & Zetterberg, H.
Adapting prescribing criteria for amyloid-targeted antibodies for adults with Down syndrome.
Alzheimer's & Dementia: The Journal of the Alzheimer's Association, 2024 May, 20(5), 3649-3656. https://doi:10.1002/alz.13778. Epub 2024 Mar 13..
Abstract: Prior authorization criteria for FDA-approved immunotherapeutics, among the class of anti-amyloid monoclonal antibodies (mAbs), established by state drug formulary committees, are tailored for adults with late-onset Alzheimer's disease. This overlooks adults with Down syndrome (DS), who often experience dementia at a younger age and with different diagnostic assessment outcomes. This exclusion may deny DS adults access to potential disease-modifying treatments. To address this issue, an international expert panel convened to establish adaptations of prescribing criteria suitable for DS patients and parameters for access to CMS registries. The panel proposed mitigating disparities by modifying CMS and payer criteria to account for younger onset age, using alternative language and assessment instruments validated for cognitive decline in the DS population. The panel also recommended enhancing prescribing clinicians' diagnostic capabilities for DS and initiated awareness-raising activities within healthcare organizations. These efforts facilitated discussions with federal officials aimed at achieving equity in access to anti-amyloid immunotherapeutics, with implications for national authorities worldwide evaluating these and other new AD disease-modifying therapeutics.

Hithersay, R., Baksh, R.A., Startin, C.M., Wijeratne, P., Hamburg, S., Carter, B., The LonDownS Consortium, & Strydom, A.
Optimal age and outcome measures for Alzheimer's disease prevention trials in people with Down syndrome.
Alzheimer's & Dementia, 2021 April, 17(4), 595-604.
https://doi.org/10.1002/alz.12222
Abstract: People with Down syndrome (DS) typically develop Alzheimer's disease (AD) neuropathology before age 40, but a lack of outcome measures and longitudinal data have impeded their inclusion in randomized controlled trials (RCTs). A cohort study was employed in which event-based and dose-response Emax models were fitted to longitudinal cognitive data, to stage AD and determine the earliest ages of decline. Results informed sample size estimations for hypothetical RCTs of disease-modifying treatments that reduced decline by 35% or 75%.. Seventy-five percent of participants progressed or remained stable in the AD staging model; effect sizes varied by age group and tests. Varied treatment effects could be detected with 50-200 people per arm when using sensitive cognitive outcome measures and targeting recruitment to ages 36 to 45 years. Efficient RCTs of AD preventative treatments can be conducted in the DS population using sensitive outcome measures to monitor early decline. Dose-response models could help tailor future RCTs.

Hithersay, R., Hamburg, S., Knight, B., & Strydom, A.
Cognitive decline and dementia in Down syndrome.
Current Opinion in Psychiatry, 2017, Mar; 30(2), 102-107.
https://doi:10.1097/YCO.0000000000000307.
Abstract: Alzheimer's disease is most likely universal in older individuals with Down syndrome, due to having three copies of the amyloid precursor protein gene, resulting in amyloid-beta plaque deposition. Down syndrome is an important population in which to consider clinical trials of treatments to prevent or delay the development of dementia. However, assessment of subtler cognitive changes is challenging due to the presence of intellectual disability. Recent research confirmed that older adults with Down syndrome often present with cognitive decline: more than 80% may experience dementia by age 65 years. Efforts have been made to improve and validate neuropsychological assessment and to describe the relationship with comorbidities such as epilepsy and haemorrhagic stroke. There have also been advances in biomarkers such as neuroimaging using amyloid PET. Clinical trials of treatments, particularly in the presymptomatic phase of Alzheimer's disease, are important to consider in individuals with Down syndrome given their high dementia burden, and may also serve as proof of concept for other forms of Alzheimer's disease. However, further work is required to improve outcome measures and better understand the biomarkers of progression of disorder and their relationship with symptom development during the presymptomatic period.

Hithersay, R,, Startin, C.M., Hamburg, S., Mok, K.Y., Hardy, J., Fisher, E.M.C., Tybulewicz, V.L.J., Nizetic, D., & Strydom, A.
Association of dementia with mortality among adults with Down syndrome older than 35 years
JAMA Neurology, 2019, Feb 1, 76(2), 152-160.
https://doi:10.1001/jamaneurol.2018.3616.
Abstract: This work quantifies the fatal burden of dementia associated with Alzheimer disease in individuals with Down syndrome (DS). To explore the association of dementia associated with Alzheimer disease with mortality and examine factors associated with dementia in adults with DS. Prospective longitudinal study in a community setting in England. Data collection began March 29, 2012. Cases were censored on December 13, 2017. The potential sample consisted of all adults 36 years and older from the London Down Syndrome Consortium cohort with 2 data times and dementia status recorded (N = 300); 6 withdrew from study, 28 were lost to follow-up, and 55 had a single data collection point at time of analysis. The final sample consisted of 211 participants, with 503.92 person-years' follow-up. Dementia status, age, sex, APOE genotype, level of intellectual disability, health variables, and living situation. Crude mortality rates, time to death, and time to dementia diagnosis with proportional hazards of predictors. Of the 211 participants, 96 were women (45.5%) and 66 (31.3%) had a clinical dementia diagnosis. Twenty-seven participants (11 female; mean age at death, 56.74 years) died during the study period. Seventy percent had dementia. Crude mortality rates for individuals with dementia (1191.85 deaths per 10 000 person-years; 95% CI, 1168.49-1215.21) were 5 times higher than for those without (232.22 deaths per 10 000 person-years; 95% CI, 227.67-236.77). For those with dementia, APOE e4 carriers had a 7-fold increased risk of death (hazard ratio [HR], 6.91; 95% CI, 1.756-27.195). For those without dementia, epilepsy with onset after age 36 years was associated with mortality (HR, 9.66; 95% CI, 1.59-58.56). APOE e4 carriers (HR, 4.91; 95% CI, 2.53-9.56), adults with early-onset epilepsy (HR, 3.61; 95% CI, 1.12-11.60), multiple health comorbidities (HR, 1.956; 95% CI, 1.087-3.519), and those living with family (HR, 2.14; 95% CI, 1.08-4.20) received significantly earlier dementia diagnoses. Dementia was associated with mortality in 70% of older adults with DS. APOE e4 carriers and/or people with multiple comorbid health conditions were at increased risk of dementia and death, highlighting the need for good health care. For those who died without a dementia diagnosis, late-onset epilepsy was the only significant factor associated with death, raising questions about potentially undiagnosed dementia cases in this group

Hobson, B., Webb, D., Sprague, L., Grizzell, M., Hawkins, C., & Benbow, S.M.
Establishing a database for proactive screening of adults with Down's syndrome: when services work together
Advances in Mental Health and Intellectual Disabilities, 6(2), 99-105.
https://doi.org/10.1108/20441281211208464
Abstract: This paper describes a service improvement project with two aims: to identify and screen all adults with Down syndrome aged over 30 years in a defined locality using a standardised instrument to establish functional baselines; and to set up a database to facilitate early diagnosis of dementia in this population. An assistant psychologist used a standardised instrument to screen participants who were identified through contact with health, social, and third sector, and housing services. Eligible people were identified and screened using an informant-based measure. Three groups were identified: group 1 showed no significant change; group 2 showed significant change but no signs of dementia; and group 3 showed significant change plus signs of dementia. People with suspected dementia were referred on for further investigation/ assessment and supportive services. Terminology is important in engaging families in a screening

project, as is the opportunity to provide information. A proactive screening project can be established by employing working partnerships between intellectual disability and older adult services to aid diagnosis. Adults with Down syndrome aged over 30 years in a defined locality can be identified through contact with health, social, and third sector, and housing services. Those identified can be screened using a standardised instrument and a database of screening results established in order to establish baselines against which future re-screening can be conducted. Partnership working between older adult mental health services and intellectual disability services can improve the diagnostic service to adults with Down syndrome.

Hoekman, J., & Maaskant, M.A.
Comparison of instruments for the diagnosis of dementia in individuals with intellectual disability
Journal of Intellectual & Developmental Disability, 2002, 27(4), 296-309. https://doi.org/10.1080/1366825021000029339
Abstract: The authors describe the agreement among the results (dementia/no dementia) of three instruments used for the potential diagnosis of dementia in persons with intellectual disability. The instruments are: the Dementia Questionnaire for Mentally Retarded Persons (DMR), the Checklist with Symptoms of Dementia (CLD) and the Delayed Match-to-Sample Test (DMTS). The results were compared with the expert opinion of a physician/educational specialist/psychologist. The participants were 329 adults affiliated with centres for people with intellectual disability in The Netherlands. It was found that the agreement among the three instruments was low (kappa < 0.5). The agreement between the expert opinion and the results of the tests was also found to be low. It was concluded that the instruments do not mutually agree upon which of the adults can be regarded as dementing or not dementing and they also provide inconsistent agreement with the expert opinion when dementia is present. It was further concluded that it is not advisable to use a single instrument when attempting to diagnose dementia in people with intellectual disability.

Holingue, C., Wise, E., Caoili, A., Klein, A., Kalb, L.G., & Beasley, J.
Screening for dementia among adults with intellectual disability: Outcomes from a pilot study
Journal of Mental Health Research in Intellectual Disabilities, 2022, 15(1), 20-36. https://doi.org/10.1080/19315864.2021.1965270
Abstract: Screening for dementia among individuals with ID is important to identify individuals in need of care and support. The objective of this pilot study was to identify obstacles associated with screening and assessment of dementia among older adults with ID in a crisis-prone population. The NTG's Early Detection Screen for Dementia (EDSD) was administered to eligible enrollees ages 50 years and older within the START (Systemic, Therapeutic, Assessment, Resources, and Treatment) program. Focus groups were carried out to understand the barriers to screening and diagnosis of dementia. Of the 95 eligible enrollees, 63 participants had dementia screening tools completed. Obstacles identified through focus groups included difficulty differentiating changes from baseline function, competing priorities in this crisis-prone population, lack of access to providers, and an unclear understanding of the benefit or purpose of screening among some caregivers. START coordinators noted that the EDSD provided a helpful way to collect information and document changes in the enrollee's functioning. The EDSD may be helpful for capturing potential dementia-associated changes over time in crisis-prone adults with ID, though obstacles remain to the access of further evaluation for dementia.

Holland, A.J.
Ageing and its consequences for people with Down's syndrome
Fact Sheet Series - Learning about intellectual disabilities and health
Accessed 24 August 2004 at
http:www.intellectualdisability.info/lifestages/ds_ageing.htm
Down Syndrome Association (UK) and the Department of Mental Health & Learning Disability at St. George's Hospital Medical School, University of London.
9 pp.
Abstract: Fact sheet outlines the evidence which suggests that ageing and t he problems of old age are particularly relevant to people with Down syndrome as some of these age-related problems develop earlier in life than would normally be the case. Topics covered include: aging and the brain, aging and dementia, behavioral features of dementia in people with Down syndrome, apparent decline in later life - cases to consider, difficulties in detecting dementia in people with intellectual disabilities, differential diagnosis - which conditions mimic dementia, common causes of decline in later life in people with Down syndrome, genetic mechanisms, treatment, supporting the individual, and the future.

Holland, A.J., Karlinsky, H. & Berg, J.M.
Alzheimer's disease in persons with Down syndrome: Diagnostic and management considerations
In J.M. Berg, H. Karlinsky, A.J. Holland (Eds.), Alzheimer's Disease, Down Syndrome, and Their Relationship, pp. 96-114
Oxford: Oxford University Press (1993)
Abstract: Book chapter that examines the implications of Alzheimer's disease for adults with Down syndrome, including assessment and diagnosis and specialty service provision. Authors note that assigning a tenable diagnosis of Alzheimer disease requires careful and comprehensive data assembly, including medical history, clinical examination, neuropsychological assessment and laboratory investigations. Once the diagnosis is established, effective ongoing management should focus on supporting not only the affected individual (including advocacy for his or her rights) but also the family and professional carers. During the course of the illness various medical, psychiatric and psychological interventions can be helpful as can changes in the environment. A wide range of services for persons with Down syndrome who develop Alzheimer's disease makes it possible for affected individuals, despite deterioration, to remain in the family home or in community residential settings. Authors proffer some general suggestions for services and adaptations.

Holland, A.J., Hon, J., Huppert, F.A., & Stevens, F.
Incidence and course of dementia in people with Down's syndrome: findings from a population-based study.
Journal of Intellectual Disability Research, 2000, 44(2), 138-146. https://doi: 10.1046/j.1365-2788.2000.00263.x
Abstract: The prevalence rate of Alzheimer's disease (AD) in people with Down's syndrome (DS) increases significantly with age. However, the nature of the early clinical presentation, course and incidence rates of dementia are uncertain. The aims of the present study were to investigate the characteristics of age-related clinical changes and incidence rates for dementia in a population-based sample of people with DS aged 30 years and older at the age of risk for dementia. A modified version of the Cambridge Examination for Mental Disorders of the Elderly informant interview was used to determine the extent and nature of changes in memory, personality, general mental functioning and daily living skill 18 months after a similar assessment. At the time of the first assessment, the initial changes reported were predominately in behavior and personality. At the second assessment, overall estimated incidence rates for frontal-like dementia were high (0.24), mainly in the younger groups, with incidence rates of AD, meeting both ICD-10 and DSM-IV criteria, of 0.04 predominately in the older groups. The present authors have hypothesized that the observed personality changes and the high estimated incidence rates of frontal-like dementia in the younger groups may indicate that functions served by the frontal lobes are the first to be compromised with the progressive development of Alzheimer-like neuropathology in people with DS.

Holst, G., Johansson, M., & Ahlstrom, G.
Signs in people with intellectual disabilities: Interviews with managers and staff on the identification process of dementia.
Healthcare (Basel)), 2018, 6(3), 103. https:// doi.org/10.3390/healthcare6030103
Abstract: The life expectancy of people with intellectual disabilities (ID) has steadily increased, which has been accompanied by an increased risk of

dementia. Staff and managers are key resources for safety diagnosis since they deliver information about people with ID behavior every day. The aim of the present study was to explore the identification process employed by staff and managers to detect signs of suspected dementia in people with an ID within intellectual disability services (ID-services). Twenty managers and 24 staff within an ID-service were interviewed and qualitative latent content analysis was applied. A model consisting of three themes on three levels of resources for the identification process of signs of suspected dementia emerged from the analysis. On the first level was the time and continuity in the care relationship, which is crucial for identifying and responding to changes in cognitive ability that indicate dementia. On the second level, the staff identify deficiencies in their own knowledge, seek support from colleagues and managers within their workplace and, on the third level, outside their workplace. Staff and managers expressed a need for early and continuous guidance and education from specialists in dementia and primary healthcare. This finding indicates an urgent need for intervention research and digital support for staff in dementia care. Only when these resources are used will it be possible for staff to identify, and react to, dementia symptoms in a safe and knowledge-based way, initiate examination, design well-adapted care, and, if necessary, consider moving the person to adapted housing. A close collaboration with specialists within dementia care may contribute to a good working environment for staff with skilled guidance and support, continuous education, and the initiation of intervention research concerning the best accommodation form and digital support for people with an ID and staff.

Hom, C.
Neuropsychological subtypes of incident mild cognitive impairment in Down syndrome.
AAIC 2020, Poster presentation, July 29, 2020.
Alzheimer's & Dementia, 2020, 16 (S6), https://doi.org/10.1002/alz.043299
Abstract: Past attempts to characterize the earliest cognitive changes as individuals with Down Syndrome (DS) transition from cognitively stable to mild cognitive impairment (MCI) have been equivocal (Garcia-Alba et al., 2019; Lautarescu et al., 2017). Difficulties identifying MCI in this population are complicated by variability in pre-morbid cognitive abilities, the use of neuropsychological tests that were created for the neurotypical population, and participants scoring at floor on the baseline assessment (Krinsky-McHale and Silverman, 2013). We examined data from 151 individuals with Down Syndrome (M age=50.25, SD age=6.94). Their pre-morbid level of intellectual impairment ranged from mild to severe. All participants received comprehensive evaluations. Following data collection, the clinical status of each participant was rated at consensus review that considered performance on a core neuropsychological test battery and the clinical data for each participant. Data from the non-demented and MCI groups are examined: Cognitive Stable (N=107, 70.9%) and MCI-DS (N=44, 29.1%). The full battery consists of 27 subtests that were hypothesized a priori to measure five cognitive domains: language, memory, executive function, visuospatial reasoning, and motor coordination. Factor analysis revealed 7 principal components that maximally discriminated between test scores in older adults with DS who have not reached clinical AD status: (1) general intelligence (2) sensorimotor, (3) memory, (4) language comprehension and expression, (5) executive function/speed, (6) attention/language expression, and (7) visuomotor. Cluster analysis for the MCI group produced 3 distinct groups: (1) dysexecutive (n=4), (2) dysnomic/visuospatial impaired (n=28), and (3) amnestic/motor impaired (n=12). Author concludes that the neuropsychological battery assesses 7 distinct cognitive functions in older adults with DS. It can also capture cognitive decline, as we were able to empirically identify three distinct neuropsychological subtypes of MCI: amnestic/visuomotor impaired, dysexecutive, and dysnomic. These subtypes are generally consistent with those that have been found within the neurotypical population (Edmonds et al., 2015; Dick et al., 2016), strengthening the evidence that AD has a similar course in the DS population and late onset AD.

Hom, C.L., Kirby, K.A., Ricks-Oddie, J., Keator, D.B., Krinsky-McHale, S.J., Pulsifer, M.B,. Rosas, H.D., Lai, F., Schupf, N., Lott, I.T., & Silverman, W.
Cognitive function during the prodromal stage of Alzheimer's disease in Down syndrome: Comparing models.
Brain Sciences, 2021, 11(9), 1220. https://doi.org/10.3390/brainsci11091220
Abstract: Accurate identification of the prodromal stage of Alzheimer's disease (AD), known as mild cognitive impairment (MCI), in adults with Down syndrome (MCI-DS) has been challenging because there are no established diagnostic criteria that can be applied for people with lifelong intellectual disabilities (ID). As such, the sequence of cognitive decline in adults with DS has been difficult to ascertain, and it is possible that domain constructs characterizing cognitive function in neurotypical adults do not generalize to this high-risk population. The present study examined associations among multiple measures of cognitive function in adults with DS, either prior to or during the prodromal stage of AD to determine, through multiple statistical techniques, the measures that reflected the same underlying domains of processing. Participants included 144 adults with DS 40-82 years of age, all enrolled in a larger, multidisciplinary study examining biomarkers of AD in adults with DS. All participants had mild or moderate lifelong intellectual disabilities. Overall AD-related clinical status was rated for each individual during a personalized consensus conference that considered performance as well as health status, with 103 participants considered cognitively stable (CS) and 41 to have MCI-DS. Analyses of 17 variables derived from 10 tests of cognition indicated that performance reflected three underlying factors: language/executive function, memory, and visuomotor. All three domain composite scores significantly predicted MCI-DS status. Based upon path modeling, the language/executive function composite score was the most affected by prodromal AD. However, based upon structural equation modeling, tests assessing the latent construct of memory were the most impacted, followed by those assessing visuomotor, and then those assessing language/executive function. Our study provides clear evidence that cognitive functioning in older adults with DS can be characterized at the cognitive domain level, but the statistical methods selected and the inclusion or exclusion of certain covariates may lead to different conclusions. Best practice requires investigators to understand the internal structure of their variables and to provide evidence that their variables assess their intended constructs.

Horovitz, M., Kozlowski, A. M., & Matson, J. L.
Compliance training in an adult with dementia of the Alzheimer's type and Down syndrome.
Clinical Case Studies, 2010, 9(2), 95-105.
https://doi.org/10.1177/1534650109357784
Abstract: The authors describe the treatment of noncompliance and public stripping in a 53-year-old man with Down syndrome and dementia. Based on a review of the relevant literature and a comprehensive functional behavioral analysis, an intervention procedure utilizing contingent reinforcement of compliance was conducted. Compliance with requests to allow assistance getting dressed was reinforced with a combination of verbal praise, physical contact, and edible reinforcers. Noncompliance had decreased by 85.22% at follow-up. Factors responsible for intervention success and implications for clinicians are discussed.

Horvath, S,. Garagnani, P., Bacalini, M.G., Pirazzini, C., Salvioli, S., Davide, G.,Di Blasio, A.M., Giuliani, C.,Tung, S., Vinters, H.V., & Franceschi, C.
Accelerated epigenetic aging in Down syndrome
Aging Cell, 2015, 1–5, eprint. doi: 10.1111/acel.12325
Abstract: Down syndrome (DS) entails an increased risk of many chronic diseases that are typically associated with older age. The clinical manifestations of accelerated aging suggest that trisomy 21 increases the biological age of tissues, but molecular evidence for this hypothesis has been sparse. Here, we utilize a quantitative molecular marker of aging (known as the epigenetic clock) to demonstrate that trisomy 21 significantly increases the age of blood and brain tissue (on average by 6.6 years, $P = 7.0\ 3\ 10^{-14}$).

Hughes, M., Hanna, K., Wiles, A., Taylor, E., & Giebel, C.
The experiences of caring for someone with dementia and a learning disability: A qualitative systematic review.
Dementia (London), 2024 Jul, 23(5), 817-849.
https://doi:10.1177/14713012231225797.
Abstract: The life expectancy of people with a learning disability is increasing and with this comes a greater risk of developing dementia. Dementia poses new challenges for both family and formal learning disability carers as they try to support dementia's progressive nature and quality of life for their care recipient. This qualitative systematic review explores the evidence base of family and formal carers' experiences and needs of caring for someone with both a learning disability and dementia. Six electronic databases (PubMed, PsycINFO, Cochrane Library, Prospero, Scopus, CINAHL), were searched in May 2022, utilizing a predefined search strategy. Thirteen papers fulfilled inclusion criteria and were included in the review. Thematic synthesis was used to explore and synthesize the qualitative findings of the studies. Four conceptual themes were identified following analysis: Knowledge and skills, Accessing support, Repercussions of dementia for carers, Influences of continuity of caring role. There are significant training and educational needs for all carers who support the dual diagnosis of dementia and learning disability. Differences between family and formal carers relate to the organizational support and process available to formal carers. Parity across services combined with sufficiently trained carers may support dementia diagnosis and improve quality of care provided. Further research is needed to address environmental, and economic barriers carers face to facilitate ageing in place for their care recipients.

Humphreys, L., Bigby, C., & Iacono, T.
Dimensions of group home culture as predictors of quality of life outcomes
Journal of Applied Research in Intellectual Disabilities, 2020 Nov, 33(6), 1284-1295. doi: 10.1111/jar.12748. Epub 2020 May 27.
Abstract: Research has shown that there is variability in quality of life (QOL) outcomes for people with intellectual disabilities who live in group homes. The aim was to examine dimensions of group home culture as predictors of QOL outcomes. The Group Home Culture Scale (GHCS) was used to measure staff perceptions of culture in 23 group homes. QOL data were available from 98 people with intellectual disabilities. Multilevel modelling was used to examine the associations between the GHCS subscales and four QOL-dependent variables. Of the GHCS subscales, Effective Team Leadership and Alignment of Staff with Organizational Values significantly predicted residents' engagement in activities. Supporting Well-Being significantly predicted residents' community involvement. None of the GHCS subscales significantly predicted domestic participation and choice making. The findings suggest that strategies to improve Effective Team Leadership and Supporting Well-Being dimensions of culture may contribute to enhancing certain QOL outcomes.

Huxley, A., Van-Schaik, P., & Witts, P.
A comparison of challenging behavior in an adult group with Down's syndrome and dementia compared with an adult Down's syndrome group without dementia.
British Journal of Learning Disabilities, 2005, 33(4), 188-193.
https://doi.org/10.1111/j.1468-3156.2005.00323.x
Abstract: This study investigated the frequency and severity of challenging behavior in adults with Down's syndrome with and without signs of dementia. Care staff were interviewed using the Aberrant Behavior Checklist-Community version (M.G. Aman & N.N. Singh, Slosson, East Aurora, NY, 1994), to investigate the frequency and severity of challenging behavior. Individuals' 'dementia status' was assessed by using the Dementia Scale for Down's Syndrome (Gedye Research and Consulting, Vancouver, 1995). The results showed that the dementia group displayed more frequent and severe forms of challenging behavior than the nondementia group. The difference in reported levels of challenging behavior of both groups with the general learning disabilities population was not considered to be clinically significant and levels fell predominantly within the 'normal range'. The findings of this study suggest that frequent and severe forms of challenging behavior in adults with Down's syndrome is more likely to be a behavioral symptom associated with the onset of a dementing illness and not due to normal aging alone.

Iacono, T., Bigby, C., Carling-Jenkins, R., & Torr, J.
Taking each day as it comes: Staff experiences of supporting people with Down syndrome and Alzheimer's disease in group homes.
Journal of Intellectual Disability Research, 2013; 58(6). DOI:10.1111/jir.12048.
Abstract: Disability staff are being increasingly required to support adults with Down syndrome who develop Alzheimer's disease. They have little understanding of the nature of care required, and may lack input from aged care and dementia services, which lack knowledge of intellectual disability. The aim of this study was to report on the experiences of disability staff in group homes supporting residents with Down syndrome and Alzheimer's disease in relation to their under understanding of what was happening to these residents, their responses to them, and how they felt about their support role. Disability support staff for nine adults with Down syndrome who had a diagnosis of Alzheimer's disease were interviewed twice, over intervals of 6-9 months. Interviews were transcribed and analyzed for themes. Authors foiund that three key themes emerged - (I) struggling to understand change, (ii) taking each day as it comes, and (iii) he's got a disability and that's our job. Staff had only limited understanding of how Alzheimer's disease impacted the adults with Down syndrome, their responses to changes were ad hoc, and they used strategies on a trial and error basis. They were committed to providing care, but at the risk of rejecting input and support. The need for collaboration across disability, and aged and disability care was evident to facilitate aging-in-place and planned care transitions.

Idris, M., Saini, F., Pape, S. E., Baksh, R. A., Cahart, M.-S., & Strydom, A.
Common mental health disorders and cognitive decline in a longitudinal Down syndrome cohort.
BJPsych Open, 2023), 9(6), e206. doi:10.1192/bjo.2023.590
Abstract: Down syndrome is the most common genetic cause of intellectual disability and Alzheimer's disease. In the general population, common mental disorders (CMDs), including anxiety, depression and obsessive-compulsive disorder, are linked to cognitive decline and higher risk for dementia. It is not known how CMDs affect longer-term cognitive outcomes in Down syndrome, and there is often diagnostic uncertainty in older people with Down syndrome and psychiatric comorbidity. The authors studied the influence of CMDs on cognitive ability and whether they are related longitudinally to development of clinical signs of Alzheimer's disease in Down syndrome. We followed 115 individuals with Down syndrome, 27 of whom were diagnosed with a CMD, over approximately 3 years. Changes in cognitive and behavioural outcomes between baseline and follow-up assessment were analysed, with comparisons made between those with and without a comorbid CMD. Age, gender, apolipoprotein E status and level of intellectual disability were included as covariates. No significant association between presence of a CMD and poorer performance on cognitive tasks or informant-rated decline over time was observed ($P > 0.05$).
Our results suggest that a diagnosis of a CMD does not have a significant negative effect on long-term cognitive or behavioural outcomes in individuals with Down syndrome. In individuals with stable or treated CMD, subsequent cognitive decline is likely indicative of Alzheimer's disease rather than a consequence of mental disorder.

Ilacqua, A., Benedict, J., Shoben, A., Skotkp, B.G., Mathews, T. Benson, B., & Allain, D.C.
Alzheimer's disease development in adults with Down syndrome: Caregivers' perspectives
American Journal of Medical Genetics, 2020, 182(1), 104-114. doi: 10.1002/ajmg.a.61390.
Abstract: Research about Alzheimer's disease (AD) in individuals with Down syndrome (DS) has predominantly focused on the underlying genetics and neuropathology. Few studies have addressed how AD risk impacts caregivers of

adults with DS. This study aimed to explore the perceived impact of AD development in adults with DS on caregivers by assessing caregiver knowledge, concerns, effect on personal life, and resource utilization via a 40-question (maximum) online survey. Survey distribution by four DS organizations and two DS clinics resulted in 89 caregiver respondents. Only 28 caregivers correctly answered all three AD knowledge questions. Caregivers gave an average AD concern rating of 5.30 (moderately concerned) and an average impact of possible diagnosis rating of 6.28 (very strong impact), which had a significant negative correlation with the age of the adult with DS (p = .009). Only 33% of caregivers reported utilization of resources to gain more information about the AD and DS association, with low levels of perceived usefulness. Our data reveal caregivers' misconceptions about AD development in DS, underutilization of available resources, and substantial concerns and perceived impacts surrounding a possible AD diagnosis. This study lays the foundation for how the medical community can better serve caregivers of aging adults with DS.

Innes, A., McCabe, L., & Watchman, K.
Caring for older people with an intellectual disability
Maturitas, 2012 Aug, 72(4), 286-295. doi: 10.1016/j.maturitas.2012.05.008.
Abstract: This review critically evaluates the available research literature on aging among people with an intellectual disability. 42 papers meeting the review inclusion criteria are presented under three themes: studies with a service user perspective (13), studies of carers of older people with ID (14) and studies of service provision for older people with ID (15). User view specific findings relate to concerns about accommodation; experiences of services; and perceptions of aging; with a common underlying finding from all user focused themes that of unmet need. Carer specific findings relate to fear of the future; experiences of older carers; and planning for the future. Services themes reflect the debate over specialist or generalist services as people age; accommodation; retirement from day services; and staff training. Overall this review reveals a lack of robust research evidence concerning the lives of older people with ID and a need for more research that directly engages with older people with ID and their carers.

Ismail, Z., Ghahremani, M., Amlish Munir, M. et al.
A longitudinal study of late-life psychosis and incident dementia and the potential effects of race and cognition.
Nature Mental Health, 2023, 1, 273-283.
https://doi.org/10.1038/s44220-023-00043-x
Abstract: Later-life psychotic symptoms are meaningful and are associated with adverse outcomes. Psychosis is an important domain in mild behavioural impairment (MBI), a syndrome that incorporates later-life emergent and persistent neuropsychiatric symptoms (NPS) in dementia-free individuals into dementia prognostication. However, MBI-psychosis-associated risk and its interaction with race has not been well quantified. Here we determined risk of incident dementia in dementia-free participants with MBI-psychosis and effect modification by race as an important factor in assessing the risk of psychosis. Data for participants with normal cognition (NC) or mild cognitive impairment (MCI) from the National Alzheimer Coordinating Centre were used. Participants with neurodevelopmental, neurological and/or longstanding psychiatric disorders were excluded. MBI-psychosis was defined by persistence of delusions and hallucinations across two consecutive visits. Kaplan-Meier curves of ten-year dementia-free survival were generated for MBI-psychosis versus no NPS before dementia diagnosis. Cox proportional hazard models were implemented to assess relative incidence rates, adjusted for cognitive status, age, sex, education, race and APOE-ε4 status. Interaction terms were included for relevant demographic variables. Similar secondary analyses utilized MBI-no-psychosis as reference. The sample consisted of 3,704 no-NPS (age = 72.8 ± 9.9; 62.7% female; 13.4% MCI) and 66 MBI-psychosis (age = 75.2 ± 9.8; 53% female; 72.7% MCI) participants. For MBI-psychosis, in reference to no NPS, the hazard ratio (HR) for incident dementia was 3.76 (CI: 2.53-5.58, p < 0.001), while for conventionally captured psychosis, the HR was 1.92 (CI: 1.58-2.33, p < 0.001). Interaction analyses revealed that in NC, those with MBI-psychosis had a 9.96-fold greater incidence of dementia than those with no NPS (CI: 3.65-27.22, p < 0.001). In MCI, the MBI-psychosis-associated dementia incidence was 3.38-fold greater than no-NPS (CI: 2.22-5.15, p < 0.001). Furthermore, MBI-psychosis-associated dementia incidence in Black participants was 7.44-fold greater than no NPS (CI: 3.54-15.65, p < 0.001), while in white participants, it was 3.18-fold greater (CI: 1.94-5.2, p < 0.001). In a secondary analysis, compared with MBI-no-psychosis (n = 2,260), MBI-psychosis had a 2.47-fold greater incidence of dementia (CI: 1.69-3.59, p < 0.001). Although psychosis is an infrequently endorsed MBI domain, when present it is associated with substantial risk for dementia. HRs differed between cognitive strata, and these differences were significantly greater when MBI-psychosis emerged in NC as opposed to MCI, emphasizing the importance of cognitive assessment at the time of symptom emergence. In addition, the relationship between MBI-psychosis and incident dementia was stronger in Black participants than in white participants. The emergence of persistent psychotic symptoms in older adults is clinically meaningful, and MBI-psychosis identifies a high-risk group for precision medicine approaches to dementia prevention.

Iulita MF, Bejanin A, Vilaplana E, Carmona-Iragui M, Benejam B, Videla L, Barroeta I, Fernández S, Altuna M, Pegueroles J, Montal V, Valldeneu S, Giménez S, González-Ortiz S, Torres S, El Bounasri El Bennadi S, Padilla C, Rozalem Aranha M, Estellés T, Illán-Gala I, Belbin O, Valle-Tamayo N, Camacho V, Blessing E, Osorio RS, Videla S, Lehmann S, Holland AJ, Zetterberg H, Blennow K, Alcolea D, Clarimón J, Zaman SH, Blesa R, Lleó A, Fortea J.
Association of biological sex with clinical outcomes and biomarkers of Alzheimer's disease in adults with Down syndrome.
Brain Communications, 2023 Mar 17;5(2):fcad074. doi: 10.1093/braincomms/fcad074. PMID: 37056479; PMCID: PMC10088472.
Abstract: The study of sex differences in Alzheimer's disease is increasingly recognized as a key priority in research and clinical development. People with Down syndrome represent the largest population with a genetic link to Alzheimer's disease (>90% in the 7th decade). Yet, sex differences in Alzheimer's disease manifestations have not been fully investigated in these individuals, who are key candidates for preventive clinical trials. In this double-centre, cross-sectional study of 628 adults with Down syndrome [46% female, 44.4 (34.6; 50.7) years], we compared Alzheimer's disease prevalence, as well as cognitive outcomes and AT(N) biomarkers across age and sex. Participants were recruited from a population-based health plan in Barcelona, Spain, and from a convenience sample recruited via services for people with intellectual disabilities in England and Scotland. They underwent assessment with the Cambridge Cognitive Examination for Older Adults with Down Syndrome, modified cued recall test and determinations of brain amyloidosis (CSF amyloid-ß 42 / 40 and amyloid-PET), tau pathology (CSF and plasma phosphorylated-tau181) and neurodegeneration biomarkers (CSF and plasma neurofilament light, total-tau, fluorodeoxyglucose-PET and MRI). We used within-group locally estimated scatterplot smoothing models to compare the trajectory of biomarker changes with age in females versus males, as well as by apolipoprotein e4 carriership. Our work revealed similar prevalence, age at diagnosis and Cambridge Cognitive Examination for Older Adults with Down Syndrome scores by sex, but males showed lower modified cued recall test scores from age 45 compared with females. AT(N) biomarkers were comparable in males and females. When considering apolipoprotein e4, female e4 carriers showed a 3-year earlier age at diagnosis compared with female non-carriers (50.5 versus 53.2 years, P = 0.01). This difference was not seen in males (52.2 versus 52.5 years, P = 0.76). Our exploratory analyses considering sex, apolipoprotein e4 and biomarkers showed that female e4 carriers tended to exhibit lower CSF amyloid-ß 42/amyloid-ß 40 ratios and lower hippocampal volume compared with females without this allele, in line with the clinical difference. This work showed that biological sex did not influence clinical and biomarker profiles of Alzheimer's disease in adults with Down syndrome. Consideration of apolipoprotein e4 haplotype, particularly in females, may be important for clinical research and clinical trials that consider this population.

Accounting for, reporting and publishing sex-stratified data, even when no sex differences are found, is central to helping advance precision medicine.

Iulita, M.F., Garzón Chavez, D., Klitgaard Christensen, M., Valle Tamayo, N., Plana-Ripoll, O., Rasmussen, S.A., Roqué Figuls, M., Alcolea, D., Videla, L., Barroeta, I., Benejam, B., Altuna, M., Padilla, C., Pegueroles, J., Fernandez, S., Belbin, O., Carmona-Iragui, M., Blesa, R., Lleó, A., Bejanin, A., & Fortea, J.
Association of Alzheimer disease with life expectancy in people with Down syndrome.
JAMA Network Open. 2022 May 2;5(5):e2212910. doi: 10.1001/jamanetworkopen.2022.12910.
Abstract: People with Down syndrome have a high risk of developing Alzheimer disease dementia. However, penetrance and age at onset are considered variable, and the association of this disease with life expectancy remains unclear because of underreporting in death certificates. The authors assessed whether the variability in symptom onset of Alzheimer disease in Down syndrome is similar to autosomal dominant Alzheimer disease and to assess its association with mortality. This study combines a meta-analysis with the assessment of mortality data from US death certificates (n = 77 347 case records with an International Classification of Diseases code for Down syndrome between 1968 to 2019; 37 900 [49%] female) and from a longitudinal cohort study (n = 889 individuals; 46% female; 3.2 [2.1] years of follow-up) from the Down Alzheimer Barcelona Neuroimaging Initiative (DABNI). A meta-analysis was conducted to investigate the age at onset, age at death, and duration of Alzheimer disease dementia in Down syndrome. PubMed/Medline, Embase, Web of Science, and CINAHL were searched for research reports, and OpenGray was used for gray literature. Studies with data about the age at onset or diagnosis, age at death, and disease duration were included. Pooled estimates with corresponding 95% CIs were calculated using random-effects meta-analysis. The variability in disease onset was compared with that of autosomal dominant Alzheimer disease. Based on these estimates, a hypothetical distribution of age at death was constructed, assuming fully penetrant Alzheimer disease. These results were compared with real-world mortality data. The authors found that in this meta-analysis, the estimate of age at onset was 53.8 years (95% CI, 53.1-54.5 years; n = 2695); the estimate of age at death, 58.4 years (95% CI, 57.2-59.7 years; n = 324); and the estimate of disease duration, 4.6 years (95% CI, 3.7-5.5 years; n = 226). Coefficients of variation and 95% prediction intervals of age at onset were comparable with those reported in autosomal dominant Alzheimer disease. US mortality data revealed an increase in life expectancy in Down syndrome (median [IQR], 1 [0.3-16] years in 1968 to 57 [49-61] years in 2019), but with clear ceiling effects in the highest percentiles of age at death in the last decades (90th percentile: 1990, age 63 years; 2019, age 65 years). The mortality data matched the limits projected by a distribution assuming fully penetrant Alzheimer disease in up to 80% of deaths (corresponding to the highest percentiles). This contrasts with dementia mentioned in 30% of death certificates but agrees with the mortality data in DABNI (78.9%). Important racial disparities persisted in 2019, being more pronounced in the lower percentiles (10th percentile: Black individuals, 1 year; White individuals, 30 years) than in the higher percentiles (90th percentile: Black individuals, 64 years; White individuals, 66 years). These findings suggest that the mortality data and the consistent age at onset were compatible with fully penetrant Alzheimer disease. Lifespan in persons with Down syndrome will not increase until disease-modifying treatments for Alzheimer disease are available

Jacinto, M., Matos, R., Gomes, B., Caseiro, A., Antunes, R., Monteiro, D., Ferreira, J.P., & Campos, M.J,
Physical fitness variables, general health, dementia and quality of life in individuals with intellectual and developmental disabilities: A cross-sectional study
Healthcare, 2023, 11(19), 2688; https://doi.org/10.3390/healthcare11192688
Abstract: The average life expectancy of individuals with intellectual and developmental disabilities (IDDs) is increasing. However, living more years does not mean living better, leading to the need for research on comorbidities associated with the aging process. Associated with this process are the physical characteristics most prevalent in an individual with IDD: low levels of all physical capacities, the accumulation of central fat, hyperglycemia, dyslipidemia, and hypertension, variables considered to be some of the main risk factors of the onset of metabolic and cardiovascular diseases, and variables that can negatively impact quality of life (QoL). Therefore, the aim of this study is to evaluate a sample of 21 institutionalized adults with IDD (42.81 ± 10.99 years old) in terms of their anthropometric characteristics, body composition, general health status, functional capacity, neuromuscular capacity, and dementia/cognitive function, and the possible associations with QoL. All assessments were performed in the laboratory of the Faculty of Sport Sciences and Physical Education—University of Coimbra. Participants, in the present study, have low levels of physical fitness and high metabolic and cardiovascular markets, which need to be improved. On the other hand, functional and neuromuscular ability seems to be associated with QoL ($p = 0.05$). This study highlights the role of primary and secondary care providers in diagnosis, prevention, and supporting individuals with IDDs to promote QoL.

Jack, C.R. Jr, Andrews, J.S., Beach, T.G., Buracchio, T., Dunn, B., Graf, A., Hansson, O., Ho, C., Jagust, W., McDade, E., Molinuevo, J.L., Okonkwo, O.C., Pani, L., Rafii, M.S., Scheltens, P., Siemers, E., Snyder, H.M., Sperling, R., Teunissen, C.E.,& Carrillo, M.C.
Revised criteria for diagnosis and staging of Alzheimer's disease: Alzheimer's Association Workgroup
Alzheimer's & Dementia, 2024; 1-27. First published: 27 June 2024 /https://doi.org/10.1002 alz.13859
Abstract: The National Institute on Aging and the Alzheimer's Association convened three separate work groups in 2011 and single work groups in 2012 and 2018 to create recommendations for the diagnosis and characterization of Alzheimer's disease (AD). The present document updates the 2018 research framework in response to several recent developments. Defining diseases biologically, rather than based on syndromic presentation, has long been standard in many areas of medicine (e.g., oncology), and is becoming a unifying concept common to all neurodegenerative diseases, not just AD. The present document is consistent with this principle. Our intent is to present objective criteria for diagnosis and staging AD, incorporating recent advances in biomarkers, to serve as a bridge between research and clinical care. These criteria are not intended to provide step-by-step clinical practice guidelines for clinical workflow or specific treatment protocols, but rather serve as general principles to inform diagnosis and staging of AD that reflect current science.

Jacobs, J., Schwartz, A., McDougle, C.J., & Skotko, B.G.
Rapid clinical deterioration in an individual with Down syndrome
American Journal of Medical Genetics, Part A 9999A:1-4 DOI 10.1002/ajmg.a.37674
Abstract: A small percentage of adolescents and young adults with Down syndrome experience a rapid and unexplained deterioration in cognitive, adaptive, and behavioral functioning. Currently, there is no standardized work-up available to evaluate these patients or treat them. Their decline typically involves intellectual deterioration, a loss of skills of daily living, and prominent behavioral changes. Certain cases follow significant life events such as completion of secondary school with friends who proceed on to college or employment beyond the individual with DS. Others develop this condition seemingly unprovoked. Increased attention in the medical community to clinical deterioration in adolescents and young adults with Down syndrome could provide a framework for improved diagnosis, evaluation, and treatment. This report presents a young adult male with Down syndrome who experienced severe and unexplained clinical deterioration, highlighting specific challenges in the systematic evaluation and treatment of these patients.

Jacobs, P., Watchman, K., Wilkinson, H., Hoyle, L., & McGenily, L.
Experiences of people with intellectual disability and dementia: A systematic review.
Journal of Applied Research in Intellectual Diabilities, 2022 Dec 23. doi: 10.1111/jar.13063. Epub ahead of print. PMID: 36562340.
Abstract: Dementia disproportionately affects people with intellectual disability. Most qualitative studies explore their experiences by utilising proxy-reports. A smaller number of studies illustrate the possibility of exploring perspectives directly from people with intellectual disability and dementia. This systematic review synthesized findings from existing studies (n = 8) that involve people with intellectual disability and dementia as participants to understand their experiences of dementia. Searches were conducted using CINAHL, PsychInfo and Social Services Abstracts. Findings include descriptions of changes in individual functioning, a narrowing of social worlds and of how people made sense of the changes despite often having no knowledge of their dementia diagnosis. Additionally, discussion focuses on how people's experiences are shaped by their environments. People's subjective experiences and views on how dementia affected their lives were more ambiguous compared to contextual data and the descriptive portrayal of how people's lives had changed. Thus, this review highlights the importance of researchers needing consider how to facilitate conversations with people about their experiences and the ethics involved in conducting qualitative research with people with intellectual disability and dementia, particularly how respond to participants not knowing about their dementia diagnosis. Spending time with participants over a longer period, getting to know how people communicate and the use of visual aids or everyday items were examples of approaches that supported research involvement. The review recognized the complexities of speaking to people with intellectual disability about dementia, challenges views that people with intellectual and dementia cannot be involved in research and makes recommendations to support inclusion in future studies.

Jamieson-Craig, R., Scior, K., Chan, T., Fenton, C., & Strydom, A.
Reliance on carer reports of early symptoms of dementia among adults with intellectual disabilities
Journal of Policy and Practice in Intellectual Disabilities, 2010, 7(1), 34 - 41. https://doi.org/10.1111/j.1741-1130.2010.00245.x
Abstract: As clinicians often rely on carer reports to identify adults with intellectual disabilities (ID) with early signs of dementia, this study focused on carer-reported symptoms to ascertain whether carer reports of decline in everyday function would be a more effective screening method to detect possible cases of dementia than reports of memory decline in older adults with ID. Subjects were 154 participants who were reassessed along with their carers two to three years after baseline. A questionnaire for carer-reported change in everyday function and the Dementia Questionnaire for Persons with Mental Retardation (DMR) were used to assess carer views of everyday function and memory. The diagnosis of dementia was confirmed by two psychiatrists working independently. Participants who developed dementia displayed both everyday function and memory decline. Overall, decline in everyday function appeared to be the best indicator of new dementia cases. Retrospective carer report of change in everyday function was as good as, if not better than, prospective ratings to identify dementia; however, in those with mild ID, memory change was a better indicator of dementia, while in those with more severe ID, decline in everyday function was a better indicator. Decline in everyday function (whether prospective change from baseline or reported retrospectively by carers) appears to be a better screening method for dementia than memory decline, particularly for participants with moderate/severe ID.

Janicki, M.P.
Health status and comorbidities of adults with dementia and ID-implications for screening and healthcare
Innovation in Aging, 2017 July, 1(suppl_1), 712. https://doi.org/10.1093/geroni/igx004.2554
Abstract: Declining health status and comorbidities are often markers for associated mild cognitive impairment or dementia. A group of community-dwelling adults with intellectual disability diagnosed with dementia (along with controls) have been tracked longitudinally with respect to their health and function. With time, the dementia-capable group home residents with dementia are showing significantly varied health status and comorbidities as well as marked behavioral changes. By tracking the health and function longitudinally, outcome information can pinpoint markers that are associated with premorbid dementia and can help health providers maintain surveillance over select functions and health conditions of those adults already affected. Screening instruments, incorporating these markers, can more precisely be used to identify at-risk adults for ADRD and aid providers design remediation programs earlier.

Janicki, M.P.
Dementia capable group homes for adults with intellectual disability: development implications
Innovation in Aging, 2018 Nov, 2(suppl_1), 532-533. https://doi.org/10.1093/geroni/igy023.1967
Abstract: Community housing for adults with intellectual disability (ID) and dementia (AD) is becoming more prevalent. An opportunistic longitudinal study (2011-2018) of group homes (GHs) for such adults examined a number of resident and administrative factors. Study followed 2 cohorts of adults with ID, 15 w/dementia and 15 matched controls (CO), over 7 years (8 time intervals). Initial cohort of 15 AD (Xage=59.1; NDS=5) resided in 3 (5-bed-each) dementia GHs. COs resided in general GHs/apartments. Over 7 years, 8 AD died, and 7 replacements (Xage=58.7; NDS=2) were added to the study. Instruments captured subject characteristics, behavior/health/function, staffing, time spent on caregiving, and administrative factors. Findings noted differences in health and function factors between the ADs and COs. Deaths occurred at age-norms. Comparative co-morbidities showed AD residents had significantly more classic cognitive and physical health issues associated with dementia and physical debilitation. An ebb and flow was observed of residents affected by dementia when an agency has multiple dementia GHs, as well as variations in staffing patterns and periods of intensity of care during the day. Over time, the 3 dementia GHs, by inter-home transfers and selective new admissions, have trended toward stage/level specific care settings. Findings can help with planning long-term use of such GHs and can providing dementia capable care for adults with ID in a community-based specialized setting. With dementia affecting an increasing number of adults w/ID (due to aging) more attention needs to be given to functional GHs providing in-community long-term mid-stage and advanced dementia capable care.

Janicki, M.P.
Quality outcomes in group home dementia care for adults with intellectual disabilities.
Journal of Intellectual Disability Research, 2011, 55(8), 763-776. [doi: 10.1111/j.1365-2788.2011.01424.x].
Abstract: Dementia, as a public health challenge, is a phenomenon vexing many care organizations providing specialized residential and family supports for older adults with intellectual disabilities. With increasing survivorship to ages when risk is greatest, expectations are that many more adults in service will present with cognitive decline and diagnosed dementia as they grow older. As persons with dementia present with new needs, there is often a call for a reorientation of services. With respect to residential supports, agencies may need to adapt current methods of care, with particular attention to providing care in small group homes. However, dementia-related care also must be quality care and applicable standards need to be met. The author reviewed relevant policy and practice organizational guidelines and applied research literature addressing components of care and service provision that were critical to quality care and that were consistent with professional practice. Examined were the nuances and contributing factors of quality dementia care and it was proposed that quality of care criteria need to be universally applicable and serve as a framework for adapting extant residential environments and make them 'dementia-capable'. It is

proposed that efforts to evaluate dementia-related care provision with respect to quality need to consider quality of care provision components such as (1) clinically relevant early and periodic assessment; (2) functional modifications in the living setting; (3) constructive staff education and functionality for stage-adapted care; and (4) flexible long-term services provision that recognizes and plans for progression of decline and loss of function.

Janicki, M.P.
Stationäre Einrichtungen der Behindertenhilfe für Me nschen mit geistiger Behinderung und dementielle Erkrankung [Group home care for adults with intellectual disabilities and Alzheimer's disease]
Lebensqualität im Alter, 2016, 10.1007/978-3-658-09976-3_14, (237-263).
Abstract: According to the WHO, prevalence and incidence rates suggest that the number of dementia cases continues to rise. This also affects people with intellectual disabilities, who require specialized care models. The focus is on optimal community care, which, in addition to a dementia-friendly environment, is characterized by individualized and person-centered care. Care in small group homes is the most common form of community care. Most such residential facilities offer two models, which can be described as linear and sequential. Both models have in common the emphasis on the need for a dementia-friendly environment and comprehensive training for caregivers.

Janicki, M.P.
On-going activities of the National Task Group on Intellectual Disabilities and Dementia Practices
Gerontologist, 2018, 56(Suppl_3), 573.
Abstract: The National Task Group on Intellectual Disabilities and Dementia Practices (NTG), organized in 2011, has been actively involved in stimulating development of services for people with intellectual disabilities (ID) affected by dementia. The NTG has created several sets of practice guidelines, a screening and early detection instrument for use by families and agencies, web-based informational materials, and a national curriculum on ID and dementia, and has undertaken the provision of workforce development workshops across the US on dementia and ID. The NTG works to compliment the activities being undertaken under the National Plan to Address Alzheimer's Disease and consults with various national organizations focusing on dementia and lifelong disabilities. The goal of the NTG is to continue to affect change and improve the quality of community dementia care provision corresponding with National Plan updates.

Janicki, M.P.
Small group homes as "dementia-capable" settings for people with intellectual disabilities and early stage dementia.
Alzheimer's & Dementia, 3(3S), Part_3, First published: 01 July 2007. https://doi.org/10.1016/j.jalz.2007.04.369
Abstract: Localities are beginning to feel the impact of the growing number of older adults with lifelong intellectual disabilities (ID) who are also affected by dementia. Many local organizations are attempting to adapt their support and residential services to help this group be served more effectively within the community. Yet, questions have been raised as to the models that may most reasonably be used and how to address the discordance between traditional ID service and "dementia-capable" services. Investigated was how localities and organizations have adapted to the onset of dementia and address early stage care demands and determine practices that are effective in promoting "dementia-capable" care. Several studies were conducted to determine how government entities and local providers are adapting services to identify models prevalent in the provider sector, and specifically to identify staff training needs, physical and environmental adaptations, and differential time spent by staff in providing dementia care. Data showed that most US states are not prepared to address growing onset of dementia in select parts of the ID population, that responses to early stage service needs have been mostly handled by local entities and service organizations, that most have not developed extensive training programs for staff and are experimenting with best practice methods to deliver care - primarily via small group homes - and that dementia care takes up a disproportionate amount of staff time in small care settings. To address early stage dementia related services in the most effective manner, a concerted effort needs to be in place to aid local service entities adapt services to dementia-related presentations among ID clientele, set up coordinated training for staff, secure funds for adapting group homes for community "dementia-capable" care, and construction of clinical support services and augmentation of family support services for parents and other kin carers.

Janicki, M.P.
Community-based housing and NPI-care practices for adults with intellectual disability and dementia
AAIC 2020 Conference, Poster presentation, July 30, 2020. *Alzheimer's & Dementia*, 16(S8), First published: 07 December 2020. https://doi.org/10.1002/alz.047061
Abstract: Aging persons with intellectual disability (ID) represent a vulnerable population with respect to cumulative neuropathological conditions, including dementia. Adults with Down syndrome (DS), a subset, have a recognized high risk for Alzheimer's disease. With dementia present, how to provide post-diagnostic supports is challenging. Dementia care group homes (GHs) along with NPIs are emerging as a mode for providing out-of-home community supports. Data from a longitudinal study provide insights on what care organizations need to consider when organizing specialty group home care. The study, begun in 2011, followed three co-located homes providing NPIs to 15 adults with dementia. Findings revealed trajectories of changes over time, housing need/function level patterning, and health status outcomes. Key findings noted 3 age-of-admission clusters (X=50.5; X=57.1; X=66.8); overall mortality (Xage-death=65.4; ID=69.3; DS=56.3) – half of original entrants died within 7 years; age at entry (X= 59.1); years from entry to death (X= 5.4 yrs); LOS (X=49.4 months/4.12 yrs); morbidities (number of co-morbidities decreased among survivors). In same period, 8/15 deaths in GHs vs 3/15 deaths in Controls. NPI-related practices included day program activities (adults in mid- to later stages were engaged in regular off-site day activities that agency provided; adults with advanced dementia remained in homes), staffing patterns differed based on level of care – more staff assigned to homes with residents with advanced dementia, and staff training included dementia capable communications, engagement, and managing daily routines. Trends showed adults with Down syndrome were admitted to homes earlier but had more life-years in the GHs than older adults admitted at later age but who succumbed earlier to disease complications. Dementia care GHs should expect varied trajectories of decline; mortality linked to complexity of pre-existing conditions and progression of dementia; changes in the focus of care needs over time (including advanced dementia and end-of-life care). Dementia care GHs can enable provision of in-community group housing and quality care in accord with stage-defined functional changes and needs if structured in a planful way (factoring in dementia-stage, dementia type, mortality expectations, health status, patterns of care needs, dementia-related behaviors, aging-related issues, and probable trajectories of decline of the residents).

Janicki,M.P., Dalton, A.J., McCallion, P., Davies Baxley, D., & Zendell, A.
Group home care for adults with intellectual disabilities and Alzheimer's disease
Dementia, 2005, 4, 361-385. https://doi.org/10.1177/1471301205055028
Abstract: The growing numbers of individuals with intellectual disabilities affected by Alzheimer disease and related dementias has raised new challenges for community care providers. This paper examines means of providing community group home-based care in a sample of care providers in five different countries. The aim is to identify trends that have emerged. Two samples of group homes for adults with intellectual disabilities affected by dementia were studied to determine: (1) what are the physical characteristics of the homes; (2) what physical environmental adaptations have been made in response to behavioral deterioration expressed by residents with dementia, and (3) what are the demands on staff resulting from dementia care. The first sample of group homes in five countries provided comparative international data on home designs, staffing, costs, and residents. The second sample, drawn from homes in the

USA and the UK, provided data on the impact of dementia. Findings revealed staffing and design of homes varied but generally abided by general practices of dementia care; homes relied on existing resources to manage changes posed by dementia care; programmatic and environmental adaptations were implemented to address progression of dementia; and residents with dementia presented more demands on staff time with respect to hygiene maintenance and behavior management when compared to other residents not affected by dementia.

Janicki, M. P., Heller, T., Seltzer, G., & Hogg, J.
Practice guidelines for the clinical assessment and care management of Alzheimer's disease and other dementias among adults with intellectual disability.
Journal of Intellectual Disability Research, 1996, 40(4), 374-382. PMID: 8884593.
Abstract: The AAMR/IASSID practice guidelines, developed by an international workgroup, provide guidance for stage-related care management of Alzheimer's disease, and suggestions for the training and education of carers, peers, clinicians, and program staff. The guidelines suggest a three step intervention activity process, that includes: (1) recognizing changes, (2) conducting assessments and evaluations, and (3) instituting medical and care management. They provide guidance for public policies that reflect a commitment for aggressive care of people with Alzheimer's disease and intellectual disability, and avoidance of institutionalization solely because of a diagnosis of dementia.

Janicki, M.P., Hendrix J.A., McCallion, P. and the Neuroatypical Conditions Expert Consultative Panel.
Examining older adults with neuroatypical conditions for MCI/dementia: Barriers and recommendations of the Neuroatypical Conditions Expert Consultative Panel.
Alzheimers & Dementia (Amst), 2022 Jul 8,14(1).e12335. doi: 10.1002/dad2.12335.
Abstract: The Neuroatypical Conditions Expert Consultative Panel composed of numerous clinical and academic experts was convened to examine barriers to the examination of cognitive impairment in adults with a variety of neuroatypical conditions. Neuroatypical conditions affect normative intellectual development and function (such as intellectual disability and intellectual disability with conjoint psychiatric conditions), thought, moods, and cognition (such as severe mental illness), communication functions (such as the autism spectrum and hearing/vision impairments), and brain and motor function (such as cerebral palsy and acquired or traumatic brain injury). The panel concluded that current federal guidance for the assessment of cognitive impairment for mild cognitive impairment (MCI) or dementia does not sufficiently include information as to how to assess such adults. In addition, it concluded that adults with these conditions (1) challenge clinicians when attempting to discern current behavior and function from that which was pre-existing; (2) often have inherent comprehension and oral communication difficulties, motor task performance impediments, and difficulty with visuals; and (3) pose difficulties when assessed with standardized dementia measures and can benefit from the use of specialized instruments. The panel recommended that federal guidance be broadened to include adaptations of assessment practices to accommodate neuroatypical conditions; that educational packs be developed for clinicians about such conditions and on detecting and diagnosing MCI or dementia; and that research be expanded to produce more evidence-based information on both assessing adults with neuroatypical conditions for later-life adult cognitive diseases/disorders and planning post-diagnostic care.

Janicki, M.P., & Jacobson, J.W.
Generational trends in sensory, physical, and behavioral abilities among older mentally retarded persons.
American Journal of Mental Deficiency, 1986, 90(5), 490-500.
https://pubmed.ncbi.nlm.nih.gov/3953681/
Abstract: An extensive client registry data base was used to examine whether there were identifiable patterns in prevalence of chronic disease conditions, in decline of sensory capacities, and in decline of behavioral abilities among 10,532 elderly mentally retarded persons born between 1890 and 1939. Data indicated identifiable patterns in the prevalence of chronic disease conditions (cardiovascular, digestive, musculoskeletal, and respiratory systems) and the decline of behavioral and physical abilities and sensory capacities with increasing age. Functional skill losses were most prominent in mobility and fundamental activities of daily-living skill areas. Some behavioral losses were noted to occur earlier than would be expected in the general population; however, as with nonretarded persons, decline was highly variable and linked to the aging process experienced by each older retarded person. Implications of the findings were discussed.

 **Janicki, M.P., McCallion, P., & Dalton, A.J.**
Supporting people with dementia in community settings.
In M.P. Janicki & A.F. Ansello (Eds.), Community Supports for Aging Adults with Lifelong Disabilities, pp. 387-413
Baltimore, Maryland: Paul H. Brookes Publishing (2000)
Abstract: Due to the "greying" of the nation's population, dementia associated with Alzheimer's disease and other causes, has become another challenge for providers of services to adults with intellectual disabilities. In this book chapter, the authors explore the factors, policies, and support structures that can help agencies provide continued "aging-in-place" dementia-capable care, develop "in-place progression" dementia specific programs, or chose alternative care settings. It also explores some features of dementia-related behaviors that may need to be taken into account in program design and makes suggestions for staff training and planning for dementia programs.

Janicki, M.P., McCallion, P., & Dalton, A.J.
Dementia-related care decision-making in group homes for persons with intellectual disabilities
Journal of Gerontological Social Work, 2002, 38(½), 179-196.
https://doi.org/10.1300/J083v38n01_04
Abstract: The number of age-associated pathologies is increasing, with the increase in the number of elderly persons. One such age-associated condition, Alzheimer's disease and related dementias, affects a significant number of adults with intellectual disability (ID), in particular those with Down syndrome. Many affected adults live in small community group homes or with their families. How to provide sound and responsive community care is becoming a challenge for agencies faced with an increasing number of affected adults. This study reports the outcome of a survey of group homes serving adults with ID and dementia, explores the onset, duration and effects of dementia and their impact on planning for community care of adults with ID. It also examines emerging community care models that provide for "dementia capable" supports and services. Two models, "aging in place," and "in place progression" are examined with regard to care practices and critical agency decision making. An approach, the ECEPS model, for responding to dementia is offered.

Janicki, M.P., McCallion, P., Jokinen, N., Larsen, F.K., Mughal, D., Palanisamy, V., Santos, F., Service, K., Shih, A., Shooshtari, S., Thakur, A., Tiziano, G., & Watchman, K.
Autism and dementia: A summative report from the 2nd International Summit on Intellectual Disabilities and Dementia.
Journal of Autism and Developmental Disorders, 2025 May 6. doi: 10.1007/s10803-025-06843-7. Epub ahead of print.
Abstract: This article synthesizes findings, from the Autism/Dementia Work Group of the 2nd International Summit on Intellectual Disabilities and Dementia, on the nature of autism/autism spectrum disorder and later-age neuropathologies, particularly dementia. The convened group of experts explored genetic, neurobiological, and environmental risk factors that may affect the lifespan and lived experiences of older adults with autism. A review of current literature indicates a lack of comprehensive information on the demographics and factors

associated with aging in autistic adults. However, our understanding of autism is evolving, challenging traditional views of it as a static, inherited neurodevelopmental disorder. The relationship between autism and other neurodevelopmental conditions-such as Down syndrome, fragile X syndrome, and tuberous sclerosis complex-reflects the complex genetic landscape of neurodevelopmental disorders. These genetic and familial factors may contribute to progressive health challenges and cognitive decline in later life. Key findings reveal a complex link between autism and dementia, despite limited research on this relationship, particularly among older adults. The overall prevalence of dementia in this population appears to be influenced by co-occurring intellectual disabilities, particularly Down syndrome. While the association between autism and specific types of dementia is still not well understood, the reviewed evidence suggests a notable connection with frontotemporal dementia, although causality has not been established. Exploration of biomarkers may offer further insights. Currently, the relationship between autism, cognitive health, and cognitive decline in older adults remains a complex and underexplored area of research.

Janicki, M.P., McCallion, P., Jokinen, N., Larsen, F.K., Service, K.P., Mughal D.T,, Watchman, K., Gomiero, T., & Keller, S.M.
Autism, diagnostics, and dementia: A consensus report from the 2nd International Summit on Intellectual Disabilities and Dementia.
International Journal of Geriatric Psychiatry, 2025 Jun, 40(6), e70110. doi: 10.1002/gps.70110.
Abstract: The second International Summit on Intellectual Disability and Dementia, held in 2023, highlighted the unique challenges of diagnosing dementia in older autistic adults, particularly those with intellectual disabilities, due to the complex interplay of cognitive, communicative, and behavioral factors. This article addresses key diagnostic issues and post-diagnostic considerations for this population. A consensus report was developed by the Summit's Autism/Dementia Working Group through background reviews, expert discussions at the Summit, and iterative draft revisions, incorporating feedback from internal and external stakeholders. Key issues were extracted from the report and abridged for this manuscript. Diagnostic challenges stem from overlapping symptoms of co-occurring neurodevelopmental and psychiatric conditions, rendering standard dementia tools insufficient. Comprehensive evaluations tailored to autism-related traits, sensory sensitivities, and alternative communication methods are essential. Building diagnostic capacity among clinicians and fostering multidisciplinary collaboration are critical. Longitudinal assessments, initiated before dementia symptoms appear, facilitate early detection of subtle changes. Emerging biomarkers and neuroimaging techniques show promise and should be incorporated where feasible. Accommodations, such as virtual assessments in familiar settings, can enhance diagnostic accuracy by reducing anxiety. Creating transition processes from diagnostics to post-diagnostic supports will aid in mitigating challenges and enhance life quality when dementia is a factor. Research and clinician education are urgently needed to improve diagnostic approaches and streamline the transition from diagnosis to tailored post-diagnostic support. An integrated framework of comprehensive efforts is vital for our better understanding of age-associated neuropathological diagnostics and enabling long-term well-being of older autistic adults with dementia.

Janicki, M.P. & Dalton A.J.
Care management, diagnostic and epidemiologic considerations in adults with intellectual disabilities and Alzheimer disease
British Journal of Developmental Disabilities, 1996, 42(Supplement), s84.
Abstract: Review of the process and outcome of the Invitational International Colloquium on Alzheimer Disease among Persons with intellectual Disabilities held in Minneapolis, Minnesota (USA) and the subsequent development of a set of international practice guidelines and reports on the assessment, epidemiology, and care management of adults with intellectual disabilities affected by dementia.

Janicki, M.P., & Dalton, A.J.
Dementia in developmental disabilities
In N. Bouras (Ed.), Psychiatric and Behavioral Disorders in Developmental Disabilities and Mental Retardation (1999)
pp. 121-153
Cambridge: Cambridge University Press
Abstract: This book chapter provides a brief overview of the current status of knowledge about dementia and its relationship to intellectual disability, touching on current developments in the evaluation of possible comorbid psychiatric, medical and age-associated conditions. The clinical presentation of dementia is examined as well as relevant contemporary issues related to diagnosis, assessment, and care management. Lastly, questions of dementia policy and suggestions for training programs on dementia and intellectual disability are addressed.

Janicki, M.P., & Dalton, A.J.
Dementia and public policy considerations
In M.P. Janicki & A.J. Dalton (eds.), Dementia, Aging, and Intellectual Disabilities (1999), pp. 388-414
Philadelphia: Brunner-Mazel
Abstract: This book chapter examines a number of the major public policy considerations related to the aging of adults with intellectual disabilities who evidence change due to dementia. Specifically addressed is the changing structure of at-risk adult populations with intellectual disabilities in service systems, the programmatic and policy issues raised by providers attempting to cope with these changes, needs for further training, education and dissemination of information on aging, and lastly, the challenges and policy imperatives to be confronted with the new millennium.

Janicki, M.P., & Dalton, A.J.
Dementia, aging, and intellectual disabilities: A handbook
488pp.
Philadelphia: Brunner-Mazel [http://www.taylorand francis.com] (1999)
Abstract: 21 chapter text on dementia issues and intellectual disabilities. Six parts: Introduction, Biomedical considerations, Assessment considerations, Clinical considerations, Program considerations, and Education and policy considerations. Text provides most up-to-date information available about Alzheimer's disease and related dementias as they affect persons with mental disabilities. Text examines biology and physiology of dementia, neurological and medical complications associated with dementia, best practices to meet the needs of aging persons with intellectual disabilities, policy issues raised by the growing number of older adults with ID, and case studies of affected individuals. Contains glossary of terms, and appendices with AAMR/IASSID practice guidelines for dementia diagnosis and care management in adults with intellectual disabilities, as well as Newroth & Newroth guidelines for coping with Alzheimer's disease in persons with Down syndrome.

Janicki, M.P., & Dalton, A.J.
Prevalence of dementia and impact on intellectual disability services
Mental Retardation, 2000, 38, 277-289. doi: 10.1352/0047-6765(2000)038<0276:PODAIO>2.0.CO;2.
Abstract: A statewide survey, conducted to ascertain the administrative prevalence of dementia in adults with an intellectual disability, found a prevalence of about 3% of the adult service population over the age of 40 years (a rate of 28/1000), 6.1% of the population over the age of 60 years, and 12.1% of the population over the age of 80 years (or rates of 68.7/1000 and 121.3/1000, respectively). The rate of dementia was consistent with that for adults in the general population, except for those adults with Down syndrome (who made up a third of the overall group) who had a much higher rate: 22.1% among adults age 40 and older and 56.4% among adults age 60 and older. Onset was observed to occur in the mid-60s (early 50s for Down syndrome). Alzheimer-type dementia was the most frequent diagnosis. Late-onset seizures were reported in about 12% of the cases. With the occurrence of dementia expected to rise

proportionately with the increase of longevity among adults with an intellectual disability, it is clear that care systems will have to raise the "index of suspicion" among staff and families, adapt to become "dementia capable," and improve their diagnostic and technical resources, as well as their community-based care management supports.

Janicki, M.P., & Dalton, A.J.
Alzheimer disease in a select population of older adults with mental retardation
The Irish Journal of Psychology, 1993, 14(1), 38-47
doi:10.1080/03033910.1993.10557913
Abstract: Twenty New York State Office of Mental Retardation and Developmental Disabilities district offices were surveyed to determine the prevalence of older adults with mental retardation who also had suspected or diagnosed Alzheimer disease (AD). The survey identified 123 individuals in this category, about 1% of an overall case roll of 10,878 people of 40 years of age and older for the 17 reporting offices. Of these 123 individuals, 64% were people with Down syndrome. Typically, a person with suspected Alzheimer disease was identified through staff observation or report of changes in behaviour leading to suspicion of Alzheimer disease. Information on training needs indicated that a workshop on AD was a primary need. Training in various content areas was indicated, including confirming suspicions, diagnosis, programming, and individual program planning.

Janicki, M.P., McCallion. P., Splaine, M., Santos, F.H., Keller, S.M., & Watchman, K.
Consensus statement of the international summit on intellectual disability and dementia related to nomenclature
Intellectual and Developmental Disabilities, 2017, 55(5), 338–346. DOI: 10.1352/1934-9556-55.5.338.
Abstract: A working group of the 2016 International Summit on Intellectual Disability and Dementia was charged to examine the terminology used to define and report on dementia in publications related to intellectual disability (ID). A review of related publications showed mixed uses of terms associated with dementia or causative diseases. Like general applications, language related to dementia in ID field often lacked precision and could lead to a misunderstanding of the condition(s) under discussion. Most articles related to ID and dementia reporting clinical or medical research generally provided a definition of dementia or related terms; social care articles tended toward term use without definition. Toward terminology standardization within studies/ reports on dementia and ID, the Summit recommended (a) gaining familiarity with dementia-related diagnostic, condition-specific, and social care terms (as identified in the working group's report), (b) creating a guidance document on accurately defining and presenting information about individuals or groups referenced, and (c) that in reports on neuropathologies or cognitive decline or impairment, definitions are used and data include subjects' ages, sex, level of ID, residential situation, basis for dementia diagnosis, presence of Down syndrome (or other risk conditions), years from diagnosis, and if available, scores on objective measures of changing function.

Janicki, M.P., Zendell, A., & DeHaven. K.
Coping with dementia and older families of adults with Down syndrome.
Dementia, 2010, 9(3), 391-407. https://doi.org/10.1177/1471301210375
Abstract: The authors studied a group of older carers of aging adults with Down syndrome (DS) to ascertain what effects such caregiving may have on them given the presence or possibility of age-associated decline or dementia. The study also examined the comparative levels of care provided, key signs noted when decline was beginning, the subjective burden experienced, and what were the key associated health factors when carers faced a changed level of care. The authors found that this group was made up of long-term, committed carers who had decided early to look after their relative with DS over their lifetime. When faced with the onset and ongoing progression of dementia, their commitment was still evident as evidenced by adopting physical accommodations and finding ways to continue to provide care at home, while also seeking help from outside sources. Most saw a family or group home environment as the place of choice for their relative with DS when they decided they could no longer offer care. The study did not ascertain any burn-out or significant health related problems associated with their continued caregiving save for their concerns about day-to-day strain and what will happen in the future.

Janicki, M.P., & McCallion, P.
A group home cluster model for providing community-based dementia care.
Paper presented at the 21st annual conference of Alzheimer Europe, Warsaw, Poland. (2011, October).
https://www.the-ntg.org/_files/ugd/8c1d0a_911c14fa5fec4f89a59dc60da1f0d390.pdf
Abstract: Paper reports on a study undertaken of an innovation group home program operated by a provider organization serving older adults with intellectual disabilities. The provider built three co-located group homes for five adults within a neighborhood setting. Each of the adults resident at the homes have some degree of diagnosed dementia. The adults were both males and females, all were age 50+, and some had Down syndrome. The homes are staffed by paid staff working 24/7. The residents were studied for health co-morbidities, program activities, and degrees of impairment and compared with a matched group of adults without dementia. The study examined administrative and programmatic factors related to the operation of the homes, as well as shifts in characteristics related to their intellectual disability and the effects of dementia
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Jaycock, S., Persaud, M. & Johnson, R.
The effectiveness of dementia care mapping in intellectual disability residential services: A follow-up study.
Journal of Intellectual Disabilities, 2006, 10(4), 365-375. doi: 10.1177/1744629506072870.
Abstract: The authors present a follow-up to exploratory work published in the Journal of Intellectual Disabilities in 2001. This article describes a study that aimed to assess the effectiveness of dementia care mapping in supporting practice improvement in intellectual disability residential services. An average of 9 hours of observational data were collected using dementia care mapping in relation to 14 adults with severe or profound intellectual disabilities (but who not have dementia). Sixteen interviews were also undertaken with staff over a 4 month period.The findings provided a detailed picture of the activities and interactions between the participants involved in the study and raised some issues about 'organizational culture' when developing person-centered approaches. These data have helped strengthen the case that care mapping has the potential to be a useful addition to the existing repertoire of tools to support effective practice improvement and person-centered planning.

Jenkins, R., Davies, R., Sardi, I., Llewellyn, P., Northway, R., O'Connor, C., Trudgeon, C. & Keeling, D.
Adults with learning disabilities presenting with dementia [Final Report - May 2009]
University of Glamorgan (Faculty of Health, Sport, and Science), 2008. 78pp.
https://www.choiceforum.org/docs/pdem.pdf
Abstract: As life expectancy improves, increasing numbers of people with intellectual (learning) disabilities are affected by dementia. Supporting someone with a dual diagnosis of intellectual disability and dementia presents unique challenges to both those responsible for delivering services and carers. In response to these challenges Gwent Healthcare NHS Trust developed a Dementia Care Pathway in 2005. The aims were: (1) to ensure early and appropriate diagnosis, (2) to provide a co-ordinated approach to assessment and intervention, (3) to develop intervention plans that will support both client and carers, (4) to provide a process for monitoring the person over time, and (5) to support, carers, clients, and professionals via the proviso of information and training. The study demonstrated that the stated aims of the Dementia Care Pathway are being met. Stage one results found that the training provided to paid support staff was received enthusiastically and had a positive impact on knowledge, confidence, and competence levels in relation to working iwth clients

who have, or may develop, dementia. In most cases the knowledge was well maintained at six month follow-up. There is some evidence, however, that although staff who attended the training reported it to be 'interesting' and 'very relevant, but they still doubted its usefulness in practical day-to-day situations when caring for their clients. In order for the potential of the pathway to be further maximized attention should be paid to increasing the level of information given to carers, widening contributions to pathway review meetings, and educating carers on the diverse range of interventions possible through the pathway.

Jensen, K.M., Bulova, P.D., Santoro, S.
Down syndrome.
Book chapter In: Kuo, A.A., Pilapil, M., DeLaet, D.E., Peacock, C., Sharma, N. (eds), *Care of Adults with Chronic Childhood Conditions.* Springer, Cham.
https://doi.org/10.1007/978-3-031-54281-7_30
Abstract: Resulting from triplication of all or a portion of chromosome 21, Down syndrome is the most common identifiable genetic cause of intellectual and developmental disability. Down syndrome occurs in 1/700-1/1000 live births and affects approximately 150,000 persons in the United States. Down syndrome is characterized by intellectual disability ranging from mild to profound and results in an increased risk of congenital and acquired conditions in most organ systems. Adult care is complicated by multiple comorbid conditions in the setting of accelerated aging. Due to dramatic improvements in their medical care during early childhood and routine surgical correction of congenital heart conditions, persons with Down syndrome now live well into adulthood with a median life expectancy into their mid-50s. Consequently, the care of adults with Down syndrome is a rapidly growing field requiring increased attention from adult primary care providers.

Jervis, G.A..
Early senile dementia in mongoloid idiocy.
American Journal of Psychiatry, 1948 Aug;105(2):102-6. doi: 10.1176/ajp.105.2.102.
Abstract: In 3 mongoloid patients aged respectively 47, 42, and 37 years, personality changes and mental deterioration were observed. Pathologically, evidence of senile changes was obtained consisting of degeneration of neurons, numerous senile plaques, and Alzheimer's neurofibrillary changes. With the exception of the age of onset, the clinical and pathological manifestations are those of senile dementia. Since mongoloid patients show a marked tendency to develop this type of reaction, it is suggested that the study of it offers some information which may contribute to a better understanding of the causes of senile dementia.

Jervis, N., & Prinsloo, L.
How we developed a multidisciplinary screening project for people with Down's syndrome given the increased prevalence of early onset dementia.
British Journal of Learning Disabilities, 2008, 36 (1), 13–21.
https://doi.org/10.1111/j.1468-3156.2007.00474.x
Abstract: Much research has identified an increased prevalence of dementia in adults with Down syndrome when compared with the general population. Neuropathological changes associated with Alzheimer's dementia in the brain have been found in most people with Down syndrome who die over the age of 35 years. Given the limitations of many assessments for dementia in relation to people with Down syndrome for a single completion, it has been recommended that all people with Down syndrome are assessed at least once in early adulthood in order that they have their own baseline which can be compared with in the future if changes in skills and abilities occur. The authors have had many requests from other services enquiring about their project and how a similar initiative could be set up. Therefore, this article focuses on the way the Manchester Learning Disability Partnership approached screening 135 adults with Down syndrome and details the assessments used, practical considerations, what has been learned and future service implications.

Jethwa, H. & Cassidy, G.
Difficulties of dealing with dementia in individuals with intellectual disabilities: the healthcare perspective
Advances in Mental Health and Intellectual Disabilities, 2010, 4(4), 48-52.
https://doi.org/10.5042/amhid.2010.0675
Abstract: Dementia is a condition that involves inevitably progressive deficits in numerous cognitive domains, including thought, language, memory, understanding and judgement. A difference in behavior may be noted, as well as overall loss of skills. Dementia is more than four times as prevalent in people with intellectual disabilities as in the general population. Diagnosis of the condition in people with intellectual disabilities, however, is often difficult due to lack of baseline skill assessment, high staff turnover in supported accommodation and low expectations of capabilities. Current National Institute for Health and Clinical Excellence (NICE) guidelines on anti-dementia medication state that treatment should not be initiated until the condition has reached moderate severity. Determining whether symptoms are at this stage in people with intellectual disabilities is difficult because their skill level is already impaired. An accurate and extensive record of baseline skill levels in people with intellectual disabilities is therefore crucial, and regular comparison with baseline is key to early diagnosis of dementia.

Johannsen, P., & Mai, J.
Alzheimer-type demens og Downs syndrom [Alzheimer-type dementia and Down syndrome] *Article in Danish*
Ugeskrirft for Laeger, 1995 Feb 20, 157(8), 1021-1024.
Abstract: Dementia is seen earlier and more often in patients with Down syndrome (DS) than in the general population. Patients with DS develop neuropathological changes similar to the changes seen in dementia of Alzheimer's type (DAT). The literature concerning DAT in DS is reviewed. The clinical course and differential diagnoses are discussed. Before the diagnosis of DAT is made, treatable causes of dementia should be excluded.

Johannsen, P., Christensen, J.E.J., & Mai, J.
The prevalence of dementia in Down syndrome
Dementia, 1996, 7(4), 221-225. doi: 10.1159/000106883.
Abstract: The authors assess the prevalence of clinical dementia in three age groups of persons with Down syndrome in the county of Aarhus, Denmark. Group 1 was composed of 14-16 year olds (n=13), group 2 was composed of 23-29 year olds (n=34), and group 3 was composed of 50-60 year olds (n=25). Of the 85 subjects, 72 (85%) participated. Carers were interviewed and a neurological examination was performed. An EEG was recorded in 50 of the Ss. Definite clinical dementia was defined as a acquired and progressive decline in 4 or more out of 17 items that are considered to indicate dementia in people with Down syndrome. Possible dementia was considered when 1-3 items were affected. Six adults (24%) in group 3 had definite clinical dementia and 6 adults in group 3 and 2 (6%) in group 2 had possible dementia. Authors note that this was the first Danish population-based study of the prevalence of dementia in people with Down syndrome.

Johansson, P.E., & Terenius, O.
Development of an instrument for early detection of dementia in people with Down syndrome
Journal of Intellectual & Developmental Disabilities, 2002, 27(4), 325-345.
https://doi.org/10.1080/1366825021000029357
Abstract: The successful detection of early signs of dementia in people with Down syndrome could form a basis for useful early support and for drug treatment. This report describes the development and preliminary application of an interview and test instrument for the assessment of dementia among people with intellectual disability, as well as a framework for diagnosis that combines the findings of an interview and a test with the diagnostic criteria of ICD-10, DSM-IV and NINCDS-ADRDA. From among the number of tests and interview questions developed, those showing the most significant differences between participants in three groups of differing levels of intellectual disability and estimated dementia were kept. Reported are the assumptions for the items used, descriptions of the process and items used, and the associations of test items with predicting the

presence of dementia. The authors conclude that a protocol combining testing and interview has promise and potential for detecting early signs of dementia in this population and could prove feasible for use in practice.

Johansson, M., Holst,G., & Ahlström, G.
Signs in people with intellectual disabilities: Interviews with managers and staff on the identification process of dementia.
Healthcare (Basel), 2018 Aug 25;6(3):103. doi: 10.3390/healthcare6030103.
Abstract: The life expectancy of people with intellectual disabilities (ID) has steadily increased, which has been accompanied by an increased risk of dementia. Staff and managers are key resources for safety diagnosis since they deliver information about people with ID behavior every day. The aim of the present study was to explore the identification process employed by staff and managers to detect signs of suspected dementia in people with an ID within intellectual disability services (ID-services). Twenty managers and 24 staff within an ID-service were interviewed and qualitative latent content analysis was applied. A model consisting of three themes on three levels of resources for the identification process of signs of suspected dementia emerged from the analysis. On the first level was the time and continuity in the care relationship, which is crucial for identifying and responding to changes in cognitive ability that indicate dementia. On the second level, the staff identify deficiencies in their own knowledge, seek support from colleagues and managers within their workplace and, on the third level, outside their workplace. Staff and managers expressed a need for early and continuous guidance and education from specialists in dementia and primary healthcare. This finding indicates an urgent need for intervention research and digital support for staff in dementia care.

Johansson, M., Holst,G., & Ahlström, G.
Signs in people with intellectual disabilities: Interviews with managers and staff on the identification process of dementia.
Journal of Intellectual Disability Research, 2019, 63(7), 649-649. DOI: 10.1111/jir.12651. Presentation at the IASSIDD conference, Glasgow, Scotland, August 6-9, 2019.
Abstract: An increasing number of people with intellectual disability (ID) are reaching older ages and an increased risk of dementia diseases. Staff and managers give support in daily living and can deliver information about residents' changes in behavior. The aim of this Swedish study was to explore the identification process employed by staff and managers to detect signs of suspected dementia in people with ID within intellectual disability services (ID-services). Twenty managers and 24 staff within ID-service were interviewed and qualitative latent content analysis was applied. A model consisting of three themes on three levels of resources for the identification process of signs of suspected dementia emerged from the analysis. On the first level was the time and continuity in the care relationship, which is crucial for identifying and responding to changes in cognitive ability that indicate dementia. On the second level, the staff identifies deficiencies in their own knowledge, seek support from colleagues and managers within their workplace and, on the third level, outside their workplace. Staff and managers expressed needs for guidance and education from specialists in dementia and primary healthcare. This finding indicates an urgent need for intervention research and digital support for staff in dementia care.

Johnson, N., Fahey, C., Chicoine, B., Chong, G., & Gitelman, D.
Effects of donepezil on cognitive functioning in Down syndrome.
American Journal on Mental Retardation, 2003,108(6), 367-372. doi: 10.1352/0895-8017(2003)108<367:EODOCF>2.0.CO;2.
Abstract: This study to determined whether donepezil, an acetylcholinesterase inhibitor, would improve cognitive functioning in 19 subjects with Down syndrome and no dementia. They were assigned to either a donepezil or placebo group. Cognitive functioning and caregiver ratings were measured at baseline, 4 weeks, and 12 weeks. With the exception of one area (language), no improvement was noted in any of the cognitive subtests, behavioral scores, or caregiver ratings. Subjects in the donepezil group showed an improvement in language scores compared to subjects in the placebo group. The results suggest that donepezil may improve language performance in subjects with Down syndrome and no dementia, but further studies need to be done on a larger group to confirm this result.

Jokinen, N.S., Janicki,M.P., Hogan,M., & Force, L.T.
The middle years and beyond: Transitions and families of adults with Down syndrome
Journal on Developmental Disabilities, 2012, 18(2), 59-69.
https://oadd.org/wp-content/uploads/2012/01/41012_JoDD_18-2_59-69_Jokinen_et_al.pdf
Abstract: Normally expected transitions connect the various periods of life. Often these transitions are prompted by life events that require adaptation to a changed circumstance and may challenge both individual and family quality of life. Such transitions may be *planful* (proactive) or *demand* (reactive). Little, however, has been written about the nature of such transitions and how they specifically affect older-aged families of adults with Down syndrome. Such families are often predominate lifelong carers of adults with Down syndrome. Drawing on research and experience, the authors examined three transition points from a family perspective. Each of these points of change requires that people adapt and may lead to various outcomes, including at times outcomes that are unexpected, stressful, and challenging. The three points of transition examined include moving away from the parental home, changes occurring within a residential service (e.g., staff changes, relocations), and the reactions to the onset and course of dementia. Vignettes and quotes illustrate the complexities of these transitions and show that, even with planful management, often such transitions can go awry and produce unpredictable outcomes.

Jokinen, N.
The content of available practice literature in dementia and intellectual disability.
Dementia: The International Journal of Social Research and Practice, 2005, 4(3), 327-339.
https://journals.sagepub.com/doi/abs/10.1177/1471301205055026?journalCode=dema
Abstract: Adults with intellectual disability are living to ages seen within the general population and they, too, are at risk of developing dementia. This review was to conducted to identify the nature and content of the literature related to adults with intellectual disability and dementia and bring together guidelines for services and staff providing care. The preponderance of work between 1995 and 2004 focuses on the biomedical, diagnosis and assessment aspects of the disease. Although guidelines exist, there is a lack of published literature on the efficacy of practice strategies to guide the provision of daily care. Future research is discussed that could support continued community living and high quality of life during all stages of the disease.

Jokinen, N., Service, K., Marsack-Topolewski, C., & Janicki, M.P.
Support-staging model for caregivers of adults with intellectual disability affected by dementia
AAIC2020,Poster presentation, July 30, 2020. *Alzheimer's & Dementia,* 16(S8), First published: 07 December 2020. https://doi.org/10.1002/alz.047274
Abstract: Adults with intellectual disability (ID) and dementia are a sub-population of persons who are often un- or underserved. Most adults with ID are integrated within the general community (living autonomously, or in apartments/group residences); but significant numbers also reside with their families, particularly adults with Down syndrome. Family help/counseling approaches, such as the New York University-Caregiver Intervention (NYUCI), might benefit from a support-staging model assessment focus on what specific aid a family requires to meet their needs. Patterns of such needs have been identified that can help with providing dementia-capable care. Objective needs include: (a) information on signs and symptoms, (b) diagnostic advice, (c) understanding behavioral changes and managing dementia-related behaviors, (d) adapting homes, (e) determining daily routines most conducive to calming, (f) planning for the future, (g) finding and navigating resources, and (h) responding to end-of-life needs. Subjective needs include: (a) being informed at time of diagnosis and throughout the course of dementia, (b) coping with a profound sense of loss from knowing

the diagnosis, (c) fearing the future [including financial concerns], (d) formulating long-term plans, (e) accessing community-based coordinated care, (f) facing difficulties from the medical community, (g) feeling overwhelmed by caregiving demands, (h) feeling a sense of isolation and abandonment, and (i) facing end-of-life issues. A working group emanating from the 2016 Glasgow Summit on Intellectual Disability and Dementia organized a schema encapsulating these concerns into a support-staging model. The schema suggested four fluid stages: Diagnostic (seeking cause of changes in function, abilities, personality), Explorative (exploring dementia capable interventions), Adaptive (coping with and managing the symptoms/changes), and Closure (resolving / relief from responsibilities). Using this schema, a process (utilizing the NYUCI) is underway to operationalize a support-staging assessment instrument which would enable counseling staff to frame the state of a family's concerns, build relationships through this knowledge of the caregiver and provide tailored services to them. The outcome will enable systematic coding and organizing both objective and subjective data so that specific interventions and counseling can be adapted to meet both intermittent and continuous caregiver needs.

Jokinen, N., Janicki, M.P., Keller, S.M., McCallion, P., Force,L.T., & National Task Group on Intellectual Disabilities and Dementia Practices
Guidelines for structuring community care and supports for people with intellectual disabilities affected by dementia
Journal of Policy and Practice in Intellectual Disabilities, 2013, 10(1), 1–24. https://doi.org/10.1111/jppi.12016
Abstract: To assist families and organizations in their planning for extended care that accompanies the diagnosis of dementia, the National Task Group on Intellectual Disabilities and Dementia Practices (NTG) in the United States adopted a set of practice guidelines covering the period from when suspicions are aroused to when care ends with eventual death. These guidelines are drawn from the research literature as well as clinical experiences and demonstrated best practices. The guidelines delineate what actions should be undertaken and are presented in a manner that reflects the progressive nature of prevalent dementias. To enable the development of the most appropriate and useful services and care management for adults with intellectual disabilities affected by dementia, the NTG adopted the staging model generally accepted for practice among generic dementia services. The staging model follows the flow from a prediagnosis stage when early recognition of symptoms associated with cognitive decline are recognized through to early, mid, and late stages of dementia, and characterizes the expected changes in behavior and function. In keeping with the National Plan to Address Alzheimer's Disease recommendations for earlier and more widespread efforts to detect possible symptoms, the guidelines cite the application of the NTG-Early Detection Screen for Dementia as a first step in documenting early signs of cognitive and functional changes among people with intellectual disabilities. The guidelines also provide information on nonpharmacological options for providing community care for persons affected by dementia as well as commentary on abuse, financial, managing choice and liability, medication, and nutritional issues.

Jokinen, N., Gomiero, T., Watchman, K., Janicki, M.P. Hogan, M., Larsen, F., Service, K., & Crowe, J .
Perspectives on family caregiving of people aging with intellectual disability affected by dementia: Commentary from the International Summit on Intellectual Disability and Dementia
Journal of Gerontological Social Work, 61(4), 411-431. DOI: 10.1080/01634372.2018.1454563
Abstract: This article, an output of the 2016 International Summit on Intellectual Disability and Dementia, examines familial caregiving situations within the context of a support-staging model for adults with intellectual disability (ID) affected by dementia. Seven narratives offer context to this support-staging model to interpret situations experienced by caregivers. The multidimensional model has two fundamental aspects: identifying the role and nature of caregiving as either primary (direct) or secondary (supportive); and defining how caregiving is influenced by stage of dementia. We propose staging can affect caregiving via different expressions: (1) the "diagnostic phase," (2) the "explorative phase," (3) the "adaptive phase," and (4) the "closure phase." The international narratives illustrate direct and indirect caregiving with commonality being extent of caregiver involvement and attention to the needs of an adult with ID. We conclude that the model is the first to empirically formalize the variability of caregiving within families of people with ID that is distinct from other caregiving groups, and that many of these caregivers have idiosyncratic needs. A support-staging model that recognizes the changing roles and demands of carers of people with ID and dementia can be useful in constructing research, defining family-based support services, and setting public policy.

Jokinen, N.S., Udell, L., Stemp, S., Berman, T.
Canadian guide for community care and supports for adults with intellectual disabilities affected by dementia.
Published by the Reena Organization, Toronto, ON Canada, 2024, 78pp. https://reena.org/wp-content/uploads/2024/04/V-5-REENA-GUIDE_April-18-final-ebook-1.pdf
Abstract: Authors note that this Guide is intended to support planning and preparation for the creation of community-based dementia-capable supports and services for adults with intellectual disabilities across Canada. It is based on knowledge and feedback gathered from the Advisory Committee, focus groups, interviews, and a survey completed by disability, seniors and health organizations, as well as a scan of research and gray literature. The Guide reflects relationship-centered and person-centred care approaches, emphasizing the importance of relationships and the inclusion of adults with intellectual disabilities and dementia as active participants in planning for their future. The hope is that it will also guide the future efforts of various levels of government and national organizations to create an environment promoting collaboration and innovation.

Jordens, W. C. S., Evenhuis, H. M., & Janssen, C. G. C.
Ageing and cognitive decline in people with Down's syndrome.
British Journal of Development Disabilities, 1997, 43(85), 79-84. https://doi.org/10.1179/bjdd.1997.009
Abstract: None available.

Kåhlin, I., Kjellberg, A., & Hagberg, J-E.
Ageing in people with intellectual disability as it is understood by group home staff
Journal of Intellectual & Developmental Disability, 2016. 41(1),1-10. https://doi.org/10.3109/13668250.2015.1094038
Abstract: The number of older residents in group homes for people with intellectual disability (ID) is increasing. This interview study was focused on how group home staff address issues of ageing and being old among people with ID. Twelve members of staff at 4 different group homes in Sweden were interviewed. Findings revealed old age as something unarticulated in the group home. Group home staff felt unprepared to meet age-related changes in residents. The study also revealed that group home staff had a one-tracked way of describing the process of ageing among people with ID, which was seemingly rooted in a medical paradigm of disability. Based on this study's findings, we suggest that there is a need to raise issues and give guidance related to ageing and ID in disability policy documents to support the development of a formal culture that addresses old age and ID in disability services.

Kalsy-Lillico, S., Adams, D., & Oliver, O.
Older adults with intellectual disabilities: Issues in ageing and dementia
In (Eds) Eric Emerson, Chris Hatton, Kate Dickson, Rupa Gone, Amanda Caine, Jo Bromley
Clinical Psychology and People with Intellectual Disabilities (2nd Edition), Chapter 17, pp. 359-392. John Wiley & Sons, Ltd. Published 30 March 2012 https://doi.org/10.1002/9781118404898.ch17
Abstract: Book chapter containing content on aging and physical health, aging and mental health needs, dementia in adults with intellectual disabilities and Down syndrome, assessment and diagnosis of dementia, and behavioral presentation of dementia in people with intellectual disabilities.

Kalsy, S., & Oliver, C.
The assessment of dementia in people with intellectual disabilities: Key assessment instruments
In J. Hogg and A. Langa (eds), *Assessing Adults with Intellectual Disabilities*. Blackwell Publishing: Oxford. (2005), Chapter 15, pp 207-219. doi: 10.1002/9780470773697.ch15
Abstract: None available. This chapter contains sections titled: Introduction, diagnostic Instruments, direct performance-based neuropsychological assessments, informant-based assessment instruments, and conclusions

Kalsy, S., McQuillan, S., Adams, D., Basra, T., Konstantinidi, E., Broquard, M., Peters, S., Lloyd, V., & Oliver, C.
A proactive psychological strategy for determining the presence of dementia in adults with Down syndrome: Preliminary description of service use and evaluation
Journal of Policy and Practice in Intellectual Disabilities, 2005, 2(2), 116-125. https://doi.org/10.1111/j.1741-1130.2005.00025.x
Abstract: The authors describe and assess the experience of providing proactive screening for dementia in older adults with intellectual disabilities (ID) through a dedicated clinical psychology service within the National Health Service in England. Subjects were the first 18 participants who were referred to the clinical service or were identified as showing early signs of probable dementia in a proactive screening strategy. The screening process involved combining neuropsychological, behavioral, and health data with information from a clinical assessment of the presenting problem in a case series approach. The process of psychological assessment and formulation is illustrated together with an outline of the psychological interventions employed for early-, mid-, and late-stage dementia. An appraisal of the service strategy showed that a dedicated psychology service for dementia assessment can be effective when offering a defined and workable psychological response to the increasing presentation of dementia-associated behaviors among people with ID. Ancillary services included supporting carers in contributing to the assessment and intervention process so as to ensure appropriately responsive and respectful care management for the person with ID and dementia. The authors recommend that a multimodal stage model of intervention founded on direct performance and informant-based assessments (within a framework of differential diagnosis) be employed in supporting people with ID and dementia.

Kalsy, S., McQuillan, S., Oliver. C., Hall, S.
Manual for the "Assessment for Adults with Developmental Disabilities" (A.A.D.S.) Questionnaire
School of Psychology, University of Birmingham, Edgbaston, Birmingham B15 2TT (2000).
Scales designed to assess behaviors associated with dementia and levels of caregiving. American version is available for download from www.uic.edu/orgs/rrtcamr/dementia.

Kalsy, S., Heath, R., Adams, D., & Oliver, C.
Effects of training on controllability attributions of behavioural excesses and deficits shown by adults with Down syndrome and dementia.
Journal of Applied Research in Intellectual Disabilities, 2007, 20(1), 64 -68. https://doi.org/10.1111/j.1468-3148.2006.00341.x
Abstract: Whereas there is a knowledge base on staff attributions of challenging behavior, there has been little research on the effects of training, type of behavior and biological context on staff attributions of controllability in the context of people with intellectual disabilities and dementia. A mixed design was used to investigate the effects of three factors on care staff attributions of the controllability of challenging behavior. Pre- and post-training measures were administered to participants (n = 97) attending training on ageing, dementia and people with intellectual disabilities. Authors found no significant effects of diagnosis or type of behavior on attributions were found. There was a significant increase in knowledge after training ($P < 0.001$) and training was found to significantly decrease the attribution of controllability ($P < 0.001$). Conclusion was that the results suggest that training that focuses on aspects of change relevant to behavior can favorably influence care staff's knowledge and attributions of controllability within the context of people with Down syndrome and dementia.

Karlinsky H.
Alzheimer's disease in Down's syndrome. A review.
Journal of the American Geriatrics Society, 1986 Oct, 34(10), 728-34. doi: 10.1111/j.1532-5415.1986.tb04304.x. PMID: 2944942.
Abstract: Alzheimer's disease is a progressive dementing illness accompanied by characteristic neuropathologic changes. Although its etiology is unknown, its risk of occurrence increases with age and in relatives of affected individuals. An additional risk factor is the presence of Down's syndrome. Almost all individuals with Down's syndrome over the age of 40 have the characteristic neuropathologic changes of Alzheimer's disease at autopsy. Although clinical evidence for Alzheimer's disease in Down's syndrome is less consistent, the association between Alzheimer's disease and Down's syndrome may contribute to an understanding of Alzheimer's disease in the general population. This article summarizes the neuropathologic and clinical observations of Alzheimer's disease in Down's syndrome and reviews the hypotheses that attempt to account for this association.

Kasanzi, K, & Tariq, S.
Social impact of ageing in people with intellectual disabilities.
Journal of Geriatric Care and Research, 2018, 5(2), 52-55. https://www.academia.edu/37369632/Social_impact_of_ageing_in_people_with_intellectual_disabilities
Abstract: People with intellectual disabilities have a longer life expectancy than ever before, which is resulting in an increase in population of the elderly in this group. There is scant information about ageing related issues in this population especial the psychosocial aspects. The objective of the review is to highlight the social impact related to ageing in the growing population of the individuals with intellectual disabilities in different areas and cultures and to suggest ways of addressing any difficulties. Relevant articles were searched from electronic databases. Authors noted that there is a lack of appropriate studies to compare the care and health status for the elderly with intellectual disabilities in developed and developing countries. In different parts of the world those growing old with intellectual disabilities are managed in different ways; some are placed in specialist care homes while others are cared for by their families. There is inadequate support for this population which is leading to social isolation and marginalization and this is a major concern. Authors conclude that appropriate emphasis for the needs of the growing ageing intellectual disability population is required. Culturally appropriate actions to deal with social isolation and to develop facilities for elderly with learning disability are to be explored and put into place. There is a need to expand support for family and community care givers. Developing and improving the strategies that would address the needs will need a multidisciplinary effort.

Keater, D.B., Phelen, M.J., Taylor, L., Doran, E., Krinsky-McHale, S., Price, J., Ballard, E.E., Kreisl, W.C., Hom, C., Nguyen, D., Pulsifer, M., Lai, F., Rosas, D.H., Brickman, A.M., Schupf, N., Yassa, M.A., Silverman, W., & Lott, I.T.
Down syndrome: Distribution of brain amyloid in mild cognitive impairment
Alzheiimer's Dementia: Diagnosis, Assessment & Disease Monitoriing, 2020, 12(1), e12013. https://doi.org/10.1002/dad2.12013
Abstract: Down syndrome (DS) is associated with a higher risk of dementia. We hypothesize that amyloid beta (Aß) in specific brain regions differentiates mild cognitive impairment in DS (MCI-DS) and test these hypotheses using cross-sectional and longitudinal data. 18F-AV-45 (florbetapir) positron emission tomography (PET) data were collected to analyze amyloid burden in 58 participants clinically classified as cognitively stable (CS) or MCI-DS and 12 longitudinal CS participants. The study confirmed our hypotheses of increased amyloid in inferior parietal, lateral occipital, and superior frontal regions as the main effects differentiating MCI-DS from the CS groups. The largest annualized amyloid increases in longitudinal CS data were in the rostral middle frontal,

superior frontal, superior/middle temporal, and posterior cingulate cortices. Authors note that this study helps us to understand amyloid in the MCI-DS transitional state between cognitively stable aging and frank dementia in DS. The spatial distribution of Aß may be a reliable indicator of MCI-DS in DS.

Keller, S.M., Janicki, M.P., & Esralew, L.
Dementia: screening, evaluation, diagnosis and management
In: Rubin I.L., Merrick J., Greydanus D.E., Patel D.R. (eds) *Health Care for People with Intellectual and Developmental Disabilities across the Lifespan. Springer*, Cham. https://doi.org/10.1007/978-3-319-18096-0_116.
Abstract: The emergent acknowledgment of the increase rate of dementia among adults with intellectual and developmental disability (IDD) has led to the recognition of a lifespan approach to securing and providing services with relevant supports. Consideration of what is most needed in older age has taken on more prominence. Service organizations are responding to the emergence of age-associated neuropathologies as coincident to lifelong conditions and are attempting to adapt their services for continued community care until death. The historical focus on aging among adults with IDD and the recent focus on dementia has heightened awareness of the latter age needs of adults with IDD - and in particular those at-risk of or already affected by dementia. The positive outcome of these public health initiatives is that more dementia-capable services are being developed, technologies are improving, and there is an increased interest in maintaining quality of life through to the end-of-life, irrespective of the nature and complexity of conditions prevalent in older age.

Kenshole AV, Gallichan D, Pahl S, Clibbens J.
Lifestyle factors and Alzheimer's disease in people with Down syndrome.
Journal of Applied Research in Intellectual Diabilities, 2017 Dec;30 Suppl 1:58-66. doi: 10.1111/jar.12369. Epub 2017 Jul 30.
Abstract: Lifestyle has previously been associated with the onset of Alzheimer's disease (AD) in the typically developing population, but research investigating this association in Down syndrome (DS) is limited. Adults with DS and AD (n = 27) were compared to adults with DS without AD (n = 30) on physical activity, diet, weight, where participants currently lived, where participants had lived for the majority of their lives, educational attainment, occupational attainment and cognitive activity. There was a significant difference between samples on where participants currently lived, with the majority of the clinical sample living in institutionalized settings and the majority of the control sample living in independent/supported living settings. This may reflect a tendency to move people once they start to deteriorate which, if correct, is contrary to clinical recommendations that people with AD should be supported to "die in place." Further research into the way in which lifestyle factors, particularly living environment, could contribute to the increased risk of AD in adults with DS is required. This may support interventions aimed at preventing or delaying the onset of the disease.

Kerins, G., Petrovic, K., Bruder, M.B., & Gruman, C.,
Medical conditions and medication use in adults with Down syndrome: A descriptive analysis.
Down Syndrome Research and Practice, 2008, 12(2), 141-147.
[http://www.down-syndrome.org/reports/2009/reports-2009.pdf]
Abstract: Authors the presence of medical conditions and medication use within a sample of adults with Down syndrome. The author employed a retrospective chart review using a sample of 141 adults with Down syndrome and age range of 30 to 65 years. They identified 23 categories of commonly occurring medical conditions and 24 categories of medications used by adults with Down syndrome. From their work, the authors concluded that approximately 75% of older adults with Down syndrome in their sample experienced memory loss and dementia. Hypothyroidism, seizures, and skin problems also occurred commonly. The prevalence of cancer (i.e., solid tumors) and hypertension was extremely low. Older adults with Down syndrome used anticonvulsants more often than younger adults with Down syndrome. The use of multivitamins and medications such as pain relievers, prophylactic antibiotics, and topical ointments was common.

Kerr, D.
Down's syndrome and dementia
76 pp.
Birmingham, UK: Venture Press (1997)
Abstract: Text providing a comprehensive review of issues and practices relative to adults with Down syndrome affected by Alzheimer's disease. Covered are a range of topics related to care management, including assessment of need, communication, creating a therapeutic environment, how to maintain skills, and dealing with challenging behaviors. Also covered are specific interventions and supporting carers.

Kerr, D., Cunningham, C., & Wilkinson, H.
Learning disability and dementia Are we prepared?
Journal of Dementia Care, 2006, May/June, 17-19
Abstract: None provided

Kerr, D., Cunningham, C. & Wilkinson, H.
Responding to the pain experiences of people with a learning difficulty and dementia
Joseph Rowntree Foundation, 2006. (The Homestead, 40 Water End, York YO30 6WP). https://www.choiceforum.org/docs/jrfpainfindings.pdf
Abstract: The report explores knowledge and practice in relation to pain recognition and management amongst direct support staff, members of community learning [intellectual] disability teams and general practitioners. It also examines the understanding and experiences of pain amongst people with a learning difficulty [intellectual disability] and dementia. It identifies the dilemmas and obstacles to effective pain management, and reports on examples of good practice. The report found that the pain experiences and management of people with a learning difficulty [intellectual disabiltiy] who have dementia mirrored findings in relation to people in the general population. It did, however, identify extra and compounding issues in relation to people with a learning difficulty [intellectual disability]. The authors proffer recommendations for practitioners and service providers.

Kiddle, H., Drew, N., Crabbe, P., & Wigmore, J.
A pilot memory cafe for people with learning disabilities and memory difficulties
British Journal of Learning Disabilities, 2016, 44(3), 175-181. https://doi.org/10.1111/bld.12135
Abstract: Memory cafés have been found to normalize experiences of dementia and provide access to an accepting social network. People with learning disabilities are at increased risk of developing dementia, but the possible benefits of attending a memory café are not known. This study evaluates a 12-week pilot memory café for people with learning disabilities in terms of adaptations required and benefits of attending. Results indicate that affect levels significantly improved across the course of the café and that communication, interaction, alertness and participation in other activities improved outside the café. Future plans for attendance at memory cafés for people with learning disabilities are discussed.

Kirk, L.J., Hick, R., & Laraway, A.
Assessing dementia in people with learning disabilities: The relationship between two screening measures.
Journal of Intellectual Disabilities, 2006, 10(4), 357-364. doi:10.1177/1744629506070053.
Abstract: As life expectancy increases for people with intellectual disabilities, the impact of dementia on people with intellectual disabilities and their families, carers and services is becoming more apparent. Psychological services for intellectual disabilities are receiving an increasing number of referrals requesting dementia assessment. Health and social care services are adapting to the diverse needs of an ageing population with intellectual disabilities. The authors describe a study investigating the relationship between two assessments for dementia in people with intellectual disabilities. Carers of people with intellectual disabilities over the age of 50 (or 40 if the individual had Down syndrome) completed the Dementia Questionnaire for Mentally Retarded People (DMR) and the Adaptive Behavior Scale–Residential and Community (ABS). Overall, the two

questionnaire measures showed significant relationships. However, results suggested that both assessments have clinical value in informing individual needs and aiding diagnosis. The authors discuss the Implications for both clinical and social care services.

Kirwan, R., Sheerin, F., McGlinchey, E., McCallion, P., & McCarron, M.
Functional loss in older adults with intellectual disabilities and dementia.
Learning Disability Practice, 2022, 25(5). doi: 10.7748/ldp.2022.e2184
Abstract: Diagnosing dementia in people with intellectual disabilities can be challenging due to pre-existing cognitive impairment. In this population, functional loss could be an early indicator of dementia, but the relationship between functional loss and dementia is not well understood. This means there is a risk of delayed diagnosis and therefore a delay in care planning. To identify and compare the prevalence and age distribution of dementia in people with intellectual disabilities, differentiating between those with Down's syndrome and those with an intellectual disability not attributable to Down's syndrome, and to identify and compare functional loss in people with intellectual disabilities who have developed dementia, differentiating between those with Down's syndrome and those with an intellectual disability not attributable to Down's syndrome. This was a secondary analysis of data from waves 1 and 3 of the Intellectual Disability Supplement to the Irish Longitudinal Study on Ageing (IDS-TILDA), a nationally representative study in Ireland of older adults with intellectual disabilities as they age. Functional loss was determined by participants' level of difficulty with activities of daily living (ADLs) and instrumental activities of daily living (IADLs). The prevalence of dementia was higher among participants with Down's syndrome than among those with an intellectual disability not attributable to Down's syndrome, and the age distribution of dementia differed markedly between the two groups. Both groups experienced similar patterns of difficulty with ADLs and IADLs. IADLs posed more difficulty than ADLs, and among ADLs self-care activities posed the most difficulty. It is important to continue to investigate functional loss in older adults with intellectual disability who have developed dementia, as this should lead to earlier assessment and care planning.

Klein, C.B.,& Klinger, L.G.
Aging well and autism: A narrative review and recommendations for future research.
Healthcare (Basel), 2024 Jun 17, 12(12),1207. doi: 10.3390/healthcare12121207
Abstract: With **autism** first recognized in the 1940s, the early cohorts of autistic children are beginning to enter older adulthood. Little is known about the experiences and outcomes of autistic older adults. In the general population, "successful aging" is a dominant model among gerontologists and is used to evaluate outcomes in older adulthood. This narrative review aims to provide a framework for understanding and supporting successful aging in older autistic adults. Using Fernández-Ballesteros' four-domain model of "aging well" we review knowledge on aging and autism by examining outcomes in health and functioning, cognitive and physical functioning, positive affect and control, and social participation and engagement. Findings indicate that outcomes in autistic older adults are generally poor, marked by increased medical conditions, low adaptive skills, elevated risk of **cognitive decline**, limited physical activity, high rates of mental health conditions, low quality of life, and reduced social or community participation. Patterns of challenges are similar across cognitive abilities and profiles of autistic traits. Challenges and next steps in aging and autism research are identified, and future directions for the field are discussed.

Knox, K., Stanley, J., Hendrix, J.A., Hillerstrom, H., Dunn, T., Achenbach, J., Chicoine, B.A., Lai, F., Lott, I., Stanojevic, S., Howlett, S.E., & Rockwood, K.
Development of a symptom menu to facilitate Goal Attainment Scaling in adults with Down syndrome-associated Alzheimer's disease: a qualitative study to identify meaningful symptoms.
Journal of Patient Reported Outcomes, 2021 Jan 11, 5(1), 5. doi: 10.1186/s41687-020-00278-7.
Abstract: : As life expectancy of people with Down syndrome (DS) increases, so does the risk of Alzheimer's disease (AD). Identifying symptoms and tracking disease progression is especially challenging whenever levels of function vary before the onset of dementia. Goal Attainment Scaling (GAS), an individualized patient-reported outcome, can aid in monitoring disease progression and treatment effectiveness in adults with DS. Here, with clinical input, a validated dementia symptom menu was revised to facilitate GAS in adults living with Down Syndrome-associated Alzheimer's disease (DS-AD). Four clinicians with expertise in DS-AD and ten caregivers of adults living with DS-AD participated in semi-structured interviews to review the menu. Each participant reviewed 9-15 goal areas to assess their clarity and comprehensiveness. Responses were systematically and independently coded by two researchers as 'clear', 'modify', 'remove' or 'new'. Caregivers were encouraged to suggest additional items and recommend changes to clarify items. Median caregiver age was 65 years (range 54-77). Most were female (9/10) with 15 years of education (10/10). Adults with DS-AD had a median age of 58 years (range 52-61) and either a formal diagnosis (6/10) or clinical suspicion (4/10) of dementia. The initial symptom menu consisted of 67 symptoms each with 2-12 descriptors (589 total). The clinicians' adaptation yielded 58 symptoms each with 4-17 descriptors (580 total). Of these 580 descriptors, caregivers identified 37 (6%) as unclear; these were reworded, and one goal area (4 descriptors) was removed. A further 47 descriptors and one goal area were added to include caregiver-identified concepts. The final menu contained 58 goal areas, each with 7-17 descriptors (623 total). A comprehensive symptom menu for adults living with DS-AD was developed to facilitate GAS. Incorporating expert clinician opinion and input from caregivers of adults with DS-AD identified meaningful items that incorporate patient/caregiver perspectives.

Kobayashi, S., Yamamoto-Mitani, N., Nagata, S., & Murashima, S
End-of-life care for older adults with dementia living in group homes in Japan
Japan Journal of Nursing Science, 2008, 5(1), 31-40. doi: 10.1111/j.1742-7924.2008.00097.x.
Abstract: The aim of this study was to elucidate the components of end-of-life care provided to older adults with dementia who live in group homes (GHs) in Japan. The number of GHs in Japan is rapidly increasing. Although GHs were originally not established to care for elderly people with advanced-stage dementia, many residents remain in the GH even after their stage of dementia advances; thus, end-of-life care is required. Interviews were conducted with seven GH administrators on their experience in providing end-of-life care to their residents. The constant comparative approach was used for data collection and analysis. Four themes emerged as essential components of end-of-life care in the GH setting: (i) maintaining a familiar lifestyle; (ii) minimizing physical and mental discomfort; (iii) proactively utilizing desirable medical care; and (iv) collaborating with family members. The combination of the four components seems to be a unique characteristic of end-of-life care in GHs in Japan. These findings may be used to establish a framework for end-of-life care at GHs.

Koehl, L.H., Harp, J., Van Pelt, K.L., Head, E., & Schmitt, F.A.
Longitudinal assessment of dementia measures in Down syndrome.
Alzheimer's & Dementia : Diagnosis, Assessment & Disease Monitoring, 2020, Nov 14, 12(1), e12075. doi: 10.1002/dad2.12075. eCollection 2020.
Abstract: Early detection of dementia symptoms is critical in Down syndrome (DS) but complicated by clinical assessment barriers. The current study aimed to characterize cognitive and behavioral impairment using longitudinal trajectories comparing several measures of cognitive and behavioral functioning. Measures included global cognitive status (Severe Impairment Battery [SIB]), motor praxis (Brief Praxis Test [BPT]), and clinical dementia informant ratings (Dementia Questionnaire for People with Learning Disabilities [DLD]). One-year reliability was assessed using a two-way mixed effect, consistency, single measurement intraclass correlation among non-demented participants. Longitudinal assessment of SIB, BPT, and DLD was completed using linear mixed effect models. One-year reliability (n = 52; 21 male) was moderate for DLD (0.69 to 0.75) and good for SIB (0.87) and BPT (0.80). Longitudinal analysis (n = 72) revealed significant age by diagnosis interactions for SIB (F(2, 115.02) = 6.06, P

= .003), BPT (F(2, 85.59) = 4.56, P = .013), and DLD (F(2, 103.56) = 4.48, P = .014). SIB progression (PR) had a faster decline in performance versus no-dementia (ND) (t(159) = -2.87; P = .013). Dementia had a faster decline in BPT performance versus ND (t(112) = -2.46; P = .041). PR showed quickly progressing scores compared to ND (t(128) = -2.86; P = .014). Current measures demonstrated moderate to good reliability. Longitudinal analysis revealed that SIB, BPT, and DLD changed with age depending on diagnostic progression; no change rates were dependent on baseline cognition, indicating usefulness across a variety of severity levels in DS.

Koenig, B.R.
Aged and dementia care issues for people with an intellectual disability: Best practices (vol. 2).
80 pp.
Brighton, South Australia: MINDA, Inc. (1995)
Abstract: Text covering a range of useful topics related to service provision for dementia among persons with intellectual disabilities. Highly detailed chapters cover health issues, physical decline, behavioral changes, and social aspects. Specific remedial information is provided on communication issues and adapting the environment. A chapter also addresses counseling strategies, examining a diverse range of approaches.

Kolanowski, A., Fortinsky, R. H., Calkins, M., Devanand, D. P., Gould, E., Heller, T., Hodgson, N. A., Kales, H. C., Kaye, J., Lyketsos, C., Resnick, B., Schicker, M., & Zimmerman, S.
Advancing research on care needs and supportive approaches for persons with dementia: recommendations and rationale.
Journal of the American Medical Directors Association, 2018,19(12), 1047-1053. https://doi.org/10.1016/j.jamda.2018.07.005
Abstract: The first National Research Summit on Care, Services, and Supports for Persons with Dementia and Their Caregivers was held on October 16-17, 2017, at the National Institutes of Health. In this paper, participants from the Summit Session on Research on Care Needs and Supportive Approaches for Persons with Dementia summarize the state of the science, identify gaps in knowledge, and offer recommendations to improve science and practice in long-term care. Recommendations cover 4 areas focused on persons living with dementia: (1) symptoms (behavioral and psychological symptoms of dementia, function, cognition, and sleep); (2) dementia care settings (physical and social environments, home, and residential care); (3) living with dementia (living well with dementia, living alone with dementia, and living with dementia and intellectual and developmental disabilities); and (4) technology as a cross-cutting theme. The unique challenges of adults with ID warrant inclusion of these individuals in general studies of dementia care. The Summit noted that the National Task Group has developed practice guidelines, and a national training curriculum drawing on the dementia and ID fields. Federal efforts can bridge the aging and disability service sectors as exemplified by the Administration on Community Living inclusion of ID in dementia funding. There is a rich literature on career caregivers and family support models in aging and ID that could inform dementia care practices, and also extensive research on general dementia caregiving that has the potential to inform practice in ID.The workgroup recommended conducting comparative effectiveness research to study different integrative support models involving aging and ID networks; increasing research on community programming that supports people living with family caregivers, as well as those living in a variety of supported living and group settings; and including persons with ID and dementia and their families in research on dementia care.

Konst, M., Rasmussen, L., & Turygin, N.
Dementia.
In: Matson, J.,& Matson, M. (eds), Comorbid conditions in individuals with intellectual disabilities. (2015). Autism and Child Psychopathology Series. (pp 237-273). Springer, Cham. https://doi.org/10.1007/978-3-319-15437-4_8
Abstract: Persons with intellectual disabilities are living longer lives, and as a result of normal aging, dementia is becoming more common among these persons. This problem is compounded by the fact that specific subgroups such as persons with Down syndrome develop dementia at much earlier ages than the general population. This chapter explores defining and describing this problem in the population of persons with intellectual disabilities, its prevalence, symptom expression, and popular interventions.

Kozma, C.
Down syndrome and dementia.
Topics in Geriatric Rehabilitation, 2008, 24(1), 41–53. doi: 10.1097/01.TGR.0000311405.01555.3b
Abstract: Down syndrome (DS) is one of the most common genetic conditions with an estimated incidence of 1 in 750 in the general population. It results from an extra chromosome 21 with the total chromosome count being 47 instead of the normal 46. The classic features of DS include hypotonia, atypical facial characteristics, an increased incidence of major and minor anomalies, vision and hearing deficits, other health problems, and intellectual disabilities. People with DS are living longer and experiencing premature aging, specifically Alzheimer disease (AD). The incidence of AD among adults with DS varies significantly according to studies averaging between 11% to 22% for people aged 40 to 49 years, 24.9% for people aged 50 to 59 years, and 25.6% to 77% for people older than 60 years. All studies indicate an early onset of AD as well as an exponential increase in prevalence with age. Furthermore, senile plaques and neurofibrillary tangles, the neuropathological characteristics of AD, are seen in the brain of all people with DS. Annual screening for AD should become part of routine medical practice of older adults with DS, because an early diagnosis is important for comprehensive care.

Kramarow, E.A.
Diagnosed dementia in adults age 65 and older: United States, 2022.
National Health Statistics Reports; no 203. Hyattsville, MD: National Center for Health Statistics. 2024.
DOI: https://dx.doi.org/10.15620/cdc/154766.
Abstract: This report presents estimates of diagnosed dementia in the U.S. civilian noninstitutionalized population age 65 and older by selected sociodemographic characteristics. Data from the 2022 National Health Interview Survey were used to estimate the percentage of noninstitutionalized older adults with a dementia diagnosis. Information was self-reported unless a knowledgeable proxy responded to questions when the respondent was physically or mentally unable to answer. Prevalence of diagnosed dementia among older adults is presented by age, sex, race and Hispanic origin, veteran status, education, family income as a percentage of the federal poverty level, urbanization, and region. Estimates of dementia reporting by proxy respondent status and interview mode also are presented. In 2022, 4.0% of adults age 65 and older reported ever having received a dementia diagnosis. The percentage of adults with a dementia diagnosis was similar for men (3.8%) and women (4.2%). The percentage of adults with a dementia diagnosis increased with age, from 1.7% in those ages 65-74 to 13.1% in those age 85 and older, and decreased with rising education level, from 7.9% in adults age 65 and older with less than a high school diploma to 2.2% in those with a college degree or higher. Overall levels of older adults with a dementia diagnosis did not vary significantly by mode of interview (telephone or in person).

Krinsky-McHale SJ, & Silverman W.
Dementia and mild cognitive impairment in adults with intellectual disability: issues of diagnosis.
Developmental Disabilities Research Reviews, 2013,18(1).31-42. doi: 10.1002/ddrr.1126.
Abstract: Individuals with intellectual disability (ID) are now living longer with the majority of individuals reaching middle and even "old age." As a consequence of this extended longevity they are vulnerable to the same age-associated health problems as elderly adults in the general population without ID. This includes dementia, a general term referring to a variety of diseases and conditions causing substantial loss of cognitive ability and functional declines; adults with Down syndrome are at especially high risk. A great deal of recent effort has focused on

the very earliest detectable indicators of decline (and even prodromal stages of dementia-causing diseases). A condition called mild cognitive impairment (MCI) has been conceptually defined as a decline in functioning that is more severe than expected with typical brain aging but not severe enough to meet criteria for a diagnosis of dementia. Consensus criteria for both dementia and MCI have been developed for typically developing adults but are of limited applicability for adults with ID, given their pre-existing cognitive impairments. Early diagnosis will continue to be of growing importance, both to support symptomatic treatment and to prevent irreversible neuropathology when interventions are developed to slow or halt the progression of underlying disease. While the intellectual and developmental disabilities field has for some time recognized the need to develop best-practices for the diagnosis of MCI and dementia, there remains a pressing need for empirically based assessment methods and classification criteria.

Krinsky-McHale, S.J., & Silverman. W.
Dementia and mild cognitive impairment in adults with intellectual disability: Issues of diagnosis
Developmental Disabilities Research Reviews, 2013, 18, 31- 42. https://doi.org/10.1002/ddrr.1126
Abstract: Individuals with intellectual disability (ID) are now living longer with the majority of individuals reaching middle and even "old age." As a consequence of this extended longevity they are vulnerable to the same age-associated health problems as elderly adults in the general population without ID. This includes dementia, a general term referring to a variety of diseases and conditions causing substantial loss of cognitive ability and functional declines; adults with Down syndrome are at especially high risk. A great deal of recent effort has focused on the very earliest detectable indicators of decline (and even prodromal stages of dementia-causing diseases). A condition called mild cognitive impairment (MCI) has been conceptually defined as a decline in functioning that is more severe than expected with typical brain aging but not severe enough to meet criteria for a diagnosis of dementia. Consensus criteria for both dementia and MCI have been developed for typically developing adults but are of limited applicability for adults with ID, given their pre-existing cognitive impairments. Early diagnosis will continue to be of growing importance, both to support symptomatic treatment and to prevent irreversible neuropathology when interventions are developed to slow or halt the progression of underlying disease. While the intellectual and developmental disabilities field has for some time recognized the need to develop best-practices for the diagnosis of MCI and dementia, there remains a pressing need for empirically based assessment methods and classification criteria.

Krinsky-McHale, S.J., Zigman, W.B., Lee, J.H., Schupf, N., Pang, D., Listwan, T., Kovacs, C., & Silverman. W.
Promising outcome measures of early Alzheimer's dementia in adult with Down syndrome
Alzheimer's & Dementia : Diagnosis, Assessment & Disease Monitoring. 2020; 12(1): e12044. Published online 2020 Jul 5. doi: 10.1002/dad2.12044
Abstract: Adults with Down syndrome (DS) are at high risk for developing Alzheimer's disease (AD) and its associated dementia, warranting the development of strategies to improve early detection when prevention is possible. Using a broad battery of neuropsychological assessments, informant interviews, and clinical record review, we evaluated the psychometrics of measures in a large sample of 561 adults with DS. We tracked longitudinal stability or decline in functioning in a subsample of 269 participants over a period of 3 years, all initially without indications of clinically significant aging-related decline. Results identified an array of objective measures that demonstrated sensitivity in distinguishing individuals with incident "mild cognitive impairment" (MCI-DS) as well as subsequent declines occurring with incident dementia. **Several instruments showed clear promise** for use as outcome measures for future clinical trials and for informing diagnosis of individuals suspected of experiencing early signs and symptoms of a progressive dementia process.

Krasny, S., Yan, C., Hartley, S.L., Handen, B.L., Wisch, J.K., Boehrwinkle, A.H., Ances, B.M., Rafii, M.S.; ABC-DS consortium.
Assessing amyloid PET positivity and cognitive function in Down syndrome to guide clinical trials targeting amyloid.
Alzheimers & Dementia, 2024 Jun 28, doi: 10.1002/alz.14068. Epub ahead of print.
Abstract: Trisomy 21, or Down syndrome (DS), predisposes individuals to early-onset Alzheimer's disease (AD). While monoclonal antibodies (mAbs) targeting amyloid are approved for older AD patients, their efficacy in DS remains unexplored. This study examines amyloid positron emission tomography (PET) positivity (A+), memory function, and clinical status across ages in DS to guide mAb trial designs. Cross-sectional data from the Alzheimer Biomarker Consortium-Down Syndrome (ABC-DS) was analyzed. PET amyloid beta in Centiloids classified amyloid status using various cutoffs. Episodic memory was assessed using the modified Cued Recall Test, and clinical status was determined through consensus processes. Four hundred nine DS adults (mean age = 44.83 years) were evaluated. A+ rates increased with age, with mean amyloid load rising significantly. Memory decline and cognitive impairment are also correlated with age. These findings emphasize the necessity of tailoring mAb trials for DS, considering age-related AD characteristics. There is rapid increase in prevalence of amyloid beta (Aß) positron emission tomography (PET) positivity in Down syndrome (DS) after the age of 40 years. Aß PET positivity thresholds have significant impact on prevalence rates in DS. There is a significant lag between Aß PET positivity and clinical symptom onset in DS.

Kruse, B., Müller, S,V., & Sappok, T.
Demenz bei Menschen mit geistiger Behinderung [Dementia in people with intellectual disabilities]
*NeuroTransmitte*r, 2019, 30(3), 36-45. doi: 10.1007/s15016-019-6594-y,
Abstract :People with a mental disability are getting older and age-typical diseases are more common in them, including dementia. The symptoms and the diagnostic process, however, differ significantly in comparison to people without intellectual disabilities. However, recognizing the diagnosis is essential for adequate treatment and support.

Kuske, B., Wolfe, C., Gövert, U., & Müller, S.V.
Early detection of dementia in people with an intellectual disability – A German pilot study
Journal of Applied Research in Intellectual Disabilities, 2017, Dec, 30(S1), 49-57. https://doi.org/10.1111/jar.12347
Abstract: This study investigated the application of a newly developed neuropsychological assessment, the Wolfenbütteler Dementia Test for Individuals with Intellectual Disabilities (WDTIM) in combination with the Dementia Screening Questionnaire for Individuals with Intellectual Disabilities (DSQIID). The instruments were evaluated in a prospective 2-year follow-up study. A total of 102 people with an intellectual disability were assessed at 6-month intervals. Data were analysed using qualitative and statistical analyses. Four groups of individuals emerged from the analysis: (1) confirmed suspicion, (2) no suspicion, (3) questionable suspicion and (4) early suspicion. Significant differences were found between groups 1 and 2. The WDTIM could be administered to 90%–100% of all participants exhibiting mild-to-moderate intellectual disability and to 50% with severe intellectual disability. The WDTIM was shown to have good applicability to people with mild-to-moderate intellectual disability and to be appropriate for detecting cognitive changes. Using the two instruments in combination achieved greater accuracy in reinforcing a dementia suspicion than did using the DSQIID alone.

Kwon C, Rafati A, Ottman R, Christensen, J., Kanner, A.M., Jetté, N., Newton, C.R.
Psychiatric comorbidities in persons with epilepsy compared with persons without epilepsy: A systematic review and meta-analysis.
JAMA Neurology. Published online November 25, 2024. doi:10.1001/jamaneurol.2024.3976

Abstract: Several psychiatric disorders have been found to occur more frequently in persons with **epilepsy** (PWE) than in persons without epilepsy. To summarize the prevalence of 20 psychiatric disorders in PWE compared with persons without epilepsy, the authors undertook a search that included records from inception to February 2024 in Ovid, MEDLINE, Embase, and PsycINFO. Published epidemiological studies examining the prevalence of psychiatric disorders among PWE compared with persons without epilepsy were systematically reviewed. There were no restrictions on language or publication date. Abstracts were reviewed in duplicate, and data were extracted using a standardized electronic form. Descriptive statistics and meta-analyses are presented. Data were recorded on the prevalence of 20 psychiatric disorders among PWE compared with persons without epilepsy. Meta-analyses were performed along with descriptive analyses. The systematic search identified 10 392 studies, 27 of which met eligibility criteria. The meta-analyses included 565 443 PWE and 13 434 208 persons without epilepsy. The odds of most psychiatric disorders studied were significantly increased in PWE compared with those without epilepsy, including anxiety (odds ratio [OR], 2.11; 95% CI, 1.73-2.58); depression (OR, 2.45; 95% CI, 1.94-3.09); bipolar disorder (OR, 3.12; 95% CI, 2.23-4.36); suicidal ideation (OR, 2.25; 95% CI, 1.75-2.88) but not suicide attempt (OR, 3.17; 95% CI, 0.49-20.46); psychotic disorder (OR, 3.98; 95% CI, 2.57-6.15); schizophrenia (OR, 3.72; 95% CI, 2.44-5.67); obsessive-compulsive disorder (OR, 2.71; 95% CI, 1.76-4.15); posttraumatic stress disorder (OR, 1.76; 95% CI, 1.14-2.73); eating disorders (OR, 1.87; 95% CI, 1.73-2.01); alcohol misuse (OR, 3.64; 95% CI, 2.27-5.83) and alcohol dependence (OR, 4.94; 95% CI, 3.50-6.96) but not alcohol abuse (OR, 2.10; 95% CI, 0.60-7.37); substance use disorder (OR, 2.75; 95% CI, 1.61-4.72); **autism spectrum disorder** (OR, 10.67; 95% CI, 6.35-17.91); and attention-deficit/hyperactivity disorder (OR, 3.93; 95% CI, 3.80-4.08). In this comprehensive study, most psychiatric comorbidities examined were significantly more prevalent in PWE than in those without epilepsy. These findings show the high burden of psychiatric comorbidities in PWE. This, in turn, underscores the need for appropriately identifying and treating psychiatric comorbidity in epilepsy to manage patients effectively and improve quality of life.

Kyprianou N, Hendrix J, Hillerstrom H, Grimm R, Kirova AM, Rubenstein E.
Caregivers' perception of adults with Down syndrome willingness to participate in research.
J Intellect Disabil Res. 2023 Apr;67(4):352-361. doi: 10.1111/jir.12999. Epub 2022 Dec 21
Abstract: Historically, individuals with Down syndrome have been excluded from clinical research. Our objective was to assess the degree of interest adults with Down syndrome have in participating in research from the perspective of the caregivers who care for them. We conducted an online survey of N = 390 caregivers of adults with Down syndrome and asked about interest in research participation and demographics. Caregivers were mostly family members, older than 55 years, and White. Caregivers reported that the adult with Down syndrome that they cared for would be more comfortable participating in research that was physiological, such as research involving fit bits (70.2% would participate), exercise (63.3%) or diet apps (53.9%), whereas they would be less likely to participate in clinical trials involving more invasive procedures such as injections (10.9%) and laboratory exams like MRIs (32.0%). We found little difference by age or gender of the adult with Down syndrome or by caregiver education level. Our survey identified high interest for less invasive studies, illustrating acceptability of observational and lifestyle studies. More effort may be needed to understand fear and barriers to participation and to create tools and methods to increase interest in more invasive studies.

Lai, F., & Williams, R.S.
A prospective study of Alzheimer disease in Down syndrome.
Archives of Neurology, 1989 Aug, 46(8), 849-7853. doi: 10.1001/archneur.1989.00520440031017. PMID: 2527024.
Abstract: Ninety-six individuals with Down syndrome over age 35 years were evaluated and followed up for evidence of nontreatable dementia. Dementia was judged to be present when a functional decline occurred in areas such as orientation, memory, verbal and motor skills, and self-care abilities. Forty-nine patients with Down syndrome fit this criterion, with an average onset of dementia at 54.2 +/- 6.1 years. The prevalence of dementia in the institutionalized Down syndrome population of our study (n = 53) was 8% (2/25 patients) between 35 and 49 years, 55% (11/20 patients) between 50 and 59 years, and 75% (6/8 patients) of those over 60 years old. Of note, 41 (84%) demented individuals with Down syndrome developed seizures. Ten (20%) had parkinsonian features. Adequately treated hypothyroidism was present in 27 (59%) of 46 demented patients with Down syndrome tested. The average duration of dementia in the 23 patients who died was 4.6 +/- 3.2 years. Computed tomographic scans in 43 patients all showed brain tissue loss, most pronounced in the temporal lobes. Brains from 12 autopsied cases showed large numbers of plaques and tangles in the same locations as in persons with Alzheimer disease.

Lai, F., Kammann, E., Rebeck, G.W., Anderson, A., Chen, Y., & Nixon, R.A.
APOE genotype and gender effects on Alzheimer disease in 100 adults with Down syndrome.
Neurology,1999 Jul 22, 53(2), 331-336. doi: 10.1212/wnl.53.2.331. PMID: 10430422.
Abstract: Alzheimer disease (AD) neuropathology is present in Down syndrome (DS) after age 35, but dementia onset varies from ages 40 to 70 years. Because of small sample sizes and nonuniform determination of dementia, previous studies produced differing results on the influence of APOE subtypes on AD in DS. The authors intended to determine the influence of the APOE genotype and gender on development of AD in adults with DS to ascertain similarities with AD in the general population. A total of 100 adults with DS (ages 35 to 79 years), almost all of whom were longitudinally assessed by neurologists, underwent APOE genotyping. Dementia onset was determined using criteria applied from the Tenth International Classification of Mental and Behavioral Disorders. This cohort contains the largest number of DS subjects with dementia (n = 57) in a single study, thus increasing reliability of the results.The epsilon2 allele frequency was 4% in those with dementia versus 13% in those without dementia (p = 0.03); epsilon4 allele frequency was 18% in those with dementia versus 13% in those without dementia (p = 0.45). Using APOE-epsilon3/3 as the reference group, the risk ratio for the development of AD at any given time was 0.34 for the APOE-epsilon2/3 group (p = 0.04) and 1.44 for the APOE-epsilon(3/4,4/4) group (p = 0.25). Women were 1.77 times as likely to dement as men at any given point in time (p = 0.04). Authors noted that the epsilon2 allele confers a protective effect, and women with DS have an increased risk for AD, as in the general population. In this sample, epsilon4 does not confer a significantly increased risk for AD in DS.

Lai, F., Mhatre, P.G., Yang, Y., Wang, M.C., Schupf, N., & Rosas, H.D.
Sex differences in risk of Alzheimer's disease in adults with Down syndrome.
Alzheimer's & Dementia : Diagnosis, Assessment & Disease Monitoring. 2020 Sep 13;12(1):e12084. doi: 10.1002/dad2.12084. PMID: 32995462; PMCID: PMC7507514.
Abstract: Adults with Down syndrome (DS) older than 40 have Alzheimer's disease (AD) neuropathology and high risk for dementia, but little is known about the relationship of sex to AD risk in this population. Using nonparametric methods and Cox proportional hazards models we analyzed differences in incidence of dementia, by sex, presence of an apolipoprotein E (APOE) ?4 or ?2 allele, and dementia duration and decline in 246 adults over 40 with DS. There was no significant sex difference in risk of AD or rate of cognitive decline. APOE ?4 allele significantly increased risk of AD irrespective of sex. No significant interactions were found between sex and APOE status on AD risk. Among those who died, dementia duration was significantly longer in women. This study showed no effect of sex nor interaction between sex and APOE for risk of AD in adults with DS; however, women had longer dementia duration.

Lamsal Lamichhane, S., Ramesh, V., Opara, C. O., Khan, F.Y., Kabiraj, G., Kauser, H., Palakeel, J.J., Ali, M., Chaduvula, P., Chhabra, S., & Mohammed, L.

Treatment modalities for dementia in Down's syndrome: A literature review. *Cureus,* 2022 Aug, 14(8): e27881. doi:10.7759/cureus.27881
Abstract: Down's syndrome (DS) is the most well-known chromosomal abnormality characterized by an extra chromosome 21 and multiple systemic issues. The higher production of amyloid precursor protein (APP), the precursor peptide of beta-amyloid, predisposes persons with DS to early Alzheimer's disease (AD). The prevalence of dementia has increased as a function of the extended life expectancy of persons with DS. This review focused on the pathophysiology and treatment options, including pharmacological and non-pharmacological options in reducing the progression of dementia in people with DS. Overexpression of APP appears to be a key component in developing Alzheimer's disease in people with DS. Pharmacological therapies such as Donepezil and Memantine may have a role in the management of dementia; however, research with more persons with DS is needed. Persons with DS are predisposed to oxidative stress and subsequent DNA damage. Vitamin E, DYRK1A inhibitor, and CoQ are antioxidants that help individuals with DS fight oxidative stress. There is a positive association between regular exercise and the maintenance of memory, personality, behavior, and conversational abilities in dementia-related cognition and function in adults with DS. Very little evidence exists for appropriate pharmacological treatment of cognitive impairment and dementia in people with DS. Therefore, further research should focus on more extensive trials with well-powered cohorts to explore the prevention of cognitive decline in the DS population.

Landes, S.D., Finan, J.M., & Turk, M.A.
Reporting Down syndrome on the death certificate for Alzheimer disease/unspecified dementia deaths.
PLoS ONE, 2023, 18(2): e0281763.
https://doi.org/10.1371/journal.pone.0281763
Abstract: Death certificates are crucial for understanding population health trends including the burden of disease mortality. Accurate reporting of causes of death on these records is necessary in order to implement adequate public health policies and fund disease research. While there is evidence that Alzheimer disease and unspecified dementia are prevalent among people with Down syndrome, a 2014 Centers for Disease Control and Prevention (CDC) rule change instructing that Down syndrome should be reported as the underlying cause of death in instances when death occurred from Alzheimer disease or unspecified dementia threatens the accuracy and the utility of death certificates for this population. This study used 15 years (2005-2019) of US death certificate data for adults with and without Down syndrome. We compare the mortality burden due to Alzheimer disease and unspecified dementia prior to and after amending death certificates that report Down syndrome as the underlying cause of death. When analyzing death certificates without addressing the reporting of Down syndrome as the underlying cause of death, rates of death due to Alzheimer disease and dementia ranked as the third leading cause of death for both adults with and without Down syndrome. After amending death certificates that reported Down syndrome as the underlying cause of death, Alzheimer disease and dementia were the leading cause of death among those with Down syndrome, occurring 2.7 times more in adults with compared to without Down syndrome. The findings of this study highlight the importance of accurate mortality data for studying and addressing population health trends. The current practice of reporting Down syndrome as the underlying cause of death rather than the disease responsible for death needs to be reconsidered and modified. If not, people with Down syndrome may be further marginalized within dementia related support and research.

Landes, S.D., Stevens, J.D., & Turk, M.A.
Cause of death in adults with intellectual disability in the United States.
Journal of Intellectual Disability Research, 2021 Jan;65(1):47-59. doi: 10.1111/jir.12790.
Abstract: Prior studies report that adults with intellectual disability (ID) have cause of death patterns distinct from adults in the general population but do not provide comparative analysis by specific causes of death. Data are from the National Vital Statistics System 2005-2017 US Multiple Cause-of-Death Mortality files. We utilised adjusted odds ratios to identify causes of death that were more common for adults whose death certificate indicated ID (N = 22 512) than for adults whose death certificate did not indicate ID (N = 32 738 229), controlling for severity level of ID. We then examine the associations between biological sex and race-ethnicity and causes of death solely among adults with ID. The leading cause of death for adults with and without ID indicated on their death certificate was heart disease. Adults with ID, regardless of the severity of the disability, had substantially higher risk of death from pneumonitis, influenza/pneumonia and choking. Adults with mild/moderate ID also had higher risk of death from diabetes mellitus. Differences in cause of death trends were associated with biological sex and race-ethnicity. Efforts to reduce premature mortality for adults with ID should attend to risk factors for causes of death typical in the general population such as heart disease and cancer, but also should be cognisant of increased risk of death from choking among all adults with ID, and diabetes among adults with mild/moderate ID. Further research is needed to better understand the factors determining comparatively lower rates of death from neoplasms and demographic differences in causes of death among adults with ID.

Landes, S.D., Stevens, J.D., & Turk, M.A.
Cause of death in adults with Down syndrome in the United States.
Disability and Health Journal, 2020, 13(4),100947.
doi:10.1016/j.dhjo.2020.100947.
Abstract: Prior studies report anomalous cause of death patterns for adults with Down syndrome, but do not provide comparison of age trends for specific causes of death between adults with and without Down syndrome, or explore biological sex and racial-ethnic differences in causes of death among adults with Down syndrome. Authors aimed to better understand cause of death trends for adults, age 18 and over, with Down syndrome. Cross-sectional data were from the 2013-2017 US Multiple Cause of Death Mortality files. Adjusted odds ratios were utilized to compare cause of death trends overall, and by age, between adults with (N = 9870) and without (N = 13,323,001) Down syndrome. We also analyzed biological sex and race-ethnic differences in cause of death solely among adults with Down syndrome. Although heart disease, dementia and Alzheimer's disease, and cancer were common among adults all adults, death from these diseases was more prominent at younger ages for adults with Down syndrome. Adults with Down syndrome were also more likely to die from influenza and pneumonia, pneumonitis, respiratory failure, and choking at all ages. Distinct biological sex and racial-ethnic differences were present in causes of death among adults with Down syndrome. While efforts to reduce premature mortality for adults with Down syndrome should attend to common risk factors such as heart disease, dementia and Alzheimer's disease, and cancer, it is imperative to afford increased attention to earlier onset of these diseases, as well as increased risk of death from respiratory and swallowing/choking related disorders at all ages.

Lane, A.M., Reed, M.B., & Hawranik, P.
Aging individuals with Down syndrome and dementia as teachers: Learnings from staff in a developmental disability program in long-term care, Journal of *Gerontological Nursing,* 2019, 45(5), 17-22. Doi: 0.3928/00989134-20190328-02
Abstract: Older adults with Down syndrome (DS) and dementia are an emerging sub-population. With much longer life spans than decades ago, issues have arisen as to where these aging adults will live and how nurses and other staff in facilities can provide effective care to these individuals. This article presents a research study that examined the learnings of nurses and staff members working within a western Canadian program for older adults with DS and dementia. These learnings include: the importance of learning from each other; importance of collaboration; how individuals with developmental disabilities communicate; older adults with DS and dementia differ from older adults with dementia; and residents' impact on staff.

Lao, P., Zimmerman, M. E., Hartley, S. L., Gutierrez, J., Keator, D., Igwe, K. C., ... & Brickman, A. M.

Obstructive sleep apnea, cerebrovascular disease, and amyloid in older adults with Down syndrome across the Alzheimer's continuum.
Sleep Advances, 2022 May 5, 3(1):zpac013. doi: 10.1093/sleepadvances/zpac013.
Abstract: We determined the extent to which obstructive sleep apnea (OSA) is associated with increased cerebrovascular disease and amyloid burden, and the relation of the two processes across clinical Alzheimer's disease (AD) diagnostic groups in adults with Down syndrome (DS). Adults with DS from the Biomarkers of Alzheimer's Disease in Down Syndrome (ADDS) study were included given available research MRI (n = 116; 50 ± 8 years; 42% women) and amyloid PET scans (n = 71; 50 ± 7 years; 39% women) at the time of analysis. Participants were characterized as cognitively stable (CS; 64%), with mild cognitive impairment-DS (MCI-DS; 23%), with possible AD dementia (5%), or with definite AD dementia (8%). OSA was determined via medical records and interviews. Models tested the effect of OSA on MRI-derived cerebrovascular biomarkers and PET-derived amyloid burden, and the moderating effect of OSA and AD diagnosis on biomarkers. OSA was reported in 39% of participants, which did not differ by clinical AD diagnostic group. OSA was not associated with cerebrovascular biomarkers but was associated with greater cortical amyloid burden. White matter hyperintensity (WMH) volume (primarily in the parietal lobe), enlarged perivascular spaces, and cortical and striatal amyloid burden were greater across clinical AD diagnostic groups (CS<MCI-DS<possible AD<definite AD). OSA increased the differences in WMH volumes across clinical AD diagnostic groups, primarily in the frontal and temporal lobes. Adults with DS and OSA had greater amyloid burden and greater cerebrovascular disease with AD. Importantly, OSA may be a modifiable risk factor that can be targeted for intervention in this population at risk for AD.

Lao, P.J., Gutierrez, J., Keator, D., Rizvi, B., Banerjee, A., Igwe, K.C., Laing, K.K., Sathishkumar, M., Moni, F., Andrews, H., Krinsky-McHale, S., Head, E., Lee, J.H., Lai, F., Yassa, M.A., Rosas, H.D., Silverman, W., Lott, I.T., Schupf, N., & Brickman, A.M.
Alzheimer-related cerebrovascular disease in Down syndrome
Annals of Neurology, 2020, Dec, 88(6), 1165-1177. doi: 10.1002/ana.25905. Epub 2020 Oct 9.
Abstract: Adults with Down syndrome (DS) develop Alzheimer disease (AD) pathology by their 5th decade. Compared with the general population, traditional vascular risks in adults with DS are rare, allowing examination of cerebrovascular disease in this population and insight into its role in AD without the confound of vascular risk factors. We examined in vivo magnetic resonance imaging (MRI)-based biomarkers of cerebrovascular pathology in adults with DS, and determined their cross-sectional relationship with age, beta-amyloid pathology, and mild cognitive impairment or clinical AD diagnostic status. Participants from the Biomarkers of Alzheimer's Disease in Down Syndrome study (n = 138, 50 ± 7 years, 39% women) with MRI data and a subset (n = 90) with amyloid positron emission tomography (PET) were included. We derived MRI-based biomarkers of cerebrovascular pathology, including white matter hyperintensities (WMH), infarcts, cerebral microbleeds, and enlarged perivascular spaces (PVS), as well as PET-based biomarkers of amyloid burden. Participants were characterized as cognitively stable (CS), mild cognitive impairment-DS (MCI-DS), possible AD dementia, or definite AD dementia based on in-depth assessments of cognition, function, and health status. There were detectable WMH, enlarged PVS, infarcts, and microbleeds as early as the 5th decade of life. There was a monotonic increase in WMH volume, enlarged PVS, and presence of infarcts across diagnostic groups (CS < MCI-DS < possible AD dementia < definite AD dementia). Higher amyloid burden was associated with a higher likelihood of an infarct. The findings highlight the prevalence of cerebrovascular disease in adults with DS and add to a growing body of evidence that implicates cerebrovascular disease as a core feature of AD and not simply a comorbidity.

Larsen, F.K., Baksh, R.A., McGlinchey, E., Langballe, E.M., Benejam, B., Beresford-Webb, J., McCarron, M., Coppus, A., Falquero, S., Fortea, J., Levin, J., Loosli, S.V., Mark, R., Rebillat, A.S., Zaman, S., & Strydom, A.
Age of Alzheimer's disease diagnosis in people with Down syndrome and associated factors: Results from the Horizon 21 European Down syndrome consortium.
Alzheimer's & Dementia, 2024 May, 20(5), 3270-3280. doi: 10.1002/alz.13779. Epub 2024 Mar 20.
Abstract: : People with Down syndrome (DS) have high risk of developing Alzheimer's disease (AD). This study examined mean ages of AD diagnosis and associations with co-occurring conditions among adults with DS from five European countries. Data were collected from 1335 people with DS from the Horizon 21 European DS Consortium were used for the analysis. Mean ages of AD diagnosis ranged between 51.4 (SD 7.0) years (United Kingdom) and 55.6 (SD 6.8) years (France). Sleep-related and mental health problems were associated with earlier age of AD diagnosis. The higher number of co-occurring conditions the more likely the person with DS is diagnosed with AD at an earlier age. Mean age of AD diagnosis in DS was relatively consistent across countries. However, co-occurring conditions varied and impacted on age of diagnosis, suggesting that improvements can be made in diagnosing and managing these conditions to delay onset of AD in DS. Mean age of AD diagnosis was relatively consistent between countries Sleep problems and mental health problems were associated with earlier age of AD diagnosis APOE ε4 carriers were diagnosed with AD at an earlier age compared to non-carriers Number of co-occurring conditions was associated with earlier age of AD diagnosis No differences between level of intellectual disability and mean age of AD diagnosis.

Lautarescu, B.A., Holland, A.J., & Zaman, S.H.
The early presentation of dementia in people with Down syndrome: A systemic review of longitudinal studies
Neuropsychology Review, 2017, Mar, 27(1), 31-45. doi: 10.1007/s11065-017-9341-9. Epub 2017 Mar 13. PMID: 28289920
Abstract: Adults with Down syndrome (DS) are at a very high risk of developing early onset Alzheimer's disease (AD) due to trisomy of chromosome 21. AD is preceded by a prolonged prodromal "pre-clinical" phase presenting with clinical features that do not fulfil the diagnostic criteria for AD. It is important to clinically characterize this prodromal stage to help early detection of the disease as neuropathology of AD is almost universal by the fifth decade in DS. There is a lack of knowledge of the trajectory of decline associated with the onset of dementia in this population and early signs may be overlooked or misdiagnosed, negatively affecting the quality of life of those affected and the use of early pharmacological or psychosocial interventions. The objective of this systematic review was to evaluate the published literature on longitudinal data in order to identify the cognitive and behavioral changes occurring during the prodromal and early stages of AD in this population. Fifteen peer-reviewed articles met the inclusion criteria, including a total number of 831 participants, with the duration between baseline and follow up varying from 1 year to 47 years. Results suggest that, compared to the general population for which short-term (episodic) memory loss is the most common indicator associated with the onset of AD, in people with DS, executive dysfunction and Behavioural and Psychological Symptoms of Dementia (BPSD) are commonly observed during pre-clinical and early stages and may precede memory loss. The review highlights the importance of using a broad spectrum of assessments in the context of heterogeneity of symptoms. Theoretical and practical implications are discussed, as well as the need for further research.

Lazenby, T.
The Impact of aging on eating, drinking, and swallowing function in people with Down's syndrome
Dysphagia, 2008, 23, 88–97. https://doi.org/10.1007/s00455-007-9096-1
Abstract: Many people with Down's syndrome (DS) experience eating, drinking, and swallowing (EDS) difficulties, which can potentially lead to life-threatening conditions such as malnutrition, dehydration, and aspiration pneumonia. As the life expectancy of people with DS continues to improve, there is an increasing need to examine how the aging process may further affect these conditions. Published research studies have yet to address this issue; therefore, this article draws on the literature in three associated areas in order to consider the

dysphagic problems that might develop in aging people with DS. The areas examined are EDS development in children and adolescents with DS, EDS changes associated with aging, and EDS changes associated with **dementia** of the Alzheimer's type (DAT) because this condition is prevalent in older adults with DS. This article concludes that unlike in the general population, the aging process is likely to cause dysphagic difficulties in people with DS as they get older. Therefore, it is suggested that longitudinal studies are needed to examine the specific aspects of EDS function that may be affected by aging and concomitant conditions in DS.

Lessov-Schlaggar, C.N., Del Rosario, O.L., Morris, J.C., Ances, B.M., Schlaggar, B.L., & Constantino, J.N.
Adaptation of the Clinical Dementia Rating Scale for adults with Down syndrome
Journal of Neurodevelopmental Disorders, 2019, Dec 16, 11(1), 39. doi:10.1186/s11689-019-9300-2.
Abstract: Adults with Down syndrome (DS) are at increased risk for Alzheimer disease dementia, and there is a pressing need for the development of assessment instruments that differentiate chronic cognitive impairment, acute neuropsychiatric symptomatology, and dementia in this population of patients. We adapted a widely used instrument, the Clinical Dementia Rating (CDR) Scale, which is a component of the Uniform Data Set used by all federally funded Alzheimer Disease Centers for use in adults with DS, and tested the instrument among 34 DS patients recruited from the community. The participants were assessed using two versions of the modified CDR-a caregiver questionnaire and an in-person interview involving both the caregiver and the DS adult. Assessment also included the Dementia Scale for Down Syndrome (DSDS) and the Raven's Progressive Matrices to estimate IQ. Both modified questionnaire and interview instruments captured a range of cognitive impairments, a majority of which were found to be chronic when accounting for premorbid function. Two individuals in the sample were strongly suspected to have early dementia, both of whom had elevated scores on the modified CDR instruments. Among individuals rated as having no dementia based on the DSDS, about half showed subthreshold impairments on the modified CDR instruments; there was substantial agreement between caregiver questionnaire screening and in-person interview of caregivers and DS adults. The modified questionnaire and interview instruments capture a range of impairment in DS adults, including subthreshold symptomatology, and the instruments provide complementary information relevant to the ascertainment of dementia in DS. Decline was seen across all cognitive domains and was generally positively related to age and negatively related to IQ. Most importantly, adjusting instrument scores for chronic, premorbid impairment drastically shifted the distribution toward lower (no impairment) scores.

Lethin, C., Rahm Hallberg, I., Karlsson, S., & Janlöv, A-C.
Family caregivers experiences of formal care when caring for persons with dementia through the process of the disease
Scandinavian Journal of Caring Sciences, 2016, 30(3), 526-534. https://doi.org/10.1111/scs.12275
Abstract: Family caregivers' experiences of formal care when caring for persons with dementia through the process of the disease is sparsely investigated. Authors investigated family caregivers' experiences of formal care when caring for a person with dementia, through the stages of the disease. A qualitative approach with focus group interviews. Four focus group interviews were conducted in October 2011 with 23 spouses and adult children of persons with dementia and analyzed with content analysis. The participants' experiences of formal care when caring for a person with dementia were captured in the theme `Family caregiving requires collaboration with formal care to get support adjusted to needs specific to the stages of dementia'. This can be broken down into the categories `The dementia diagnosis - entry into formal care as a novice family caregiver', `Needing expanded collaboration with formal care to continue care at home' and `Being dependent on a nursing home and trying to maintain involvement'. Family caregiving requires collaboration with formal care to get support adjusted to the individual's needs, specific to the stages of dementia. Caregivers experience a transition process with three main turning points: the dementia diagnosis; when they realize increased need for formal care to continue caring at home; and when the person with dementia is moved into a nursing home. The interviewed caregivers experience formal care reactive to their needs and this often promoted unhealthy transitions. Formal care needs to be proactive and deliver available care and support early on in the dementia trajectory. Interventions should focus on facilitating a healthy transition for family caregivers through the trajectory of the dementia disease to ensure their well-being.

Lewis, F., Hinson-Raven, M., Saraswat, C., Cruden-Smith, L., Blunstone, R., & Bonnici-Mallia, A.M.
Dementia in people with Down syndrome.
InnovAiT: Education and Inspiration for General Practice, 2023, 16(5), 236-241. doi:10.1177/17557380231153674
Abstract: Down syndrome (trisomy 21) is one of the most common causes of intellectual disability. It is associated with characteristic physical features and multiple comorbidities, due to the process of accelerated ageing. Improved recognition and access to healthcare in recent decades has dramatically increased life expectancy. However, as a result, people with the syndrome are more at risk of developing conditions associated with ageing, significantly Alzheimer's dementia. The diagnosis of dementia remains a challenge in the context of intellectual disability, leading to delayed diagnosis and poor outcomes. The importance of recognizing early signs is a key to improving management and prognosis.

Li, R.S.Y., Kwok, H.W.M., Deb, S., Chui, E.M.C., Chan, L.K., & Leung, D.P.K.
Validation of the Chinese version of the dementia screening questionnaire for individuals with intellectual disabilities (DSQIID-CV).
Journal of Intellectual Disability Research, 2015, Apr, 59(4), 385-95. doi: 10.1111/jir.12173.
Abstract: An increasing number of people with intellectual disabilities (ID) are at risk of developing age-related disorders such as dementia because of a dramatic increase in life expectancy in this population in the recent years. There is no validated dementia screening instrument for Chinese people with ID. The Dementia Screening Questionnaire for Individuals with Intellectual Disabilities (DSQIID) was reported to be a valid, user-friendly, easy-to-use observer-rated instrument. It was developed in the UK and has good psychometric properties. Validation of a Chinese version of the DSQIID will facilitate its application among the Chinese population. The DSQIID was translated into the Chinese version (DSQIID-CV). By purposive sampling, service users with ID aged 40 years or over were recruited through two large centres serving adults with ID in Hong Kong. Carers who had taken care of the participants continuously for the past 6 months were invited to complete the DSQIID-CV. All participants were examined by qualified psychiatrists to determine the presence or absence of dementia.. Two hundred people with ID whose age ranged between 40 and 73 years (mean 51 years, SD=7.34 years) were recruited to the study. A clinical diagnosis of dementia was established in 13 participants. An overall total score of 22 as a screening cut-off provided the optimum levels of specificity (0.995) and sensitivity (0.923). The DSQIID-CV showed good internal consistency (alpha=0.945) for all its 53 items, and excellent test-retest reliability (0.978, n=46) and inter-rater reliability (1.000, n=47). Exploratory factor analysis resulted in a four-factor solution explaining 45% of the total variance. The DSQIID-CV is shown to have robust psychometric properties. It is the first valid and reliable dementia screening instrument for Chinese adults with ID.

Lifshitz, H., & Merrick, J.
Aging among persons with intellectual disability in Israel in relation to type of residence, age, and etiology.
Research in Developmental Disabilities, 2004 Mar-Apr, 25(2), 193-205. https://doi.org/10.1016/J.RIDD.2003.05.002
Abstract: This study was conducted to compare aging phenomena of persons with intellectual and developmental disability (ID) aged 40 years and older living in community residence (N = 65) with those living with their families (N = 43) in Jerusalem, Israel. All 108 persons and care givers were interviewed to ascertain health problems, sensory impairment, activity of daily living (ADL), cognitive

skills, and leisure activities. Health problem had already developed by age 40 years. The most frequent were visual (33%), hearing impairments (20%) and dental problems (30%). The community residence group displayed more medical problems, whereas individuals living at home had more dental problems. Health problems in persons with Down syndrome were significantly higher. ADL functioning for all participants was high, but persons with Down syndrome and cerebral palsy had more dependence. A decline in functioning in both residential groups was observed concerning leisure time, but scores for social life leisure activities were better for the community residential group. The data provided in this study can serve as information to develop geriatric services for persons with ID and provide a basis for comparison with peers in the general population in Israel. Dental service to persons with ID living at home should be improved

Lin, J-D., Lin, L-P., Hsia, Y-C., Hsu, S-W., Wu, C-L., & Chu, C.M.
A national survey of caregivers' perspective of early symptoms of dementia among adults with an intellectual disability based on the DSQIID scale
Research in Autism Spectrum Disorders, 2014, 8(3), 275-280.
Abstract: As life expectancy increases for persons with an intellectual disability, concerns have been raised that there will be an increased demand for health or social services, particularly to address the challenges posed by the problems of dementia. To plan services for people with an intellectual disability who might experience the consequences of aging, an important first step is to obtain epidemiological data on the prevalence of dementia in this vulnerable population. This study aimed to investigate the dementia prevalence rate and its associated demographical factors in adults with an intellectual disability in Taiwan. A national survey was conducted to recruit 460 community residents of at least 45 years of age with an intellectual disability. The Dementia Screening Questionnaire for Individuals with Intellectual Disabilities (DSQIID) was administered to caregivers to determine the symptoms of dementia in adults with an intellectual disability. The results indicated that 16.5% of the adults with an intellectual disability might have dementia conditions (DSQIID score ? 20). After controlling for other factors in a multiple logistic regression analysis, the older adults with intellectual disability (?55 vs. 45–54, OR = 2.594, 95% CI = 1.438–4.679) and those individuals with a comorbid diagnosis of mental illness or neurological disease (with vs. without, OR = 2.826, 95% CI = 1.593–5.012) had a higher risk of dementia than their counterparts. This study suggests that further longitudinal studies are needed to examine the specific aspects of the functions of living and morbidity that might be affected by aging and concomitant conditions in adults with an intellectual disability.

Lin, L.P., Hsu, S.W., Hsia, Y.C., Wu, C.L., Chu, C., & Lin, J.D.
Association of early-onset dementia with activities of daily living (ADL) in middle-aged adults with intellectual disabilities: the caregiver's perspective.
Research in Developmental Disabilities, 2014, 35(3), 626-631. doi: 10.1016/j.ridd.2013.12.015. Epub 2014 Jan 24.
Abstract: Few studies have investigated in detail which factors influence activities of daily living (ADL) in adults with intellectual disabilities (ID) comorbid with/without dementia conditions. The objective of the present study was to describe the relation between early onset dementia conditions and progressive loss of ADL capabilities and to examine the influence of dementia conditions and other possible factors toward ADL scores in adults with ID. This study was part of the "Healthy Aging Initiatives for Persons with an Intellectual Disability in Taiwan: A Social Ecological Approach" project. We analyzed data from 459 adults aged 45 years or older with an ID regarding their early onset symptoms of dementia and their ADL profile based on the perspective of the primary caregivers. Results show that a significant negative correlation was found between dementia score and ADL score in a Pearson's correlation test ($r=-0.28$, $p<0.001$). The multiple linear regression model reported that factors of male gender ($ß=4.187$, $p<0.05$), marital status ($ß=4.79$, $p<0.05$), education level (primary: $ß=5.544$, $p<0.05$; junior high or more: $ß=8.147$, $p<0.01$), Down's syndrome ($ß=-9.290$, $p<0.05$), severe or profound disability level ($ß=-6.725$, $p<0.05$; $ß=-15.773$, $p<0.001$), comorbid condition ($ß=-4.853$, $p<0.05$) and dementia conditions ($ß=-9.245$, $p<0.001$) were variables that were able to significantly predict the ADL score ($R(2)=0.241$) after controlling for age. Disability level and comorbidity can explain 10% of the ADL score variation, whereas dementia conditions can only explain 3% of the ADL score variation in the study. The present study highlights that future studies should scrutinize in detail the reasons for the low explanatory power of dementia for ADL, particularly in examining the appropriateness of the measurement scales for dementia and ADL in aging adults with ID.

Liou JJ, Lou J, Nakagiri J, Yong W, Hom CL, Doran EW, Totoiu M, Lott I, Mapstone M, Keator DB, Brickman AM, Wright S, Nelson B, Lai F, Xicota L, Dang LT, Li J, Santini T, Mettenburg JM, Ikonomovic MD, Kofler J, Ibrahim T, Head E.
Alzheimer Biomarker Consortium - Down syndrome. A neuropathology case report of a woman with Down syndrome who remained cognitively stable.
medRxiv [Preprint]. 2024 Jun 3:2024.06.01.24308050. doi: 10.1101/2024.06.01.24308050
Abstract: In this neuropathology case report, we present findings from an individual with Down syndrome (DS) who remained cognitively stable despite Alzheimer's disease (AD) neuropathology. Clinical assessments, fluid biomarkers, neuroimaging, and neuropathological examinations were conducted to characterize her condition. Notably, her ApoE genotype was E2/3, which is associated with a decreased risk of dementia. Neuroimaging revealed stable yet elevated amyloid profiles and moderately elevated tau levels, while neuropathology indicated intermediate AD neuropathologic change with Lewy body pathology and cerebrovascular pathology. Despite the presence of AD pathology, the participant demonstrated intact cognitive functioning, potentially attributed to factors such as genetic variations, cognitive resilience, and environmental enrichment. The findings suggest a dissociation between clinical symptoms and neuropathological changes, emphasizing the complexity of AD progression in DS. Further investigation into factors influencing cognitive resilience in individuals with DS, including comorbidities and social functioning, is warranted. Understanding the mechanisms underlying cognitive stability in DS could offer insights into resilience to AD neuropathology in people with DS and in the general population and inform future interventions.

Lischka, A.R., Mendelsohn, M., Overend, T., & Forbes, D.
A systematic review of screening tools for predicting the development of dementia.
Canadian Journal on Aging / La Revue canadienne du vieillissement. 2012;31(3):295-311. doi:10.1017/S0714980812000220
Abstract: Early detection of dementia is essential to guide front-line health care practitioners in further clinical evaluations and treatments. There is a paucity of literature assessing the effectiveness of screening tools to predict the development of dementia, thus we conducted a systematic review to fill this gap. The purpose of the systematic review was to make recommendations to health care practitioners on which screening tool best predicts the development of dementia and is most feasible in the primary care setting. Ten databases were searched for relevant articles, yielding 751 papers. Of these, 12 met relevance criteria for inclusion. Screening tools were assessed for test accuracy, cognitive domain coverage, predictive ability, and feasibility. Four screening tools were recommended. Addenbrooke's Cognitive Examination (ACE) was considered to be the ideal tool. A revised version of this tool is now used in clinical practice but the psychometric properties of the ACE-R remain to be established.

Listwan, T.A., Krinsky-McHale, S.J., Kovacs, C.M., Lee, J.H., Pang, D.I., Schupf, N., Tycko, B., Zigman, W.B., & Silverman. W.
Prodromal Alzheimer's disease can affect activities of daily living for adults with Down syndrome.
Alzheimers Dement (Amst), 2024 Mar 11,16(1), e12562. doi: 10.1002/dad2.12562
Abstract: Alzheimer's disease (AD) affecting adults with Down syndrome (DS-AD), like late-onset AD (LOAD) in the neurotypical population, has preclinical, prodromal, and more advanced stages. Only tasks placing high demands on cognition are expected to be affected during the prodromal stage, with activities of daily living (ADLs) typically being spared. However, cognitive

demands of ADLs could be high for adults with DS and may be affected during prodromal DS-AD. Cognitively stable cases that subsequently developed prodromal DS-AD were identified within a set of archived data from a previous longitudinal study. Measures of ADLs and multiple cognitive domains were examined over time. Clear declines in ADLs accompanied cognitive declines with prodromal DS-AD while stability in all measures was verified during preclinical DS-AD. Operationally defining prodromal DS-AD is essential to disease staging in this high-risk population and for informing treatment options and timing as new disease-modifying drugs become available. Cognitive and functional stability were demonstrated prior to the onset of prodromal DS-AD.ADL declines accompanied cognitive declines as adults with DS transitioned to prodromal AD. Declines in ADLs should be a defining feature of prodromal AD for adults with DS. Better characterization of prodromal DS-AD can improve AD diagnosis and disease staging.Improvements in DS-AD diagnosis and staging could also inform the timing of interventions.

Liu, L., Saba, A., Pascual, J.R., Miller, M.B., Hennessey, E.L., Lott, I.T., Brickman, A.M., Wilcock, D.M., Harp, J.P., Schmitt, F.A., Selkoe, D.J., Chhatwal, J.P., & Head, E.
Lecanemab and vascular-amyloid deposition in brains of people with Down syndrome.
JAMA Neurology, 2024 Oct 1, 81(10), 1066-1072, doi:10.1001/jamaneurol.2024.2579.
Abstract: Anti-ß-amyloid immunotherapy using lecanemab is becoming increasingly available to patients with Alzheimer disease (AD). Individuals with Down syndrome (DS) develop AD neuropathology by age 40 years, representing a significant cohort of genetically determined AD. To investigate the binding properties of lecanemab in the brains of people with DS, in anticipation of their inclusion in clinical trials or access to antiamyloid immunotherapies. The study included cases of postmortem brain tissue analysis from 15 individuals with DS aged 43 to 68 years that were acquired from Alzheimer Disease research centers at the University of California, Irvine and the University of Kentucky from 2008 to 2021. Data were analyzed from August 2023 through May 2024. The binding properties of lecanemab were assessed in brain tissue. The primary outcome was the extent of lecanemab binding to amyloid plaques and brain blood vessels. Tissue from 15 people (8 were female [53%]) with DS ranging in age from 43 to 68 (mean, 56.6) years were included in the study. Lecanemab-labeled amyloid plaques appeared in all 15 DS cases studied, indicating potential target engagement. However, extensive binding of lecanemab to brain blood vessels in DS was observed, raising significant safety concerns. These findings underscore the necessity for clinical trials of lecanemab in people with DS to evaluate both safety and efficacy, particularly in individuals older than 43 years. These findings suggest significant binding of lecanemab to cerebral amyloid angiopathy in DS. Lecanemab should be rigorously tested in clinical trials for AD in the DS population to determine its safety and efficacy, especially in those older than 43 years.

Livingston, G., & Strydom, A.
Improving Alzheimer's disease outcomes in Down's syndrome.
Lancet, 2012 Feb 11, 379(9815), 498-500. doi: 10.1016/S0140-6736(11)61929-6.
Abstract: A comment on Hanney et al. article. 'More than 40% of people with Down's syndrome in the UK are aged 40 years or older,1 and one in 10 of these individuals develops Alzheimer's disease. This rate rises to one in three of those who are older than 50 years and more than half of those older than 60 years.2 This progression is very distressing for parents and family carers because it can come at a time when-facing the physical and social challenges of ageing-they are least able to provide care, resulting in high caregiver burden.3 Thus, the MEADOWS trial4 reported by Marisa Hanney and colleagues in The Lancet is an important attempt to improve outcomes of adults with Down's syndrome and Alzheimer's disease.'

Llewellyn, P.
The needs of people with learning disabilities who develop dementia: A literature review
Dementia (London), 2011, 10(2), 235-247. https://doi.org/10.1177/1471301211403457
Abstract: People with [intellectual] isabilities are living longer and are increasingly developing age related conditions including dementia. If this occurs, their medical and social needs pose many challenges for services. A literature review was undertaken of articles published between 1996— 2006. Data was collected relating to the needs of people with [intellectual] disabilities and dementia, their carers and their peers. The primary medical need is for timely and accurate diagnosis. There is a multitude of diagnostic tools and advice is available as to which are most suitable for different client groups. The needs of carers are intertwined with those of people with [intellectual] disabilities and dementia and meeting their needs for education, training and increased staff numbers, has proved beneficial. Although multiple services will be responsible for the needs of this client group, there is a consensus that [intellectual] disability services should be at the heart of service provision.

Lloyd, V. , Kalsy, S., & Gatherer, A.
The subjective experience of individuals with Down syndrome living with dementia
Dementia, 2007, 6(1), 63-88. https://doi.org/10.1177/1471301207075633
Abstract: An increasing number of studies have begun to explore the subjective experience of individuals with dementia. However, despite the increased prevalence of dementia in individuals with Down syndrome, no such published research has been undertaken within this population. The aim of this study was to explore the perspectives and subjective experiences of six individuals with Down syndrome and dementia. Semi-structured interview accounts were analyzed using Interpretative Phenomenological Analysis, in order to gain a level of understanding concerning the impact of dementia upon respondents' lives and sense of self. Five main themes emerged: (1) Self-image, (2) The Relational Self, (3) Making Sense of Decline,(4) Coping Strategies and (5) Emotional Experience. Whilst the process of adjusting to dementia appeared comparable to the general population, the content of this was influenced by multiple levels of context specific to having a concomitant intellectual disability.

Lloyd, V., Kalsy, S., & Gatherer, A.
Impact of dementia upon residential care for individuals with Down syndrome
Journal of Policy and Practice in Intellectual Disabilities, 2008, 5(1), 33-38. https://doi.org/10.1111/j.1741-1130.2007.00137.x
Abstract Despite the increased prevalence of dementia in individuals with Down syndrome, relatively little is known about its impact upon care provision. Carers may be familiar with the demands of assisting a person with Down syndrome, but generally have little knowledge about the course or impact of dementia. This dissonance may lead to stress, which can have a detrimental effect on the carer and the quality of care for the recipient. In this exploratory study, the authors examined the objective and subjective impact of dementia upon paraprofessional paid carers of individuals with Down syndrome working in residential settings. The study used the Caregiver Activities Scale—Intellectual Disabilities (CAS-ID), the Caregiver Difficulties Scale—Intellectual Disabilities (CDS-ID), and the Maslach Burnout Inventory (MBI). Responses given for these measures by paraprofessional carers of individuals with Down syndrome and dementia (n = 9) were compared with responses from those caring for recipients with Down syndrome and no additional cognitive decline (n = 11). No significant differences were found in the responses from these sets of carers on measures of objective (CAS-ID) or subjective burden (CDS-ID). However, the MBI revealed that carers of individuals with Down syndrome and dementia reported significantly increased levels of emotional exhaustion. Findings suggested that, while even when there is little difference in the level of caregiving tasks or the subjective difficulties of caregiving, the onset of dementia in individuals with Down syndrome resulted in increased emotional exhaustion for carers. Additional factors not considered within this study, such as challenging behavior, may also be pertinent to carer burden.

Loi, S.M., Cations, M., & Velakoulis, D.
Young-onset dementia diagnosis, management and care
Medical Journal of Australia, 2023, 218(4), 182-189.
https://doi.org/10.5694/mja2.51849
Abstract: Young-onset dementia comprises a heterogeneous range of dementias, with onset at less than 65 years of age. These include primary dementias such as Alzheimer disease, frontotemporal and vascular dementias; genetic/familial dementias; metabolic disorders; and secondary dementias such as those that result from alcohol use disorder, traumatic brain injury, and infections. The presentation of young-onset dementia is varied and may include cognitive, psychiatric and neurological symptoms. Diagnostic delay is common, with a frequent diagnostic conundrum being, "Is this young-onset dementia or is this psychiatric?". For assessment and accurate diagnosis, a thorough screen is recommended, such as collateral history and investigations such as neuroimaging, lumbar puncture, neuropsychology, and genetic testing. Identification of dementia in people with Down syndrome and other intellectual disabilities can be difficult without premorbid cognitive assessment data. Neuropsychological assessment of a person with intellectual disability in early adulthood can be useful for detecting cognitive decline,30 and mid- to late-life behaviour changes reported by family and carers can be important early signs of neurodegeneration. The management of young-onset dementia needs to be age-appropriate and multidisciplinary, with timely access to services and consideration of the family (including children).

Loosli, S.V., Schmidt ,L., Nübling, G., Wlasich, E., Prix, C., Danek, A., & Levin, J.
Kognition bei Down-Syndrom: Entwicklung über die Lebensspanne und neuropsychologische Diagnostik im Erwachsenenalter [Cognition in Down Syndrome: Development across the Life Span and Neuropsychological Assessment in Adults].
Fortschritte der Neurologie-Psychiatrie, 2021 Sep, 89(9), 433-444. German. doi: 10.1055/a-1362-9868. Epub 2021 Mar 1.
Abstract: Down's syndrome is the most frequent genetic cause of intellectual disability. As the risk for developing Alzheimer's disease is increased in Down's syndrome, comprehensive cognitive examination is essential, both in young adults (for baseline evaluation), as well as later for diagnosing dementia. So far, there are only a few recommendations for neuropsychological assessment in Down syndrome. Here, we review the development of cognition across the life span, various causes of cognitive change in adults with Down syndrome, and procedures available for their evaluation. Furthermore, we provide recommendations for the assessment and interpretation of diagnostic findings in adults with intellectual disabilities. We conclude with recommendations for cognitive assessment in intellectual disability in general.

Loosli, S. V., Neumann, L. C., Wlasich, E., Prix, C., Koll, L., Weidinger, E., & Levin, J.
Measurement properties of the German version of the Cambridge examination for mental disorders of older people with Down syndrome and others with intellectual disabilities (CAMDEX-DS).
Journal of Intellectual & Developmental Disability, 2024, 1-13.
https://doi.org/10.3109/13668250.2024.2317794
Abstract: The CAMDEX-DS is an instrument to diagnose Alzheimer's disease (AD) in Down syndrome consisting of an informant interview and a cognitive test battery (CAMCOG-DS). Measurement properties of the German CAMDEX-DS were investigated. Fifty-five adults with Down syndrome (19-58 years) participated in this observational study. "Dementia" and "Alzheimer's dementia" (Alzheimer's disease) were diagnosed clinically and operationalised CAMDEX-ICD-10 criteria were applied to evaluate criterion validity. Validity and reliability of the CAMCOG-DS were analysed. Specificity of the interview was 69-93%; sensitivity 0-80% for "dementia"; and 0-20% for Alzheimer's disease. A complete CAMCOG-DS score was obtained in 85% (item difficulty 0.11-0.96). Construct validity and retest-reliability were low to moderate (τ = .04-.79), inter-rater reliability excellent (τ = .70-.89), internal consistency and selectivity acceptable to excellent. Currently, the CAMDEX-DS including the CAMCOG-DS are the outcome assessments for assessing dementia in Down syndrome with the best psychometric properties; however, revision is recommended.

Lott, J.D.
The rate of decline of social skills across dementing and non-dementing individuals with intellectual disabilities: A longitudinal study.
Dissertation Abstracts International: Section B: The Sciences and Engineering, 2007, 67(8-B), 2007, 4715.
https://repository.lsu.edu/gradschool_dissertations/1484/
The author sought to establish rate of decline of adaptive skills in a population of individuals with intellectual disability (ID) with dementia compared to similar adults with ID and without dementia, as well as examining the variability of positive and negative social behaviors across diagnostic classes. The participants were matched for age, sex, Down syndrome, and level of ID. The control group was screened for the presence of dementia with the Early Signs of Dementia Checklist. Rate of decline within groups was assessed by the Vineland Adaptive Behavior Scales and changes in positive and negative behaviors were measured by the Matson Evaluation of Social Skills for the Severely Retarded. (MESSIER) Prior to a diagnosis of dementia groups were equivalent. No significant differences were found for adaptive behaviors. Visual analysis of plotted means supports predicted decline in skills for both groups. Significant differences were found across time for positive social skills. Significant correlations were observed between the VABS and the MESSIER Positive domains. The findings provide support for the diagnostic utility of the MESSIER with dementia. However, no support was observed for different variances of negative behaviors across diagnostic groups. This would suggest that the measure of negative behaviors is not supported as a diagnostic tool at this time.

Lott, I.T., & Head, E.
Dementia in Down syndrome: unique insights for Alzheimer disease research
Nature Reviews Neurology, 2019, 15, 135–147.
https://doi.org/10.1038/s41582-018-0132-6
Abstract: Virtually all adults with Down syndrome (DS) show the neuropathological changes of Alzheimer disease (AD) by the age of 40 years. This association is partially due to overexpression of amyloid precursor protein, encoded by APP, as a result of the location of this gene on chromosome 21. Amyloid-ß accumulates in the brain across the lifespan of people with DS, which provides a unique opportunity to understand the temporal progression of AD and the epigenetic factors that contribute to the age of dementia onset. This age dependency in the development of AD in DS can inform research into the presentation of AD in the general population, in whom a longitudinal perspective of the disease is not often available. Comparison of the risk profiles, biomarker profiles, and genetic profiles of adults with DS with those of individuals with AD in the general population can help to determine common and distinct pathways as well as mechanisms underlying increased risk of dementia. This review evaluated the similarities and differences between the pathological cascades and genetics underpinning DS and AD with the aim of providing a platform for common exploration of these disorders.

Lott, I.Y., Doran, E., Nguyen, V.Q., Tournay, A., Movsesyan, N., & Gillenc, D.L.
Down syndrome and dementia: Seizures and cognitive decline
Journal of Alzheimer's Disease, 2012, 29(1),177-185.
doi:10.3233/JAD-2012-111613
Abstract: The objective of this study was to determine the association of seizures and cognitive decline in adults with Down syndrome (DS) and Alzheimer's-type dementia. A retrospective data analysis was carried out following a controlled study of antioxidant supplementation for dementia in DS. Observations were made at baseline and every 6 months for 2 years. Seizure history was obtained from study records. The primary outcome measures comprised the performance-based Severe Impairment Battery (SIB) and Brief Praxis Test (BPT). Secondary outcome measures comprised the informant-based Dementia Questionnaire for Mentally Retarded Persons and Vineland Adaptive Behavior Scales. Because a large proportion of patients with seizures had such severe

cognitive decline as to become untestable on the performance measures, time to "first inability to test" was measured. Adjustments were made for the potentially confounding co-variates of age, gender, APOE44 status, baseline cognitive impairment, years since dementia onset at baseline, and treatment assignment. The estimated odds ratio for the time to "first inability to test" on the SIB comparing those with seizures to those without is 11.02 (95% CI: 1.59, 76.27), a ratio that is significantly different from 1 (p = 0.015). Similarly, we estimated an odds ratio of 9.02 (95% CI: 1.90, 42.85) on the BPT, a ratio also significantly different than 1 (p = 0.006). Results from a secondary analysis of the informant measures showed significant decline related to seizures. We conclude that there is a strong association of seizures with cognitive decline in demented individuals with DS. Prospective studies exploring this relationship in DS are indicated.

Lott, I.T., & Lai, F.
Dementia in Down's syndrome: observations from a neurology clinic.
Applied Research in Mental Retardation, 1982, 3, 233–239. doi: 10.1016/0270-3092(82)90017-0.
Abstract: Clinical manifestations of dementia were reviewed in 15 Down's syndrome (DS) patients referred to a neurological clinic over a 24-month period for mental deterioration. The ages ranged from 32-64 years. One hundred percent showed personality changes and loss of independent daily living skills, the presenting symptoms in two-thirds of the cases. Other manifestations included seizures (53%), gait deterioration (73%), sphincteric incontinence (40%), and pathological release reflexes (67%). All 7 patients with CT-scans showed moderate or severe central and peripheral cortical atrophy. Detailed clinical information is presented for two patients, one of whom showed a temporary remission with imipramine. A characteristic dementia syndrome appears to be present in a subpopulation of aging DA patients with radiographic findings of Alzheimer's disease.

Lynggard, H., & Alexander, N.
'Why are my friends changing?' Explaining dementia to people with learning disabilities
British Journal of Learning Disabilities, 2004, 32(1), 30-34.
https://doi.org/10.1111/j.1468-3156.2004.00246.x
Abstract: Many publications seek to explain the causes and effects of dementia to the general population and there is evidence of the benefit of supporting carers and of establishing support groups. However, there is much less published material aimed at people with intellectual disabilities, and little focus on the specific needs of people who share their homes and lives with other people with learning disabilities who develop dementia. This article, based on group work, describes residents who had expressed bewilderment at the gradual changes they were witnessing in two of their housemates with dementia with whom they had shared a home and friendships over many years. Employing a wide range of visual aids, equipment, role plays and exercises, we sought to make the explanation of dementia as accessible and concrete as possible. The group also provided a forum for the residents to talk about the effects of living with others who develop dementia. Evaluation showed how a relatively short intervention can result in positive changes for both the people with learning disabilities who develop dementia and their peers.

Lyons, V., Nickels, J., Harrison Dening, K.
Recognising and assessing for dementia in people with learning disabilities.
Learning Disability Practice, 2024, 27(3), online. doi: 10.7748/ldp.2024.e2241
Abstract: People with [intellectual] disabilities are living longer and are therefore at risk of developing age-related conditions such as dementia. They also tend to develop dementia at a younger age than the general population. However, people with [intellectual] disabilities experience a range of health inequalities and barriers to accessing healthcare services, including dementia assessment and diagnostic services. Furthermore, the early signs and symptoms of dementia in this group tend to differ from those in the general population. It is vital, therefore, that those working with people with [intellectua] disabilities are able to recognise the early signs and symptoms of dementia in this population and know how to refer the person for assessment. The authors discuss dementia in people with [intellectual] disabilities, including early signs and symptoms, and uses a case study to explore an `ideal' dementia diagnostic pathway for this population.

Lyons, V., Oliver, E., Knifton, & Molesworth, S.
Role of Admiral Nurses in supporting people with learning disabilities and dementia
Learning Disability Practice. 2022, 25(5), ??-?? doi: 10.7748/ldp.2022.e2180
Abstract: The average age of people with learning disabilities is increasing, meaning that the number of people with learning disabilities and dementia is also rising. The care trajectory for people with learning disabilities and dementia is complex, starting with challenges in obtaining an appropriate diagnosis through to receiving appropriate and high-quality end of life care. The charity Dementia UK recognises the issues that families experience when someone in their family has a learning disability and dementia, and has developed a model of care in which Admiral Nurses, who are specialist dementia nurses, work in learning disability services. This article explores the role of the Admiral Nurse in learning disability services and examines the areas in which these specialist nurses provide tailored support. The article also outlines the expected outcomes of the service provided by these nurses.

MacDonald, S., & Summers, S.J.
Psychosocial interventions for people with intellectual disabilities and dementia: A systematic review
Journal of Applied Research in Intellectual Disabilities, 27 February 2020
https://doi.org/10.1111/jar.12722
Abstract: People with intellectual disability experience a higher prevalence of dementia, at an earlier age, than the general population. The aim of this review was to establish the psychological interventions and outcomes for individuals with intellectual disability and dementia. A search of eight electronic databases and reference lists of all included articles was conducted using PRISMA guidelines. Data were synthesized using an integrative method. Initial searching produced 2,331 papers. Twenty-one studies met the inclusion criteria. Interventions were deductively categorized into behavioural, systemic and therapeutic. All studies reported positive findings for individuals and for the systems which support them, but limited by methodological issues and neglect of the direct experience and impact on individuals themselves. The lack of a synthesis of psychosocial interventions within clinical practice, and the associated evidence, has invariably led to a lack of knowledge in practice. This is clearly evidenced by the omission of psychosocial interventions within established intellectual disability and dementia care pathways. The findings are discussed in relation to the wider literature and evidence base. Future research should aim to adopt methodologically robust designs that are inclusive of the individual experience of people with intellectual disability. The authors also posit that this review will provide essential knowledge for enacting policies that all individuals diagnosed with dementia and their carers have access to meaningful post-diagnostic care, including social and psychological care and support.

Mahon, A., Tilley, E., Randhawa, G., Pappas, Y., & Vseteckova, J.
Ageing carers and intellectual disability: a scoping review
Quality in Ageing and Older Adults, 2019, 20(4), 162-178.
https://doi.org/10.1108/QAOA-11-2018-0057
Abstract: Individuals with intellectual disability(ies) are living longer contributing to an overall increase in the average age of caregivers. The purpose of this paper is to review the literature on the physical, social and psychological needs of ageing carers of individuals with intellectual disability(ies) in the UK. A scoping review framework was used to identify literature from eleven databases, the grey literature and the references lists of relevant studies. Only primary research studies that discussed the needs of non-professional carers, aged 65+ years old, of individuals with intellectual disability(ies) in the UK were included. No date restrictions were applied. Thematic analysis was used to narratively synthesize findings. Six studies were included. Five key themes were identified: Living with fear, lack of information, rebuilding trust, proactive professional involvement and

being ignored. Housing and support information is not communicated well to carers. Professionals require more training on carer needs and trust must be rebuilt between carers and professionals. Proactive approaches would help identify carer needs, reduce marginalization, help carers feel heard and reduce the risk of care crisis. Greater recognition of mutual caring relationships is needed.

Mahmoudi, E., Lin, P., Kamdar, N., Gonzales, G., Norcott, A., & Peterson, M.D.
Risk of early- and late-onset Alzheimer disease and related dementia in adults with cerebral palsy.
Developmental Medicine and Child Neurology, 2022 Mar; 4(3), 372-378. doi: 10.1111/dmcn.15044.
Abstract: To examine the risk of Alzheimer disease and related dementia (ADRD) among adults with **cerebral palsy** (CP). Using administrative insurance claims data for 2007 to 2017 in the USA, we identified adults (45y or older) with a diagnosis of CP (n=5176). Adults without a diagnosis of CP were included as a typically developing comparison group (n=1 119 131). Using age, sex, ethnicity, other demographic variables, and a set of chronic morbidities, we propensity-matched individuals with and without CP (n=5038). Cox survival models were used to estimate ADRD risk within a 3-year follow up. The unadjusted incidence of ADRD was 9 and 2.4 times higher among cohorts of adults 45 to 64 years (1.8%) and 65 years and older (4.8%) with CP than the respective unmatched individuals without CP (0.2% and 2.0% among 45-64y and 65y or older respectively). Fully adjusted survival models indicated that adults with CP had a greater hazard for ADRD (among 45-64y: unmatched hazard ratio 7.48 [95% confidence interval {CI} 6.05-9.25], matched hazard ratio 4.73 [95% CI 2.72-8.29]; among 65y or older: unmatched hazard ratio 2.21 [95% CI 1.95-2.51], matched hazard ratio 1.73 [1.39-2.15]). Clinical guidelines for early screening of cognitive function among individuals with CP need updating, and preventative and/or therapeutic services should be used to reduce the risk of ADRD.

Makary, A.T., Testa, R., Tonge, B.J., Einfeld, S.L., Mohr, C., & Gray, K.M.
Association between adaptive behaviour and age in adults with Down syndrome without dementia: examining the range and severity of adaptive behaviour problems.
Journal of Intellectual Disability Research, 2015 Aug, 59(8), 689-702. doi:10.1111/jir.12172.://doi.org/10.1111/jir.12172
Abstract: Studies on adaptive behavior and ageing in adults with Down syndrome (DS) (without dementia) have typically analyzed age-related change in terms of the total item scores on questionnaires. This research extends the literature by investigating whether the age-related changes in adaptive abilities could be differentially attributed to changes in the number or severity (intensity) of behavioral questionnaire items endorsed. The Adaptive Behavior Assessment System-II Adult (ABAS-II Adult) was completed by parents and caregivers of 53 adults with DS aged between 16 and 56 years. Twenty adults with DS and their parents/caregivers were a part of a longitudinal study, which provided two time points of data. In addition 33 adults with DS and their parents/caregivers from a cross-sectional study were included. Random effects regression analyses were used to examine the patterns in item scores associated with ageing. Increasing age was found to be significantly associated with lower adaptive behavior abilities for all the adaptive behaviour composite scores, expect for the practical composite. These associations were entirely related to fewer ABAS-II Adult items being selected as present for the older participants, as opposed to the scores being attributable to lower item severity. This study provides evidence for a differential pattern of age-related change for various adaptive behavior skills in terms of range, but not severity. Possible reasons for this pattern will be discussed. Overall, these findings suggest that adults with DS may benefit from additional support in terms of their social and conceptual abilities as they age.

Manji, S.W.L.U.
Aging with dementia and an intellectual disability: A case study of supported empowerment in a community living home.
Dissertation Abstracts International Section A: Humanities and Social Sciences, 2009, 70(1-A), 352. https://core.ac.uk/download/pdf/143682577.pdf
Abstract: Case study explored the qualitative experience of 4 adults with intellectual disability (ID) and dementia residing in a specializing dementia support group home. Participant observation, daily living log notes, and interviews with family/friend carers, direct-care staff, and administrators were used to obtain data. The three study questions were: (I) how the onset of dementia in people with ID changes their needs, what adjustments have to be made in the support practices, and what service barriers and successes are experienced; (ii) how adults with ID and dementia experience living in a home specializing in dementia support and how stakeholders perceive this model of support; and (iii) what are the ways policymakers can better respond to the changing needs of people with ID and dementia. Two social processes were identified: 'marginalization' and 'supported empowerment'. Marginalization depicted how dementia affected adults with ID as they incurred multiple losses in ability, home, and community. Despite losses, the adults maintained their 'selfhood' with good health support, decision-making, self-agency, and autonomy as the home provided an individualized transition process, consistent and person-centered support, and elevated empathy to facilitate freedom of choice. Supported empowerment was found as an empowering social model with micro-practices that harnessed elements of empowerment necessary to support people with dual disabilities. Seven policy considerations that prevent premature placement in nursing homes, enable aging in place, and maintain a participatory life in community were recommended.

Mann, D.M.
Alzheimer's disease and Down's syndrome.
Histopathology, 1988 Aug,13(2),125-37. doi: 10.1111/j.1365-2559.1988.tb02018.x.
Abstract: The neuropathology of Down's syndrome at middle age is compared with that of Alzheimer's disease at that age, through a review of the published literature and from the author's personal observations. The pathological changes of Down's syndrome at middle age, i.e. the form and distribution of senile plaques and neurofibrillary tangles, and the pattern of involvement (atrophy) of neuronal systems are qualitatively the same as those of Alzheimer's disease at that age. Quantitative differences do occur and these may relate to biological or sociological variations inherent to the two parent populations. It is concluded that, in pathological terms, patients with Down's syndrome at middle age do indeed have Alzheimer's disease. Some ways in which a study of patients with Down's syndrome can give insight into the nature and development of the pathological changes of Alzheimer's disease are put forward and discussed

Mansell, J., Beadle-Brown, J., Whelton, B., Beckett, C., & Hutchinson, A.
Effect of service structure and organization on staff care practices in small community homes for people with intellectual disabilities
Journal of Applied Research in Intellectual Disabilities, 2008, Sep, 21(5), 398-413. https://doi.org/10.1111/j.1468-3148.2007.00410.x
Abstract: An important question in community living is what factors influence the extent to which staff provide 'active support'. Engagement, care practices and a range of staff and organizational characteristics were studied in 72 residential homes serving 359 adults with intellectual disabilities. Managers in 36 settings were trained in person-centred active support (PCAS). A group comparison design and multivariate analysis was used to investigate the relationship between variables. The PCAS group showed more active support, assistance, other contact from staff and engagement in meaningful activity but no difference in choice-making or assessment of participation in activities of daily living. The PCAS group had more staff with a professional qualification, were more likely to think that challenging behaviour was caused by lack of stimulation, had attitudes more in line with a policy of community care, rated most care tasks as less difficult, and were more organized to deliver active support. The comparison group were more likely to think that challenging behaviour was learned negative behaviour, showed more teamwork and were more satisfied. Multivariate analysis identified a range of staff and organizational variables associated with engagement and active support. The results suggest that some variables which

have not hitherto been studied in relation with active support are associated with it. Professional qualification, knowledge and experience appear to be important as do some staff attitudes, clear management guidance, more frequent supervision and team meetings, training and support for staff to help residents engage in meaningful activity.

Margallo-Lana M.L., Moore, P.B., Kay, D.W., Perry, R.H., Reid, B.E., Berney, T.P., Tyrer, S.P.
Fifteen-year follow-up of 92 hospitalized adults with Down's syndrome: incidence of cognitive decline, its relationship to age and neuropathology
Journal of Intellectual Disability Research, 2007, 51(6), 463-477.
doi: 10.1111/j.1365-2788.2006.00902.x.
Abstract: The clinical and neuropathological features associated with dementia in Down's syndrome (DS) are not well established. To examine clinico-pathological correlations and the incidence of cognitive decline in a cohort of adults with DS. A total of 92 hospitalized persons with DS were followed up from 1985 to December 2000. At outset, 87 participants were dementia-free, with a median age of 38 years. Assessments included the Prudhoe Cognitive Function Test (PCFT) and the Adaptive Behavior Scale (ABS), to measure cognitive and behavioral deterioration. Dementia was diagnosed from case records and caregivers' reports. Eighteen (21%) patients developed dementia during follow-up, with a median age of onset 55.5 years (range 45-74). The PCFT demonstrated cognitive decline among those with a less severe intellectual disability (mild and moderate) but not among the profoundly disabled people (severe and profound). Clinical dementia was associated with neuropathological features of Alzheimer's disease, and correlated with neocortical neurofibrillary tangle densities. At the age of 60 years and above, a little more than 50% of patients still alive had clinical evidence of dementia. Authors concluded that clinical dementia associated with measurable cognitive and functional decline is frequent in people with DS after middle age, and can be readily diagnosed among less severely intellectually disabled persons using measures of cognitive function such as the PCFT and behavioral scales such as the ABS. In the more profoundly disabled people, the diagnosis of dementia is facilitated by the use of behavioral and neurological criteria. In this study, the largest prospective DS series including neuropathology on deceased patients, the density of neurofibrillary tangles related more closely to the dementia of DS than senile plaques. In people with DS surviving to middle and old age, the development of dementia of Alzheimer type is frequent but not inevitable, and some people with DS reach old age without clinical features of dementia.

Markar, T.N., Cruz, R., Yeoh, H., & Elliott, M.
A pilot project on a specialist memory clinic for people with learning disabilities
British Journal of Developmental Disabilities, 2006, 52(102), 37-46.
https://doi.org/10.1179/096979506799103640
Abstract: The aim of this pilot project was to evaluate the usefulness of establishing aspecially designed " Memory Clinic" for the assessment and diagnosis of dementia in people with learning disabilities. This pilot memory clinic was set up by re-organization of existing resources, especially in terms of professional time. The core team members were a psychiatrist, psychologist and a community nurse from the specialist learning disability team with a special interest in dementia and its treatment. Over a period of 8 months a total of 12 assessments were carried out. Seven females and 4 males were assessed, the age range being 41 years – 83 years; one being a repeat assessment. In 3 individuals a definitive diagnosis of dementia was made. Six of these service users alsohad associated Down's syndrome. In the Downs syndrome group 2 had a definitive diagnosis of dementia and for1 the diagnosis was inconclusive at the initial assessment. Authors note that the professionals involved in the assessment process need to be specially trained in dealing with the issues related to identifying and interpreting the significance of changes in function, behavior, and personality in adults with learning disabilities

Marler, R., & Cunningham, C.
Down's Syndrome and Alzheimer's Disease: A Guide for Carers.
39 pp.
London: Down's Syndrome Association [155 Mitcham Road, London, UK SW17 9PG] (1994).
Abstract: This booklet for community carers and agency staff covers some of the fundamentals concerning adults with Down syndrome and Alzheimer's disease, including information on obtaining diagnoses, approaches to care management, and securing services in the UK. Contains some vignettes and a small glossary and references.

Marrus, N., Koth, K. A., Hellings, J. A., McDonald, R., Gwynette, M. F., Muhle, R., Lohr, W. D., & Vasa, R. A..
Psychiatry training in autism spectrum disorder and intellectual disability: Ongoing gaps and emerging opportunities.
Autism, 2023 Apr, 27(3), 679-689Ã). https://doi.org/10.1177/13623613221112197
Abstract: Autism spectrum disorder and intellectual disability are associated with psychiatric comorbidities, yet a 2009 study of US child and adolescent psychiatry program directors indicated that psychiatry residents receive insufficient training in autism spectrum disorder/intellectual disability. This follow-up study surveyed child and adolescent psychiatry and general psychiatry program directors to assess (1) the current extent of residency training in autism spectrum disorder/intellectual disability, (2) program director perceptions of educational topics and resident competency in autism spectrum disorder/ intellectual disability, and (3) preferred resources to strengthen autism spectrum disorder/intellectual disability training. As in 2009, many child and adolescent psychiatry program directors reported few lecture hours, although current child and adolescent psychiatry residents saw slightly more patients with autism spectrum disorder but not intellectual disability. General psychiatry program directors reported fewer lecture hours in autism spectrum disorder/intellectual disability and fewer patients with autism spectrum disorder than child and adolescent psychiatry program directors. Both child and adolescent psychiatry and general psychiatry program directors recognized the importance of a range of educational topics in autism spectrum disorder/intellectual disability. Child and adolescent psychiatry program directors reported higher resident competency, and lecture hours and patients seen moderately correlated with resident competency. Program directors indicated that online videos and other resources would help improve autism spectrum disorder/intellectual disability training in their programs. Collectively, these findings suggest minimal improvements in autism spectrum disorder/intellectual disability training over the past decade and highlight the urgent need to advance psychiatry training in this field through dissemination of resources.

Marsack-Topolewski, C.N. & Sameul, P.S.
Caregivers' perceptions of family quality of life of individuals with developmental disabilities comorbid with dementia: A pilot study.
Journal for ReAttach Therapy and Developmental Diversities, 2020 Dec 25; 3(2), 56-70. https://doi.org/10.26407/2020jrtdd.1.38
Abstract: Although individuals with intellectual/ developmental disabilities (I/DD) are living longer than in the past, they also are exposed to age-related changes in health and well-being. They are prone to acquire dementia that often manifests earlier and more frequently than in the general population. However, there is sparse knowledge on the daily challenges that affect the quality of life of the individuals with I/DD and comorbid dementia and their family caregivers.
This pilot study examined strengths and challenges of individuals with dual diagnoses of I/DD and dementia using the family quality of life (FQOL) framework. Cross-sectional data was gathered from a convenience sample of family caregivers using a web-based electronic survey. This study aims to identify the common and differential elements of the DLD (SLI) and LD through a quantitative and qualitative analysis. The variables of interest in this study were the levels of importance and satisfaction attributed to the nine FQOL domains, and overall FQOL. The mean level of importance was higher than the associated ratings of satisfaction in eight of the nine domains, with an overall importance mean of 4.15 and satisfaction mean of 3.28. Analysis of the open-ended comments indicated that the negative impact of social isolation, compound caregiving, and dynamically changing caregiving needs on overall FQOL was

balanced by participants' values and beliefs. A statistical analysis (Student's t test) was conducted in order to compare the children in LD and DLD groups. The data obtained from this analysis along with LSA indicate that the language skills differ between the two groups in the following aspects: lexical, pragmatic, semantic, syntactic, morphological and phonological. Significant differences ($p < .05$) occur for the start of speech therapy age, phonological disorder, passive vocabulary and language psychological age. Results for active vocabulary did not indicate a statistical difference between LD and DLD children. The discrepancies in the FQOL domains pertaining to formal and informal services and social supports elucidate a need to empower families with high caregiving needs through research, practice and policy. Providers should be cognizant of the needs of individuals with I/DD and dementia comorbid, as well as the needs of their family caregivers.

Martínez-Cué, C., & Rueda, N.
Signalling pathways implicated in Alzheimer's disease neurodegeneration in individuals with and without Down syndrome.
International Journal of Molecular Sciences, 2020 Sep 20, 21(18), 6906. doi: 10.3390/ijms21186906
Abstract: Down syndrome (DS), the most common cause of intellectual disability of genetic origin, is characterized by alterations in central nervous system morphology and function that appear from early prenatal stages. However, by the fourth decade of life, all individuals with DS develop neuropathology identical to that found in sporadic Alzheimer's disease (AD), including the development of amyloid plaques and neurofibrillary tangles due to hyperphosphorylation of tau protein, loss of neurons and synapses, reduced neurogenesis, enhanced oxidative stress, and mitochondrial dysfunction and neuroinflammation. It has been proposed that DS could be a useful model for studying the etiopathology of AD and to search for therapeutic targets. There is increasing evidence that the neuropathological events associated with AD are interrelated and that many of them not only are implicated in the onset of this pathology but are also a consequence of other alterations. Thus, a feedback mechanism exists between them. In this review, we summarize the signalling pathways implicated in each of the main neuropathological aspects of AD in individuals with and without DS as well as the interrelation of these pathways.

Mason, D., Stewart, G.R., Capp, S.J., & Happé, F.
Older age autism research: A rapidly growing field, but still a long way to go.
Autism Adulthood, 2022 Jun 1, 4(2), 164-172. doi: 10.1089/aut.2021.0041
Abstract: There is a paucity of research involving older autistic people, as highlighted in a number of systematic reviews. However, it is less clear whether this is changing, and what the trends might be in research on autism in later life. We conducted a broad review of the literature by examining the number of results from a search in three databases (PubMed, Embase, PsycINFO) across four age groups: childhood, adolescence, adulthood, and older age. We also examined the abstracts of all the included articles for the older age group and categorized them under broad themes. Our database search identified 145 unique articles on autism in older age, with an additional 67 found by the authors (hence, the total number of articles in this review is 212). Since 2012, we found a 392% increase in research with older autistic people, versus 196% increase for childhood/early life, 253% for adolescence, and 264% for adult research. We identify 2012 as a point at which, year-on-year, older age autism research started increasing, with the most commonly researched areas being cognition, the brain, and genetics. However, older adult research only accounted for 0.4% of published autism studies over the past decade. This increase reflects a positive change in the research landscape, although research with children continues to dominate. We also note the difficulty of identifying papers relevant to older age autism research, and propose that a new keyword could be created to increase the visibility and accessibility of research in this steadily growing area.

Mattheys, K., Boustead, I., Doyle, A., & Watchman, K.
'Life through a lens': understanding the impact of dementia, a participatory action project led by people with intellectual disability
Journal of Intellectual Disability Research, 2019, 63(8), 650.
Abstract: Co-researchers with an intellectual disability are part of a team looking at the effects of non-drug interventions with people who have dementia, including people with Down syndrome. Photovoice is a method of data collection and analysis combining photography with social action supporting the inclusion of people typically excluded from research. Three co-researchers with intellectual disabitly working on the 'Life through a Lens' research project attended training in photovoice methodology and use of the camera, followed by a series of practice exercises. Each engaged in participant observation to understand the impact of non-drug interventions on peers with an intellectual disabitly and dementia. Photographs were then taken that represented their feelings about the intervention followed by a group discussion with wider research team. Two of the co-researchers believed that their peers benefitted from the non-drug interventions. For example, after observing changes to the home environment of one participant, the co-researcher discussed the relaxing and calming effect this created and how it helped her to be safer at home; his photography reflected the security he observed. Such photography can help with reflecting perceptions of the effect of non–drug interventions in dementia care, offering greater authority to co-researchers with intellectual disability.

Maure-Blesa, L., Carmona-Iragui, M., Lott, I., Head, E., Wisniewski, T., Rafii, M.S., Espinosa, J., Flórez, J., Mobley, W.C., Holland, A., Strydom, A., Zaman, S., & Fortea J.
The history of Down syndrome-associated Alzheimer's disease; past, present, and future.
Alzheimer's & Dementia, 2025 Jun, 21(6), e70158. doi: 10.1002/alz.70158.
Abstract: The landscape of Down syndrome-associated Alzheimer's disease (DSAD) research reflects decades of scientific endeavor and collaborative effort, charting a remarkable journey from initial observations to the elucidation of complex genetic and molecular mechanisms. This perspective article chronicles key milestones and breakthroughs, paying homage to the pioneering scientists and advancements that have shaped the field. A thorough review of historical and contemporary literature offers a comprehensive narrative, highlighting the evolution of knowledge surrounding DSAD, from early recognition to the characterization of clinical presentation and natural history. The unique challenges and ethical considerations associated with DSAD populations are also examined, underscoring the importance of tailoring research and clinical approaches. By reflecting on the field's trajectory, this work celebrates past achievements while emphasizing the critical need for sustained research efforts. As part of a special issue, this article provides a foundation for appreciating the challenges and opportunities that lie ahead in advancing DSAD understanding and care. This article provides a comprehensive overview of Down syndrome-associated Alzheimer's disease (DSAD) history, from early descriptions to its recognition as a genetic form of AD. It reflects on historical challenges faced by individuals with intellectual disabilities in achieving inclusion in scientific research. This historical perspective highlights the critical contributions of individuals with DS in advancing understanding of AD natural history. It explores pivotal milestones and efforts that have driven progress in DSAD research. Finally, it provides context to understand challenges and opportunities in DSAD research and its future directions.

May, H.L., Fletcher, C., Alvarez, N., Zuis, J., & Cavallari, S.G.
Alzheimer's disease and Down syndrome: A manual of care
Wrentham, Mass.: Alzheimer's Committee of Wrentham Developmental Center (1996)
89 pp.
Abstract: A 9-chapter staff training manual covering the basic issues related to the occurrence of Alzheimer's disease in adults with Down syndrome. Chapters include an introduction, Alzheimer's disease and Down syndrome, assessment, family and guardian considerations, early Alzheimer's disease, mid-stage Alzheimer's disease, feeding and nutrition concerns, and understanding difficult behaviors. Appendix contains a "Level of Capacity Scale," and table outlining implications and treatment suggestions for persons with intellectual disabilities affected by dementia.

McBrien, J., Whitwham, S., Olverman, K., & Masters, S.
Screening adults with Down's syndrome for early signs of Alzheimer's disease.
Tizard Learning Disability Review, 2005, 10(4), 23-32.
https://www.emerald.com/insight/content/doi/10.1108/13595474200500035/full/html
Abstract: Given the now well-recognized risk of Alzheimer's Disease (AD) for adults with Down's Syndrome (DS) as they reach middle age, services for people with learning disability (LD) need to meet this new challenge. Good practice guidance from the Foundation for People with Learning Disabilities recommended that every service for people with learning disability should set up a register of adults with DS, conduct a baseline assessment of cognitive and adaptive functioning before the age of 30 years, develop specialist skills in this area, offer training to other professionals, front-line staff and carers, and seek high-quality co-ordination between agencies. This article reports the progress of one LD service in meeting these challenges, highlighting the successes and difficulties that may guide other teams considering such a development.

McCallion, P.
Maintaining communication
In M.P. Janicki & A.J. Dalton (Eds.), Dementia, Aging, and Intellectual Disabilities
pp. 261-277
Philadelphia: Brunner-Mazel (1999)
Abstract: This book chapter is based on the premise that progression of dementia among persons with intellectual disabilities appears to be similar to that in the general population. Therefore, it explores how existing service models and programs may be adapted for the population with intellectual disabilities. A five part program, Maintaining Communication and Independence (MCI), is proposed which adapts an existing program for persons with dementia to better meet the needs of persons with intellectual disabilities. The five parts to MCI are: (1) strengths identification and deficit assessment, (2) environ-mental modification, (3) good communication, (4) memory aids, and (5) taking care of the carer.

McCallion, P., & Janicki, M.P.
Intellectual disabilities and dementia (Computer-based Course)
2 CD-Rom set
Center for Excellence in Aging Services, School of Social Welfare, Richardson 208, University at Albany, Albany, New York 12222 (2002)
Abstract: 2 disk set - usable on Windows 9.X/2000 on 233 MHZ Pentium or faster with audio/video playback. Instructional course on aging, intellectual disabilities and dementia. Contains digital video version of "Dementia and People with Intellectual Disabilities– What Can We Do?"

McCallion, P., McCarron, M., & Force, L.T
A measure of subjective burden for dementia care: the Caregiving Difficulty Scale – Intellectual Disability
Journal of Intellectual Disability Research, 2005, 49(5),
365-371.https://doi.org/10.1111/j.1365-2788.2005.00670.x
Abstract: It has been suggested in the literature on family caregiving for persons with Alzheimer's dementia (AD) that levels of objective and subjective burden among carers often predict institutionalization of the persons with AD. There is a paucity of measures to assess whether perceived burden among formal caregivers may also predict movement to more restrictive settings for persons with intellectual disabilities (ID) and AD. This study focused upon the development of a measure of subjective burden, The Caregiving Difficulty Scale – Intellectual Disability (CDS-ID) as a first step in addressing this measurement deficit. An existing caregiver subjective burden scale, the Caregiving Hassles Scale (CHS) was adapted for use with 203 staff caregivers of persons with ID and AD. Preliminary testing of existing CHS items and proposed new items was carried out in two countries, Ireland and the USA. Confirmatory factor analysis with the existing items and exploratory factor analysis with existing and proposed new items for the scale was used to establish the content and test the psychometric properties of a revised scale, the CDS-ID. On the existing CHS items, staff carers appeared to experience greater subjective burden than has been reported for family caregivers. However, the psychometric properties of the CHS found with this population were poor. Factor analysis produced a revised scale, the CDS-ID with three subscales with Cronbach alphas ranging from 0.75 to 0.93 and 38 items overall. This new scale when used with objective burden and other scales offers an opportunity to more systematically measure the difficulties staff experience when caring for persons with ID who present with symptoms of AD.

McCallion, P., Nickle, T., & McCarron, M.
A comparison of reports of caregiver burden between foster family care providers and staff caregivers in other settings: A pilot study
Dementia (London), 2005, 4(3), 401-412
https://doi.org/10.1177/1471301205055034
Abstract: There has been increasing concern about the impact of dementia symptoms on the lives and on the care being provided for persons with intellectual disability (ID) in out-of-home settings. One such setting that has received little attention is foster family care homes. These settings in the USA replicate family living and while some supports and resources are provided, they are not designed to meet intensive care needs. As a preliminary step in understanding family experiences and to expand the range of interest in Alzheimer's disease (AD) in persons with ID beyond traditional out-of-home settings, a pilot study was initiated that included aging persons with ID and symptoms of AD who were living in foster family care settings in two regions of New York State as well as more traditional out-of-home care subjects. Comparisons of matched samples on subjective and objective burden measures suggest that there are few differences in experiences. The limitations of these findings are considered and recommendations made for future, related research.

McCarron, M.
Some issues in caring for people with the dual disability of Down's syndrome and Alzheimer's dementia
Journal of Learning Disabilities for Nursing, Health and Social Care, 1999, 3(3), 123-129. https://doi.org/10.1177/174462959900300302
Abstract: Virtually all individuals with Down's syndrome over the age of 35 years have neurological changes characteristic of Alzheimer's disease. It has become increasingly recognized that people with Down's syndrome and dementia have very special needs, and those who care for them require specialist knowledge and skills. This paper aims to explore some important issues in caring for persons with this dual disability. It commences with a brief outline on the prevalence of dementia in this population. Diagnostic issues and the clinical presentation of dementia in persons with Down's syndrome are reviewed. In an attempt to help staff respond to the opportunities and challenges they encounter, issues discussed, include: promoting well-being, developing a shared vision on which to build practice, mealtimes -- a therapeutic event, reality orientation and validation therapy, communication, activity and entertainment.

McCarron, M., Gill, M., Lawlor, B., & Begley, C.
Time spent caregiving for persons with the dual disability of Down's syndrome and Alzheimer's dementia: Preliminary findings
Journal of Learning Disabilities, 2002, 6(3), 263-279.
https://doi.org/10.1177/1469004702006003036
Abstract: Persons with Down's syndrome (DS) are at increased risk of Alzheimer's type dementia (AD) compared with the general population. Little attention has been paid to the current and future impact of AD on caregivers and clients in residential and community settings. This study sought to test if the Caregiver Activity Survey-Intellectual Disability (CAS-ID) would be useful in measuring time spent by professional caregivers aiding persons with DS and AD. Preliminary findings suggest that staff caregiving time increases significantly when a person with DS experiencessymptoms of dementia. No significant

differences were reported in time spent caregiving for subjects at mid-stage versus end-stage dementia; however, the nature and tasks of caregiving change as dementia progresses. This study supports the utility of the CAS-ID in measuring time spent caregiving for persons with AD and DS. Care providers must plan appropriate models of health and social care to effectively address these needs.

McCarron, M., Gill, M., Lawlor, B., & Beagly, C.
A pilot study of the reliability and validity of the Caregiver Activity Survey – Intellectual Disability (CAS-ID)
Journal of Intellectual Disability Research, 2002, 46, 605-612. doi: 10.1046/j.1365-2788.2002.00437.x.
Abstract: Authors undertook to amend the Caregiver Activity Survey (Davis et al., 1997) and apply it for use with caregivers of persons with intellectual disabilities. Under this study, the CAS-ID was tested with 30 adults and convergent validity was assessed by comparing the CAS-ID with other measures of cognitive and functional impairment of adults with intellectual disabilities. Final version of the CAS-ID contains 8 items: dressing, bathing/showering, grooming, toileting, eating and drinking, housekeeping, nursing care-related activities, and supervision/behavior management. Authors content that the CAS-ID has the potential for identifying and measuring care and resource requirements for people experiencing decline associated with dementia.

McCarron, M., Gill, M., McCallion, P., & Begley, C.
Health co-morbidities in ageing persons with Down syndrome and Alzheimer's dementia.
Journal of Intellectual Disability Research, 2005, 49(7), 560-566. doi:10.1111/j.1365-2788.2005.00704.x.
Abstract: Consideration of the relationship between physical and mental health co-morbidities in ageing persons with Down syndrome (DS) and Alzheimer's dementia (AD) is of clinical importance both from a care and resource perspective. To investigate and measure health co-morbidities in ageing persons with Down syndrome with and without AD. Recorded physical and mental health needs were ascertained for 124 persons with DS >35 years through a systematic and detailed search of individual medical and nursing case records. Differences in persons with and without AD were investigated, by stage of dementia and by level of intellectual disability (ID). A summed score for health co-morbidities was created and compared using r-tests. Persons with AD had significantly higher co-morbidity scores than persons without AD ® = -8.992, d.f. = 121, P<0.0001). There was also a significant difference in summed co-morbidity scores for persons at end-stage vs. persons at midstage AD (t = -6.429, d.f. = 56, P < 0.0001). No differences were found by level of ID. Increasing health co-morbidities in persons with DS and AD have important implications for care and resources. Appropriate environmental supports combined with competent skilled staff are crucial and will have an important impact on the quality of life for this increasingly at risk population.

McCarron, M., & Lawlor, B.A.
Responding to the challenge of ageing and dementia in intellectual disability in Ireland
Aging and Mental Health, 2003, 7(6), 413-417. doi:10.1080/13607860310001594655.
Abstract: The intellectual disability (ID) population in Ireland is ageing and the number of older persons with the dual disability of ID and dementia is increasing. In spite of these demographic trends, as in other countries adequate policy and service provision for this population are lacking. This paper draws upon data available on the population with ID and dementia, reviews both generic and ID specific literature, considers the policy context and argues for a specific model of service provision. A service model is proposed for the development of multidisciplinary specialist teams within ID, delivered through mobile regional ID dementia clinics.

McCarron, M., Gill, M., Mccallion, P., Begley, C.
Alzheimer's dementia in persons with Down's syndrome: predicting time spent on day-to-day caregiving.
Dementia, 2005, 4(4), 521-538. https://doi.org/10.1177/1471301205058305
Abstract: The aim of this study was to investigate the amount of time formal caregivers spend addressing activities of day-to-day care activities for persons with Down's syndrome (DS) with and without Alzheimer's dementia (AD). Caregivers completed for 63 persons with DS and AD, and 61 persons with DS without AD, the Caregiving Activity Survey-Intellectual Disability (CAS-ID). Data was also gathered on co-morbid conditions. Regression analysis was used to understand predictors of increased time spent on day-to-day caregiving. Significant differences were found in average time spent in day-to-day caregiving for persons with and without AD. Mid-stage and end-stage AD, and co-morbid conditions were all found to predict increased time spent caregiving. Nature and tasks of day-to-day caregiving appeared to change as AD progressed. The study concluded that staff time to address day-to-day caregiving needs appeared to increase with onset of AD and did so most dramatically for persons with moderate intellectual disability. Equally, while the tasks for staff were different, time demands in caring for persons at both mid-and end-stage AD appeared similar.

McCarron, M., McCallion, P., Fahey-McCarthy, E., Connaire, K., & Dunn-Lane, J.
Supporting persons with Down syndrome and advanced dementia: Challenges and care concerns
Dementia, 2010, 9, 285-298. https://doi.org/10.1177/1471301209354025
Abstract: To understand staff perceptions of critical issues in caring for persons with intellectual disability (ID) and advanced dementia. There has been growing interest in addressing resource, training, and service redesign issues including an increase in collaborative practices in response to the growing incidence of dementia among persons with ID. Most recently this has included consideration of the specific issues in advanced dementia. Thirteen focus group interviews were held involving staff in six ID services and one specialist palliative care provider in Ireland. A qualitative descriptive approach was taken to analysis. Staff identified three key themes: (1) readiness to respond to end of life needs, (2) the fear of swallowing difficulties, and (3) environmental concerns and ageing in place. Four underlying issues that emerged in this study offer clues to solutions: (a) differences in staff preparation associated with settings, (b) lack of understanding and lack of collaboration with palliative care services, © uncertainties about the ability to transfer existing palliative care models to persons with ID and dementia and (d) the need to develop training on end stage dementia and related care approaches

McCarron, M. McCallion, P., Fahey-McCarthy, E., & Connaire, K.
Staff perceptions of essential prerequisites underpinning end-of-life care for persons with intellectual disability and advanced dementia.
Journal of Policy and Practice in Intellectual Disabilities, 2010, 7(2), 143–152. https://doi.org/10.1111/j.1741-1130.2010.00257.x
Abstract To better address palliative care and end-of-life issues for persons with intellectual disability (ID) and dementia, work was undertaken to understand the perspectives of agency staff in both the ID services and specialist palliative care fields. A qualitative descriptive design composed of 13 focus group interviews involved 50 participants drawn from six ID service providers and seven participants from one specialist palliative care service. Analysis was an iterative process; codes were identified and through thematic analysis, collapsed into two core themes: building upon services' history and personal caring—offering quality and sensitive care, and supporting comfort and optimal death in persons with ID and advanced dementia. Challenges were raised for service systems in the areas of aging in place, person-centered care, and interservice collaboration. Authors recommend both more practice relationship based and collaborative approaches to care and a stronger evidence-based research program on the timing and the efficacy of palliative care for persons with ID and dementia.

McCarron, M., McCallion, P., Fahey-McCarthy, E., & Connaire, K.
The role and timing of palliative care in supporting persons with intellectual disability and advanced dementia.
Journal of Applied Research in Intellectual Disabilities, 2011, 24, 189–198. https://doi.org/10.1111/j.1468-3148.2010.00592.x
Abstract: To better describe the role and timing of palliative care in supporting persons with intellectual disabilities and advanced dementia (AD). Specialist palliative care providers have focused mostly on people with cancers. Working with persons with intellectual disabilities and AD offers opportunities to expand such palliative care to other populations and disease conditions and to better understand the timing and role of palliative care delivery. Thirteen focus group interviews were held involving staff in six intellectual disability services and one specialist palliative care provider in Ireland. A qualitative descriptive approach was taken to analysis. Specialist palliative care staff recognized that person-centered care delivered in intellectual disability services was consistent with palliative approaches, but staff in intellectual disability services did not consider advanced dementia care as 'palliative care'. Both groups were unsure about the role of palliative care at early stage of dementia but appreciated specialist palliative care contributions in addressing pain and symptom management challenges. Successful extension of palliative care principles, philosophy and services to persons with intellectual disabilities and AD will require in-depth understanding of prevailing care philosophies and agreement regarding timing and the unique contributions of specialist palliative care services.

McCarron, M., McCallion, P., Reilly, E., & Mulryan, N.
Responding to the challenges of service developmental to address dementia needs for people with an intellectual disability and caregivers
Watchman, K. (ed.), 2014, *Intellectual Disability and Dementia: Research into Practice*, Jessica Kingsley Publishers, London, pp. 241-70.
Abstract: Book chapter addressing the association between intellectual disability and dementia; the experiences of dementia in people with intellectual disabilities; and service planning. This book chapter pertains to the latter. It acknowledges the challenges faced by traditional intellectual disability services and explains the approach adopted by one such service in ROI to supporting and accommodating people with an intellectual disability who develop dementia. It exemplifies the move to an in place progression model in traditional intellectual disability services, whereby services are adapted so that people with an intellectual disability can be supported and accommodated as they progress through dementia.

McCarron, M., McCallion, P., Reilly, E., Dunne, P., Carroll, R., & Mulryan, N.
A prospective 20-year longitudinal follow-up of dementia in persons with Down syndrome.
Journal of Intellectual Disability Research, 2017 Sep;61(9):843-852. doi: 10.1111/jir.12390.
Abstract: Authors sought to examine dementia characteristics, age at onset and associated co-morbidities in persons with Down syndrome. A total of 77 people with Down syndrome aged 35 years and older were followed up from 1996 to 2015. The diagnosis of dementia was established using the modified ICD 10 Criteria and a combination of objective and informant-based tests. Cognitive tests included the Test for Severe Impairment and the Down Syndrome Mental Status Examination; adaptive behaviour was measured using the Daily Living Skills Questionnaire, and data from the Dementia Questionnaire for People with Intellectual Disabilities have been available since 2005. Over the 20-year period, 97.4% (75 of 77) persons developed dementia with a mean age of dementia diagnosis of 55 years (SD = 7.1, median = 56 years). Clinical dementia was associated with cognitive and function decline and seizure activity. Risk for dementia increased from 23% in those aged 50 years to 80% in those aged 65 years and above. There were no differences by level of ID. The previously reported high risk levels for dementia among people with Down syndrome were confirmed in this data as was the relationship with late onset epilepsy. The value of the instruments utilized in tracking decline and helping to confirm diagnosis is further highlighted.

McCarron, M., & Riley, E.
Supporting persons with intellectual disability and dementia: Quality dementia care standards - A guide to practice
39 pp.
Dublin, Ireland: Trinity College Dublin (2010)
Source: http://www.docservice.ie/includes/documents/Dementia%20Publication%202011.pdf
Abstract: Document contains a series of six standards covering a range of areas concerned with care affecting adults with intellectual disabilities affected by dementia. Drawn from standards affecting the general population, this document groups together focal areas under six main categories reflecting person-centered dementia care. The standards consist of statements, indicators, and criteria for assessing evidence. The standards cover (1) appropriately trained staff and service development, (2) memory assessment services, (3) health and personal care, (4) communication and behavior, (5) promoting well-being and social connectedness, and (6) supporting persons with advanced dementia.

McCarron, M., Reilly, E., & Dunne, P.
Achieving quality environments for person centred dementia care
45 pp.
Dublin, Ireland: Daughters of Charity Service, (2013).
Abstract: Provides an overview of principles and practices designed to enable the operation of small group homes, including covering the planning process, design of private and public spaces, as well as therapeutic uses. Illustrated by two Daughters of Charity homes established for dementia specific care for people with ID. One home offers care for people with moderate dementia and includes 4 permanent beds and 2 respite beds for people both living with their families in the community and community group homes. Home also has a 6 bed step-down palliative care unit for people with ID in the later stages of dementia. These purpose built facilities were designed to be responsive to the changing needs of persons across the continuum of dementia. The home-like environments support people with dementia and staff to participate and complete tasks together, as well as informal impromptu unplanned activities. The homes are designed so that each resident has his or her own bedroom, with numerous communal areas including sitting rooms and garden areas.

MacDonald, S. & Summers, S.J.
Psychosocial interventions for people with intellectual disabilities and dementia: A systematic review.
Journal of Applied Research in Intellectual Disabilities, 2020 Sep,33(5), 839-855. doi: 10.1111/jar.12722. Epub 2020 Feb 27.
Abstract: People with intellectual disability experience a higher prevalence of dementia, at an earlier age, than the general population. The aim of this review was to establish the psychological interventions and outcomes for individuals with intellectual disability and dementia. A search of eight electronic databases and reference lists of all included articles was conducted using PRISMA guidelines. Data were synthesized using an integrative method. Initial searching produced 2,331 papers. Twenty-one studies met the inclusion criteria. Interventions were deductively categorized into behavioral, systemic, and therapeutic. All studies reported positive findings for individuals and for the systems which support them, but limited by methodological issues and neglect of the direct experience and impact on individuals themselves. The findings are discussed in relation to the wider literature and evidence base. Future research should aim to adopt methodologically robust designs that are inclusive of the individual experience of people with intellectual disability.

McGlinchey, E., Iulita, M.F., & Fortea, J.
Compounded inequality: racial disparity and Down syndrome.
Lancet Public Health, 2023 Nov, 8(11), e836. doi: 10.1016/S2468-2667(23)00213-X.

Abstract: (none provided). People with Down syndrome continue to face health inequities. We recently reported vulnerability to racial inequalities in people with Down syndrome, with vast disparities in the mortality rates between Black people with Down syndrome and White people with Down syndrome, particularly for infants and middle aged adults; this difference was greater than that observed in the general population. For Black people with Down syndrome, disparities due to both race and intellectual disability further compound inequity. Alzheimer's disease is the most frequent comorbidity in people with Down syndrome. Alzheimer's disease is currently incurable, but this might change with disease-modifying therapies. Equitable access to these medications is, therefore, critical for the whole population of people with Down syndrome, but particularly for Black individuals. Without equitable access, we risk further widening the gap between Black people and White people with Down syndrome. (Extract from Correspondence)

McGuire, B. E.; Whyte, N. , & Hardardottir, D.
Alzheimer's disease in Down Syndrome and intellectual disability: A review.
The Irish Journal of Psychology, 2006, 27(3-4), 114-129.
https://doi.org/10.1080/03033910.2006.10446235
Abstract: The authors review the literature on Alzheimer's disease (AD) in persons with general intellectual disabilities and those with Down syndrome. It focuses on the prevalence, clinical manifestations, diagnosis and management of AD in these populations. The literature indicates that people with Down syndrome have a greatly increased risk of dementia from their early 40s, while people with general intellectual disabilities have similar rates of AD to the general population. Taking into account the life expectancy of people with intellectual disabilities and those with Down syndrome, guidelines are provided for estimating the proportion of service users in a population that are at risk of developing dementia. The difficulties around diagnosis are reviewed and a particular emphasis is placed on the range of psychometric measures that may contribute to assessment and diagnosis. The management of service users who develop dementia is also reviewed and the implications for service providers are highlighted.

McKenzie, K., Metcalfe, D., Murray, G., & Michie A.
Service provision in Scotland for people with an intellectual disability and dementia: adherence to good practice guidelines,
Learning Disability Practice, 2019, 22(2), 26-33. doi:10.7748/ldp.2019.e1968.
Abstract: Good practice guidelines aim to promote equity and quality in service provision to improve the outcomes, experiences and quality of life for service users. The aim of the study was to evaluate the extent to which the practices of NHS services in Scotland for people with an intellectual disability and dementia are consistent with a range of quality indicators that cover screening, assessment and intervention. Staff from ten intellectual disability services in Scotland completed an online survey rating the extent to which their service met quality indicators adapted from the British Psychological Society, Division of Clinical Psychology, Royal College of Psychiatrists' Good Practice Standards - Self-Assessment Checklist. Areas most commonly `fully met' related to assessment and diagnosis and those most commonly `not/only partially met' related to areas, such as flexible funding, service development and monitoring and location of out-of-area placements. There is disparity in the extent to which the participating services have practices that are consistent with the quality indicators.

McKenzie, K., Harte, C., Patrick, S., Matheson, E., & Murray, G.C.
The assessment of behavioural decline in adults with Down's syndrome
Journal of Learning Disabilities, 2002, 6, 175-184.
https://doi.org/10.1177/146900470200600206
Abstract: Article reports study the examined two methods of using the Vineland Adaptive Behavioral Scales (VABS) to measure behavioral change in adults with Down syndrome who were surmised to be at-risk of Alzheimer's disease. The first approach used the VABS within a semi-structured interview and all areas of behavioral change identified by staff were noted. The second approach used the basal rule of the VABS as indicated in the Scales' manual. Comparison of the two approaches indicated that using the second approach highlighted significant decline in scores (for adults meeting the criteria for "probable Alzheimer's disease) on a number of domains between baseline and 12-24 months. One limitation of this approach that was noted was that this scoring method appeared to miss more subtle changes on behavior, which may be indicative of early Alzheimer's disease – which were picked up by the first approach. Authors recommend flexibility in using the VABS for assessment purposes and caution researchers to be explicit in reporting how the VABS was used in studies assessing dementia.

McKenzie, K., Metcalfe, D., Michie, A., & Murray, G.
Service provision in Scotland for people with an intellectual disability who have, or who are at risk of developing, dementia.
Dementia (London), 2020, 19(3), 736-749. doi: 10.1177/1471301218785795.
Abstract: This research aimed to identify current national provision by health services in Scotland in relation to proactive screening and reactive assessment for people with an intellectual disability in Scotland who have, or are at risk of developing, dementia. Staff from 12 intellectual disability services, representing the 11 health board areas in Scotland, completed an online questionnaire which asked about proactive screening and reactive assessment for people with intellectual disability who had, or were at risk of developing, dementia as well as suggested areas for improvement. All of the areas provided services for people with intellectual disability who have, or are at risk of developing, dementia, but differed as to whether this was reactive, proactive or both. Nine services offered intervention following diagnosis. The most common elements used across both proactive screening and reactive assessment were conducting a health check, using a general dementia questionnaire designed for people with an intellectual disability and direct assessment with the person. Clinical psychology and community learning disability nurses were the professions most likely to be involved routinely in both proactive screening and reactive assessments. The psychometric properties of the most commonly used assessments of cognitive and behavioral functioning were mixed. The areas of improvement suggested by practitioners mainly related to ways of improving existing pathways. This research represents the first step in providing an overview of service provision in Scotland. There was some inconsistency in relation to the general and specific components which were involved in proactive screening and reactive assessment. Implications for service provision are discussed.

McKenzie, K., Metcalfe, D., & Murray, G.
A review of measures used in the screening, assessment and diagnosis of dementia in people with an intellectual disability.
Journal of Applied Research in Intellectual Disabilities, 2018, 31(5), 725-742. doi: 10.1111/jar.12441.
Abstract: The increasing number of individuals with an intellectual disability who are at risk of developing dementia highlights the need to use measures with strong psychometric properties as part of the screening, assessment and diagnostic process. Searches were made of clinical and good practice guidelines and English language journal articles sourced from Proquest, Web of Science and Scopus databases (up to July 2017) for tools which were designed or adapted for the purpose of helping to diagnose dementia in people with intellectual disability. Based on a detailed review of 81 articles and guidelines, the present authors identified 22 relevant tools (12 cognitive, 10 behaviour). These were reviewed in terms of their psychometric properties. A number of tools were found to be available for use with people with intellectual disability; however, few were specifically standardized for this purpose which also had comprehensive information about reliability and validity.

McLaughlin, K., & Jones, D.
'It's all changed: 'carers' experiences of caring for adults who have Down's syndrome and dementia.
British Journal of Learning Disabilities, 2011, 39(1), 57-63.
https://doi.org/10.1111/j.1468-3156.2010.00618.x

Abstract: A qualitative interview study was undertaken to determine the information and support needs of carers of adults who have Down's syndrome and dementia. The data were analysed thematically. People who care for someone with Down's syndrome and dementia were asked about what it was like being a carer. Four of the carers were brothers or sisters of the person who had dementia, and two were paid carers not family members. All carers said that they wanted more information about some of the health and social problems that come with having dementia. Carers said that they wanted to know more information and meet other carers and other people who have dementia. Carers' information and support needs were seen to change at pre-diagnosis, diagnosis, and post-diagnosis. Helping carers to manage the changing nature of the adult with dementia is seen to be an essential part of the health professional's role.

McQuillan, S., Kalsy, S., Oyebode, J., Millichap, D., Oliver, C., & Hall, S.
Adults with Down's syndrome and Alzheimer's disease
Tizard Learning Disabilities Review, 2003, 8(4), 4-13.
https://www.emerald.com/insight/content/doi/10.1108/13595474200300032/full/html?skipTracking=true
Abstract: Adults with Down's syndrome are at risk of developing Alzheimer's disease in later life. This paper gives an overview of the current research in the area and discusses the implications it raises for individuals, carers, and service providers. Information on the link between Down's syndrome and Alzheimer's disease and prevalence rates are given. The clinical symptoms of Alzheimer' disease and a stage model documenting the progression of the disease are presented. Attention is drawn to the problems inherent in assessing and diagnosing Alzheimer's disease in a person with a pre-existing intellectual disability. Also discussed are the management of Alzheimer's disease, a focus on care management practices, and recommendations for service provision (including guidelines for supporting individuals which include maintaining skills, adapting a person-centered approach, implementing psychosocial interventions, and multi-disciplinary care management. Recommendations for the future include increasing education and awareness, implementing screening services, improving assessment methods, and developing appropriate services.

McVicker, R.W., Shanks, O.E., & McClelland, R.J.
Prevalence and associated features of epilepsy in adults with Down's syndrome
British Journal of Psychiatry, 1994, Apr;164(4), 528-532.
doi:10.1192/bjp.164.4.528.
Abstract: The aim of this study was to establish the prevalence of epilepsy in persons with Down's syndrome aged 19 years and over. A total of 191 adults with Down's syndrome were identified, giving a prevalence of 0.76/1000 (95% CI 0.75 to 0.77). Of these, 18 had epilepsy, giving a prevalence of 9.4% (95% CI 5.3% to 13.5%). The prevalence of epilepsy increased with age, reaching 46% in those over 50. The neurophysiological (EEG) findings of the epilepsy group were compared with those of a control group of Down's syndrome adults without epilepsy. Paroxysmal abnormalities consistent with a diagnosis of epilepsy were found in 80% of the epilepsy group, compared with only 13% of controls ($P < 0.001$). Epilepsy of late onset was associated with diffuse EEG abnormalities and clinical evidence of dementia. The age distribution and EEG findings suggest two independent processes in the causation of epilepsy: late-onset epilepsy associated with clinical evidence of dementia, and early-onset epilepsy in the absence of dementia.

Melville, C. A., Hamilton, S., Hankey, C.R., Miller, S., Boyle, S.
The prevalence and determinants of obesity in adults with intellectual disabilities
Obesity Reviews, 2007 (May), 8(3), 223-230. doi:
10.1111/j.1467-789X.2006.00296.x.
Abstract: People with intellectual disabilities experience significant health inequalities compared with the general population, including a shorter life expectancy and high levels of unmet health needs. Another accepted measure of health inequalities, the prevalence of obesity, has been shown to be higher in adults with intellectual disabilities than in the general population. While the factors contributing to the increased prevalence among adults with intellectual disabilities are not well understood, the high rates of obesity among younger adults highlight the need for further research involving children and adolescents with intellectual disabilities. To take forward the priorities for research and the development of effective, accessible services, there is a need for collaboration between professionals working in the fields of intellectual disabilities and obesity.

Menéndez M.
Down syndrome, Alzheimer's disease and seizures.
Brain Development, 2005, 27(4), 246-252. doi:10.1016/j.braindev.2004.07.008.
Abstract: Neuropathologically, Alzheimer-type abnormalities are demonstrated in patients with Down syndrome (DS), both demented and nondemented and more than a half of patients with DS above 50 years develop Alzheimer's disease (AD). The apolipoprotein E epsilon4 allele, oestrogen deficiency, high levels of Abeta1-42 peptide, elevated expression of BACE2, and valine polymorphism of prion protein gene are associated with earlier onset of dementia in DS individuals. Advanced AD alone may be an important risk factor for new-onset seizures in older adults and age above 60 years is a recognized risk factor for poor outcome from convulsive and nonconvulsive status epilepticus. DS patients aged over 45 years are significantly more likely to develop Alzheimer's disease than those less than 45 years and up to 84% demented individuals with DS develop seizures. Late-onset epilepsy in DS is associated with AD, while early-onset epilepsy is associated with an absence of dementia. In AD patients with a younger age of dementia onset are particularly susceptible to seizures. DS adults with epilepsy score significantly higher overall on the adaptive behaviour profile. Language function declined significantly more rapidly in AD patients with seizures and there is a good correlation between the severity of EEG abnormalities and cognitive impairment whereas in DS slowing of the dominant occipital rhythm is related to AD and the frequency of the dominant occipital activity decreases at the onset of cognitive deterioration.

Millichap, D., Oliver, C., McQuillan, S., Kalsy, S., Lloyd, V., & Hall, S.
Descriptive functional analysis of behavioral excesses shown by adults with Down syndrome and dementia.
International Journal of Geriatric Psychiatry, 2003, 18, 844-854. doi: 10.1002/gps.930
Abstract: The study examined the hypothesis that a functional relationship exists between social environmental events and behavioral excesses in individuals with Down syndrome and dementia. A case-series design was employed (n = 4) using an direct observation-based descriptive functional assessment procedure. Observations were conducted in the natural environments of four participants over periods ranging from 11 to 15.4 hours. Data were collected on non-verbal and verbal behavioral excesses, appropriate engagement and verbal interaction with others. Social environmental events observed including both staff and peer behavior. Analysis of co-occurrence for behavioral excesses and social environmental events indicated significant relationships for some behaviors consistent with operant reinforcement processes. Sequential analysis showed that changes in the probability of social contact occurred in the period directly preceding and following verbal behaviors. Results support the hypothesis that, consistent with literature for older adults with dementia in the general population, some behavioral excesses were functional in nature and not randomly occurring events. No relationship was found between appropriate engagement and staff.

Mircher, C., Cieuta-Walti, C., Marey, I., Rebillat, A-S., Cretu, L., Milenko, E., Conte, M., Sturtz, F., Rethore, M-O, & Ravel, A.
Acute regression in young people with Down syndrome.
Brain Science, 2017 Jun, 7(6), 57. Published online 2017 May 27. doi: 10.3390/brainsci7060057
Abstract: Adolescents and young adults with Down syndrome (DS) can present a rapid regression with loss of independence and daily skills. Causes of regression are unknown and treatment is most of the time symptomatic. We did a retrospective cohort study of regression cases: patients were born between 1959 and 2000, and were followed from 1984 to now. We found 30 DS patients aged 11 to 30 years old with history of regression. Regression occurred regardless of

the cognitive level (severe, moderate, or mild intellectual disability (ID)). Patients presented psychiatric symptoms (catatonia, depression, delusions, stereotypies, etc.), partial or total loss of independence in activities of daily living (dressing, toilet, meals, and continence), language impairment (silence, whispered voice, etc.), and loss of academic skills. All patients experienced severe emotional stress prior to regression, which may be considered the trigger. Partial or total recovery was observed for about 50% of them. In our cohort, girls were more frequently affected than boys (64%). Neurobiological hypotheses are discussed as well as preventative and therapeutic approaches.

Mizen, L.A.M.
Intellectual disability.
Medicine, 2024, 52(8), 506-511. https://doi.org/10.1016/j.mpmed.2024.05.012. (https://www.sciencedirect.com/science/article/pii/S1357303924001300)
Abstract: All doctors should expect to have some patients with intellectual disability. People with intellectual disability have high rates of physical and mental ill-health problems, and co-morbidity is typical. Frequently associated conditions such as epilepsy, aspiration and choking can cause avoidable death, and problems such as gastro-esophageal reflux disorder, sensory impairments and injuries can cause recurrent symptoms, persistent disability and distressed behavior. Other developmental conditions, psychosis and behavioral problems are common. Be aware of readily treatable associated physical conditions such as hypothyroidism in Down syndrome. Problem behavior can be a sign of distress, which can be the result of physical ill-health, mental ill-health, environmental factors or a combination of these. Medical assessment requires well-developed communication skills and access to multiple sources of information, which must involve relatives and paid carers as well as the person with intellectual disability, so sufficient time should be allocated. Use a bio-psychosocial-developmental framework. Avoid attributing symptoms of medical conditions to the person's developmental disabilities (`diagnostic overshadowing'), which results in illness going untreated. People with intellectual disability face many barriers in accessing healthcare, and proactive approaches are required. Specialist intellectual disability teams, where available, are excellent resources for specialist multidisciplinary assessment and advice.

Mohan, M., Bennet, C., & Carpenter, P.K.
Rivastigmine for dementia in people with Down syndrome
Cochrane Systemic Review - Intervention, 2009, 1.
https://doi.org//10.1002/14651858.CD007658
Abstract: Alzheimer's dementia (AD) is the most common form of dementia in people with Down Syndrome (DS). Acetylcholine is a chemical found in the brain that has an important role in memory, attention, reason and language. Rivastigmine is a “pseudo-irreversible” inhibitor of acetylcholinesterase, which is thought to maintain levels of acetylcholine. Rivastigmine can improve cognitive function and slow the decline of AD in the general population over time. It is important to note that people with DS tend to present with AD at a much younger age than the normal population as well as having subtle differences in physiology (e.g. metabolism and heart rate) and may therefore have different requirements from the general population. The authors sought to determine the effectiveness and safety of rivastigmine for people with DS who develop AD by using the following search methods, CENTRAL, MEDLINE, EMBASE, CINAHL, PsycINFO, BIOSIS, SCI, SSCI and the NRR, up to October 2008. They also contacted the manufacturers of rivastigmine as well as experts in the field, to ask about reports of unpublished or ongoing trials. Selection criteria included randomised controlled trials of participants with DS and AD in which treatment with rivastigmine was administered compared with a placebo group. Authors found that no study was identified which met inclusion criteria for this review and concluded that as there are no included trials, recommendations cannot be made about rivastigmine for AD in DS. Well-designed, adequately powered studies are required.

Molnar, F.J., Benjamin, S., Hawkins, S.A., Briscoe, M., & Ehsan, S.
One size does not fit all: Choosing practical cognitive screening tools for your practice.
JAGS (Journal of the American Geriatrics Society), 2020, 68(10), 2207-2213. doi: 10.1111/jgs.1671
Abstract: Every year, millions of patients worldwide undergo cognitive testing. Unfortunately, new barriers to the use of free open access cognitive screening tools have arisen over time, making accessibility of tools unstable. This article is in follow-up to an editorial discussing alternative cognitive screening tools for those who cannot afford the costs of the Mini-Mental State Examination and Montreal Cognitive Assessment (see www.dementiascreen.ca). The current article outlines an emerging disruptive“free-to-fee”cycle where free open access cognitive screening tools are integrated into clinical practice and guidelines, where fees are then levied for the use of the tools, resulting in clinicians moving on to other tools. This article provides recommendations on means to break this cycle, including the development of tool kits of valid cognitive screening tools that authors have contracted not to charge for (i.e., have agreed to keep free open access).The PRACTICAL.1 Criteria (PRACTIcing Clinician Accessibility and Logistical Criteria Version 1) are introduced to help clinicians select from validated cognitive screening tools, considering barriers and facilitators, such as whether the cognitive screening tools are easy to score and free of cost. It is suggested that future systematic reviews embed the PRACTICAL.1 criteria, or refined future versions, as part of the standard of review. Methodological issues, the need for open access training to insure proper use of cognitive screening tools, and the need to anticipate growing ethnolinguistic diversity by developing tools that are less sensitive to educational, cultural, and linguistic bias are discussed in this opinion piece.

Molsa, P.
Dementia and intellectual disability (in Finnish).
Finnish Medical Journal, 2001,13, 1495-1497.
Abstract: Unavailable

Moran, J.A., Rafii, M.S., Keller, S.M., Singh, B.K., Janicki, M.P.
The National Task Group on Intellectual Disabilities and Dementia Practices consensus recommendations for the evaluation and management of dementia in adults with intellectual disabilities.
Mayo Clinic Proceedings, 2013 Aug;88(8):831-840. doi: 10.1016/j.mayocp.2013.04.024. Epub 2013 Jul 10.
Abstract: Adults with intellectual and developmental disabilities (I/DD) are increasingly presenting to their health care professionals with concerns related to growing older. One particularly challenging clinical question is related to the evaluation of suspected cognitive decline or dementia in older adults with I/DD, a question that most physicians feel ill-prepared to answer. The National Task Group on Intellectual Disabilities and Dementia Practices was convened to help formally address this topic, which remains largely under-represented in the medical literature. The task group, comprising specialists who work extensively with adults with I/DD, has promulgated the following Consensus Recommendations for the Evaluation and Management of Dementia in Adults With Intellectual Disabilities as a framework for the practicing physician who seeks to approach this clinical question practically, thoughtfully, and comprehensively.

Moriconi, C., Schlamb, C., & Harrison, B.
Down syndrome and dementia: Guide to identification, screening, and managment.
Journal for Nurse Practitioners, 2015, 11(8), 812-818.
https://www.npjournal.org/article/S1555-4155(15)00602-9/pdf
Abstract: Down syndrome (DS) is an intellectual disability due to the genetic disorder trisomy21. Many individuals with DS are living into middle and older adulthood,experiencing chronic health problems, and are at risk for dementia. This article describes the primary care management of adults with DS, the relationship between DS and Alzheimer’s dementia, and screening protocols for primary care. [Extract follows] The family of the person with DS and AD needs a

team to navigate the changes that are inevitable during the course of this disease. An open discussion with both the NP and social worker will assist most families in providing the best quality of life for aging adults with DS. Maintaining social contact with family and friends may be difficult for aging DS adults, but is vital for well-being. Expectations for self-care and new learning must be readjusted and give way to an emphasis on a positive approach to the adult with DS. Families and caregivers need to proactively create a safe and calming home environment to promote quality of life.As AD progresses, assessment of nonverbal communication is essential in the care of adults with DS, but it may be challenging. Communication may be based on nonverbal cues and gestures. Additional response time is needed to allow for verbal or nonverbal communication of needs. Utilizing communication boards and pictures of common persons or objects is beneficial in this population. Safe relationships grounded in familiarity and trust are paramount in providing a secure environment for adults with DS and AD.

Moro, T.T., Savage, T.A., & Gehlert, S.
Agency, social and healthcare supports for adults with intellectual disability at the end of life in out-of-home, non-institutional community residences in Western nations: A literature review.
Journal of Applied Research in Intellectual Disabilities, 2017 Nov, 30(6), 1045-1056. doi: 10.1111/jar.12374. Epub 2017 Jun 6.
Abstract: The nature and quality of end-of-life care received by adults with intellectual disabilities in out-of-home, non-institutional community agency residences in Western nations is not well understood. A range of databases and search engines were used to locate conceptual, clinical and research articles from relevant peer-reviewed journals. The authors present a literature review of the agency, social and healthcare supports that impact end-of-life care for adults with intellectual disabilities. More information is needed about where people with intellectual disabilities are living at the very end of life and where they die. The support needs for adults with intellectual disabilities will change over time, particularly at the end of life. There are some areas, such as removing barriers to providing services, staff training, partnerships between agencies and palliative care providers, and advocacy, where further research may help to improve the end-of-life care for adults with intellectual disabilities.

Moss, S., Lambe, L., & Hogg, J.
Physical and mental health
Ageing Matters - Pathways for Older People with Learning Disabilities: Manager's Reader.
pp. 41-60
Kidderminster: British Institute of Learning Disabilities [Wolverhampton Road, Kidderminster, Worcestershire DY10 3PP United Kingdom] (1998)
Abstract: This unit, one of six that is used for training staff, covers briefly some of the key issues related to physical and mental health, and touches on dementia. Although not specifically developed for care management of adults with dementia, the text, in total, can be a useful resource for staff working in care settings when one or more of the adults in the setting are affected by dementia.

Moss, S., & Patel, P.
Psychiatric symptoms associated with dementia in older people with learning disability.
British Journal of Psychiatry,1995 Nov,167(5), 663-667. doi: 10.1192/bjp.167.5.663.
Abstract: This paper describes a study of non-cognitive features of dementia in a population of 105 people over 50 years of age, with learning disability. The study involved psychiatric assessment using the Psychiatric Assessment Schedule for Adults with a Developmental Disability (PAS-ADD), a semi-structured psychiatric interview developed specifically for people who have learning disability. Dementia was diagnosed using a combination of informant interviewing and observation of cognitive change over a three year period. Sample members with definite dementia had higher levels of sleep difficulty, hypersomnia, irritability, inefficient thought, loss of interest and anhedonia. Discriminant function analysis showed that non-cognitive features could help discriminate between definite and probable cases. Results support the previously reported observation for the general population that cognitive and non-cognitive features are only weakly related. The study suggests that psychiatric symptom information may be useful in screening for dementia in people with learning disability

Moss, S., & Patel, P.
Dementia in older people with intellectual disability: symptoms of physical and mental illness, and levels of adaptive behavior.
Journal of Intellectual Disability Research, 1997, 41(1), 60-69. doi: 10.1111/j.1365-2788.1997.tb00677.x.
Abstract: Detailed data on health and functional ability of 101 people with intellectual disability over 50 years of age are presented. Using a combination of informant interviewing, observation and measurement of cognitive change over a 3-year period, 12 of these individuals were identified as suffering from dementia. Their data are compared to those of the non-dementia sufferers. The people suffering from dementia had a greater number of chronic physical health problems and chronic disability resulting from physical health problems. Their capacity for self-directed activity was lower. The subjects had a reduced capacity to enjoy things, and were more irritable and more prone to violence. However, the outlook is somewhat different from a strategic perspective. The population of people with intellectual disability shows considerable epidemiological changes across the lifespan because of the effects of differential survival. The interaction of these factors tends to mask the impact of dementia-related skill loss in this population.

Motegi, N., Yamaoka, Y., Moriichi, A., & Morisaki, N.
Causes of death in patients with Down syndrome in 2014-2016: A population study in Japan.
American Journal of Medical Genetics Part A, 2022 Jan, 188(1), 224-236. doi: 10.1002/ajmg.a.62526. Epub 2021 Oct 7.
Abstract: Despite the higher mortality rates in patients with Down syndrome compared with the general Japanese population, the life span has dramatically increased in Japan and other countries. We aimed to clarify recent causes of death in patients with Down syndrome in Japan. We calculated proportionate mortality and standardized mortality odds ratios (SMORs) among all deaths registered with Down syndrome as the cause of death (ICD-10 code, Q90) in the Japanese National Death Registry Database in 2014-2016. In the study period, 762 in patients with Down syndrome died. The main causes of death were pneumonia/respiratory infections (20.5%), congenital malformations of the circulatory system (11.2%), other diseases of the circulatory system (9.2%), and aspiration pneumonia (8.4%). The SMORs (95% confidence intervals) were higher for natural death, defined as death of an elderly person with no other cause of death to be mentioned (55.73 [36.92-84.12]), **early-onset Alzheimer's disease, defined as Alzheimer's disease with onset <65 years of age (29.36 [16.44-52.44])**, aspiration pneumonia (18.33 [14.03-23.96]), pneumonia/respiratory infections (8.11 [6.76-9.73]), congenital malformations of the circulatory system (8.07 [5.98-10.88]), and leukemia/lymphoma (2.16 [1.55-2.99]) but lower for malignant solid tumors (0.04 [0.02-0.06]) in patients with Down syndrome. Patients with Down syndrome had the greatest relative risk of dying from natural death, early-onset Alzheimer's disease, and respiratory illnesses, highlighting the need for appropriate medical, health, and welfare services.

Müller, S. V., & Kuske, B.
DTIM - Demenztest für Menschen mit Intelligenzminderung [Dementia test for people with a reduction in intelligence: Early detection and gradient diagnosis].[Original in German]
Testzentral, Hogrefe Verlag, Göttingen, Deutschland 2020.
https://www.testzentrale.de/shop/demenztest-fuer-menschen-mit-intelligenzminderung.html

Abstract: The DTIM helps to determine whether there is also a dementia in addition to a reduction in intelligence. In addition to neuropsychological functional diagnostics, an external assessment is obtained. The diagnosis requires consideration of the course. Therefore, at least two survey times are required. Benefit from this innovative approach and clarify in suspected diagnoses. The test is for adults (from approx. 40 years) with the reduction of intelligence of different etiology, in which there is evidence of the existence of dementia. A caregiver should be present when it comes to implementation. This test procedure can also be used for people without language skills.

Mullins, D., Daly, E., Simmons, A., Beacher, F., Foy, C.M.L., Lovestone, S.,, Hallahan, B., Murphy, K.C., & Murphy, D. G.
Dementia in Down's syndrome: an MRI comparison with Alzheimer's disease in the general population.
Journal of Neurodevelopmental Disorders, 2013, 5, 19. doi:10.1186/1866-1955-5-19.
Abstract: Down's syndrome (DS) is the most common genetic cause of intellectual disability. People with DS are at an increased risk of Alzheimer's disease (AD) compared to the general population. Neuroimaging studies of AD have focused on medial temporal structures; however, to our knowledge, no *in vivo* case–control study exists comparing the anatomy of dementia in DS to people with AD in the general population. We therefore compared the *in vivo* brain anatomy of people with DS and dementia (DS+) to those with AD in the general population. Using MRI in 192 adults, we compared the volume of whole brain matter, lateral ventricles, temporal lobes and hippocampus in DS subjects with and without dementia (DS+, DS-), to each other and to three non-DS groups. These included one group of individuals with AD and two groups of controls (each age-matched for their respective DS and general population AD cohorts). AD and DS+ subjects showed significant reductions in the volume of the whole brain, hippocampus and temporal lobes and a significant elevation in the volume of the lateral ventricle, compared to their non-demented counterparts. People with DS+ had a smaller reduction in temporal lobe volume compared to individuals with AD. DS+ and AD subjects have a significant reduction in volume of the same brain regions. We found preliminary evidence that DS individuals may be more sensitive to tissue loss than others and have less 'cognitive reserve'.

Muralidhara, M., Spectorb, A., Huia, E.K., Liua, L., & Alia, A.
A systematic review of psychosocial interventions for people with intellectual disabilities and dementia.
Aging & Mental Health, 2023, 28(3), 385-395.
https://doi.org/10.1080/13607863.2023.2265322
Abstract: As the life expectancy of individuals with intellectual disabilities (ID) continues to increase, there is an increased risk of developing dementia. While psychosocial interventions are gaining prominence, evidence is limited for people with both dementia and ID. This review discusses the effectiveness of direct psychosocial interventions and adaptations to facilitate delivery within this population. The review followed the PRISMA guidelines. Five electronic databases, grey literature, and reference lists of included articles were searched for relevant studies. 10 eligible studies were appraised and analyzed by narrative synthesis. Ten distinct interventions were identified and categorized as (1) behavioral, (2) structured (manualized), and (3) personalized interventions, based on their purpose and delivery. All interventions were beneficial in improving a range of outcomes, though some studies were of low quality and most had small samples. Common adaptations included simplification of tasks and material, higher staff-to-client ratio, and alternative communication methods. There is emerging evidence for several psychosocial interventions for people with ID and dementia, though further research is required on effectiveness and generalizability. The adaptations discussed may guide implementation into routine care and contribute to current policies and guidelines on improving ID and dementia care.

Nadeau, P.A., Jobin, B., & Boller, B.
Diagnostic sensitivity and specificity of cognitive tests for mild cognitive impairment and Alzheimer's disease in patients with Down syndrome: A systematic review and meta-analysis.
Journal of Alzheimer's Disease, 2023, 95(1), 13-51. doi: 10.3233/JAD-220991. Erratum in: J Alzheimers Dis. 2024;97(1):521.
Abstract: Improved health care for people with Down syndrome (DS) has resulted in an increase in their life expectancy therefore increasing comorbidities associated with age-related problems in this population, the most frequent being Alzheimer's disease (AD). To date, several cognitive tests have been developed to evaluate cognitive changes related to the development of mild cognitive impairment (MCI) and AD in people with DS. The authors aimed to identify and evaluate available cognitive tests for the diagnosis of MCI and AD in people with DS. A systematic search of the Pubmed and PsycInfo databases was performed to identify articles published from January 1, 2000 and July 1, 2022. Keysearch terms were DS, AD or MCI, cognition, and assessment. Relevant studies assessing the diagnostic accuracy of cognitive tests for AD or MCI with standard clinical evaluation were extracted. Risk of bias was assessed using the QUADAS 2. We identified 15 batteries, 2 intelligence scales, 14 memory tests, 11 executive, functioning tests, 11 motor and visuospatial functioning tests, 5 language tests, 3 attention tests, and 2 orientation tests. Analysis showed that the CAMCOG-DS present a fair to excellent diagnostic accuracy for detecting AD in patients with DS. However, for the diagnosis of MCI, this battery showed poor to good diagnostic accuracy. The findings highlight important limitations of the current assessment available for the screening of mild cognitive impairment and AD in patients with DS and support the need for more clinical trials to ensure better screening for this highly at-risk population.

Nagdee, M.
Dementia in intellectual disability: a review of diagnostic challenges.
African Journal of Psychiatry (Johannesburg), 2011, 14, 194-199. doi: 10.4314/ajpsy.v14i3.1.
Abstract: The evaluation of dementia in individuals with intellectual disability, which will guide subsequent intervention, care and management depends on the systematic review of a number of factors: (1) the individual historical context, obtained from multiple sources, (2) evaluation of the pre-existing cognitive, behavioral, psychiatric, medical and adaptive skill profile, (3) the constellation, and pattern of evolution, of presenting signs and symptoms, (4) results of focused investigations, and (5) refinement of the differential diagnosis. In patients with ID, standard clinical methods need to be supplemented by careful, longitudinal behavioral observations, and individually tailored assessment techniques. Co-morbidity, multiple biological, psychological and socio-environmental factors, and complex interactions among events, are the reality for many ageing people with ID. Determining the various influences is often a formidable clinical task, but should be systematically carried out using medical, cognitive, behavioral, neuropsychiatric and psycho-social frameworks.

National Institute for Health and Care Excellence.
Dementia: Assessment, management and support for people living with dementia and their carers
[NICE Guideline No. 97]. (2018). https://www.nice.org.uk/guidance/ng97/chapter/Recommendations
Abstract: This guideline covers diagnosing and managing dementia (including Alzheimer's disease). It aims to improve care by making recommendations on training staff and helping carers to support people living with dementia.

National Task Group on Intellectual Disabilities and Dementia Practices.
My thinker's not working': A national strategy for enabling adults with intellectual disabilities affected by dementia to remain in their community and receive quality supports.
42pp.
National Task Group on Intellectual Disabilities and Dementia Practices [www.the-ntg.org]. (2012).

Abstract: 'My Thinker's Not Working' is the short title for the 42-page summative report issued by the National Task Group on Intellectual Disabilities and Dementia Practices, a planning and advocacy group organized to produce a national plan on dementia and intellectual disabilities. The report offers 20 recommendations for the improvement of services nationally and locally and suggests that its findings and recommendations be considered and integrated into the reports and plans being developed by the federal Advisory Council on Alzheimer's Research, Care, and Services -- under the National Alzheimer's Project Act. The document reviews the main issue facing adults with intellectual disabilities as they age when they are affected by dementia, as well as their families and provider organizations. The document is composed of 7 sections (Charge and Purpose, The Population, Challenges Facing the Population, Community Services, Education and Training, Financing, and Possible Solutions) and the National Dementia and Intellectual Disabilities Action Plan.

NAMHI
Alzheimer's Dementia in persons with intellectual disabilities: Some common questions and concerns
NAMHI, 5 Fitzwilliam Place, Dublin 2, Ireland
Abstract: 28 page booklet with 18 sections/question areas outlining basic information about Alzheimer's disease and people with ID, diagnostic resources, and service to help cope with the course of the disease. Developed by Dr. Mary McCarron of Trinity College Dublin.

Nelson, L., Lott, I., Touchette, P., Satz, P., & D'Elia, L.D.
Detection of Alzheimer disease in individuals with Down syndrome
American Journal of Mental Retardation, 1995 May, 99(6), 616-622. PMID: 7632429.
Abstract:A comprehensive baseline of emotional functioning was established for adults with Down syndrome. Five emotional factors were studied using groups of (a) adults with Down syndrome (n = 30), (b) clinical control subjects with dementia of the Alzheimer type (n = 18), and (c) elderly control subjects without intellectual disability (n = 25). Results of planned statistical comparisons showed indifference, pragnosia, and inappropriateness as primary emotional factors separating Down syndrome and Alzheimer disease groups from elderly control subjects without intellectual disabiity. Indifference was also shown to covary with cognitive mental state, whereby increased levels of indifference were associated with decreased levels of cognitive functioning. There is the possibility of noncognitive variables signaling dementia of the Alzheimer type in individuals with Down syndrome.

Nelson L.D., Orme, D., Osann, K., & Lott, I.T.
Neurological changes and emotional functioning in adults with Down Syndrome.
Journal of Intellectual Disability Research, 2001, 45, 450-456. doi: 10.1046/j.1365-2788.2001.00379.x.
Abstract: Study examined emotional changes in adults with Down Syndrome (DS) over time and to determine whether changes in these psychological variables were associated with brain atrophy on MRI scan and the presence of pathological reflexes on the neurological examination. Participants were 26 adults with DS and their caregivers. Caregivers completed a measure of emotional functioning about individuals with DS at two different time points (1 year apart). Levels of cognitive functioning were measured and neurological and MRI examinations were performed on all subjects at initial testing. Significant group effect separated those with and without pathological findings on MRI and neurological exam across three different scales: depression, indifference, and pragmatic language functioning. Problems of poor pragmatic language functioning appeared later in the course of suspected Alzheimer's disease (AD), as demonstrated by a significant group effect at time 2, but not at initial testing. In these subjects, the primary emotional change was a decline in social discourse (e.g. conversational style, literal understanding, verbal expression in social contexts). These emotional levels were stable over time, regardless of degree of cognitive decline. Specific emotional changes occur during the course of AD which were associated with abnormal findings from MRI and from neurological examination. These results, along with abnormalities in brain imaging and the presence of pathological reflexes, suggested that frontal lobe dysfunction is likely to be an early manifestation of Alzheimer's Disease in Down Syndrome.

New York State Developmental Disabilities Planning Council
When people with developmental disabilities age
18 minutes
New York State Developmental Disabilities Planning Council [155 Washington Avenue, Albany, New York 12222] (1992).
Abstract: A 18-minute video outlining the major physical and social change issues affecting adults with intellectual and developmental disabilities as they age, including a brief mention of Alzheimer's disease and Down syndrome. Available in VHS and CD-Rom format.

New York State Developmental Disabilities Planning Council
Dementia and people with intellectual disabilities – What can we do?
23 minutes
New York State Developmental Disabilities Planning Council [155 Washington Avenue, Albany, New York 12222] (2001).
Abstract: An instructional video which covers the basics of how dementia affects adults with intellectual disabilities, and provides information on diagnostics and suggestions on providing supports and services in community care settings. Produced by the University at Albany, this video can serve as primer on dementia and intellectual disabilities and provides information on basic design and service issues. Available in VHS and CD-Rom format.

Newroth, S., & Newroth, A.
Coping with Alzheimer disease: a growing concern.
28 pp.
Downsview: Ontario: National Institute on Mental Retardation (Kinsmen NIMR Building, York University Campus, 4700 Keele Street, Ontario, Canada, M3J IP3) (1981)
Abstract: Monograph describing one residential program's experience in caring for persons with Down syndrome who developed Alzheimer's disease; includes a chart of observations and guidelines for care. The guidelines are reproduced as an appendix in Janicki & Dalton (1999).

Ng, N., Flygare Wallen, E., & Ahlstrom, G.
Mortality patterns and risk among older men and women with intellectual disability: a Swedish national retrospective cohort study
BMC Geriatrics, 2017, 17(1), 269. doi: 10.1186/s12877-017-0665-3.
Abstract: Sweden has closed all institutions and imposed legislation to ensure service and support for individuals with intellectual disability (ID). Understanding mortality among older individuals with ID is essential to inform development of health promotion and disease control strategies. We investigated patterns and risk of mortality among older adults with ID in Sweden. This retrospective cohort study compared older adults aged 55 years and older with ID with a control population. Participants were followed during 2002-2015 or death, and censored if they moved out of Sweden. Individuals with ID were identified from two national registers: one covering all specialist health-care visits (out-patient visits and hospitalization) and the other covering people accessing social/support services. Individuals with ID (n = 15,289) were matched with a control population by sex, birth year, and year of first hospitalisation/out-patient visit/access to LSS services. Cause-of-death data were recorded using International Classification of Diseases, Tenth Revision. Cox proportional hazards regression were conducted to assess if overall and cause-specific mortality rate among individuals with ID was higher than in the Swedish population. The overall mortality rate among individuals with ID was 2483 per 100,000 people compared with 810 in the control population. Among those who died, more individuals with ID were younger than 75 years and unmarried. Leading causes of death among individuals with ID

were circulatory diseases (34%), respiratory diseases (17%) and neoplasms (15%). Leading causes of death in a sub-sample with Down syndrome (DS) were respiratory diseases (37%), circulatory diseases (26%) and mental/behavioural disorders (11%). Epilepsy and pneumonitis were more common among individuals with ID than controls. **Alzheimer's disease was common in the control population and individuals with DS, but not among those with ID** when DS was excluded. Individuals with ID had a higher overall mortality risk (hazard ratio [HR] 4.1, 95% confidence interval [CI] 4.0-4.3) and respiratory disease death risk (HR 12.5, 95% CI 10.9-14.2) than controls. Older adults with ID in Sweden carry a higher mortality risk compared with the general population, mainly attributable to respiratory, nervous and circulatory diseases. Care for this group, particularly during the terminal stage of illness, needs **to be tailored based on understanding of their main health problem.**

Nicholas, D. B., Shafai, F., Edelson, S. M., Bal, V., Nelson, H., Lawson, W., Doherty, M., Geurts, H. M., Sullivan, W. F., Braden, B. B., Wallace, G. L., Share, M., St. John, L., Amaral, D. G., Whitaker, A. H., Watters, L., Robson, T., Gauthier, J., Moulanier, S., Perry, L., Stemp, S., Wilkinson, E., Jokinen, N., Bauman, M. L., Hendren, R., Anagnostou, A., & Trollor, J. N.
Advancing care priorities for health and quality of life among older adults in the autism and/or intellectual disabilities communities: Proceedings of an international Think Tank.
BMC Proceedings, 2025, 19(Suppl 11), 15.
https://doi.org/10.1186/s12919-025-00330-8
Abstract: As the number of older autistic adults and adults with intellectual disabilities grows, expanding capacity to meet their needs is crucial. On November 23-25, 2023, a Think Tank on aging in autism and/or intellectual disabilities was convened, with national and international delegates within this field. The Think Tank consisted of presentations focusing on key issues as well as a series of panel presentations addressing first-person lived experience, family caregiving, service provision in the community, and physical and mental health-based care. Discussion reflected lived experiences and care needs in this population, with an ultimate aim of advancing healthcare and community support. Delegates, who represent perspectives as self-advocates, family caregivers, service providers, clinicians and researchers, ranked guidelines and areas of focus identified in the literature in terms of the most important priorities for the near-term development of capacity building resources.

Nieuwenhuis-Mark, R.E.
Diagnosing Alzheimer's dementia in Down syndrome: Problems and possible solutions.
Research in Developmental Disabilities, 2009, 30(5), 827-838. doi:10.1016/j.ridd.2009.01.010.
Abstract: It is widely accepted that people with Down syndrome are more likely than the general population to develop Alzheimer's dementia as they age. However, the diagnosis can be problematic in this population for a number of reasons. These include: the large intra-individual variability in cognitive functioning, the different diagnostic and methodological procedures used in the field and the difficulty in obtaining baseline levels of cognitive functioning in this population with which to assess cognitive and behavioral change. Recent researchers have begun to suggest ways around these difficulties. This review explores these recent developments and provides recommendations which may aid clinicians in their attempts to diagnose Alzheimer's dementia in the early stages in the Down syndrome population.

Nijhof, K., Boot, F.H., Naaldenberg, J., Leusink, G.L., & Bevelander, K.E.
Health support of people with intellectual disability and the crucial role of support workers.
BMC Health Services Research, 2024 Jan 2, 24(1), 4. doi: 10.1186/s12913-023-10206-2
Abstract: People with intellectual disability have a poorer health status than the general population. In The Netherlands, support workers play a key role in meeting health support needs of people with intellectual disability. Research on how people with intellectual disability and their support workers experience the support worker's role in preventing, identifying, and following up health needs of people with intellectual disability is scarce. To enhance health support of people with intellectual disability it is crucial that we understand how health support is delivered in everyday practice. Therefore, this study investigated experiences of people with intellectual disability and support workers with the health support of people with intellectual disability. Data collection consisted of six focus group (FG) discussions with between four and six participants (N = 27). The FGs consisted of three groups with support workers (n = 15), two groups with participants with mild to moderate intellectual disability (n = 8), and one group with family members as proxy informants who represented their relative with severe to profound intellectual disability (n = 4). The data was analysed thematically on aspects relating to health support. We identified three main themes relevant to the health support of people with intellectual disability: 1) dependence on health support, 2) communication practices in health support, and 3) organizational context of health support. Dependence on health support adresses the way in which support workers meet a need that people with intellectual disability cannot meet themselves, and communication practices and organizational context are identified as systems in which health support takes place. This study investigated experiences with the health support of people with intellectual disability from the perspectives of people with intellectual disability and support workers. We discuss the dependence of people with intellectual disability and the complexity of health support in everyday practice. We provide practical implications that can strengthen support workers in the provision of health support for people with intellectual disability in everyday practice. The findings of this study emphasize the need for intellectual disability care-provider organizations to establish policies around consistency in support staff to make it easier to identify and follow up health needs, and an environment where support staff can develop their expertise concerning communication practices, lifestyle choices, and identifying and following up health needs.

📖 **Noelker, E.A. & Somple, L.C.**
Adults with Down syndrome and Alzheimer's
In K.A. Roberto (Ed.), The Elderly Caregiver: Caring for Adults with Developmental Disabilities.
pp. 81-92
Newbury Park: SAGE Publications (1993)
Abstract: Book chapter providing a brief summary of significant assessment and care issues affecting adults with Down syndrome who have Alzheimer's disease. Noted are the needs for education of carers and families, as well as specialty care provision and community services.

Northway, R., Holland-Hart, D., & Jenkins, R.
Meeting the health needs of older people with intellectual disabilities: exploring the experiences of residential social care staff.
Health and Social Care in the Community, 2017 May, 25(3), 923-931. doi: 10.1111/hsc.12380. Epub 2016 Aug 31.
Abstract: Older people with intellectual disabilities often experience high levels of health needs and multiple morbidities but they may be supported by residential care staff with little or no previous experience of identifying and meeting health needs. Little is known regarding how they undertake this health-related role and this exploratory study seeks to address this gap. A purposive sample of 14 managers of supported living accommodation in Wales were interviewed in 2014 to determine their experiences of supporting tenants in relation to age-related health needs. The semi-structured interviews were transcribed and thematically analysed. Three of the emerging themes are reported in this paper: meeting health needs, the consequences of ageing and relationships. Findings indicate that residential care staff support older people with intellectual disabilities with complex and multiple health needs: they monitor health status, support access to healthcare, provide additional support arising from changing health needs and advocate for tenants in the context of healthcare. However, their role is often not understood by healthcare professionals. The importance of staff having a

long-term relationship with those they support was identified as being important to identifying any health-related changes. The need to develop effective relationships with healthcare staff was also noted. It is concluded that there is a need for better understanding among health staff of the role of residential social care workers and for further research regarding health-related communication.

Nübling, G., Loosli, S.V., Wlasich, E., Prix, C., Schönecker, S., Freudelsperger, L., Smrzka, N., Strydom, A.M., Zaman, S.H, Benejam, B., Missios, J., Meister, R., Danek, A., & Levin, J.
Eine deutsche Fassung der Cambridge Examination for Mental Disorders of Older People with Down's Syndrome and Others with Intellectual Disabilities : Ein Diagnoseverfahren zur Erfassung von Demenz bei Menschen mit einem Down-Syndrom [A German version of the Cambridge examination for mental disorders of older people with Down's syndrome and others with intellectual disabilities: A diagnostic procedure for detecting dementia in people with Down's syndrome]. [Original in German]
Zeitschrift fur Gerontologie und Geriatrie, 2020 Oct, 53(6), 546-551. German. doi: 10.1007/s00391-019-01591-7. Epub 2019 Aug 9. PMID: 31399752.
Abstract: Although people with Down's syndrome (DS) are at a high risk of developing an Alzheimer type dementia (AD) due to a triplication of the amyloid precursor gene, there are practically no internationally available test procedures to detect cognitive deficits in this at risk population in the German language. The aim was to provide a German translation and intercultural adaptation of the Cambridge examination for mental disorders of older people with Down's syndrome and others with intellectual disabilities (CAMDEX-DS), which is available in English and Spanish. This instrument for diagnostics and monitoring consists of a psychological test examination (CAMCOG-DS) and a caregiver interview. The translation and adaptation of the CAMDEX-DS were achieved through a multistep translation process, whereby two independent forward and back translations were provided by professional translators and a consensus version was finalized and tested. The final version of the caregiver interview was applied to 11 subjects and the CAMCOG-DS was conducted with 28 patients. The German version of the CAMDEX-DS proved to be easily administered. The CAMCOG-DS could be fully administered to 21 out of 28 patients (75%). The CAMCOG-DS values were much lower for older patients aged ≥45 years than for younger patients (46/109 vs. 73.5/109; p = 0.033). The German version of the CAMDEX-DS provides an internationally recognized tool for the diagnostics and monitoring of cognitive decline in Down's syndrome. Furthermore, the German version can standardize medical care of these patients. In particular it provides a means of participation in international research trials for this at risk population.

Nuebling, G., Wagemann, O., Deb, S., Wlasich, E., Loosli, S. V., Sandkühler, K., Stockbauer, A., Prix, C., & Levin, J.
Validation of a German version of the dementia screening questionnaire for individuals with intellectual disabilities (DSQIID-G) in Down's syndrome.
Journal of Intellectual Disability Research, 2024, 68(10), 1146-1155. https://doi.org/10.1111/jir.13144
Abstract: People with Down's syndrome (DS) are at high risk of developing Alzheimer dementia (DS-AD) due to a triplication of the amyloid precursor protein gene. While several tools to diagnose and screen for DS-AD, such as the dementia screening questionnaire for individuals with intellectual disabilities (DSQIID), are available in English, validated German versions of such instruments are scarce. A German version of the DSQIID questionnaire (DSQIID-G) was completed by caregivers before attending our specialist outpatient department for DS-AD. All participants were assessed blind to DSQIID-G scoring using clinical and neuropsychological examinations, including the Cambridge Examination for Mental Disorders of Older People with Down's Syndrome and Others with Intellectual Disabilities (CAMDEX-DS). ICD-10 and amyloid/tau/ neurodegeneration (A/T/N) criteria were applied to detect and categorise cognitive decline. Of 86 participants, 43 (50%) showed evidence of cognitive decline. A definite diagnosis of DS-AD was reached in 17 (19.8%) and mild cognitive impairment in seven (8.3%) participants. Secondary causes of cognitive decline were determined among 13 (15.1%) participants, and in six (7%) cases, the diagnosis remained unclassifiable due to co-morbidities. Compared with cognitively stable individuals, participants with cognitive decline (n = 43) displayed higher DSQIID-G total scores [median (range): 3 (0-21) vs. 19 (0-48), P < 0.001]. A total score of >7 provided a sensitivity of 0.94 against a specificity of 0.76, to discriminate DS-AD and participants without cognitive decline according to ROC analysis. The convergent validity against the CAMDEX-DS interview score was good (r = 0.74), and split-half reliability (r = 0.96), internal consistency (Cronbach's α r = 0.96), test-retest reliability (r = 0.88) (n = 25) and interrater reliability (r = 0.81) (n = 31) were excellent. The DSQIID-G showed excellent psychometric properties, including concurrent and internal validity and reliability. The cut-off value for screening was lower than in the original English validation study. For a screening instrument like DSQIID-G, a lower cut-off is preferable to increase case detection.

O'Bryant, S.E., Zhang, F., Silverman, W., Lee, J.H., Krinsky-McHale, S.J., Pang, D., Hall, J., & Schupf, N.
Proteomic profiles of incident mild cognitive impairment and Alzheimer's disease among adults with Down syndrome
Alzheimer's & Dementia: Diagnosis, Assessment & Disease Monitoring, 12, 1, https://doi.org/10.1002/dad2.12033
Abstract: We sought to determine if proteomic profiles could predict risk for incident mild cognitive impairment (MCI) and Alzheimer's disease (AD) among adults with Down syndrome (DS). In a cohort of 398 adults with DS, a total of n = 186 participants were determined to be non-demented and without MCI or AD at baseline and throughout follow-up; n = 103 had incident MCI and n = 81 had incident AD. Proteomics were conducted on banked plasma samples from a previously generated algorithm. The proteomic profile was highly accurate in predicting incident MCI (area under the curve [AUC] = 0.92) and incident AD (AUC = 0.88). For MCI risk, the support vector machine (SVM)-based high/low cut-point yielded an adjusted hazard ratio (HR) = 6.46 (P < .001). For AD risk, the SVM-based high/low cut-point score yielded an adjusted HR = 8.4 (P < .001). on The current results provide support for our blood-based proteomic profile for predicting risk for MCI and AD among adults with DS.

O'Caoimh, R., Clune, Y., & Molloy, D.W.
Screening for Alzheimer's disease in Downs syndrome
Journal of Alzheimers Disease & Parkinsonism, 2013, S7; 001. http://dx.doi.org/10.4172/2161-0460.S7-001
Abstract: Down syndrome (DS), is associated with an increased incidence of Alzheimer's disease (AD). Although pathological changes are ubiquitous by 60 years of age, prevalence rates are lower. The diagnosis of AD in persons with DS is challenging, complicated by atypical presentations, baseline intellectual disability and normal age associated cognitive decline. Effective screening is limited by a paucity of diagnostic criteria, cognitive screening instruments and screening programs. Both observer-rated questionnaires and direct neuropsychological testing are suggested to screen for cognitive impairment, each with different strengths and weaknesses. This paper reviews commonly used screening instruments and explores the unique challenges of screening for AD in persons with DS. It concludes that single, one-dimensional screening tools and opportunistic evaluations are insufficient for detecting dementia in this population. These should be replaced by batteries of tests, incorporating informant questionnaires, direct neuropsychological testing, assessment of activities of daily living and behaviors, measured at baseline and reassessed at intervals. Developing these strategies into organized screening programs should improve diagnostic efficiency and management.

O'Dwyer, M., Finnerty, S., Henman, M., Carroll, R., McCallion, P., & McCarron, M.
Prevalence and treatment of dementia in older adults with intellectual disability in Ireland
Journal of Intellectual Disability Research, 2019, 63(8), 645.

Abstract: High rates of dementia have been reported among older adults with intellectual disability (ID), particularly those with Down Syndrome. As the use of dementia drugs in this patient group lacks an evidence base, their rates of use are of interest. Incidence and prevalence rates were determined using a combined dementia variable for three waves of the IDS-TILDA study, a nationally representative study of older adults with ID in Ireland. Incidence of dementia was defined as participants newly reporting a diagnosis and/or newly receiving dementia drug(s) at each wave. Prevalence of dementia was defined those who had reported a diagnosis at a previous wave and/or received dementia drug(s) at a previous wave. Drugs for dementia were included as a proxy for dementia diagnosis, in those with no diagnosis. Dementia incidence remained similar across Waves: 5.0% at Wave 1, 4.3% at Wave 3. Prevalence increased, 5% at Wave 1, to 9.6% by Wave 3. Those receiving receiving dementia drug(s) decreased, from 54.1% of those with dementia at Wave 1 to 28.8% at Wave 3. Three dementia drugs were reported: donepezil, memantine and rivastigmine. It was found that use of drugs for dementia decreased, despite an increased incidence. Further research into efficacy of use of a drugs is needed.

O'Leary, L., Cooper, S-A., & Hughes-McCormack, L.
Early death and causes of death of people with intellectual disabilities: A systematic review
Journal of Applied Research in Intellectual Disabilities, 2018, 31(3), 325-342. https://doi.org/10.1111/jar.12417
Abstract: Death of people with intellectual disabilities is considered to be earlier than for the general population. Databases were searched for key words on intellectual disabilities and death. Strict inclusion/exclusion criteria were used. Information was extracted from selected papers, tabulated and synthesized. Prospero registration number: CRD42015020161. Of 19,111 retrieved articles, 27 met criteria. Death was earlier by 20 years. It has improved in recent decades; however, the same inequality gap with the general population remains. More severe intellectual disabilities, and/or additional comorbidities rendered it shortest. Standardized mortality rates showed a greater inequality for women than men. Respiratory disease and circulatory diseases (with greater congenital and lesser ischemic disease compared with the general population) were the main causes of death. Cancer was less common, and cancer profile differed from the general population. Some deaths are potentially avoidable. All research is from high-income countries, and cause of death is surprisingly little investigated. Authors concluded that improved health care, including anticipatory care such as health checks, and initiatives addressing most relevant lifestyle behaviors and health risks are indicated.

Oliver, C,, Adams, D,, Holland, A,J,, Brown, S,S,G,, Ball, S,, Dodd, K,, & Carr, J.
Acquired mild cognitive impairment in adults with Down syndrome: Age-related prevalence derived from single point assessment data normed by degree of intellectual disability.
International Journal of Geriatric Psychiatry, 2022 Feb;37(2):10.1002/gps.5674. doi: 10.1002/gps.5674.
Abstract: Individuals with Down syndrome (DS) are at significant risk for early onset Alzheimer's disease (AD), likely due to the triplication of genes on chromosome 21 that facilitate AD neuropathology. To aid the effective early diagnosis of dementia in DS, we demonstrate the strategy of using single point assessment of cognitive performance with scoring normed for degree of intellectual disability to generate age related prevalence data for acquired mild cognitive impairment (AMCI). Four hundred and twelve adults with DS were assessed using the Neuropsychological Assessment of dementia in adults with Intellectual Disability. Normative data, banded by degree of intellectual disability, allowed identification of AMCI by atypical deviation from expected performance. AMCI was evident in approximately 20% of adults with DS aged 40 and under, 40% aged 41-50 and 45% aged 51 and over. Relative risk increased significantly in those aged 46 and over. Analysis of prevalence by 5-year age bands revealed two peaks for higher prevalence of AMCI. Psychometric data indicate single point assessment of AMCI is possible for the majority of adults with DS. Two peaks for age-related prevalence of AMCI suggest the risk for onset of AD conferred by trisomy of chromosome 21 is moderated by another factor, possibly ApoE status.

Oliver, C., & Holland, A.J.
Down's syndrome and Alzheimer's disease: a review.
Psychological Medicine, 1986, 16(2), 307-322. doi: 10.1017/s0033291700009120.
Abstract: Neuropathological change found in nearly all individuals with Down syndrome over the age of 35 years closely resembles that of Alzheimer's disease. The extent to which dementia occurs as a result of this change is unclear, and the studies which have investigated presumed cognitive deficits are reviewed. The theories put forward to explain the association between these two disorders and their possible significance to the understanding of the aetiology of Alzheimer's disease are discussed.

Oliver, C., Crayton, L., Holland, A., & Hall, S.
Cognitive deterioration in adults with Down syndrome: effects on the individual, caregivers, and service use
American Journal on Mental Retardation, 2000, 103, 455-465. doi: 10.1352/0895-8017(2000)105<0455:CDIAWD>2.0.CO;2.
Abstract: Individuals with Down syndrome (N = 49) who had participated in serial neuropsychological assessments were assigned to one of three groups comparable in level of premorbid intellectual disability: (1) those showing cognitive deterioration, (2) those comparable in age but not showing cognitive deterioration and (3) those not showing cognitive deterioration but younger. Those experiencing cognitive deterioration were less likely to receive day services, had more impoverished life experiences, and required more support compared to groups without cognitive deterioration. When age was controlled for, cognitive deterioration was significantly positively associated with carer difficulties and service use and negatively associated with life experiences for the individual. Results suggest a potential role for carer difficulties in influencing life experiences of adults with Down syndrome showing cognitive decline.

Oliver, C. & Kalsy, S.
The assessment of dementia in people with intellectual disabilities: Context strategy and methods.
In J. Hogg and A. Langa (eds), *Assessing Adults with Intellectual Disabilities*. Blackwell Publishing: Oxford. (2005), chapter 7, pp 98-107. doi: 10.1002/9780470773697.ch7
Abstract: Unavailable. This chapter contains sections titled: introduction, problems of diagnosis and the need for formal assessment, problems associated with formal assessments, retrospective and prospective assessment strategies, assessment methods, and conclusion.

Oliver, C., Kalsy, S., McQuillan, S., & Hall, S.
Behavioural excesses and deficits associated with dementia in adults who have Down syndrome.
Journal of Applied Research in Intellectual Disabilities, 2011, 24, 208–216. https://doi.org/10.1111/j.1468-3148.2010.00604.x
Abstract: Informant-based assessment of behavioral change and difference in dementia in Down syndrome can aid diagnosis and inform service delivery. To date few studies have examined the impact of different types of behavioral change. The Assessment for Adults with Developmental Disabilities (AADS), developed for this study, assesses behavioral excesses (11 items) and deficits (17 items) associated with dementia. Inter-informant reliability, internal consistency and concurrent validity were evaluated and found to be robust. A comparison of the AADS subscale scores for three groups (n = 12) of adults with Down syndrome demonstrated more frequent deficits and excesses and greater management difficulty and effects on the individual in a dementia group than age comparable and younger groups. The AADS is a promising dementia specific measure for people with intellectual disability. Further research should evaluate

change as dementia progresses and the nature of management difficulty and effects on the individual.

Olsen, R.V., Ehrenkrantz, E., & Hutchings, B.L.
Creating the movement-access continuum in home environments for dementia care
Topics in Geriatric Rehabilitation, 1996, 12(2),1-8.
DOI:10.1097/00013614-199612000-00003
Abstract: Since the majority of people with Alzheimer's disease receive some care at home, the environment of that home must be safe and supportive. In-depth interviews of 90 "seasoned" caregivers identified tactics for creating these settings through home modifications and technology. A successful modification strategy follows a three-stage movement-access continuum that responds to the disease course -- assistance, restriction with compensation, and wheelchair accessibility. Approaching home modifications along this continuum encourages independence and movement when appropriate while providing safety and control. With a sensitive and ongoing modification strategy, the home environment can become an asset rather than a liability for caregiving.

Olsen, R.V., Ehrenkrantz, E., & Hutchings, B.
Creating supportive environments for people with dementia and their caregivers through home modifications
Technology and Disability, 1993, 2(4), 47-57.
https://content.iospress.com/articles/technology-and-disability/tad2-4-08
Abstract: Article examines what caregivers did to enhance or modify their homes when a spouse or other family member had dementia. Authors address controlling access (using locking techniques, blocking access with gates and partial doors, and the like, as examining modifications to kitchens, bathrooms, and furniture. Data showed that many built ramps, double railings, hand grips, as well as extending landings for ease of wheelchair use, reducing riser heights, removing steps, and installing electric chair lifts. Home owners also reconfigured space and rooms. Authors conclude that home owners modified spaces to increase access and independence in some life areas and to limit or curtail access in others. Article is a good source of information for how the process and outcome of families tackle home modifications

Olsen, R.V., Ehrenkrantz, E., & Hutchings, B.
Homes that help: Advice from caregivers for creating a supportive home (Alzheimer's and Related Dementias)
77 pp.
Newark, New Jersey: New Jersey Institute of Technology [Architecture and Building Science Research Group, School of Architecture, NJIofT, University Heights, Newark, New Jersey 07102-1982] (1993)
Abstract: Manual that details examples of how to adapt a home for persons affected by dementia, covering care management techniques, physical adaptations, and personal monitoring strategies.

Orth, J., Flowers, D., Betzj G., & Cagle, J.G.
A systematic review of specialty dementia care units in long-term care settings.
International Journal of Geriatric Psychiatry, 2023 Apr, 38(4), e5907. doi: 10.1002/gps.5907.
Abstract: Alzheimer's disease and related dementias (ADRD) are common among nursing home residents. Yet, conclusive evidence regarding best care practices among this population is lacking. Objectives of this systematic review were to explore features of dementia specialty care units (DSCUs) in long-term care settings and examine benefits for residents, staff, families, and facilities. PubMed, CINAHL, and PsychINFO were searched to identify articles involving DSCUs in long-term care settings published in English with full text available between 01.01.2008 and 06.03.2022. Articles containing empirical data about ADRD special care in long-term care settings were included in the review. Articles focused on clinic-based or out-patient dementia care programs (e.g., adult day care) were excluded. Articles were categorized based on geography (U.S. vs. international) and study design: interventions, descriptive studies, or comparison studies (traditional vs. specialty ADRD care). Our review included 38 U.S. articles and 54 articles from 15 international countries. In the U.S., 12 intervention, 13 descriptive, and 13 comparison studies met the inclusion criteria. Articles from international countries included 22 intervention, 20 descriptive, and 12 comparison studies. Results were mixed in terms of the efficacy of DSCUs. Promising DSCU features include small-scale settings, dementia-educated staff, and multidisciplinary approaches to care. Overall, **our review did not find conclusive evidence regarding the benefits of DSCUs in long-term care settings.** No rigorous study designs were found examining 'special' features of DSCUs and associations with outcomes among residents, family, staff, and the facility. Randomized clinical trials are needed to disentangle the 'special' features of DSCUs.

Owens, D., Dawson, J. C., & Losin, S.
Alzheimer's disease in Down's syndrome.
American Journal of Mental Deficiency, 1971, 75, 606–612.
https://psycnet.apa.org/record/1971-25354-001
Abstract: Although neuropathologists describe Alzheimer's changes in the brains of all victims of Down's syndrome over 35 yr. of age, only 3 cases of clinical dementia in such individuals are described in the literature. In order to establish clinical correlates of Alzheimer's disease, psychiatric and neurologic findings obtained from a middle-aged group were compared to those of Down's syndrome patients in their early 20s. The older group exhibited significantly greater incidence of abnormality in (a) object identification, (b) snout reflex, (c) Babinski sign, and (d) palmomental sign. Both groups displayed mild hypertonia rather than hypotonia, and face-hand test was abnormal in 75% of Ss tested. While dementia is uncommon, subtle neurological changes reflect neuropathological findings present in aging sufferers of Down's syndrome.

Paiva, A.F., Nolan, A., Thumser, C., & Santos, F.H.
Screening of cognitive changes in adults with intellectual disabilities: A systematic review
Brain Sciences, 2020, 10(11), 848; https://doi.org/10.3390/brainsci10110848
Abstract: Screening and assessment of cognitive changes in adults with Intellectual Disabilities (ID), mainly Down Syndrome (DS), is crucial to offer appropriate services to their needs. Authors present a systematic review of the existing instruments assessing dementia, aiming to support researchers and clinicians' best practice. Searches were carried out in the databases Web of Science; PubMed; PsycINFO in March 2019 and updated in October 2020. Studies were selected and examined if they: (1) focused on assessing age-related cognitive changes in persons with ID; (2) included adults and/or older adults; (3) included scales and batteries for cognitive assessment. Forty-eight cross-sectional studies and twenty-seven longitudinal studies were selected representing a total sample of 6451 participants (4650 DS and 1801 with other ID). In those studies, we found 39 scales, questionnaires, and inventories, and 13 batteries for assessing cognitive and behavioural changes in adults with DS and other ID. It was noted that the most used instrument completed by an informant or carer was the Dementia Questionnaire for Learning Disabilities (DLD), and its previous versions. The authors explore the strengths and limitations of the instruments and outline recommendations for future use.

Pape, S.
Dementia diagnostic criteria in persons with IDD.
Journal of Intellectual Disability Research, 2019, 63(8), 641.
(Conference presentation abstract)
Abstract: Dementia is a common clinical presentation among older adults with IDD, particularly those with Down syndrome. The presentation of dementia may differ compared with typical Alzheimer's disease, and criteria thus require validation in IDD populations. Data from memory assessments in individuals with Down syndrome were presented to expert raters who rated the case as dementia or no dementia using ICD-10, DSM-IV-TR and DSM-5 criteria and their own clinical judgement. Estimates were then made of the concurrent validity and

reliability of clinicians' diagnoses of dementia against these manualised diagnoses. Validity of clinical diagnoses were explored by establishing the stability of diagnoses over time. Similar data from previous studies in other individuals with intellectual disabilities were compared. It was found that clinical diagnoses of dementia in Down syndrome were valid and reliable and could be used as the standard against which new criteria such as the DSM-5 are measured. Criteria had good inter-rater reliability but concurrent validity varied. Author cautions that clinicans should consider the reliability and validity of dementia diagnostic criteria when applying these in clinical settings.

Pape, S.E., Baksh, R.A., Startin, C., Hamburg, S., Hithersay, R., & Strydon, A.
The association between physical activity and CAMDEX-DS changes prior to the onset of Alzheimer's disease in Down syndrome
Journal of Clinical Medicine, 2021, Apr 27, 10(9), 1882. doi: 10.3390/jcm10091882.
Abstract: People with Down syndrome are at ultra-high risk of developing Alzheimer's dementia. At present, there are no preventative or curative treatments. Evidence from sporadic Alzheimer's disease literature suggests that lifestyle factors including physical activity may help maintain cognitive and functional skills and reduce dementia risk. Our study aimed to explore the association between regular exercise undertaken by participants with Down syndrome and changes in dementia-related domains of cognition and function. This was to consider whether physical activity may be a protective measure to delay cognitive decline and dementia in Down syndrome. Demographic, lifestyle, and health information was collected at baseline and at a two year follow up from 214 adults with Down syndrome without dementia, who also underwent assessment using the Cambridge Examination for Mental Disorders of Older People with Down Syndrome and Others with Intellectual Disabilities (CAMDEX-DS) and genetic analysis. Logistic regression models were used to examine the potential associations between decline in CAMDEX-DS domains and exercise whilst controlling for key variables. At baseline, engaging in moderate intensity exercise was associated with a 47% reduced risk of everyday skills decline and engaging in high intensity exercise was associated with a 62% reduced risk of decline in personality and behaviour. At follow-up, high levels of exercise were associated with an 87% reduced risk of decline in personality and behaviour. Moderate intensity exercise at baseline was associated with a 62% reduction in risk of decline during the follow-up period in memory and orientation. Based on our data it appears that regular moderate and high intensity exercise could reduce the risk of clinically detectable decline in a Down syndrome population with possible long-term benefits. People with Down syndrome may engage in less physical activity than their peers, and barriers remain which can prevent people with Down syndrome engaging in exercise. Our work highlights how important it is that people with Down syndrome are supported to be physically active, and to promote exercise as part of a healthy ageing plan. Clinical trials in this area would be justified to determine if engaging in exercise can lead to realistic improvements in maintaining functioning and delaying dementia onset in Down syndrome and to help develop guidance in this area.

Paton, J., Johnston, K., Katona, C., & Livingston, G
What causes problems in Alzheimer's disease: Attributions by caregivers. A qualitative study
International Journal of Geriatric Psychiatry, 2004, Jun, 19(6), 527-532. doi:10.1002/gps.1118.
Abstract: Authors soiught to gain insight into caregivers' understanding of the causes of behaviors they find problematic in people with Alzheimer's disease in order to inform the development of educational strategies. A qualitative, semi-structured interview was used. Participants were 205 caregivers for a person with Alzheimer's disease, all of whom were aware of the diagnosis and who had been recruited as part of a larger longitudinal study. Participants were from inner-city and suburban London/semi-rural Essex. The main outcome measures were caregivers' understanding of: the cause of problematic behavior; the ability of the person with dementia to control this behavior; the prognosis of the illness. Most carers attribute the cognitive, behavioral and psychological symptoms of dementia to causes other than dementia; many believe that the person with dementia has control over their behavior and substantial numbers believe the person with dementia will return to normal. This study suggests that providing facts about the illness to caregivers is not enough, as caregivers may not understand that the symptoms they observe are related to the diagnosis. Education by clinicians should focus on the understanding of caregivers and in particular explore the caregivers' attributions of the symptoms which are present in the person for whom they care.

Patti, P., Amble, K. & Flory, M.
Placement, relocation and end of life issues in aging adults with and without Down's syndrome: A retrospective study.
Journal of Intellectual Disability Research, 2010, 54(6), 538-546. doi: 10.1111/j.1365-2788.2010.01279.x.
Abstract: Aging adults with Down's syndrome (DS) experience more relocations and other life events than adults with intellectual disabilities aged 50 and older without DS. Age-related functional decline and the higher incidence of dementia were implicated as the contributing factors that led to relocation and nursing home placement. A retrospective study of adults with intellectual disabilities who were born prior to the year 1946 was conducted to analyze the number of relocations experienced over a 5- and 10-year period. The cohort consisted of 140 individuals (61 with DS between ages 50–71 years, and 79 without DS between ages 57–89 years) who had been referred to a diagnostic and research clinic. Analyses revealed the number of relocations over a 5- and 10-year period were significantly greater in the DS group. Placement in a nursing home for end of life care was significantly higher in the DS group whereas the majority (90%) in the non-DS group remained in a group home setting. Mortality was significantly earlier in the DS group with the mean age at death to be 61.4 years compared with 73.2 years in the non-DS group. The authors concluded that the present results suggest that aging adults with DS encounter more relocations, and ar e more likely to have their final placement for end of life care in a nursing home. In contrast, the adults without DS were subjected to less relocation and remained in the same group home setting.

Pary, R.J.
Differential diagnosis of functional decline in Down's syndrome
Habilitative Mental Healthcare Newsletter, 1992, 11(6), 37-41. https://www.worldcat.org/title/habilitative-mental-healthcare-newsletter/oclc/21277851
Abstract: (non provided - extract from article) Breif overview of examining adults with Down syndrome (DS) for functional decline. Provides brief summary of Alzheimer's disease and the prevalence and occurrence of AD in persons with DS. Provides two tables, Table 1: Disorders which are common in DS and Miscellaneous cerebral conditions with no apparent predilections for individuals with DS; Table 2: Work-up for functional decline in DS. Discusses complicating factors such as thyroid disease, major depression, sensory impairment, infection, and other conditions. Provides brief description of factors comprising a diagnostic workup. References (n-49) provide historical glimpse into extant topical literature prior to 1992.

Parajuá-Pozo, J.L., & Casis-Arguea, S.
Síndrome de Down y demencia [Down's syndrome and dementia].
Revisita de Neurologia, 2000 Jul 16-31;31(2):126-8. Spanish. DOI: https://doi.org/10.33588/rn.3102.2000075
Abstract: The authors reviewed the relationship between Down's syndrome and Alzheimer's disease. Down's syndrome is the commonest autosomopathy. Its clinical features permit easy diagnosis. Control of the more severe complications, such as cardiac malformations have allowed prolonged survival. The presence of neuropathological findings similar to those in Alzheimer's disease is almost constant in the brains of patients aged over 40 years with Down's syndrome. Dementia appears in a third of these patients. In this article we comment on the diagnostic difficulties, possible risk factors and the importance of findings related

to chromosome 21. They also evaluated the results of new treatments with cholinesterase inhibitors. The study of the close relationship between Down's syndrome and Alzheimer's disease should allow us to advance understanding of both of these conditions.

Pendl, D., Glatz, M., & Gasteiger-Klicpera, B.
Intellectual disabilities and dementia: New tasks and experiences of Austrian formal caregivers.
Journal of Applied Research in Intellectual Disability, 2024, 37(1), e13165. doi: 10.1111/jar.13165. .
Abstract: In Austria, due to its history, only relatively little research on the topic of intellectual disabilities and dementia has been conducted to date. The present study thus aims to explore the challenges and tasks currently facing formal caregivers, together with assessing their wishes for further development.
Ten semi-structured interviews were held with formal caregivers. Interviews were transcribed and analysed by means of structured qualitative content analysis. Caregivers must deal with conflicts between residents, and with increasing demands for care and emotional support. Education and training on dementia and intellectual disability are mostly of high quality, but still remain insufficient. Caregivers would like to see suitable adaptation of care premises, smaller groups, more staff and better training on dementia and intellectual disability. To ensure quality care and `ageing in place', caregivers and providers need to pay greater attention to dementia-related changes when planning and adapting services.

Perera, B., Kamieniarz, L., Iftikhar, M. & Solomou, S.
Screening and diagnosing dementia in people with Down's syndrome: implications of using the DLD questionnaire
Advances in Mental Health and Intellectual Disabilities, 2022, 16(4), 239-248. https://doi.org/10.1108/AMHID-04-2022-0015
Abstract: The Dementia Questionnaire for People with Learning Disabilities (DLD) is one of the main screening and monitoring tools for dementia in people with Down's syndrome (DS). As part of a quality improvement project to improve the care for people with DS and dementia in an intellectual disability service, the authors studied the screening and monitoring process by retrospectively investigating the use of DLD and exploring clinicians' experience of using it. DLDs completed in the service was retrospectively assessed. Changes in DLD scores were matched against people who received a clinical diagnosis of dementia. Data were analyzed to estimate sensitivity, specificity and predictive values of DLD. A questionnaire was used to assess clinicians' experience. Data for 20 service users was collected. DLD cognitive scores showed 80% sensitivity and 60% specificity for the diagnosis of dementia, with a positive predictive value of 40% and negative predictive value of 90%. Staff found DLD to be easy to perform but time consuming. This led to the preparation of a decision tool for appropriateness of performing a DLD. **The results show that a negative DLD helps to exclude dementia where there is concern over cognitive decline, but a positive result is not specific enough to suggest the possibility of dementia**. This shows that DLD may have limitations if used as a screening tool alone but could be used for the monitoring of the disease trajectory of those with a confirmed diagnosis as well as to establish a baseline DLD when a person is screened for dementia first.

Persaud, M., & Jaycock, S.
Evaluating care delivery: the application of dementia care mapping in learning disability residential services
Journal of Learning Disabilities, 2001, 5(4), 345-352. https://doi.org/10.1177/146900470100500
Abstract: Measurement and evaluation in intellectual disability services is still in its infancy. This report explores how good practice in relation to quality of care initiatives in dementia care transpose into intellectual disability settings. The authors applied dementia care mapping (DCM) to evaluate its effectiveness and efficiency in generic intellectual disability settings. Results showed that the application of the method to be partially successful. The data produced compared favorably in quality, quantity and detail with those collected in dementia care areas. Analysis of data revealed great potential for the method; however, result indices and coding frameworks need to be modified and adapted in future studies. No subject had dementia.

Percy, M.E., & Lukiw, W.J.
Is heart disease a risk factor for low dementia test battery scores in older persons with Down syndrome? Exploratory, pilot study, and commentary.
International Journal of Developmental Disabilities, 2020, 66(1), 22-35. doi: 10.1080/20473869.2017.1301023. Epub 2017 Apr 9.
Abstract: Certain heart conditions and diseases are common in Down syndrome (DS; trisomy 21), but their role in early onset dementia that is prevalent in older adults with DS has not been evaluated. To address this knowledge gap, we conducted a study of risk factors for low neurocognitive/behavioral scores obtained with a published dementia test battery (DTB). Participants were adults with DS living in New York (N = 29; average age 46 years). We asked three questions. 1. Does having any type of heart disease affect the association between DTB scores and chronological age? 2. Does thyroid status affect the association between heart disease and DTB scores? 3. Are the E4 or E2 alleles of apolipoprotein E (APOE) associated with DTB scores or with heart disease? The study was retrospective, pilot, and exploratory. It involved analysis of information in a database previously established for the study of aging in DS. Participants had moderate intellectual disability on average. Information for each person included: sex, age, a single DTB score obtained by combining results from individual subscales of the DTB, the presence or absence of heart disease, thyroid status (treated hypothyroidism or normal), and APOE genotype. Trends were visualized by inspection of graphs and contingency tables. Statistical methods used to evaluate associations included Pearson correlation analysis, Fisher's exact tests (2-tailed), and odds ratio analysis. P values were interpreted at the 95% confidence level without Bonferroni correction. P values >.05<.1 were considered trends. The negative correlation between DTB scores and age was significant in those with heart disease but not in those without. Heart disease was significantly associated with DTB scores >1 SD below the sample mean; there was a strong association between heart disease and low DTB scores in those with treated hypothyroidism but not in those with normal thyroid status. The APOE genotype was weakly associated with heart disease (E4, predisposing; E2, protective) in males. On the basis of the potentially important findings from the present study, large prospective studies are warranted to confirm and extend the observations. In these, particular heart conditions or diseases and other medical comorbidities in individuals should be documented.

Peterson, M.E., & O'Bryant, S.E.
Blood-base biomarkers for Down syndrome and Alzheimer's disease: A systematic review
Developmental Neurobiology, 2019, 79(7), 699-710. https://doi.org/10.1002/dneu.22714
Abstract: Down syndrome (DS) occurs due to triplication of chromosome 21. Individuals with DS face an elevated risk for development of Alzheimer's disease (AD) due to increased amyloid beta (Aß) resulting from the over-expression of the amyloid precursor protein found on chromosome 21. Diagnosis of AD among individuals with DS poses particular challenges resulting in an increased focus on alternative diagnostic methods such as blood-based biomarkers. The aim of this review was to evaluate the current state of the literature of blood-based biomarkers found in individuals with DS and particularly among those also diagnosed with AD or in prodromal stages (mild cognitive impairment [MCI]). A systematic review was conducted utilizing a comprehensive search strategy. Twenty-four references were identified, of those, 22 fulfilled inclusion criteria were selected for further analysis with restriction to only plasma-based biomarkers. Studies found Aß to be consistently higher among individuals with DS; however, the link between Aß peptides (Aß1-42 and Aß1-40) and AD among DS was inconsistent. Inflammatory-based proteins were more reliably found to be elevated leading to preliminary work focused on an algorithmic approach with predominantly inflammatory-based proteins to detect AD and MCI as well as

predict risk of incidence among DS. Separate work has also shown remarkable diagnostic accuracy with the use of a single protein (NfL) as compared to combined proteomic profiles. This review serves to outline the current state of the literature and highlights the potential plasma-based biomarkers for use in detecting AD and MCI among this at-risk population.

Petronis, A.
Alzheimer's disease and down syndrome: from meiosis to dementia
Experimental Neurology, 1999, Aug, 158(2), 403-413.
doi:10.1006/exnr.1999.7128.
Abstract: Several molecular and clinical similarities have been detected in Alzheimer's disease (AD) and Down syndrome (DS). The most remarkable feature is abnormal accumulation of beta-amyloid in the brains of both individuals affected with AD and aging DS patients followed by dementia. In addition, AD patients exhibit dermatoglyphic patterns similar to those in DS, and late maternal age is a risk factor in both diseases. AD and DS could be related genetically because AD families exhibit a higher rate of DS cases and vice versa. Although numerous discoveries have been made in the elucidation of the etiopathogenic factors in AD and DS, little progress has been achieved in understanding the origin of the common features of the two diseases. This article reviews clinical and molecular similarities in DS and AD and also chromosome 21 studies in both diseases. A new hypothesis explaining the association between AD and DS is suggested, and this hypothesis is based on the poorly understood molecular phenomenon of aberrant meiotic recombination. Aberration in meiotic recombination has been consistently detected in chromosomal diseases including trisomy 21 and sex chromosomes. There are no studies dedicated to meiotic recombination in genetic diseases; however, evidence for disturbed recombination has been documented in several neurological diseases such as Huntington's disease, myotonic dystrophy, and fragile X syndrome. Interestingly, the rate of trisomic XXY children born to mothers transmitting fragile X mutation is higher than expected. This finding suggests that AD could be associated with DS in a similar way to which fragile X syndrome is related to trisomy of sex chromosomes. Based on analogy with fragile X syndrome, it can be predicted that AD should demonstrate aberrant meiotic recombination in chromosome 21, most likely in the region D21S1/S11-D21S16 which is linked to early onset familial AD. Based on the same rationale, different patterns of meiotic recombination in the nondisjunct chromosome 21 within DS patients grouped according to the concomitant disease are predicted.

Pollack, H.A., Tabman, B., Brown, D.S., Wen, H., Berg, K.L., Peterson, L., Cliff, B.Q., Ances, B.M., & Johnston, K.J.
Neuropsychiatric disorders among adult emergency department patients with intellectual and developmental disabilities.
Psychiatric Services, 2025 Sep 4:appips20240604. doi: 10.1176/appi.ps.20240604. Epub ahead of print.
Abstract: This study aimed to compare rates of psychiatric and neurologic diagnoses on emergency department (ED) visit records of adults with versus without intellectual and developmental disabilities (IDDs). This cross-sectional study used the 2019 Nationwide Emergency Department Sample of U.S. hospital ED visit discharges. Validated codes were used to compare psychiatric and neurologic diagnoses of patients ages ≥18 with versus without diagnosed IDDs. Diagnosed psychiatric and neurologic conditions included depression, anxiety, schizophrenia or psychosis, suicidality, seizure, dementia, and sleep disorder. The analysis identified 558,408 and 112,593,527 (nationally weighted) ED visits by adults with and without IDDs, respectively. Compared with the general population, adults with IDDs were twice as likely to have a mental disorder as the principal visit diagnosis, with higher probabilities of principal visit suicidality (1.6 times higher), neurologic disorder (5.6 times higher), and seizure (8.1 times higher) diagnoses. Compared with the general population, adults with intellectual disability were nearly **twice as likely** to have a **dementia diagnosis**, and patients with **Down's syndrome were six times likelier** to have a **dementia diagnosis.** More than **one in five ED visit records of patients ages 50-54 with Down's syndrome included a dementia diagnosis**; the dementia diagnosis rate for such patients ages ≥70 was 2.7 times higher than that of the general population. Adult ED patients with IDDs were more likely than those without IDDs to have co-occurring mental and neurologic disorders. Findings underscore the need to provide neuropsychiatric services across the lifespan to address the distinctive care needs of individuals with IDDs.

Pollack, H.A., Johnston, K., & Chicoine, B.
Persons with Down syndrome face a 90% lifetime dementia risk - A reality of aging that must be addressed
The Milbank Quarterly Opinion, August 11, 2025.
https://doi.org/10.1599/mqop2025.0812
Abstract: [None provided - extract from narrative]. Our team recently analyzed 133 million adult ED patient encounters across the United States. More than 500,000 of these encounters involved persons living with intellectual disability (ID). Indicated dementias among patients with Down syndrome between the ages 45 and 49 were more common than was observed in ED encounters in the broader population over age 70. More than one-third of ED visits among patients with Down syndrome ages 55-64 include dementia indications. We must ensure that primary care providers are properly trained to help families when adults with ID present with challenges. This requires funding, quality incentives, and other strategies to increase provider uptake for pertinent training. The need for incentives is particularly great given the reality that older persons with Down syndrome and co-occurring dementia are a small fraction of most providers' medical practice. Most Americans with Down syndrome, and most Americans living with other ID, can now be expected to outlive their parents and other primary caregivers. Most will experience significant, often-accelerated infirmities that accompany human aging. Many will experience these infirmities while living at home with their parents or other older-adult caregivers. Americans with disabilities and their families require better supports to meet these challenges. This requires our culture, our care systems, and our public policies to keep pace with the realities of human aging.

Post, S.G.
Down syndrome and Alzheimer disease: defining a new ethical horizon in dual diagnosis.
Alzheimer's Care Quarterly, 2002, 3(3), 215-224.
https://www.researchgate.net/publication/232173830_Down_Syndrome_and_Alzheimer_Disease_Defining_a_New_Ethical_Horizon_in_Dual_Diagnosis
Abstract:
Within the context of our demographic transition to an aging society, special emphasis must be placed on the somewhat hidden topic of cognitive developmental disabilities and Alzheimer's disease (AD). This article focuses on end-of-life issues and the phenomenology of caring, although not at the expense of broader concerns. Drawing on the work of Jean Vanier, Tom Kitwood, and others, foundational continuities between caring for persons with retardation and persons with irreversible progressive dementia are articulated. In [intellectual disabiity], cognitive development never reaches its full heights; in dementia, the heights once reached are now radically diminished. In either case, we are dealing with cognitive disabilities. Regrettably, many persons with Down syndrome (DS) also show symptoms of AD as they reach their 40s and 50s, making this population unique in representing the interests of two otherwise separate domains of care and policy. This article presents some background demographic information and clinical epidemiology as prelude to a broad discussion of the ethics of caregiving and of a set of pressing ethical issues.

Pounds, O., Theodore, K., & Dodd, K.
Factors associated with Alzheimer's dementia diagnosis and survival in Down syndrome.
Journal of Intellectual Disability Research, 2025 Jun, 69(6), 489-501. doi: 10.1111/jir.13230. Epub 2025 Mar 12
Abstract: People with Down syndrome (DS) have an increased risk for Alzheimer's disease. Identifying factors associated with dementia onset and

subsequent survival will support in understanding the disease profile, improving timely diagnosis, management, and care planning. Variables associated with age at dementia onset and survival times were assessed in 279 adults with DS who accessed a community learning disability service. After outliers were removed, regression and hazard regression models were used for disease onset (n = 265) and survival times (n = 180), respectively. Earlier age at first assessment and living with family predicted earlier age at diagnosis, which led to longer survival, post-diagnosis. Epilepsy and living in a long-stay hospital were associated with earlier mortality. Implications for clinical practice include reflections on the importance of early baseline assessments and caregiver awareness. Suggestions for future research include investigating intersectionality of social factors with genetics to better understand ad trajectories.

Prasher, V.
End-stage dementia in adults with Down syndrome
International Journal of Geriatric Psychiatry, 1995, 10(12), 1067-1069. https://doi.org/10.1002/gps.930101213
Abstract: End-stage dementia in adults with Down syndrome has not been fully investigated. Available information, 6 months prior to death, for 20 adults with Down syndrome who had died with Alzheimer's disease was reviewed. A terminal stage of severe intellectual deterioration, marked personality and mood changes, loss of sphincter control, seizure activity, immobility with hypertonia and complete loss of self-care skills was found. These findings have important clinical and service implications.

Prasher, V.P.
Review of donepezil, rivastigmine, galantamine and memantine for the treatment of dementia in Alzheimer's disease in adults with Down syndrome: implications for the intellectual disability population
International Journal of Geriatric Psychiatry, 2004, 19, 509 - 515. doi: 10.1002/gps.1077.
Abstract: The management of dementia in Alzheimer's disease has dramatically changed since the development of anti-dementia drugs. However, there is limited information available regarding the bio-medical aspects of the differing drugs; particularly relating to adults with intellectual disability. Indeed the information available for the intellectual disabled population is limited to adults with Down syndrome. This review highlights the important pharmacological and clinical aspects of donepezil, rivastigmine, galantamine and memantine and supports the view that such drugs play an important part in the management of dementia in adults with intellectual disability. Future clinical and research issues are discussed.

Prasher, V. P.
Psychotic features and effect of severity of learning disability on dementia in adults with down syndrome: review of literature.
British Journal of Development Disabilities, 1997, 43(85), 85-92. https://doi.org/10.1179/bjdd.1997.010
Abstract: None available.

Prasher, V.P., & Corbett, J.A.
Onset of seizures as a poor indicator of longevity in people with down syndrome and dementia
International Journal of Geriatric Psychiatry,1993, 8(11), 923-927. https://doi.org/10.1002/gps.930081106
Abstract: An association between Down syndrome and dementia of Alzheimer type is well established. This study demonstrates that late onset seizures in people with Down syndrome are a strong indicator of a dementing process. Further, late onset seizures in people with Down syndrome may be used as a prognostic indicator, indicating life expectancy of less than a further 2 years, probable death within 3 years and death almost invariably within 5 years of onset.

Prasher, V., Farooq, A. & Holder, R.
The Adaptive Behavior Dementia Questionnaire (ABDQ): screening questionnaire for dementia in Alzheimer's disease in adults with Down syndrome
Research in Developmental Disabilities, 2004, 25(4), 385-397. doi: 10.1016/j.ridd.2003.12.002.
Abstract: The diagnosis of dementia in Alzheimer's disease remains at times problematic in adults with intellectual disability. The analysis of 5-year consecutive data developed a researched-based clinical screening tool for dementia in Alzheimer's disease in adults with Down syndrome. The Adaptive Behavior Dementia Questionnaire (ABDQ) is a 15-item questionnaire, which is used to detect change in adaptive behavior. The scale has good reliability and validity, with an overall accuracy of 92%. It is one of the first clinical tools designed specifically to screen for dementia in Alzheimer's disease in adults with Down syndrome.

Prasher, V.P.,& **Filer, A.**
Behavioural disturbance in people with Down's syndrome and dementia.
Journal of Intellectual Disabilities Research, 1995, 39(5), 432-436. doi: 10.1111/j.1365-2788.1995.tb00547.x.
Abstract: Behavioral disturbance associated with dementia in people with Down syndrome has not been fully researched. This study investigated such problems in subjects with Down syndrome and dementia and controls with Down syndrome but free of dementia. Changes in mood, difficulty with communication, gait deterioration, loss of self-care skills, sleep disturbance, day-time wandering and urinary incontinence were found to be associated with dementia. Problems giving the greatest cause for concern to carers were restlessness, loss of communication skills, urinary incontinence and wandering. Care provision specifically focused on management of behavioral disturbance in individuals who develop dementia is recommended.

Prasher, V.P. & Krishnan, V.H.R.
Age of onset and duration of dementia in people with Down syndrome: Integration of 98 reported cases in the literature
International Journal of Geriatric Psychiatry, 1993 Nov, 8(11), 915-922 https://doi.org/10.1002/gps.930081105
Abstract: The published case reports of 98 people with Down Syndrome were studied with respect to the age of onset and duration of clinically diagnosed dementia. The incidence of dementia was unimodal, increasing rapidly from 40 years to a peak of 30% incidence in the fifth decade of life, followed by a further rapid decline. Females with Down syndrome had an earlier onset. Duration of dementia decreased with increasing age of onset for both males and females.

Prasher, V.P., Janicki, M.P., Jozsval, E., Berg, J.M., Lovering, J.S., Rashid, A., Fung, W.L.A., & Percy, M.
Alzheimer's disease and dementia: Implications for people with Down syndrome and other intellectual and developmental disabilities.
Chapter 49 (pp. 709-733) in Wehmeyer, M., Brown, I., Percy, M., Shogren, K.A., Fung, W.L.A., (Eds.), *A Comprehensive Guide to Intellectual and Developmental Disabilities*. Baltimore, MD: Paul Brookes Publishing.
Abstract: Chapter provides comprehensive information in its first three sections on Alzheimer's disease and dementias and how these disorders affect people with Down syndrome and other intellectual disabilities, including assessment and early detection of dementia. The fourth section covers the multidisciplinary management of dementia as occuring in people with intellectual disabilities. Subsections cover pharmacological approaches and nonpharmacological interventions, as well as environmental considerations for safe living, and services and resources needed by people with dementia and their caregivers.

Prasher, V.P., Mahmood, H., & Mitra, M.
Challenges faced in managing dementia in Alzheimer's disease in patients with Down syndrome.
Degenerative Neurological and Neuromuscular Disease, 2016, 6, 85-94. doi: 10.2147/DNND.S91754. eCollection 2016.

Abstract: Dementia in Alzheimer's disease (DAD) is more common in adults with Down syndrome (DS), with characteristically an earlier onset. The treatment of DAD is not too dissimilar in the general population and in people with intellectual disabilities. However, the underlying intellectual disability can make the management of DAD more challenging in older adults with DS. This literature review aimed to look at the management of DAD in people with DS. The management of dementia is holistic. This includes treating reversible factors, aiming to slow the cognitive decline, psychological therapies, ensuring that the environment is appropriate, and use of psychotropic medication when necessary to manage behavioral problems, psychotic symptoms, depressive symptoms, and sleep difficulty. Antidementia medications have a role to play but remain limited. The management of DAD in the DS population can be at times challenging, but good clinical practice should involve accurate diagnosis of dementia, treating any reversible additional factors, consideration of psychological and behavioral management, use of antidementia medication, and a multidisciplinary team approach.

Prasher, V.P., Metseagharun, T., & Haque, S.
Weight loss in adults with Down syndrome and with dementia in Alzheimer's disease.
Research in Developmental Disabilities, 2004, Jan-Feb, 25(1), 1-7. doi: 10.1016/j.ridd.2003.04.005.
Abstract: An association between weight loss and Alzheimer's disease has been established in the general population but little information is available regarding this association in people with intellectual disabilities. A 4-year longitudinal study of adults with Down syndrome with and without Alzheimer's disease was undertaken. Age-associated weight loss was seen in virtually all older adults with Down syndrome. A significant association between weight loss and Alzheimer's disease was found for older adults with Down syndrome. This study highlights important research and clinical issues regarding weight loss and nutrition in Down syndrome adults with dementia.

Prasher, V.P., Sachdeva, N., & Tarrant, N.
Diagnosing dementia in adults with Down's syndrome.
Neurodegenerative Disease Management, 1015, 5(3), 249-256. doi: 10.2217/nmt.15.8.
Abstract: Individuals with Down's syndrome (DS) are living longer and many will survive into their fifth or sixth decade of life. Among the DS population, the prevalence of dementia in Alzheimer's disease increases from 9.4% in age group 30-39 years to 54.5% age group 60-69 years. The psychopathology of dementia in Alzheimer's disease is similar to that seen in the general population although differences are apparent due to the underlying intellectual disability in DS and on the reliance on collateral information from informants. The diagnostic workup follows accepted practice although neuropsychological tests and neuroimaging will only be adjuncts to the clinical assessment; such investigations have limited diagnostic value. Presently, research is focused on identifying genetic and biological measures of Alzheimer's disease in DS.

Prasher, V.P., Percy, M,, Janicki, M.P., Jozsvai, E., Fung, W.L.A., & Brown, I.
Implications of dementia for adults with developmental disabilities.
Chapter (pp. 699-721) in Brown, I. & Percy, M. (2020). *Developmental Disabilities in Ontario* (4th Ed.), Toronto, Ontario, Canada: Delphi Graphic Communications. https://oadd.org/publications/textbook/
Abstract: Chapter provides an introduction to the topic of dementia in persons with developmental disabilities. Dementia, as a worldwide public health concern, is increasing in prevalence markedly because the world's population if living longer and aging in greater numbers. Chapter covers the physiology of dementia, options for services, mechanisms for multidisciplinary management, and advances in advocacy, dementia prevention, and dementia research.

Proveda, B., & Broxholme, S.
Assessments for dementia in people with learning disabilities: Evaluation of a dementia battery developed for people with mild to moderate learning disabilities
Learning Disability Practice, 2016, 19(1), 31-40. doi.org/10.7748/ldp.19.1.31.s23
Abstract: An intellectual disabilities' dementia battery was developed to assess cognitive abilities in individuals referred to the intellectual disabilities service because of concerns of possible dementia. The present study aimed to establish concurrent validity with previously validated measures of cognitive ability and its clinical effectiveness in detecting dementia in this population. Fifty-five individuals aged 29 and over (range: 29 to 71), received a baseline and a follow-up assessment using the dementia battery between 2000 and 2010. Differences in performance between individuals allocated to 'probable', 'unsure' and 'no' dementia groupings were investigated at domain and subtest level, as well as overall performance. Results on the battery were compared with clinically relevant measures of dementia also included in the local assessment protocol. Significant differences in overall performance were found between the 'probable' and 'no' dementia groups as well as cognitive domain-specific differences. No differences were found at subtest level. Good concurrent validity was found between the battery and comparable measures of change within the dementia assessment protocol, namely the VABS, DMR and BPVS II. The intellectual disabilities' dementia battery appears to be a good measure, which can be used longitudinally, to detect change in individuals and help establish a diagnosis of dementia. It is also comparable with other measures of change incorporated in the dementia assessment protocol. Subtests included in the language domain appear to be the most relevant at detecting significant changes between baseline and follow up. Future studies should attempt to standardize this measure and establish cut-off scores.

PTAC [PASRR Technical Assistance Center]
How does a categorical determination for dementia and intellectual disability affect the PASRR process?
PTAC, April 10, 2018;
https://www.pasrrassist.org/resources/How-does-a-categorical-determination-for-dementia-and-intellectual-disability-affect-the-PASRR-process%3F
Abstract: 42 CFR 483.130(h) provides that the State intellectual disability authority may make categorical determinations that individuals with dementia, which exists in combination with an intellectual disability (ID) or a related condition, do not need specialized services. A categorical determination for dementia and ID can be applied at the Level I screening, but the categorical must be determined at the Level II evaluation phase of PASRR. Beyond the specialized services determination, there is no basis for ending a Level II evaluation for an individual with an intellectual disability diagnosis, as the evaluation still determines if nursing facility services are needed. Under 42 CFR § 483.128(j), findings must be issued in the form of an abbreviated written evaluative report which— (1) Identifies the name and professional title of the person applying the categorical determination and the data on which the application was made; (2) Explains the categorical determination(s) that has (have) been made and, if only one of the two required determinations can be made categorically, describes the nature of any further screening which is required; (3) Identifies, to the extent possible, based on the available data, NF services, including any mental health or specialized psychiatric rehabilitative services, that may be needed; and (4) Includes the bases for the report's conclusions. A categorical determination for dementia and intellectual disability also requires the issuing of a written determination notice per the following 483.130(k)(l) requirements: (k) Notice of determination. The State mental health or intellectual disability authority must notify in writing the following entities of a determination made under this subpart: 1. The evaluated individual and legal representative; 2. The admitting or retaining NF; 3. The individual or resident's attending physician; and 4. The discharging hospital, unless the individual is exempt from preadmission screening as provided at §483.106(b)(2). (l) Contents of notice. Each notice of the determination made by the State mental health or intellectual disability authority must include: 1. Whether a NF level of service is needed; 2. Whether specialized services are needed; 3. The placement options

that are available to the individual consistent with these determinations; and 4. The rights of the individual to appeal the determination

Puri, B.K., Ho, K.W., & Singh, I.
Age of seizure onset in adults with Down's syndrome.
International Journal of Clinical Practice, 2001, 55(7), 442-444.
https://pubmed.ncbi.nlm.nih.gov/11594252/
Abstract: In a cohort of 68 adults (35 males and 33 females) with Down's syndrome aged 29-83 years, a history of seizures was found in 26.5%. The overall mean age of onset of seizures was 37 years, males (22 years) being significantly younger than females (51 years). The age of onset was bimodally distributed, with the first peak occurring in the first two decades, and a late-onset peak occurring in the fifth and sixth decades. A strong association between Alzheimer's disease and seizures was confirmed. Of those with a history of seizures, those aged over 45 years were significantly more likely to develop Alzheimer's disease than those younger than 45. It is suggested that late-onset epilepsy in Down's syndrome is associated with Alzheimer's disease, while early-onset epilepsy is associated with an absence of dementia.

Pulsifer, M.B., Evans, C.L., Hom, C., Krinsky-McHale, S.J., Silverman, W., Lai, F., Lott, I., Schupf, N., Wen, J ., Rosas, H.D.
Language skills as a predictor of cognitive decline in adults with Down syndrome
Alzheimers Dement (Amst), 2020, Aug 25, 12(1), e12080.
doi: 10.1002/dad2.12080. eCollection 2020.
Abstract: Adults with Down syndrome (DS) are at high risk for early onset Alzheimer's disease (AD), characterized by a progressive decline in multiple cognitive domains including language, which can impact social interactions, behavior, and quality of life. This cross-sectional study examined the relationship between language skills and dementia. A total of 168 adults with DS (mean age = 51.4 years) received neuropsychological assessments, including Vineland Communication Domain, McCarthy Verbal Fluency, and Boston Naming Test, and were categorized in one of three clinical groups: cognitively stable (CS, 57.8%); mild cognitive impairment (MCI-DS, 22.6%); and probable/definite dementia (AD-DS, 19.6%). Logistic regression was used to determine how well language measures predict group status. Vineland Communication, particularly receptive language, was a significant predictor of MCI-DS. Semantic verbal fluency was the strongest predictor of AD-DS. Assessment of language skills can aid in the identification of dementia in adults with DS. Clinically, indications of emerging language problems should warrant further evaluation and monitoring.

Rafii, M.S.
Tau PET imaging for staging of Alzheimer's disease in Down syndrome
Developmental Neurobiology, 2019, 79(7), 711- 715.
https://doi.org/10.1002/dneu.22658
Abstract: Alzheimer's disease (AD) pathology and early-onset dementia develop almost universally in Down syndrome (DS). AD is defined neuropathologically by the presence of extracellular plaques of aggregated amyloid ß protein and intracellular neurofibrillary tangles (NFTs) of aggregated hyperphosphorylated tau protein. The development of radiolabeled positron emission tomography (PET) ligands for amyloid plaques and tau tangles enables the longitudinal assessment of the spatial pattern of their accumulation in relation to symptomatology. Recent work indicates that amyloid pathology develops 15–20 years before neurodegeneration and symptom onset in the sporadic and autosomal dominant forms of AD, while tau pathology correlates more closely with symptomatic stages evidenced by cognitive decline and dementia. Recent work on AD biomarkers in DS illustrates similarities between DS and sporadic AD. It may soon be possible to apply recently developed staging classifications to DS to obtain a more nuanced understanding of the development AD in DS and to provide more accurate diagnosis and prognosis in the clinic.

Rafii, M.S.
Alzheimer's disease in Down syndrome: Progress in the design and conduct of drug prevention trials.
CNS Drugs, 2020 Aug, 34(8), 785-794. doi: 10.1007/s40263-020-00740-6.
Abstract: Individuals with Down syndrome (DS) are at high risk for developing Alzheimer's disease (AD) pathology and this has provided significant insights into our understanding of the genetic basis of AD. The present review summarizes recent clinical, neuropathologic, imaging, and fluid biomarker studies of AD in DS (DSAD), highlighting the striking similarities, as well as some notable differences, between DSAD and the more common late-onset form of AD (LOAD) in the general population, as well as the much rarer, autosomal-dominant form of AD (ADAD). There has been significant progress in our understanding of the natural history of AD biomarkers in DS and their relationship to clinically meaningful changes. Additional work is needed to clearly define the continuum of AD that has been described in the general population, such as the preclinical, prodromal, and dementia stages of AD. Multiple therapeutic approaches, including those targeting not only ß-amyloid but also tau and the amyloid precursor protein itself, require consideration. Recent developments in the field are presented within the context of such efforts to conduct clinical trials to treat and potentially prevent AD in DS.

Rafii, M.
The amyloid clock: mapping Alzheimer's disease in Down syndrome
Lancet Neurology, 2024, Dec., 23, 1173-1174.
https://doi.org/10.1016/S1474-4422(24)00437-X
Abstract: [Not provided]. A Lancet Neurology [Schworer et al., 2024] report noted the findings of a longitudinal study from the Alzheimer's Biomarker Consortium-Down Syndrome (ABC-DS) cohort, which provides novel insights into the Alzheimer's disease of individuals with Down syndrome (DS).The authors describe the concept of amyloid age, which is based on centiloid magnitude (0 years equates to 18 centiloids) and offers a new metric for quantifying the progression of Alzheimer's disease (AD) in people with DS. The findings also highlight the importance of early amyloid accumulation and its association with subsequent tau pathology and cognitive decline. One of the key findings of the study is the stability in cognitive performance-for approximately 3·5 years-in people with DS who are amyloid ß-PET positive, after which a marked decline begins. The lag between amyloid positivity and cognitive decline, specifically in episodic memory, defines the so-called preclinical DS-AD and underscores the crucial window for early intervention in clinical trials. The mean amyloid age was 7·4 years (SD 6·6; equating to 62·1 centiloids) for individuals with mild cognitive impairment and 12·7 years (5·6; 99·3 centiloids) for those with dementia. This finding suggests an accelerated clinical progression in people with DS, compared with individuals with late-onset AD, in whom mild cognitive impairment typically occurs for 15·5 years after the initial detection of amyloid ß positivity. The study showed the alignment of tau pathology with cognitive decline-specifically, that increases in neurofibrillary tangle pathology occur at approximately 2·7 years after the onset of amyloid ß positivity. This finding provides another key target for trial interventions aimed at slowing the progression of tau pathology and is vital for staging interventions and for evaluating long-term therapeutic outcomes. These findings also align with the revised criteria for diagnosis and staging of AD, which emphasizes its biological
underpinnings, particularly the sequential accumulation of amyloid and tau pathologies. This alignment reinforces the importance of biomarker-based staging of AD, particularly for clinical trial design and therapeutic interventions.

Rafii, M., & Fortea, J.
Down syndrome in a new era for Alzheimer disease.
JAMA. 2023 Dec 12;330(22):2157-2158. doi: 10.1001/jama.2023.22924.
Abstract: [Not provided] US Food and Drug Administration-approved disease-modifying treatments for Alzheimer disease (AD) are now available by prescription; the approval is based on evidence from large randomized, placebo-controlled trials of amyloid-lowering immunotherapies, which have demonstrated that reducing amyloid plaques results in improved outcomes in patients with early-stage AD. However, adults with Down syndrome (DS) have

not been included in any of these trials. The approximately 6 million individuals with DS worldwide are the largest population with genetically determined AD in the world. Virtually all individuals with DS develop amyloid plaques by the age of 40 years, and more than 95% will develop AD dementia by their seventh decade, with average age of dementia onset being 54 years. With an ultra-high risk for AD and a shortened life expectancy, persons with DS who experience AD-related cognitive decline still have limited access to expert clinical evaluation for dementia, or to newly approved therapeutics. This has caused a quandary in that drugs routinely administered to individuals with DS do not necessarily have level I evidence from clinical trials performed in this population. Adults with DS-AD, as well as those with autosomal-dominant Alzheimer disease, show a higher burden of amyloid angiopathy compared with adults with sporadic AD -- which lead to higher risk for adverse events related to ARIA. However, in the absence of data in adults with DS, this theoretical risk has not materialized so far in autosomal-dominant AD. Ensuring equitable access should therefore be the priority, while balanced with safety. In this era of diversity, equity, and inclusion, now is the time to bring these new, disease-modifying treatments to individuals who have been left behind for far too long.

Rafii, M., & Santoro, S.L.
Prevalence and severity of Alzheimer disease in individuals with dementia.
JAMA Neurology, 2019, 76, 142-143. doi:10.1001/jamaneurol.2018.3443
Abstract: The median life expectancy for a child with Down syndrome (DS) born in the 1950s was less than 10 years of age, with congenital heart defects being the main cause of death. With advances in medical care and improvements in the overall health of individuals with DS, life expectancy has increased dramatically; for children with DS born in 2010, median life expectancy is estimated to be 65 years. However, along with this longer lifespan comes the prospect of a considerable increase in the risk of developing dementia associated with Alzheimer disease (AD), with a prevalence of nearly 80% for those with DS who are older than 65 years. A commentary/editorial on the work published by Hithersay et al. in JAMA Neurol.
doi:10.1001/jamaneurol.2018.3616

Rafii, M.S., Ances, B.M., Schupf, N., Krinsky-McHale, S.J., Mapstone, M., Silverman, W., Lott, I....O'Bryant, S.
The AT(N) framework for Alzheimer's disease in adults with Down syndrome.
Alzheimer's & Dementia (Amst), 2020,Oct 27,12(1),e12062. doi: 10.1002/dad2.12062. eCollection 2020.
Abstract: The National Institute on Aging in conjunction with the Alzheimer's Association (NIA-AA) recently proposed a biological framework for defining the Alzheimer's disease (AD) continuum. This new framework is based upon the key AD biomarkers (amyloid, tau, neurodegeneration, AT[N]) instead of clinical symptoms and represents the latest understanding that the pathological processes underlying AD begin decades before the manifestation of symptoms. By using these same biomarkers, individuals with Down syndrome (DS), who are genetically predisposed to developing AD, can also be placed more precisely along the AD continuum. The A/T(N) framework is therefore thought to provide an objective manner by which to select and enrich samples for clinical trials. This new framework is highly flexible and allows the addition of newly confirmed AD biomarkers into the existing AT(N) groups. As biomarkers for other pathological processes are validated, they can also be added to the AT(N) classification scheme, which will allow for better characterization and staging of AD in DS. These biological classifications can then be merged with clinical staging for an examination of factors that impact the biological and clinical progression of the disease. Here, we leverage previously published guidelines for the AT(N) framework to generate such a plan for AD among adults with DS.

Rafii, M.S., Wishnek, H., Brewer, J.B., Donohue, M.C., Ness, S., Mobley, W.C., Aisen, P.S., & Rissman, R.A.
The Down syndrome biomarker initiative (DSBI) pilot: Proof of concept for deep phenotyping of Alzheimer's disease biomarkers in Down syndrome
Frontiers in Behavioral Neuroscience, 2015, Sep 14, 9, 239. doi: 10.3389/fnbeh.2015.00239. eCollection 2015.
Abstract: To gain further knowledge on the preclinical phase of Alzheimer's disease (AD), we sought to characterize cognitive performance, neuroimaging and plasma-based AD biomarkers in a cohort of non-demented adults with down syndrome (DS). The goal of the down syndrome biomarker Initiative (DSBI) pilot is to test feasibility of this approach for future multicenter studies. We enrolled 12 non-demented participants with DS between the ages of 30-60 years old. Participants underwent extensive cognitive testing, volumetric MRI, amyloid positron emission tomography (PET; 18F-florbetapir), fluorodeoxyglucose (FDG) PET (18F-fluorodeoxyglucose) and retinal amyloid imaging. In addition, plasma beta-amyloid (Aß) species were measured and Apolipoprotein E (ApoE) genotyping was performed. Results from our multimodal analysis suggest greater hippocampal atrophy with amyloid load. Additionally, we identified an inverse relationship between amyloid load and regional glucose metabolism. Cognitive and functional measures did not correlate with amyloid load in DS but did correlate with regional FDG PET measures. Biomarkers of AD can be readily studied in adults with DS as in other preclinical AD populations. Importantly, all subjects in this feasibility study were able to complete all test procedures. The data indicate that a large, multicenter longitudinal study is feasible to better understand the trajectories of AD biomarkers in this enriched population.

Rafii, M.S., Zaman, S., & Handen, B.L.
Integrating biomarker outcomes into clinical trials for Alzheimer's disease in Down syndrome.
Journal of Prevention of Alzheimer's Disease, 2021, 8(1), 48-51. doi: 10.14283/jpad.2020.35.
Abstract: The NIH-funded Alzheimer's Biomarker Consortium Down Syndrome (ABC-DS) and the European Horizon 21 Consortium are collecting critical new information on the natural history of Alzheimer's Disease (AD) biomarkers in adults with Down syndrome (DS), a population genetically predisposed to developing AD. These studies are also providing key insights into which biomarkers best represent clinically meaningful outcomes that are most feasible in clinical trials. This paper considers how these data can be integrated in clinical trials for individuals with DS. The Alzheimer's Clinical Trial Consortium - Down syndrome (ACTC-DS) is a platform that brings expert researchers from both networks together to conduct clinical trials for AD in DS across international sites while building on their expertise and experience.

Raj,S., Stanley, M., Mackintosh,S., & Fryer, C.
Scope of occupational therapy practice for adults with both Down syndrome and dementia: A cross-sectional survey
Australian Occupational Therapy Journal, 2022, 67(3), 218-228. https://doi.org/10.1111/1440-1630.12645
Abstract: Dementia in adults with Down syndrome causes a progressive decline in daily occupations impacting both persons with Down syndrome and their informal caregivers. This study aimed to explore the scope of occupational therapy practice for adults with both Down syndrome and dementia and their informal caregivers living in their homes. A survey was conducted with occupational therapists having clinical experience in providing interventions for adults with Down syndrome. A web-based survey was developed to explore occupational therapy practice for this group of people with Down syndrome and their informal caregivers. Responses to closed-ended questions were analysed descriptively, and inductive content analysis was used for open-ended questions. Forty-three occupational therapists from Australia, Canada, United Kingdom and the United States of America participated in the survey. Two-thirds were from the United Kingdom, most of whom were employed in the public sector and had at least 10 years of clinical experience. Over 90% of respondents received one or more referrals in a typical month for adults with Down syndrome having dementia, 68% of which were for a decline in activities of daily living. Home environment and activities of daily living were frequently assessed areas, and the commonest interventions were compensatory strategies and environmental modifications. Only half the respondents provided interventions for informal caregivers. Risk and safety and manual handling were frequently addressed

domains for informal caregivers. Collaboration and developing clinical expertise were the two key perceived enablers for providing effective occupational therapy services. Fragmentation of services and a lack of client-centred care were the common perceived barriers. Occupational therapists often address decline in activities of daily living for individuals with both Down syndrome and dementia. To support participation in meaningful occupations for these people and support the needs of their informal caregivers, it is essential that services are offered in a collaborative approach.

Raj, S.E., Mackintosh,S., Kernot,J., Fryer, C., & Stanley, M.
Development and feasibility testing of an evidence-based occupational therapy program for adults with both Down syndrome and dementia
Journal of Policy and Practice in Intellectual Disabilities, 2022, First published: 28 June 2022. https://doi.org/10.1111/jppi.12435
Abstract: This paper describes the development of a home-based occupational therapy intervention program for people with Down syndrome who experience early on-set dementia causing a decline in their performance skills and increasing care dependency on their informal caregivers. A six-step methodological process adapted from the Medical Research Council framework for developing and evaluating complex interventions was formulated to develop an evidence-based occupational therapy program for people with both Down syndrome and dementia and their informal caregivers. The first two steps gathered evidence through systematic reviews of the literature and determined the scope of current occupational therapy practice. The gathered evidence was synthesised in step three to develop a client-centred occupational therapy intervention program for persons with both Down syndrome and dementia and their informal caregivers. In steps four and five, opinions were sought from occupational therapists working in this area of practice on the content of the developed program and its feasibility within the Australian disability services context. The final testing step can be conducted in the future using a single-case experimental design study. It is important to use rigorous frameworks and gather comprehensive evidence using multiple methods to develop interventions for small heterogeneous populations. The developed occupational therapy program for persons with both Down syndrome and dementia and their informal caregivers appears feasible to be implemented within the Australian disability services; however, funding limitations imposes barriers for its implementation in clinical practice.

Rebillat, A-S., Hiance-Delahaye, A., Falquero, S., Radice, G., & Sacco, S.
The French translation of the dementia screening questionnaire for individuals with intellectual disabilities is a sensitive tool for screening for dementia in people with Down Syndrome.
Research in Developmental Disabilities, 2021 Nov; 118, 104068. doi: 10.1016/j.ridd.2021.104068. Epub 2021 Aug 28.
Abstract: People with Down syndrome (DS) are at an increased risk of developing Alzheimer's disease (AD) relatively early in life. The dementia screening questionnaire for individuals with intellectual disabilities (DSQIID) has been developed for people with intellectual disabilities and was shown to have high discriminative power to distinguish between people with and without dementia. The objective of this study was to verify if the French version of the DSQIID (DSQIID-F) had a good diagnostic specificity and to determine the optimal cut-off for screening people with DS for dementia. This was a single-centre, retrospective, medical chart review study in people with DS aged =40 years. Demographics, level of intellectual disability, DSQIID-F data and clinical assessment of dementia were extracted from medical records. Sensitivity and specificity for different DSQIID-F cut-offs were calculated to determine the optimal cut-off. Some 151 people with DS were included with a median age of 51 years. The optimal DSQIID-F cut-off was 19, sensitivity was 0.940 (95 % CI: 0.830; 0.985) and specificity was 0.941 (95 % CI: 0.873; 0.975). Results were comparable to those for the English DSQIID (cut-off: 20; sensitivity: 0.92; specificity: 0.97). However, the psychometric qualities of the DSQIID-F, used for clinical follow-up, have not been verified. The DSQIID-F has good discriminative power and represents a useful tool to screen people with DS for dementia

Reppermund, S., & Trollor, J.N.
Successful ageing for people with an intellectual disability
Current Opinion in Psychiatry, 2016, March, 29(2), 149-154. doi: 10.1097/YCO.0000000000000228
Abstract: Successful ageing has not yet been defined in people with an intellectual disability. The purpose of this review is to discuss and define successful ageing in the context of intellectual disability and to propose strategies to improve health and well-being for this population. People with an intellectual disability experience higher rates of diabetes, hypertension, obesity and cardiovascular disease, and higher rates of mental disorders than people without an intellectual disability. People with an intellectual disability engage in more passive leisure activities because many active leisure activities require the participation of or assistance by others. Health promotion programs tailored to people with an intellectual disability consisting of exercise and health education can result in more positive attitudes toward exercise and improvements in psychosocial outcomes. With modifications for people with an intellectual disability, the concept of successful ageing can be used as a template for development of strategies to improve health and well-being for people with an intellectual disability as they age. Targeted programs focused on health promotion and prevention of age-related morbidities is required. There is a need for policies addressing positive ageing, including social participation and maximizing community participation. Appropriate and ongoing education for people with an intellectual disability and their carers on healthy living in areas of physical, social, and cognitive activity, nutrition and avoidance of risk factors is essential.

Reid, A. H., & Aungle, P. G.
Dementia in ageing mental defectives: A clinical psychiatric study. Journal of *Mental Deficiency Research,* 1974, 18, 15–23. Doi: 10.1111/j.1365-2788.1974.tb01214.x
Abstract: Review of literature on dementia and Down syndrome to date [no published abstract]

Robertson, J., Hatton, C., Emerson, E., Baines, S.
Prevalence of epilepsy among people with intellectual disabilities: A systematic review.
Seizure, 2015, 29, 46-62. doi: 10.1016/j.seizure.2015.03.016.
Abstract: Epilepsy is more common in people with intellectual disabilities than in the general population. However, reported prevalence rates vary widely between studies. This systematic review aimed to provide a summary of prevalence studies and estimates of prevalence based on meta-analyses. Studies were identified via electronic searches using Medline, Cinahl and PsycINFO and cross-citations. Information extracted from studies was tabulated. Prevalence rate estimates were pooled using random effects meta-analyses and subgroup analyses were conducted. A total of 48 studies were included in the tabulation and 46 studies were included in meta-analyses. In general samples of people with intellectual disabilities, the pooled estimate from 38 studies was 22.2% (95% CI 19.6-25.1). Prevalence increased with increasing level of intellectual disability. For samples of people with Down syndrome, the pooled estimate from data in 13 studies was 12.4% (95% CI 9.1-16.7), decreasing to 10.3% (95% CI 8.4-12.6) following removal of two studies focusing on older people. *Prevalence increased with age in people with Down syndrome and was particularly prevalent in those with Alzheimer's/dementia.* Epilepsy is highly prevalent in people with intellectual disabilities. Services must be equipped with the skills and information needed to manage this condition.

Robinson, A., Spencer, B., & White, L.
Understanding difficult behaviors: Some suggestions for coping with Alzheimer's disease and related illnesses
80 pp.

Geriatric Education Center of Michigan (Alzheimer's Education Program, Eastern Michigan University, P.O. Box 981337, Ypsilanti, MI 48198-1337; www.emich.edu/public/alzheimers) (1999 rev.)
Abstract: Manual format publication providing detailed information on addressing difficult behaviors and understanding their causes and environmental relationships. Specific detailed sections on angry, agitated behavior; hallucinations and paranoia; incontinence; problems with bathing, dressing, eating, sleeping and wandering; repetitive actions, screaming and verbal noises, and wanting to go home. Appendix contains selected readings, and audio-visual materials. Does not specifically focus on intellectual disabilities, but is good generic resource.

Rodríguez-Hidalgo, E., García-Alba, J., Buxó, M., Novell, R., & Esteba-Castillo, S.
The Pictorial Screening Memory Test (P-MIS) for adults with moderate intellectual disability and Alzheimer's disease.
International Journal of Environmental Research and Public Health, 2022 Aug 30,19(17), 10780. doi: 10.3390/ijerph191710780.
Abstract: In this study, we examined normative data and diagnostic accuracy of a pictorial screening test to detect memory impairment for mild cognitive impairment (MCI) and Alzheimer's disease (AD) in Spanish-speaking adults with intellectual disability (ID). A total of 94 volunteers with ID (60 controls, 17 MCI, and 17 AD), were evaluated by neuropsychological tests including the PMIS-ID in a cross-sectional validation study. Discriminative validity between the MCI, AD, and control group was analyzed by the area under the ROC curve. A cut-off score of 4.5 on the immediate recall trial had a sensitivity of 69% and a specificity of 80% to detect memory impairment (AUC = 0.685; 95% CI = 0.506-0.863) in the AD group. The PMIS-ID is a useful screening test to rule out a diagnosis of memory decline in people with moderate level of ID and AD, and it shows good psychometric properties.

Rodríguez-Hidalgo E, García-Alba J, Novell R, Esteba-Castillo S.
The Global Deterioration Scale for Down Syndrome Population (GDS-DS): A rating scale to assess the progression of Alzheimer's disease.
International Journal of Environmental Research and Public Health, 2023 Mar 14;20(6):5096. doi: 10.3390/ijerph20065096.
Abstract: The aim of this study is to adapt and validate the global deterioration scale (GDS) for the systematic tracking of Alzheimer's disease (AD) progression in a population with Down syndrome (DS). A retrospective dual-center cohort study was conducted with 83 participants with DS (46.65 ± 5.08 years) who formed the primary diagnosis (PD) group: cognitive stability (n = 48), mild cognitive impairment (n = 24), and Alzheimer's disease (n = 11). The proposed scale for adults with DS (GDS-DS) comprises six stages, from cognitive and/or behavioral stability to advanced AD. Two neuropsychologists placed the participants of the PD group in each stage of the GDS-DS according to cognitive, behavioral and daily living skills data. Inter-rater reliability in staging with the GDS-DS was excellent (ICC = 0.86; CI: 0.80-0.93), and the agreement with the diagnosis categories of the PD group ranged from substantial to excellent with κ values of 0.82 (95% CI: 0.73-0.92) and 0.85 (95% CI: 0.72, 0.99). Performance with regard to the CAMCOG-DS total score and orientation subtest of the Barcelona test for intellectual disability showed a slight progressive decline across all the GDS-DS stages. The GDS-DS scale is a sensitive tool for staging the progression of AD in the DS population, with special relevance in daily clinical practice.

Rosewarne, M.
Learning disabilities and dementia: a pilot therapy group.
Journal of Dementia Care, 2001, 9(4), 18-19.
Abstract: None available.

Ross, W.T., & Olsen, M.
Care of the adult patient with Down syndrome
Southern Medical Journal, 2014, 107(11), 715-721. doi:10.14423/SMJ.0000000000000193.
Abstract: Individuals with Down syndrome have an increased risk for many conditions, including cardiovascular disease, cancer, infections, and osteoporosis, and endocrine, neurological, orthopedic, auditory, and ophthalmic disorders. They also are at increased risk for abuse and human rights violations and receive fewer screenings and interventions than the population without Down syndrome. In this literature review, the most common health conditions associated with Down syndrome are examined, along with the topics of sexual abuse, menstrual hygiene, contraception, and human rights. Clinical guidelines for this population are summarized in an effort to assist practicing physicians in improving their provision of health care to the adult patient with Down syndrome.

Rösner, P., Berger, J., Tarasova, D., Birkner, J., Kaiser, H., Diefenbacher, A., & Sappok, T.
Assessment of dementia in a clinical sample of persons with intellectual disability
Journal of Applied Research in Intellectual Disability, 2021, 34(6):1618-1629. doi:10.1111/jar.12913
Abstract: Assessment of age-associated disorders has become increasingly important. In a clinical setting, people with intellectual disability with and without dementia were assessed retrospectively using the Neuropsychological Test Battery (NTB) and the Dementia Questionnaire for People with Learning Disabilities (DLD) at two different times to analyse neuropsychological changes and diagnostic validity. One group (n = 44) was assessed with both instruments, while the DLD was applied in 71 patients. In the NTB (n = 44), only patients with dementia (n = 26) showed a decline in the NTB total score and three subscales. Receiver operating characteristic analysis revealed a diagnostic sensitivity of .67, a specificity of .81, and an area under the curve (AUC) of .767. In the DLD group (n = 71), only those with dementia displayed a decrease in the cognitive and social scale; diagnostic sensitivity and specificity values were low (.61/.63) and the AUC was .704. Neuropsy-chological assessment was sensitive to detect cognitive changes over time. Sensitivity values of both instruments suggest a reassessment at a later time point

Rowe, M.
Will general practitioners be adequately prepared to meet the complexities of enhanced dementia screening for people with learning disabilities and Down syndrome: key considerations.
British Journal of Learning Disabilities, 2014, 44(1), 43-48. https://doi.org/10.1111/bld.12108
Abstract: This article provides a timely response in regard to the UK's Department of Health's current initiative to financially reward general practitioners (GPs) to prioritise and undertake dementia screening for people with learning disabilities over the age of 50 years and for people with Down syndrome over the age of 40 years. Whilst GPs are becoming increasingly aware of their responsibility to care for the complex needs of people with learning disabilities, the implementation of dementia screening poses a multitude of challenges. Research has continued to suggest how difficult it is to detect the early and often ambiguous signs of dementia for someone who has pre-existing cognitive impairments and may present with atypical symptomology. And it continues to be a difficult process even for those who specialise within this area. However, GPs who choose to opt into this financially incentivised scheme will now be offering dementia screening. This article outlines the main GP aims within the dementia screening process and the difficulties that may be encountered, with specific focus upon (1) offering a dementia screen, (2) obtaining consent, (3) undertaking screening within the most appropriate setting, (4) choosing a dementia screening tool and developing a baseline, and (5) detecting early dementia signs.

Ryozo, K.
A cross-sectional study of dementia in people with intellectual disability in Japan. Asahigawa Medical Welfare Center. (2023). https://center6.umin.ac.jp/cgi-open-bin/ctr_e/ctr_view.cgi?recptno=R000043616

Abstract: A cross-sectional study of dementia in people with intellectual disability in Japan. This study aimed to investigate the prevalence of dementia among people with intellectual disability.The caregivers of the subjects answer a questionnaire to evaluate the symptoms associated with dementia. The purpose is valuating the usefulness of the dementia screening tool for people with intellectual disability. Doctors performed a medical examination and diagnose the presence or absence of dementia. Inclusion criteria included (1) Patients using the supporting facilities for people with intellectual disabilities that collaborate this research in November 1, 2019; (2) Patients with intellectual disability; and (3) Legal representatives of patients fully understand this study and give written informed consent for the participation in this study. (If the patient has ability to understand this study and to give written informed consent to participate in this study, both the patient and his/her legal representative give written informed consent.) Exclusion criteria included patients who are not appropriate for participation in this study because of their health condition as judged by the doctor.

Rubenstein, E., Hartley, S., & Bishop, L.
Epidemiology of dementia and Alzheimer disease in individuals with Down syndrome
JAMA Neurology, 2020, 77(2), 262-264. doi:10.1001/jamaneurol.2019.3666
Abstract: We describe prevalence and incidence of dementia and AD in DS in a full Medicaid population of adults with DS in Wisconsin from 2008 through 2018. We assessed Medicaid claims for adults (=21 years) who ever had 2 DS claims over their lifetime (based on International Classification of Diseases, Ninth Revision and Tenth Revision codes) on 2 separate days during Medicaid enrollment. Dementia claims were extracted from codes for any dementia (with AD as a subset) from the Centers for Medicare & Medicaid Services Chronic Conditions Data Warehouse. We required 3 or more years of Medicaid enrollment for adults with DS to ensure validity of dementia claims, therefore, beneficiaries entered the cohort at any point between 2008 and 2015. We categorized age at first and last claims (<40 years, 40-54 years, and =55 years) to account for confounding by age. A total of 2,968 individuals were included, of whom 1,507 (50.8%) were male. The median (interquartile range) age at first claim was 39 (25-48) years. In the category of individuals aged 55 years or older, 490 of 938 had dementia claims (52.2%), 307 of 938 had AD claims (32.7%), and dementia incidence was 102 (95% CI, 87-119) cases per 1000 person-years. Among individuals aged 40 to 54 years, 190 of 1013 had dementia claims (18.8%), and dementia incidence was 49 (95% CI, 44-53) cases per 1000 person-years. The probability of an incident dementia claim was 40% (95% CI, 41%-47%) over 11 years of enrollment for adults with DS who were aged 40-54 years at cohort entry and 67% (95% CI, 60%-74%) for those 55 years and older at cohort entry (Figure). There were no sex differences for dementia among individuals younger than 40 years (prevalence ratio, 1.07 [95% CI, 0.63-1.81]) or among those 55 years and older (prevalence ratio, 0.94 [95% CI, 0.69-1.29]). Dementia prevalence was higher in female individuals than male individuals aged 40 to 54 years (prevalence ratio, 1.23 [95% CI, 1.02-1.50]). Findings from a statewide health system confirm that both dementia and AD in individuals with DS present in claims data at rates similar to those ascertained from clinical samples. The hypothesized causative mechanism and similar eligibility requirements between state Medicaid programs for people with DS likely mean that other state Medicaid systems experience high incidence and prevalence of dementia and AD in individuals with DS. Dementia and AD prevalence and incidence in Medicaid beneficiaries with DS highlight the need to identify prodromal presentations and develop dementia services and supports for adults with DS as they age and continue to rely on Medicaid and Medicaid-funded assisted living or skilled nursing facilities.

Rubenstein, E., Tewolde, S., Levine, A.A., Droscha, L., Meyer, R.M., Michals, A.& Skotko, B.
Medicare, Medicaid, and dual enrollment for adults with intellectual and developmental disabilities.
Health Services Research, 2024 Jun, 59(3), e14287. doi: 10.1111/1475-6773.14287. Epub 2024 Jan 24.
Abstract: Given high rates of un- and underemployment among disabled people, adults with intellectual and developmental disabilities rely on Medicaid, Medicare, or both to pay for healthcare. Many disabled adults are Medicare eligible before the age of 65 but little is known as to why some receive Medicare services while others do not. We described the duration of Medicare enrollment for adults with intellectual and developmental disabilities in 2019 and then compared demographics by enrollment type (Medicare-only, Medicaid-only, dual-enrolled). Additionally, we examined the percent in each enrollment type by state, and differences in enrollment type for those with Down syndrome. Data sources and study setting included 2019 Medicare and Medicaid claims data for all adults (≥18 years) in the US with claim codes for intellectual disability, Down syndrome, or autism at any time between 2011 and 2019. Study design was an administrative claims cohort. Data were collected from the Transformed Medicaid Statistical Information System Analytic Files and Medicare Beneficiary Summary files. It was found that in 2019, Medicare insured 582,868 adults with identified intellectual disability, autism, or Down syndrome. Of 582,868 Medicare beneficiaries, 149,172 were Medicare only and 433,396 were dual-enrolled. Most Medicare enrollees were enrolled as child dependents (61.5%) Medicaid-only enrollees (N = 819,256) were less likely to be white non-Hispanic (58.5% white non-Hispanic vs. 72.9% white non-Hispanic in dual-enrolled), more likely to be Hispanic (19.6% Hispanic vs. 9.2% Hispanic in dual-enrolled) and were younger (mean 34.2 years vs. 50.5 years dual-enrolled). There is heterogeneity in public insurance enrollment which is associated with state and disability type. Action is needed to ensure all are insured in the program that works for their healthcare needs.

Rubenstein, E., Michals, A., Wang, N., Scott, A., Tewolde, S., Levine, A.A., Tripodis, Y., & Skotko, B.G.
Medicaid enrollment and service use among adults with Down syndrome.
JAMA Health Forum, 2023 Aug 4, 4(8), e232320. doi: 10.1001/jamahealthforum.2023.2320.
Abstract: Down syndrome is the leading genetic cause of intellectual disability and automatically qualifies individuals for Social Security Insurance. Therefore, Medicaid is the major health insurance provider for a population at high risk for dementia, obesity, and premature mortality. Despite the importance of Medicaid for adults with Down syndrome, little is known about how this population uses Medicaid. Authors described enrollment in, health care use in, and cost to Medicaid for adults with Down syndrome compared with adults with intellectual disability and a random sample of adults enrolled in Medicaid. In this cohort study, the data are from a claims cohort of adults aged 18 years or older enrolled in Medicaid at any point between January 1, 2011, and December 31, 2019. Participants were enrollees with 1 or more inpatient claim or 2 or more other claims with an International Classification of Diseases, Ninth Revision code or an International Statistical Classification of Diseases and Related Health Problems, Tenth Revision code for Down syndrome or intellectual disability as well as a random sample of those without developmental disability. Analyses were conducted from June 2022 to February 2023. Data were linked across 2 data reporting systems. Main outcomes were enrollee demographic characteristics, enrollment characteristics, cost, and service use. This cohort study included 123 024 individuals with Down syndrome (820 273 person-years of coverage; mean [SD] age, 35 [14.7] years; median age, 33 years [IQR, 21-48 years]; 51.6% men; 14.1% Black individuals; 16.7% Hispanic individuals; and 74.6% White individuals), 1 182 246 individuals with intellectual disability (mean [SD] age, 37.1 [16.8] years; median age, 33 years [IQR, 22-50 years]; 56.5% men; 22.0% Black individuals; 11.7% Hispanic individuals; and 69.5% White individuals), and 3 176 371 individuals with no developmental disabilities (mean [SD] age, 38 [18.6] years; median age, 33 years [IQR, 21-52 years]; 43.8% men; 23.7% Black individuals; 20.7% Hispanic individuals; and 61.3% White individuals). Median enrollment in Medicaid for a person with Down syndrome was 8.0 years (IQR, 5.0-9.0 years; mean [SD], 6.6 [2.6] years). Costs were higher for the Down syndrome group (median, $26 278 per person-year [IQR, $11 145-$55 928 per

person-year]) relative to the group with no developmental disabilities (median, $6173 per person-year [IQR, $868-$58 390 per person-year]). Asian, Black, Hispanic, Native American, and Pacific Islander adults with Down syndrome had fewer costs and claims per person-year compared with White adults with Down syndrome. This cohort study of individuals with Down syndrome enrolled in Medicaid found consistent enrollment and high use of health care in a population with high health care needs. Results were similar comparing individuals with Down syndrome and those with intellectual disability, with both groups differing from a sample of Medicaid enrollees with no developmental disabilities.

Rubenstein, E., Tewolde, S., Michals, A., Weuve, J., Fortea, J., Fox,.M.P., Pescador Jimenez, M., Scott, A.,Tripodis, Y., & Skotko, B.G.
Alzheimer dementia among individuals with Down syndrome.
JAMA Netw Open. 2024;7(9):e2435018.
doi:10.1001/jamanetworkopen.2024.35018
Abstract: With the advancement of administrative data as a research tool and the reliance on public health insurance for individuals with Down syndrome, population-level trends in Alzheimer dementia in this population are beginning to be understood. Authors studied/described the epidemiology of Alzheimer dementia in adults with Down syndrome in a full US Medicare and Medicaid sample by analyzing a cohort 32720 adults aged 18 years or older with Medicaid and/or Medicare claims data with an International Statistical Classification of Diseases and Related Health Problems code for Down syndrome. Data were collected from January1, 2011, to December 31, 2019, and analyzed from August 2023 to May 2024. The main outcome was prevalence of Alzheimer dementia in
each calendar year and during the 9-year period. Alzheimer dementia incidence rates by calendar year and age and stratified for race or ethnicity as well as time to death after Alzheimer dementia diagnosis were also assessed. There were 132,720 unique adults with Down syndrome from 2011 to 2019: 79,578 (53.2%) were male, 17,090 (11.7%) were non-Hispanic Black, 20,777 (15.7%) were Hispanic,101,120 (68.8%) were non-Hispanic White, and 47,692 (23.3%) had ever had an Alzheimer dementia diagnosis. Incidence was 22.4 cases per 1000 person-years. The probability of an incident Alzheimer dementia diagnosis over 8 years was 0.63 (95% CI, 0.62-0.64) for those entering the study between ages 55 to 64 years. **Mean (SD) age at incident diagnosis was 54.5 (7.4) years and median (IQR) age was 54.6 (9.3) years. Mean (SD) age at death among those with Alzheimer dementia was 59.2 (6.9) years (median [IQR], 59.0 [8.0] years)**.**The mean (SD) age at onset for the Hispanic group was 54.2 (9.2) years, 52.4 (7.8) years for the American Indian or Alaska Native group, and 52.8 (8.2) years for the mixed race groups compared with 55.0 (7.8) years for the White non-Hispanic group**. For age at death, there were no differences by sex. The mean (SD) age at death was later for the White non-Hispanic group (59.3 [6.8] years) compared with the Hispanic group (58.5 [7.8] years), Native American group (57.8 [7.1] years), and mixed race group (58.2 [7.0] years). In this cohort study of adults with Down syndrome who were enrolled in Medicaid and Medicare, Alzheimer dementia occurred at high rates. Consistency with clinical studies of dementia in Down syndrome supports the use of administrative data in Down syndrome - Alzheimer dementia research.

Rubenstein, E., Tewolde, S., Skotko, B.G., Michals, A., & Fortea, J.
Occurrence of mosaic Down syndrome and prevalence of co-occurring conditions in Medicaid enrolled adults, 2016-2019.
American Journal of Medical Genetics Part C: Seminars in Medical Genetics, 2024 Jun 25, e32097. doi: 10.1002/ajmg.c.32097. Epub ahead of print.
Abstract: : Mosaic Down syndrome is a triplication of chromosome 21 in some but not all cells. Little is known about the epidemiology of mosaic Down syndrome. We described prevalence of mosaic Down syndrome and the co-occurrence of common chronic conditions in 94,533 Medicaid enrolled adults with any Down syndrome enrolled from 2016 to 2019. We identified mosaic Down syndrome using the International Classification of Diseases and Related Health Problems, tenth edition code for mosaic Down syndrome and compared to those with nonmosaic Down syndrome codes. We identified chronic conditions using established algorithms and compared prevalence by mosaicism. In total, 1966 (2.08%) had claims for mosaic Down syndrome. Mosaicism did not differ by sex or race/ethnicity with similar age distributions. Individuals with mosaicism were more likely to present with autism (13.9% vs. 9.6%) and attention deficit hyperactivity disorder (17.7% vs. 14.0%) compared to individuals without mosaicism. In total, 22.3% of those with mosaic Down syndrome and 21.5% of those without mosaicism had claims for Alzheimer's dementia (Prevalence difference: 0.8; 95% Confidence interval: -1.0, 2.8). The mosaic group had 1.19 times the hazard of Alzheimer's dementia compared to the nonmosaic group (95% CI: 1.0, 1.3). Mosaicism may be associated with a higher susceptibility to certain neurodevelopmental and neurodegenerative conditions, including Alzheimer's dementia. Our findings challenge previous assumptions about its protective effects in Down syndrome. Further research is necessary to explore these associations in greater depth.

Rubenstein, E., Toth, M., & Tewolde, S.
Autism among adults with Down syndrome: Prevalence, medicaid usage, and co-occurring conditions.
Journal of Autism and Developmental Disorders, 2024 Jul 24. doi: 10.1007/s10803-024-06484-2. Epub ahead of print.
Abstract: Our objective was to examine occurrence of both conditions in Medicaid; and compare Medicaid service use and cost, and chronic conditions among adults with Down syndrome and autism to those with Down syndrome alone and those with autism alone. We used ICD9 and ICD10 codes in Medicaid claims and encounters from 2011 to 2019 to identify autism and Down syndrome in adults > 18 years. We then calculated costs, claims, hospitalizations, long term care days, and chronic conditions, and compared by group- autism alone, Down syndrome alone, Down syndrome + autism. Between 2011 and 2019, there were 519,450 adult Medicaid enrollees who met our criteria for autism (N = 396,426), Down syndrome (N = 116,422), or both Down syndrome and autism (N = 6,602). In 2011, 4.1% of enrollees with Down syndrome had co-occurring autism; by 2011 it was 6.6%. The autism group had the fewest claims and inpatient hospitalizations, followed by the Down syndrome group, then the Down syndrome + autism group. After age adjustment, those with Down syndrome alone and Down syndrome + autism had elevated prevalence of atrial fibrillation, **dementia**, heart failure, kidney disease, and obesity compared to the autism alone group. Both groups also had decreased occurrence of depression and hypertension compared to the autism alone group. Prevalence of autism is higher among people with Down syndrome than in peers. The increased costs and service use for those with both conditions highlight the extent to which this population need health care and signal the need for more effective preventative care and therapies.

Ryan, A., Taggart, L., Truesdale-Kennedy, M. & Slevin, E.
Issues in caregiving for older people with intellectual disabilities and their ageing family carers: a review and commentary
International Journal of Older People Nursing, 2014, Sep, 9(3), 217-226. doi: 10.1111/opn.12021.
Abstract: In keeping with worldwide demographic changes and an ageing population, people with intellectual disabilities are living longer and all the evidence suggest that this trend will continue. This 'new' population of older people and their carers will pose challenges for health and social care providers. This paper presents a review of the literature on key issues influencing caregiving for older people with intellectual disabilities and their ageing family carers. The review was undertaken using a framework adapted from the NHS Centre for Reviews and Dissemination. Papers were identified through the use of databases including CINAHL, Science Direct, PsychoInfo, Blackwell Synergy, the Cochrane Library and MEDLINE. The key themes which emerged from the literature and which consequently form the basis of this review include: ageing family carers, future planning and support services. In the context of family caregiving, older people with intellectual disabilities represent a unique group insofar as they are unlikely to be married and therefore have no spouse or dependents to care for them in later life. As a result, parents (usually mothers) have to continue caring

for their son or daughter with an intellectual disability as they both grow older, often resulting in a mutually dependent relationship. The caregiving situation is further complicated by poor emergency and future planning and by a lack of appropriate services for this group of individuals. In light of the emergence of a 'new' population of older people with intellectual disabilities, there is an urgent need to develop services and support structures which will enable these individuals and their ageing carers to 'age in place' and when this is no longer possible, to have appropriate alternatives that recognise the duality of their needs as older people and as people with intellectual disabilities.

Ryan, A., & Dodd, K.
Outcomes of use of the quality outcome measure for individuals with dementia (QOMID) with people with Down's syndrome and dementia
Advances in Mental Health and Intellectual Disabilities, 2023, 17(3), 161-172. https://doi.org/10.1108/AMHID-01-2023-0002
Abstract: Authors describe the outcomes of the use of the Quality Outcome Measure for individuals with dementia (QOMID) within a dementia pathway for people with Down's syndrome (DS). The QOMID is a tool for clinicians to use with 17 outcome domains, each with descriptors for early, mid and late stage dementia, rated on a four point scale from rarely achieved for the person to consistently and completely achieved for the person. Data from first time use of the QOMID with 49 people with DS and dementia was analysed to determine if specific outcome domains are more achievable than others, if this differs by stage of dementia and what recommendations are suggested. Results suggest that there were significant differences in achievability across the domains but that achievability did not vary between early and mid-stages of dementia. The themes with most recommendations were: quality of paperwork and documentation; improving communication with the person with DS and dementia; and working together with health and social care professionals. Results suggest that outcomes which fall outside of the typical learning disabilities skillset and are dementia specific are being achieved less often. Primarily, recommendations focused on practical solutions such as using visual timetables to communicate with the person and adapting their environment. Further work on integrating the QOMID and stage-related team training for care staff is underway.

Ryan, C., MacHale, R., & Hickey, E.
"Forgetting familiar faces": Staff perceptions of dementia in people with intellectual disabiliities
British Journal of Learning Disabilities, 2018, 46(3), 155-162. [On-line version of 29 May 2018]; https://doi.org/10.1111/bld.12233
Abstract: Living with dementia is challenging, but poses unique difficulties for adults with an intellectual disability. The demands of dementia are also challenging for family, carers, and friends. The authors explored the impact of dementia on direct care staff using a focus group methodology. Thematic analysis was used to investigate the staff narratives. There were four key themes that emerged: (a) the difficulty of recognizing symptoms of dementia in people with intellectual disability, (b) the process of diagnosis, (c) the challenge of dementia for the person, (d) the emotional impact of dementia for other people. The authors concluded that the themes identified a number of important potential targets for supporting staff and peers when dementia is present in an adult witih an intellectual disability.

Ryan, J., & Carey, E..
Developing person-centred planning in dementia care.
Learning Disability Practice. 12(5), 24-28. doi:10.7748/ldp2009.06.12.5.24.c7064
Abstract: This article provides an overview of the current literature on person-centered planning, health action planning and associated concepts such as facilitation and advocacy. It explores how flexible approaches to person-centered planning and health action planning were introduced and adapted to meet the individual daily changing needs of Anna, an older person with Down syndrome and dementia.

Ryan, K., Guerin, S., Dodd, P., & McEnvoy, J.
End-of-life care for people with intellectual disabilities: Paid carer perspectives
Journal of Applied Research in Intellectual Disability, 2011, 24(3), 199-207. https://doi.org/10.1111/j.1468-3148.2010.00605.x
Abstract: Little is known of paid carers' perspectives when caring for people with intellectual disabilities at the end-of-life. Sixty four individuals from intellectual disability services took part in 12 focus groups. Interviews were analysed using framework analysis. Participants wanted to provide palliative care and felt the experience enriched practice. However, they were inadequately prepared to meet need and this often led to staff stress. A number of issues appeared to heighten stress: situations when end-of-life care decision making was challenging, when staff felt 'pushed out' by relatives and when staff did not have sufficient support or time to provide care or mourn the loss of service users. The study describes issues which contribute to the development of staff stress when providing palliative care and draws attention to areas where strategies should be developed in order to improve the quality of care provided to people with intellectual disabilities.

Sabbagh, M., & Edgin, J.
Clinical assessment of cognitive decline in adults with Down syndrome
Current Alzheimer Research, 2015 Sept, 12.(999). doi:10.2174/1567205012666150921095724
Abstract: Down syndrome is an intellectual disability requiring periodic monitoring of cognition given the near universal presence of Alzheimer's Disease related neuropathology and high rates of dementia in middle adulthood. We review current approaches to detecting decline in this population, including informant-based measures, dementia screening tools, and neuroimaging techniques. The challenges for detecting decline in this group are discussed, including the need to take into account premorbid cognitive function as well as medical comorbidity.

Salehi, A., Ashford, J.W., & Mufson, E.J.
The link between Alzheimer's disease and Down syndrome. A historical perspective
Current Alzheimer Research, 2016, 13(1), 2–6. https://doi.org/10.2174/1567205012999151021102914
Abstract: Approximately 40-80% of persons with Down syndrome (DS) develop Alzheimer's disease (AD)-like dementia by the fifth to sixth decade of life, a much younger age than is typically seen in sporadic AD. The onset of dementia symptoms in DS parallels the development of classic brain neuropathological lesions (i.e., amyloid plaques) similar to that evident in AD. Both disorders appears to have a similar genetic linkage, which is supported by the triplication of the gene that codes for amyloid beta (A4) precursor protein (APP) in persons with DS and an extra copy of the APP gene causes familial AD in persons without DS. Despite an overlap in the genetics of these disorders, the clinical presentation of dementia differs between persons with DS and AD. Whereas `forgetfulness' is a typical symptom of early-phase dementia for persons in the general population, behavioral problems and personality changes are early signs of dementia for persons with DS. There are indications that amyloid burden begins in the frontal cortex before spreading to other brain regions in those with DS-AD, something that is not always the case in sporadic AD suggesting pathological variances between these disorders. These differences beg the question: Are the genetic and neuropathological commonalities found in DS- and AD-related dementia an associated similarity or do these disorders share a common pathogenesis? To address this query, we briefly review the clinical, histopathological, and genetic research supporting a putative link between dementia in DS and AD.

Salem, L.C., & Jørgensen, K.
Demens hos personer med Downs syndrom [Dementia in people with Down syndrome]
Ugeskrift for Læger, 2014, Jun 23, 176(26), V04120217 https://pubmed.ncbi.nlm.nih.gov/25294572/

Abstract: In developed countries the population of elderly people with Down syndrome expands resulting in an increasing incidence of age-related diseases, including dementia. The assessment of dementia in individuals with intellectual disability is often complicated due to large intra-individual variability in cognitive functioning prior to dementia and to lack of standardised measures to detect dementia. Structured observations of symptoms of dementia and assessment techniques tailored for people with intellectual disability are increasingly needed.

Santoro, S.L., Campbell, A., Balasubramanian, A., Haugen, K., Schafer, K., & Mobley, W.
Specialty clinics for adults with Down syndrome: A clinic survey
American Journal of Medical Genetics A, 2021 Jun;185(6):1767-1775. doi: 10.1002/ajmg.a.62169.
Abstract: Specialty centers improve care for patients with Down syndrome. The cohort of adults with Down syndrome is increasing, but the capacity for specialty centers to meet their medical care needs is unknown. Electronic survey of staff of specialty clinics for adults with Down syndrome was conducted. Review of online clinic list ings, and calculation of the number of adults with Down syndrome were performed. Analysis identified the percent of adults with Down syndrome who could have their medical care needs met in a current specialty clinic. Fourteen specialty clinics report providing care for 4038 adults with Down syndrome. Respondents reported gaps in care including: limitations of existing clinics, need for additional clinics, and knowl edgeable health professionals in Down syndrome. Survey-respondent clinic capacity would meet needs of 3% of adults with Down syndrome. Twenty-five clinics for adults with Down syndrome were listed online with capacity to care for 6517 adults with Down syndrome meeting the needs of 5% of the population. Additional clinic capacity is needed to meet the needs of adults with Down syndrome. Survey of existing clinics provides guidance to create additional clinics, including: must-have team members, current sources of clinic financial support, and gaps in current clinical care.

Santoro, J..D, Patel, L., Kammeyer, R., Filipink, R.A., Gombolay, G.Y., Cardinale, K.M., Real de Asua, D., Zaman, S., Santoro, S.L., Marzouk, S.M., Khoshnood, M., Vogel, B.N., Tanna, R., Pagarkar, D., Dhanani, S., Ortega, M.D.C., Partridge, R., Stanley, M.A., Sanders, J.S, Christy, A., Sannar, E.M., Brown, R., McCormick, A.A., Van Mater, H., Franklin, C., Worley, G., Quinn, E.A., Capone, G.T., Chicoine, B., Skotko, B.G., & Rafii, M.S.
Assessment and diagnosis of Down syndrome regression disorder: International expert consensus.
Frontiers of Neurology, 2022 Jul 15,13, 940175. doi: 10.3389/fneur.2022.940175.
Abstract: To develop standardization for nomenclature, diagnostic work up and diagnostic criteria for cases of neurocognitive regression in Down syndrome. There are no consensus criteria for the evaluation or diagnosis of neurocognitive regression in persons with Down syndrome. As such, previously published data on this condition is relegated to smaller case series with heterogenous data sets. Lack of standardized assessment tools has slowed research in this clinical area. The authors performed a two-round traditional Delphi method survey of an international group of clinicians with experience in treating Down syndrome to develop a standardized approach to clinical care and research in this area. Thirty-eight potential panelists who had either previously published on neurocognitive regression in Down syndrome or were involved in national or international working groups on this condition were invited to participate. In total, 27 panelists (71%) represented nine medical specialties and six different countries reached agreement on preliminary standards in this disease area. Moderators developed a proposed nomenclature, diagnostic work up and diagnostic criteria based on previously published reports of regression in persons with Down syndrome. During the first round of survey, agreement on nomenclature for the condition was reached with 78% of panelists agreeing to use the term Down Syndrome Regression Disorder (DSRD). Agreement on diagnostic work up and diagnostic criteria was not reach on the first round due to low agreement amongst panelists with regards to the need for neurodiagnostic testing. Following incorporation of panelist feedback, diagnostic criteria were agreed upon (96% agreement on neuroimaging, 100% agreement on bloodwork, 88% agreement on lumbar puncture, 100% agreement on urine studies, and 96% agreement on "other" studies) as were diagnostic criteria (96% agreement). The authors present international consensus agreement on the nomenclature, diagnostic work up, and diagnostic criteria for DSRD, providing an initial practical framework that can advance both research and clinical practices for this condition.

Santoro, J.D., Khoshnood, M.M., Nguyen, L., Vogel, B.N., Boyd, N.K., Paulsen, K.C., & Rafii, M.S.
Alternative diagnoses in the work up of Down syndrome regression disorder.
Journal of Autism and Developmental Disorders, 2023 Aug 16, doi: 10.1007/s10803-023-06057-9.
Abstract: Down Syndrome Regression Disorder (DSRD) is a diagnosis of exclusion. Psychiatric and neuroimmunologic etiologies have been proposed although the exact etiology remains unknown. This study sought to review non-DSRD diagnoses at a large quaternary medical center specializing in the diagnosis of DSRD and compare clinical characteristics between those diagnosed with DSRD and those with non-DSRD diagnoses. The authors performed a single-center retrospective, chart-based, review of referrals for developmental regression in individuals with Down syndrome. Two hundred and sixty-six individuals were evaluated for DSRD and of these, 54 (20%) ultimately had alternative diagnoses. Individuals with DSRD were more likely to have shorter nadir to clinical symptoms ($p = 0.01$, 95% CI: 0.36-0.47) and have preceding triggers ($p < 0.001$, 95% CI: 1.13-1.43) compared to those with alternative diagnoses. Individuals with non-DSRD diagnoses were more likely to be born premature ($p = 0.01$, 95% CI: 0.51-0.87) and have a history of epilepsy ($p = 0.01$, 95% CI: 0.23-0.77) but were also less likely to have a history of cytokine abnormalities on bloodwork ($p < 0.001$, 95% CI: 1.19-1.43) and have catatonia ($p < 0.001$, 95% CI: 1.54-2.17). The majority of alternative diagnoses (41/54, 76%) were autism spectrum disorder. In these cases, symptoms were more likely to be longstanding (symptoms > 12 months) and earlier onset (median 8 years, IQR: 6-11). Other diagnoses included epilepsy (5/54, 9%), Celiac disease (5/54, 9%), cerebrovascular disease (3/54, 6%). This study identifies that 20% of individuals referred with concerns for DSRD have alternative diagnoses. The majority of these diagnoses were autism, but rare treatable conditions were also identified, highlighting the importance of a thorough neurodiagnostic assessment

Santos, F.H., Watchman, K., Janicki, M.P. and the Summit on Intellectual Disability and Dementia.
Highlights from the International Summit on Intellectual Disability and Dementia Implications for Brazil
Dementia & Neuropsychologia, 2018, 12(4), 329-336. doi:10.1590/1980-57642018dn12-040001
Abstract: In October of 2016, an interdisciplinary group representing North and South American and European countries met in Glasgow, Scotland, to scrutinize universal issues regarding adults with intellectual disability (ID) affected by dementia and to produce recommendations and guidelines for public policy, practice, and further research. The aim of this paper is to apprise relevant outcomes of the Summit targeting Brazilian researchers, clinicians, and nongovernmental organizations in the field of ageing and dementia that are committed to developing the Brazilian national dementia plan. Three core themes were covered by the Summit: i) human rights and personal resources, ii) personalized services and caregiver support, and iii) advocacy and public impact. The exploration of the themes highlighted variations across countries, and revealed consensual views on matters such as international networks, guidance for practices, and advocacy on behalf of both people with ID affected by dementia, and their families. The authors outline the challenges Brazil must confront regarding ageing and dementia and proffer recommendations to address the needs of adults with ID affected by dementia within this scenario; both of which would help in developing the Brazilian national dementia plan.

Sauna-Aho, O., Bjelogrlic-Laakso, N., Siren, A., & Arvio, M.
Signs indicating dementia in Down, Williams, and Fragile X syndromes.
Molecular Genetics and Genomic Medicine, 2018, 6(5), 855-860. https://doi.org/10.1002/mgg3.430
Abstract: Intellectual disability (ID) and dementia reflect disturbed cortical function during and after developmental age, respectively. Due to the wide heterogeneity of ID population the decline in cognitive and adaptive skills may be different in distinct genetic subgroups. Using the British Present Psychiatric State-learning Disabilities assessment (PPS-LD) questionnaire the dementia signs were screened in 62, 22 and 44 individuals (> 35 year of age) with Down (DS, OMIM number 190685), Williams (WS, OMIM number, 194050), and Fragile X syndrome (FXS, OMIM number 309550), respectively. The median age of those with FXS (59 years) was higher than of those with DS (50 years) and WS (53 years). Most study participants with DS (80%) and FXS (89%) were or had been moderately or severely intellectually disabled while most participants with WS (73%) were or had been mildly or moderately disabled at adolescent age. The adolescent (premorbid) level of ID did not correlate with the dementia score. The median scores were 11/27, 1/27, and 0/27 in DS, WS, and FXS subgroups, respectively. Dementia that was confirmed by brain imaging, manifested as Alzheimer disease and as moya-moya disease associated vascular dementia in DS and as vascular dementia in WS. This survey suggests that the risk of dementia varies depending on the cause of ID and that the severity of ID in adolescence does not predict the development of dementia at a later age. Consequently, the ID and dementia should be understood as separate clinical entities that need to be taken into account in the health management of intellectually disabled people. This is important for the arrangement of appropriate and timely interventions, which can be expected to delay the need for institutionalization.

Scattoni ML, Micai M, Ciaramella A, Salvitti T, Fulceri F, Fatta LM, Poustka L, Diehm R, Iskrov G, Stefanov R, Guillon Q, Rogé B, Staines A, Sweeney MR, Boilson, A.M., Leósdóttir, T., Saemundsen, E., Moilanen, I., Ebeling, H., Yliherva, A., Gissler, M., Parviainen, T., Tani, P., Kawa, R., Vicente, A., Rasga, C., Budisteanu, M., Dale, I., Povey, C., Flores, N., Jenaro, C., Monroy, M.L., Primo, P.G., Charman, T., Cramer, S., Warberg, C.K., Canal-Bedia, R., Posada, M., & Schendel, D.
Real-world experiences in autistic adult diagnostic services and post-diagnostic support and alignment with services guidelines: Results from the ASDEU study.
Journal of Autism and Developmental Disorders, 2021 Nov, 51(11), 4129-4146. doi: 10.1007/s10803-021-04873-5.
Abstract: Research providing an evidence-base for autistic adult services is sparse. The Autism Spectrum Disorders in the European Union (ASDEU) network implemented an on-line survey to determine gaps in autistic adult diagnostic evaluation and post-diagnostic support services. More than 55% in all groups experienced most of the recommended features for diagnostic evaluation for autistic adults. In contrast, < 2% of adults or carers, and < 21% of professionals experienced each of the recommended features for post-diagnostic support. In contrast to 61% of professionals, only about 30% of autistic adults and carers had knowledge of good local services models for autism diagnosis in adulthood. There are major differences between good practice guidelines for diagnostic and post-diagnostic care for autistic adults, and what is actually experienced by services users and professionals.

Schaap, F.D., Dijkstra, G.J., Finnema, E.J., & Reijneveld, S..A.
The first use of dementia care mapping in the care for older people with intellectual disability: a process analysis according to the RE-AIM framework
Aging & Mental Health, 22(7), 912-919. DOI: 10.1080/13607863.2017.1401582
Abstract: The aging of the population with intellectual disability (ID), with associated conseqences as dementia, creates a need for evidence-based methods to support staff. Dementia Care Mapping (DCM) is perceived to be valuable in dementia care and promising in ID-care. The aim of this study was to evaluate the process of the first use of DCM in ID-care. DCM was used among older people with ID and care-staff in 12 group homes of six organisations. We obtained data on the first use of DCM in ID-care via focus-group discussions and face-to-face interviews with: care-staff (N = 24), managers (N = 10), behavioural specialists (N = 7), DCM-ID mappers (N = 12), and DCM-trainers (N = 2). We used the RE-AIM framework for a thematic process-analysis. All available staff (94%) participated in DCM (reach). Regarding its efficacy, staff considered DCM valuable; it provided them new knowledge and skills. Participants intended to adopt DCM, by continuing and expanding its use in their organisations. DCM was implemented as intended, and strictly monitored and supported by DCM-trainers. As for maintenance, DCM was further tailored to ID-care and a version for individual ID-care settings was developed, both as standards for international use. To sustain the use of DCM in ID-care, a multidisciplinary, interorganisational learning network was established. DCM tailored to ID-care proved to be an appropriate and valuable method to support staff in their work with aging clients, and it allows for further implementation. This is a first step to obtain an evidence-based method in ID-care for older clients.

Schaap, F.D., Dijkstra, G.J., Stewart, R.E., Finnema, E.J., & Reijneveld,J S..A.
Effects of Dementia Care Mapping on well-being and quality of life of older people with intellectual disability: A quasi-experimental study
Journal of Applied Research in Intellectual Disabilities, 2019, July, 32(4), 849-860. https://doi.org/10.1111/jar.12576
Abstract: The ageing of people with intellectual disability, accompanied with consequences like dementia, challenges intellectual disability-care staff and creates a need for supporting methods, with Dementia Care Mapping (DCM) as a promising possibility. This study examined the effect of DCM on the quality of life of older people with intellectual disability. We performed a quasi-experimental study in 23 group homes for older people with intellectual disability in the Netherlands, comparing DCM (n = 113) with care-as-usual (CAU; n = 111). Using three measures, we assessed the staff-reported quality of life of older people with intellectual disability. DCM achieved no significantly better or worse quality of life than CAU. Effect sizes varied from 0.01 to -0.22. Adjustments for covariates and restriction of analyses to people with dementia yielded similar results. The finding that DCM does not increase quality of life of older people with intellectual disability contradicts previous findings and deserves further study.

Schaap, F.D., Finnema, E.J., Dijkstra, G.J., & Reijneveld, M.
What can we learn from dementia care in the care of older people with intellectual disability?
Journal of Intellectual Disability Research, 2019, 63(8), 645-646.
Abstract: The ageing of people with intellectual disability (ID) increases rates of dementia, starting earlier and are more prevalent than in the general population. ID-care staff call for methods, knowledge, and skills to support their older residents. Person-centred methods derived from dementia care can fill this gap, but are often used unsystematically, and not adapted to ID-care. Moreover, their effectiveness in ID-care is not yet clear. One person-centred method adapted to ID-care, is Dementia Care Mapping (DCM). The aim of this study is to examine the experiences of care staff with DCM. We assessed this after two applications of DCM in twelve group homes for older people with ID, with a qualitative study (N = 24) and a quantitative study on care-staff (N = 136). Our study showed that DCM provided better understanding of the behaviour of their residents with and without dementia, more reflection and awareness of their own professional behaviour, and new knowledge and (dementia-care) skills. Furthermore, relating the needs and interpretation of the behaviour of residents to the theory of person-centred care provided care-staff a rationale and significance in daily care. Finally, DCM led to more team coordination of care. Authors concluded that evidence from dementia care can improve the quality of care for older people with ID, if adequately embedded in ID-care.

Schäper, S. & Graumann, S.
Alter(n) als wertvolle Lebensphase erleben [Aging and quality of life: challenges and opportunities for people with intellectual disabilities]

Zeitschrift für Gerontologie und Geriatrie, 2021, Oct, 45(7), 630-636. doi:10.1007/s00391-012-0388-1.
Abstract: In the coming years, a growing number of people with an intellectual disability will reach retirement age. In line with the change of paradigms, the leading ideas of participation, inclusion and self-determination have become the principles of the ideological and conceptual framework in social services for people with disabilities. However, in many places convincing concepts and arrangements of support for elderly people with intellectual disabilities are lacking, particularly beyond institutionalized concepts. The research project "Lebensqualität inklusiv(e)" (quality of life included) tries to bridge this gap. On the base of an estimation of the demographic development for this group of people, models of best practice have been documented and evaluated focusing on living conditions and the special requirements for elderly people with intellectual disabilities in order to gather ideas for the development of arrangements of support. The results show that an interdisciplinary cooperation is indispensable

Schlamb, C.D., & Moriconi, C.D.
Betsy: A case study of a client with Down's syndrome and dementia
Advancing Care Excellence for Seniors. 8pp. (2014).
https://digitalcommons.wcupa.edu/cgi/viewcontent.cgi?article=1000&context=nurs_facpub
Abstract: This case study is about an aging woman experiencing Down syndrome (DS) and dementia. People with Down syndrome are living longer than ever before. Since the 1980s their life expectancy has doubled and many now live into their 60s, most likely because of advances in medical treatment and improved living conditions. Adults with DS and dementia typically experience several residential relocations during their lifetime and these may be traumatic events for these individuals. This study explores the complex needs of aging clients with intellectual disabilities. Target students for this teaching strategy have completed medical-surgical or geriatric nursing.

Schupf, N., Kapell, D., Lee, J.H., Ottman, R. & Mayeux, R.
Increased risk of Alzheimer's disease in mothers of adults with Down's syndrome
Lancet,1994, Aug 6, 344(8919), 353-356. doi: 10.1016/s0140-6736(94)91398-6.
Abstract: Most adults with Down's syndrome (DS) develop neuropathology characteristic of Alzheimer's disease (AD) by the age of 40. Most of the non-dysjunction events in DS are of maternal origin. We postulated therefore that a shared genetic susceptibility to DS and AD would be associated with an increased frequency of AD among mothers, but not fathers, of individuals with DS. We further hypothesised that the shared susceptibility could involve an accelerated ageing process, leading to the birth of a child with DS to a relatively young mother and to an increased risk of dementia in the mother and her relatives. Families of 96 adults with DS and of 80 adults with other forms of mental retardation were ascertained through the New York State Developmental Disabilities services network. A semi-structured interview was used to obtain information on the presence or absence of non-stroke-related dementia and other disorders in parents. There was an increase in risk of dementia among mothers of DS probands compared with control mothers (risk ratio 2.6 [95% CI 0.9-7.3]). The risk of dementia among mothers who were 35 or younger when their DS children were born was 5 times that of control mothers (4.9 [1.6-15.4]). There was no increase in risk of dementia among mothers who were older (> 35 years) at the proband's birth (0.8 [0.2-3.4]). There was no difference in risk of dementia between fathers of DS cases and fathers of controls (1.2 [0.4-3.9]) and no discernible influence of age on this risk. Familial aggregation of dementia among mothers of adults with DS supports the hypothesis of a shared genetic susceptibility to DS and AD.

Schupf, N., Kapell, D., Nightingale, B, Rodriguez, A., Tycko, B., & Mayeux, R.
Earlier onset of Alzheimer's disease in men with Down syndrome.
Neurology, 1998, 50(4), 991-995. https://doi.org/10.1212/WNL.50.4.991
Abstract: Virtually all individuals with Down syndrome (DS) have neuropathologic changes characteristic of Alzheimer's disease (AD) beginning at 40 years of age. Few studies have examined factors that influence age at onset of AD in DS. We investigated whether sex differences in age at onset and risk of AD among adults with DS are similar to those observed in the general population and whether the effect of sex on risk of AD is modified by apolipoprotein E(*APOE*) genotype. A community-based sample of 111 adults with cytogenetically confirmed DS (34 to 71 years of age) was ascertained through the New York State Developmental Disabilities system. A semi-structured interview with caregivers and review of medical records was used to ascertain the presence or absence of AD. *APOE* genotyping was carried out without knowledge of the subject's medical history or clinical diagnosis. Both male gender and the presence of an *APOE* ?4 allele were associated with an earlier onset of AD. Compared with women, men with DS were three times as likely to develop AD. Compared with those with the *APOE* 3/3 genotype, adults with DS with the 3/4 or 4/4 genotypes were four times as likely to develop AD. No individual with an *APOE* ?2 allele developed AD. No evidence of interaction of sex and *APOE* genotype was found in risk of AD. The higher risk of AD in men may be related to differences in hormonal function between men and women with DS that are distinct from those in the general population.

Schupf, N., Winsten, S., Patel, B., Pang, D., Ferin, M., Zigman, W.B., Silverman, W., & Mayeux R.
Bioavailable estradiol and age at onset of Alzheimer's disease in postmenopausal women with Down syndrome.
Neuroscience Letter, 2006, Oct 9, 406(3), 298-302.
Abstract: Several lines of evidence suggest that loss of estrogen after menopause may play a role in the cognitive declines associated with Alzheimer's disease (AD). Women with Down syndrome (DS) experience early onset of both menopause and AD. This timing provides a model to examine the influence of endogenous estrogen deficiency on risk of AD. We hypothesized that low serum levels of bioavailable estradiol (E2) would be associated with increased risk of AD. One hundred and nineteen postmenopausal women with DS, 42-59 years of age, were ascertained through the New York State developmental disability service system and followed at 18-month intervals. Information from cognitive assessments, caregiver interviews, medical record review and neurological examination was used to establish the diagnosis of dementia. Women with DS who developed AD had lower levels of bioavailable E2, lower levels of total estradiol, higher levels of sex-hormone binding globulin, and lower levels of dehydroepiandrosterone sulfate at baseline than women who remained dementia free over the course of follow-up. Women who had low levels of bioavailable E2 at baseline were four times as likely to develop AD (HR=4.1, 95% CI: 1.2-13.9) and developed AD, on average, 3 years earlier, than those with high levels of bioavailable E2, after adjustment for age, level of mental retardation, ethnicity, body mass index, history of hypothyroidism or depression and the presence of the apolipoprotein varepsilon4 allele. Our findings support the hypothesis that reductions in estrogen following menopause can contribute to the cascade of pathological processes leading to AD.

Schupf, N., Pang, D., Patel, B.N., Silverman, W., Schubert, R., Lai, F., Kline, J.K., Stern, Y., Ferin, M., Tycko, B., & Mayeux, R.
Onset of dementia is associated with age at menopause in women with Down's syndrome.
Annuals of Neurology, 2003, 54(4), 433-438. https://doi.org/10.1002/ana.10677
Abstract: Women with Down's syndrome experience early onset of both menopause and Alzheimer's disease. This timing provides an opportunity to examine the influence of endogenous estrogen deficiency, indicated by age at menopause, on risk of Alzheimer's disease. A community-based sample of 163 postmenopausal women with Down's syndrome, 40 to 60 years of age, was ascertained through the New York State Developmental Disability service system. Information from cognitive assessments, medical record review, neurological evaluation, and caregiver interviews was used to establish ages for

onset of menopause and dementia. We used survival and multivariate regression analyses to determine the relation of age at menopause to age at onset of Alzheimer's disease, adjusting for age, level of mental retardation, body mass index, and history of hypothyroidism or depression. Women with early onset of menopause (46 years or younger) had earlier onset and increased risk of Alzheimer's disease (AD) compared with women with onset of menopause after 46 years (rate ratio, 2.7; 95% confidence interval [CI], 1.2-5.9). Demented women had higher mean serum sex hormone binding globulin levels than nondemented women (86.4 vs 56.6 nmol/L, p = 0.02), but similar levels of total estradiol, suggesting that bioavailable estradiol, rather than total estradiol, is associated with dementia. Our findings support the hypothesis that reductions in estrogens after menopause contribute to the cascade of pathological processes leading to AD.

Schweber, M.S.
Alzheimer's disease and Down syndrome
Progress in Clinical and Biological Research, 1989, 317, 247-267.
https://pubmed.ncbi.nlm.nih.gov/2532369/
Abstract: This report contains a summary of an extensive survey of autopsy data for persons with intellectual disability. Among adults with Down syndrome (DS), the brain neuropathology of AD was universal in those age 37 and over; claimed exceptions were indefensible. The behavioral evaluations of the DS adults, however, could be classified into three divisions: 1. "quiescent" (neither seizures nor dementia, 2). "partial" (seizures but no dementia), and 3). "active" (dementia +/- seizures). Thus, it is reasonable to argue that all persons with DS develop AD itself upon aging. However, DS cannot be used uncritically as an AD model since no increased incidence of active AD was found in DS with aging beyond the critical threshold age (mid-30's). Improved accurate quantification of Southern blots produced 100% accuracy in decoding blind samples of DS and non-DS samples. Using this system, DNA levels similar to those of DS have been demonstrated for all categories of AD at a small subsection of chromosome 21 near to, or within the DS DNA location on chromosome 21. Increased amounts of a complete, structural gene sequence were not found (or expected). The results provide evidence for a unitary hypothesis for DS and all forms of AD.

Schworer, E., Zammit, M.D., Wang, J., Handen, B.L., Betthauser, T., Laymon, C.M.,Tudorascu, D.L. ... Zhang, F.
Timeline to symptomatic Alzheimer's disease in people with Down syndrome as assessed by amyloid-PET and tau-PET: a longitudinal cohort study
The Lancet Neurology, 2024, 23(12),1214 - 1224. doi: 10.1016/S1474-4422(24)00426-5
Abstract: Adults with Down syndrome are at risk for Alzheimer's disease. Natural history cohort studies have characterized the progression of Alzheimer's disease biomarkers in people with Down syndrome, with a focus on amyloid ß-PET and tau-PET. In this study, we aimed to leverage these well characterized imaging biomarkers in a large cohort of individuals with Down syndrome, to examine the timeline to symptomatic Alzheimer's disease based on estimated years since the detection on PET of amyloid ß-positivity, referred to here as amyloid age, and in relation to tau burden as assessed by PET. In this prospective, longitudinal, observational cohort study, data were collected at four university research sites in the UK and USA as part of the Alzheimer's Biomarker Consortium-Down Syndrome (ABC-DS) study. Eligible participants were aged 25 years or older with Down syndrome, had a mental age of at least 3 years (based on a standardised intelligence quotient test), and had trisomy 21 (full, mosaic, or translocation) confirmed through karyotyping. Participants were assessed twice between 2017 and 2022, with approximately 32 months between visits. Participants had amyloid-PET and tau-PET scans, and underwent cognitive assessment with the modified Cued Recall Test (mCRT) and the Down Syndrome Mental Status Examination (DSMSE) to assess cognitive functioning. Study partners completed the National Task Group-Early Detection Screen for Dementia (NTG-EDSD). Generalised linear models were used to assess the association between amyloid age (whereby 0 years equated to 18 centiloids) and mCRT, DSMSE, NTG-EDSD, and tau PET at baseline and the 32-month follow-up. Broken stick regression was used to identify the amyloid age that corresponded to decreases in cognitive performance and increases in tau PET after the onset of amyloid ß positivity. Some 167 adults with Down syndrome, of whom 92 had longitudinal data, were included in our analyses. Generalised linear regressions showed significant quadratic associations between amyloid age and cognitive performance and cubic associations between amyloid age and tau, both at baseline and at the 32-month follow-up. Using broken stick regression models, differences in mCRT total scores were detected beginning 2·7 years (95% credible interval [CrI] 0·2 to 5·4; equating to 29·8 centiloids) after the onset of amyloid ß positivity in cross-sectional models. Based on cross-sectional data, increases in tau deposition started a mean of 2·7-6·1 years (equating to 29·8-47·9 centiloids) after the onset of amyloid ß positivity. Mild cognitive impairment was observed at a mean amyloid age of 7·4 years (SD 6·6; equating to 56·8 centiloids) and dementia was observed at a mean amyloid age of 12·7 years (5·6; equating to 97·4 centiloids). There is a short timeline to initial cognitive decline and dementia from onset of amyloid ß positivity and tau deposition in people with Down syndrome. This newly established timeline based on amyloid age (or equivalent centiloid values) is important for clinical practice and informing the design of Alzheimer's disease clinical trials, and it avoids the limitations of timelines based on chronological age.

Scottish Down's Syndrome Association
What is dementia? - A booklet about dementia for adults who have a learning disability.
14pp
Edinburgh: Scottish Down's Syndrome Association [158-160 Balgreen Road, Edinburgh, Scotland EH11 3AU; e/m: info@sdsa.org.uk; www.sdsa.org.uk] [n.d.] [Source: http://www.rrtcadd.org/TA/Dementia_Care/Resources/Info.html]
Abstract: Written for the Scottish Down's Syndrome Association by Diana Kerr and Mo Innes this A4 size booklet is designed to explain dementia and its nuances to persons with intellectual disabilities (termed "learning disabilities in Scotland). Using drawings and easy language this booklet covers many of the symptoms and behaviors classically associated with Alzheimer's disease.

Seishi, T.
A cross-sectional study of dementia in people with intellectual disability.
Okayama University Graduate School of Medicine, Dentistry, and Pharmaceutical Sciences. (2023).
International Clinical Trials Registry Platform ICTRP.
https://trialsearch.who.int/Trial2.aspx?TrialID=JPRN-UMIN000028708
Abstract: Unavailable - Research study involving doctors who perform a medical examination and diagnose the presence or absence of dementia to derive data on prevalence and characteristics of cohort of adults in Japan with intellectual disability. Data were derived on demographics (age, sex, diagnosis); diagnosis of dementia; type of dementia; and dementia symptoms (Memory, Executive Function, Behavioral and Psychological Symptoms of Dementia, Activity of Daily Living, Adaptability)

Sekijima,Y., Ikeda, S., Tokuda, T., Satoh, S., Hidaka, H., Hidaka, E., Ishikawa, M., & Yanagisawa, N.
Prevalence of dementia of Alzheimer type and apolipoprotein E phenotypes in aged patients with Down's syndrome.
European Neurology, 1998, 39(4), 234-7. doi: 10.1159/000007940.
Abstract: Authors investigated the exact prevalence of dementia of Alzheimer type (DAT) and apolipoprotein E (ApoE) phenotypes in 106 Japanese Down's syndrome (DS) patients. Among these patients 16 were diagnosed as having DAT. The prevalence of DAT was 0% in the 30- to 39-year-old group, 16% in the 40- to 49-year-old group, and 38% in those over 50 years old. The frequency of the epsilon4 allele in DS patients with DAT was 18.8%, which was considerably higher than that of nondemented DS patients (4.5%) and Japanese nondemented controls (6.7%). Especially, the frequency of the epsilon4 allele in DS patients who developed DAT under 50 years was significantly higher (28.6%). DS patients

certainly develop DAT at earlier ages but the prevalence of DAT in each group of patients was lower than previously recognized. It is very likely that the ApoE epsilon4 is a risk factor for DAT even in DS patients with a genetic predisposition to Alzheimer's disease.

Seritan, A.L., Nguyen, D.V., Farias, S.T., Hinton, L., Grigsby, J., Bourgeois, J.A., & Hagerman, R.J.
Dementia in fragile X-associated tremor/ataxia syndrome (FXTAS): comparison with Alzheimer's disease.
American Journal of Medical Genetics: B Neuropsychiatric Genetics, 2008 Oct 5, 147B(7), 1138-44. doi: 10.1002/ajmg.b.30732.
Abstract: Neurocognitive deficits in fragile X-associated tremor/ataxia syndrome (FXTAS) involve attentional control, working memory, executive functioning, and declarative and procedural learning. To date, no studies comparing FXTAS with other dementias have been done. We characterize the dementia in FXTAS, comparing it with Alzheimer's disease. Retrospective chart review of 68 adults (50 men, 18 women) with FXTAS. 20 men with FXTAS dementia were matched by age, gender, and education to patients with mild Alzheimer's dementia (AD). Neuropsychological measures were compared between the two groups: Boston Naming Test (BNT), phonemic fluency (Controlled Oral Word Association Test), digit span forward (DSF) and backward (DSB). Comparisons were based on analysis of covariance and t-tests to assess significant differences between groups with respect to the neuropsychological measures. Some 50% of men with FXTAS and no women were cognitively impaired. On mean scores of verbal fluency (22.83 in FXTAS vs. 28.83 in AD, p = 0.112), working memory (DSB, 4.80 in AD vs. 5.41 in FXTAS, p = 0.359), and language (BNT, 48.54 in AD vs. 54.20 in FXTAS, p = 0.089), there were no significant differences. Digit span forward, measuring attention, was significantly higher in subjects with FXTAS dementia (8.59, vs. 7.10 in AD, p = 0.010). Individuals with FXTAS have significant cognitive deficits, on the order of those in AD although the cognitive profiles in these **dementias** are not similar. Further research is needed to outline the neuropsychiatric profile in FXTAS and the correlation of genetic markers with the progression and severity of cognitive loss.

Service, K.P.
Considerations in care for individuals with intellectual disability with advanced dementia
Journal of Gerontological Social Work, 2002, 38, 213-224.
https://doi.org/10.1300/J083v38n01_06
Abstract: A number of physical, psychosocial, or ethical issues related to the care of the individual with advanced dementia are reviewed and related to individuals with intellectual disabilities. The sources used include the published literature and illustrations drawn from personal observations. The author notes that through anticipation and early planning, advanced directives and service planning (which looks to adaptation of services and other care management interventions), can effectively impact care at the end. Areas that need to be addressed include technical information, including a review of and, as appropriate, adaptation of general advanced dementia resources, relief, rest, support, reassurance, receipt of on-going information, participation in planning, a sense of humor, and appreciation. Also noted, are the differences experienced because of the presence of paid staff as carers and residence outside of the family home. It is concluded that, although the goals of quality care is the same for all people with advanced dementia, the process by which to reach these goals often needs further consideration and adaptation for people with intellectual disabilities.

Service, K..P., & Clifford, C.J.
What do I really need? Assessment of caregiver supports for people with intellectual and developmental disability and dementia.
AAIC 2020 Conference (Amsterdam, NL - virtual), Poster presentation, July 30. 2020. Alzheimer's & Dementia, 16(S7), First published: 07 December 2020.
https://doi.org/10.1002/alz.047106
Abstract: The increased needs of people with an intellectual and developmental disability (IDD) and a dementia related disorder can strain caregivers and existing community support systems. The project team, comprised of IDD and Aging experts and funded, in part, through a federal grant, conducted a needs assessment on the awareness and use of typical community-based resources such as senior centers. The assessment consisted of both telephone interviews and home visits with group home, shared living, and family caregivers. Nurse Practitioner (NP) conducted a total of 95 interviews with 54 site visits, and the evaluator completed 40 interviews with caregivers of people with IDD and dementia diagnosis. Analysis included both qualitative and quantitative data. Caregivers were asked about the following: functional and health status of person with IDD since dementia diagnosis, receipt of dementia specific caregiving training, care confidence levels, perceived barriers to care, and access to community-based aging resources. Caregiver's most frequent concerns included lack of suitable day programming, planning for the person's future, and caregiver burnout and stress. 78% reported feeling confident providing care currently and 68% were confident about providing care in the future. Most caregivers are aware of local community resources such as senior centers, Alzheimer's Association Counseling and an on-line training series on aging, but rarely used the resources. Authors note that caregivers generally relied on support from provider agencies indicating a need for increased collaboration across the IDD and Aging human service support systems. Trainings, delivered to both aging advocates and caregivers of people with IDD, and designed to improve communication and collaboration focused on dementia capable care, state systems, and available community resources. In addition, a series of web-based resources were developed with a focus on IDD and dementia. Results of the assessment will continue to guide resource and training development to improve collaboration and support the relationship between the Aging and IDD communities.

📖 **Service, K.P., Lavoie, D. Herlihy, J.E.**
Coping with losses, death and grieving
In M.P. Janicki & A.J. Dalton (eds.), Dementia, Aging, and Intellectual Disabilities. pp. 330-351
Philadelphia: Brunner-Mazel (1999)
Abstract: This book chapter uses a composite case to demonstrate strategies to address the issues related to losses and death for people with intellectual disability and the diagnosis of dementia and for their families and staff. Dealing with the diagnosis and the changes are explained in the framework of the stages of death and dying as developed by Kubler-Ross. The responses to the losses of dementia which are manifested by affected individuals and members of their personal networks are reflective of a number of factors. The dilemma related to personal value systems, professional roles, and philosophies of care is explored in the context of ethical concerns. The impact of program considerations such as rules, regulations, policies, and economics is examined. Bereavement work for peers and housemates can be further developed for carers, family, and staff. Recommendations for research and interventions for public policy are given.

Service, K., Watchman, K., Hogan, M., Janicki, M.P., Cadovius, N., & Beránková, A.
Dying well with an intellectual disability and dementia
Journal of Dementia Care, 2017, 25(4), 25-31.
https://www.researchgate.net/publication/318723617_Dying_well_with_an_intellectual_disability_and_dementia
Abstract: An international summit on intellectual disability and dementia identified three areas where the added complexity of advanced dementia warrants particular attention around end-of-life services in people with an intellectual disability. The three areas were: (a) ascertainment of advanced stage of dementia, (b) place of care, and (c) active support. The authors discuss each of these three issues and note the particular challenges that arise when someone with dementia also has an intellectual disability. The summit proffered a series of recommendations that included ongoing exchange of experiences and skills across professions, development of tools and scales that facilitate understanding

of the progression of dementia, and more equitable access to palliative care and hospice services with increased and timely referral

Sheehan, R., Ali, A., & Hassiotis, A.
Dementia in intellectual disability
Current Opinion in Psychiatry, 2014, March, 27(2), 143-148.
doi:10.1097/YCO.0000000000000032
Abstract: Dementia is emerging as a significant condition in the population with intellectual disability. This review is aimed at clinicians working in the field. We revisit what is known on the subject and expand on this with results from recent research. The emphasis of this review is on the clinical research rather than laboratory or molecular research. Research has encompassed all aspects of dementia in intellectual disability, from epidemiology, assessment and diagnosis, through to management. There remains a lack of evidence concerning both pharmacological and nonpharmacological treatment of dementia in people with intellectual disability. Recent research has tended to focus on dementia in Down syndrome. More research is necessary in order to translate improvements in the understanding of the neuropathology of intellectual disability and dementia into effective treatments. There is also a need to investigate the optimum environment in which to provide holistic care for individuals affected.

Sheehan, R., Sinai, A., Bass, N., Blatchford, P., Bohnen, I,, Bonell, S., Courtenay, K., Hassiotis, A., Markar, T., McCarthy, J., Mukherji, K., Naeem, A,. Paschos, D., Perez-Achiaga, N., Sharma, V., Thomas, D., Walker, Z., Strydom, A.
Dementia diagnostic criteria in Down syndrome.
International Journal of Geriatric Psychiatry, 2015, 30(8), 857-863. doi: 10.1002/gps.4228.
Abstract: Dementia is a common clinical presentation among older adults with Down syndrome. The presentation of dementia in Down syndrome differs compared with typical Alzheimer's disease. The performance of manualised dementia criteria in the International Classification of Diseases (ICD)-10 and Diagnostic and Statistical Manual of Mental Disorders-IV-Text Revision (DSM-IV-TR) is uncertain in this population. We aimed to determine the concurrent validity and reliability of clinicians' diagnoses of dementia against ICD-10 and DSM-IV-TR diagnoses. Validity of clinical diagnoses were also explored by establishing the stability of **diagnoses** over time. The authors used clinical data from memory assessments of 85 people with Down syndrome, of whom 64 (75.3%) had a diagnosis of dementia. The cases of dementia were presented to expert raters who rated the case as dementia or no dementia using ICD-10 and DSM-IV-TR criteria and their own clinical judgement. The authors found that clinician's judgement corresponded best with clinically diagnosed cases of dementia, identifying 84.4% cases of clinically diagnosed dementia at the time of diagnosis. ICD-10 criteria identified 70.3% cases, and DSM-IV-TR criteria identified 56.3% cases at the time of clinically diagnosed dementia. Over time, the proportion of cases meeting ICD-10 or DSM-IV-TR diagnoses increased, suggesting that experienced clinicians used their clinical knowledge of dementia presentation in Down syndrome to diagnose the disorder at an earlier stage than would have been possible had they relied on the classic description contained in the diagnostic systems. The authors concluded that clinical diagnosis of dementia in Down syndrome is valid and reliable and can be used as the standard against which new criteria such as the DSM-5 are measured.

Sheth, A.J.
Intellectual disability and dementia: perspectives on environmental influences.
Quality in Ageing and Older Adults, 2019, 20(4), 179-189.
https://doi.org/10.1108/QAOA-11-2018-0060.
Abstract: The purpose of this paper was to improve understanding of environmental influences on participation in routine and familiar activities for people with intellectual disability and dementia from first-person and caregiver perspectives. The methodology involved four adults with intellectual disability and dementia participating in 2 nominal group technique sessions and 12 family and staff caregivers participating in 5 standard focus groups. Transcripts were analyzed utilizing thematic analysis centering the findings from nominal group technique sessions and an ecological systems lens. The findings revealed that participants with intellectual disability and dementia identified six important themes: activity access, caregiver assistance, social interactions, responsibilities, privacy, and health and wellness. Their perspectives focused primarily at an immediate environment level, while caregiver input added additional understandings from broader ecological systems levels. This study provides a beginning point to establishing a framework for creating supports and addressing barriers to participation for adults with intellectual disability and dementia based on direct input from potential service consumers and their caregivers. People with intellectual disabilities and dementia provide valuable insights into their experiences through engagement in accessible research.

Sheth, A.J., Kramer, J.M., Magasi, S., Heller, T., Nishida, A., & Hammel, J.
"It's not the same without you:" Exploring the experience and perception of transition for people with intellectual disabilities and dementia
British Journal of Learning Disabilities, 2021 (Sep), 49(3), 365-372.
https://doi.org/10.1111/bld.12412
Abstract: For people with intellectual disabilities and dementia, transitions are likely to become increasingly common as they age. While transitions experienced by people with intellectual disabilities in young adulthood are frequently studied, less is known about transitions in older adults, including residential, vocational and leisure changes. This article aims to explore the experiences of transition from the perspectives of people with intellectual disabilities and dementia, including the impact on their daily lives. Three women with intellectual disabilities and dementia living in residential settings participated in participant observations and informal interviews across a variety of environments and activities. Field notes and interview transcripts underwent a thematic analysis focusing on transitions. Participants experienced the impact of transitions in their residential placements, day programming, leisure activities and relationships. Themes related to their experiences of transitions included making sense of transitions, utilising peer care networks for support and tackling the looming threat of loss and transition. Peer care networks and friendships are crucial in supporting people during and after transitions. Receiving effective supports to maintain relationships, roles and activities, even in seemingly minor ways, is an important right for people with intellectual disabilities and dementia, particularly as care needs increase.

Shimizu, E., Goto-Hirano, K., Motoi, Y., Aria, M., Hattori, N.
Symptoms and age of prodromal Alzheimer's disease in Down syndrome: a systematic review and meta-analysis.
Neurological Sciences, 2024 Jan 16. doi: 10.1007/s10072-023-07292-9.
Abstract: The diagnostic criteria for adult-onset Alzheimer's disease (AD) in patients with Down syndrome (DS) have not been standardized. This study investigated the specific symptoms of AD in the prodromal stage of DS, the mean age at diagnosis at each stage of dementia, and the relationship between intellectual disability (ID) and dementia. PubMed, Web of Science, and Embase were searched for studies on DS, AD, early-stage disease, initial symptoms, and prodromal dementia registered between January 2012 and January 2022. We also performed a meta-analysis of the differences between the mean age at prodromal symptoms and AD diagnosis and the proportion of mild cognitive impairment in patients with mild and moderately abnormal ID. We selected 14 articles reporting the behavioral and psychological symptoms of dementia (BPSD) and memory- and language-related impairments as early symptoms of AD in patients with DS. **The specific symptoms of BPSD were classified into five categories: irritability (agitation), apathy, abnormal behavior, adaptive functioning, and sleep disturbance**. **The mean age at the diagnosis of prodromal symptoms and AD dementia was 52.7 and 56.2 years**, respectively (mean difference, + 3.11 years; 95% CI 1.82–4.40) in the meta-analysis. The diagnosis of mild dementia tended to correlate with ID severity (odds ratio [OR], 1.38; 95% CI 0.87–2.18). The features of behavior-variant frontotemporal dementia may be clinically confirmed in

diagnosing early symptoms of DS-associated AD (DSAD). Moreover, age-appropriate cognitive assessment is important. Further studies are required to evaluate DSAD using a combination of biomarkers and ID-related data.

Shirai, Y., Bishop, K., & Kushner, M.
National dementia capable care training: A model implementation and evaluation
Intellectual and Developmental Disabilities, 2021, 59 (5), 422–435.
https://doi.org/10.1352/1934-9556-59.5.422
Abstract: With a growing need for specialized training for direct caregivers and support staff of persons with intellectual and developmental disabilities (IDD) affected by dementia, the National Task Group on Developmental Disabilities and Dementia Practices (NTG) developed a comprehensive evidence-informed Dementia Capable Care Training (DCCT). To overcome the challenge of the training length and cost, and to extend its dissemination, the Sonoran Center developed a shorter version of the NTG-DCCT while retaining its core components, and implemented it in seven cities in the U. S. Southwest (N = 368). The pre- and post-training evaluation (n =260) demonstrated that the short version of the NTG-DCCT is effective in significantly improving participants' knowledge and/or confidence in dementia capable care. The follow-up semi-structured interviews of participants (n = 7) provide some insights.

Shooshtari, S., Stoesz, B.M., Udell, L., Fenez, L., Dik, N., Burchill, C., Sachs, E., Menec, V.
Aging with intellectual and developmental disabilities and dementia in Manitoba
Advances in Mental Health and Intellectual Disabilities, 11(4), 134-144.
https://doi.org/10.1108/AMHID-03-2017-0007
Abstract: Information on the risk of dementia in aging persons with intellectual and/or developmental disability(IDD) in Manitoba, Canada is lacking. The purpose of this paper is to estimate dementia prevalence in adults with IDD. Anonymized population-level health and non-health administrative data (1979-2012) contained in the Population Health Research Data Repository of the Manitoba Centre for Health Policy (MCHP) were linked to identify adults with IDD, and estimate the prevalence of dementia based on the presence of ICD codes. Prevalence of dementia was estimated for persons aged 18-55 years and 55+ years, and was reported by sex, type of residence, region of residence, neighbourhood income quintiles, and IDD diagnostic category.Of the 8,655 adults with IDD identified, 8.1 per cent had an indication of dementia in their medical records; an estimate three times greater than that found for those without IDD (2.6 per cent). More than 17 per cent of Manitobans with IDD aged 55+ years had an indication of dementia, which was nearly twice the rate reported previously. Of those with IDD and dementia, 34.7 per cent lived in long-term care facilities. Health and social support services are typically available to individuals with dementia aged 65+ years; thus, younger adults with IDD and dementia may not be eligible for those supports. To promote equity in health and access to care, dementia screening and increased supports for aging individuals with IDD are recommended.

Shultz, J.M., Aman, M.G., & Rojahn, J..
Psychometric evaluation of a measure of cognitive decline in elderly people with mental retardation.
Research in Developmental Disabilities, 1998, 19, 63-71. doi: 10.1016/s0891-4222(97)00029-2.
Abstract: Forty elderly persons with mental retardation were assessed by their care providers on a modified version of the Short Informant Questionnaire on Cognitive Decline in The Elderly (IQCODE) an instrument designed to quantify cognitive decline in elderly people in the general population. They were also assessed for IQ, aberrant behavior, and current mental status; test-retest and interrater reliability were evaluated as well. Internal consistency, as assessed by coefficient alpha, was moderately high (alpha = .86). Test-retest reliability was mediocre and interrater reliability levels did not reach statistical significance. The Short IQCODE was not correlated with a variety of demographic features or with behavior ratings, showing evidence of divergent validity. However, the Short IQCODE was only weakly (nonsignificantly) correlated with a measure of current mental status, which challenges its concurrent validity. The Short IQCODE probably needs to be modified further for satisfactory psychometric performance in people with mental retardation. However, some features of this study may have resulted in suboptimal estimates of the Short IQCODE's psychometric characteristics.

Sigal, M.J,, & Levine, N.
Down's syndrome and Alzheimer's disease.
Journal of the Canadian Dental Association, 1993, 59(10), 823-5, 829.
https://pubmed.ncbi.nlm.nih.gov/8221282/
Abstract: Individuals with Down's syndrome (DS) who live to be 40 years of age will demonstrate neuropathological changes that are consistent with Alzheimer's disease (AD). Due to modern medical intervention, we are now observing an aging DS population. Middle-aged Down's syndrome adults are actually considered to be "very old," and it is not uncommon to observe a progressive loss of cognitive function and a decline in the ability to perform daily tasks consistent with that seen in Alzheimer's disease. At this stage, the DS individual will not be able to perform daily preventive dental care and may be unable to cooperate for professional dental care. Clinicians who care for DS adults must be aware of this problem when preparing their dental treatment plans, which must emphasize preventive care prior to the onset of dementia and the maintenance of that program during their patients' cognitive decline. In the latter stages of AD, it may be necessary to extract all the remaining teeth due to the inability of the individual or care giver to provide adequate oral hygiene to prevent dental caries or periodontal disease.

Simard, M., & van Reekum, R.
Dementia with Lewy bodies in Down's syndrome.
International Journal of Geriatric Psychiatry, 2001, Mar;16(3), 311-20.
Abstract: The association between Down's syndrome (DS) and Alzheimer's disease is well established. This paper presents a review of the literature, suggesting a possible association between DS and the more recently recognized dementia with Lewy bodies (DLB). Patients with DLB frequently present with changes in affect and behavior, and in particular with psychotic symptoms. The literature suggests a possible role for atypical neuroleptics in the management of psychosis in DLB.

Sinai, A., Mokrysz, C., Bernal, J., Bohnen, I., Bonell, S., Courtney, K., Dodd, K., ... Strydom, A.
Predictors of age of diagnosis and survival of Alzheimer's disease in Down syndrome.
Journal of Alzheimer's Disease, 2018, 61(2), 717-728.
doi: 10.3233/JAD-170624.
Abstract: People with Down syndrome (DS) are an ultra-high risk population for Alzheimer's disease (AD). Understanding the factors associated with age of onset and survival in this population could highlight factors associated with modulation of the amyloid cascade. This study aimed to establish the typical age at diagnosis and survival associated with AD in DS and the risk factors associated with these. Data was obtained from the Aging with Down Syndrome and Intellectual Disabilities (ADSID) research database, consisting of data extracted from clinical records of patients seen by Community Intellectual Disability Services (CIDS) in England. Survival times when considering different risk factors were calculated. **The mean age of diagnosis was 55.80 years**, *SD 6.29. Median survival time after diagnosis was 3.78 years, and median age at death was approximately 60 years.* Survival time was associated with age of diagnosis, severity of intellectual disability, living status, anti-dementia medication status, and history of epilepsy. Age at diagnosis and treatment status remained predictive of survival time following adjustment. This study provides the best estimate of survival in dementia within the DS population to date, and is in keeping with previous estimates from smaller studies in the DS population. This study provides important estimates and insights into possible predictors of survival and age of diagnosis of AD in adults with DS, which will inform selection of participants for treatment trials in the future.

Silverman W, Krinsky-McHale SJ, Kovacs C, Lee JH, Listwan T, Pang DI, Zigman WB, Schupf N.
Individualized estimated years from onset of Alzheimer's disease- related decline for adults with Down syndrome.
Alzheimers Dement (Amst). 2023 Jun 27;15(2):e12444. doi: 10.1002/dad2.12444.
Abstract: Adults with Down syndrome (DS) are at increased risk for Alzheimer's disease (AD) and vary in their age of transition from AD preclinical to prodromal or more advanced clinical stages. An empirically based method is needed to determine individual "estimated years from symptom onset (EYO)," the same construct used in studies of autosomal dominant AD. Archived data from a previous study of > 600 adults with DS were examined using survival analysis methods. Age-specific prevalence of prodromal AD or dementia, cumulative risk, and EYOs were determined. Individualized EYOs for adults with DS ranging in age from 30 to 70+ were determined, dependent upon chronological age and clinical status. EYOs can be a useful tool for studies focused on biomarker changes during AD progression in this and other populations at risk, studies that should contribute to improved methods for diagnosis, prediction of risk, and identification of promising treatment targets.

Silverman, W., Krinsky-McHale, S.J., Lai, F., Rosas, H.D., Hom, C., Doran, E., Pulsifer, M., Lott. I., Schupf, N, and Alzheimer's Disease in Down Syndrome (ADDS) Consortium
Evaluation of the National Task Group-Early Detection Screen for Dementia: Sensitivity to 'mild cognitive impairment' in adults with Down syndrome
Journal of Applied Research in Intellectual Disabilities, 2020, Dec 13. doi:10.1111/jar.12849.
Abstract: The accuracy of the National Task Group-Early Detection Screen for Dementia (NTG-EDSD) was evaluated in a sample of 185 adults with Down syndrome (DS), emphasizing 'mild cognitive impairment (MCI-DS)'. Knowledgeable informants were interviewed with the NTG-EDSD, and findings were compared to an independent dementia status rating based on consensus review of detailed assessments of cognition, functional abilities and health status (including physician examination). Results indicated that sections of the NTG-EDSD were sensitive to MCI-DS, with one or more concerns within the 'Memory' or 'Language and Communication' domains being most informative. The NTG-EDSD is a useful tool for evaluating dementia status, including MCI-DS. However, estimates of sensitivity and specificity, even for detecting frank dementia, indicated that NTG-EDSD findings need to be supplemented by additional sources of relevant information to achieve an acceptable level of diagnostic/screening accuracy.

Silverman, W., Schupf, N., Zigman, W., Devenny, D., Miezejeski, C., Schubert, R., & Ryan, R.
Dementia in adults with mental retardation: Assessment at a single point in time.
American Journal of Mental Retardation, 2004, 109, 111-125.
https://doi.org/10.1352/0895-8017(2004)109<111:DIAWMR>2.0.CO;2
Abstract: Dementia status of 273 adults with mental retardation was rated based upon two extensive evaluations conducted 18 months apart. Overall, 184 individuals did not have dementia, 33 had possible or definite dementia, and 66 had findings suggesting uncertain or questionable status. These ratings were compared to binary classifications (dementia vs. no dementia) generated from the Dementia Questionnaire for Persons With Mental Retardation (Evenhuis, 1995) and the IBR Mental Status Examination (Wisniewski & Hill, 1985). When performance was referenced to IQs (established earlier in adulthood), quantitative criteria effectively distinguished between individuals with and without dementia based upon assessment at a single point in time. Findings suggest that procedures of this type could soon contribute to more accurate and rapid diagnoses of dementia.

Smith, D., & Chicoine, B.
Difficulties of diagnosing and managing dementia in people with Down syndrome
British Journal of Psychiatry, 2018, Nov, 213(5), 668-669. doi:10.1192/bjp.2018.199.
Abstract: In our experience as the directors of Down syndrome clinics for adults, the big issue is how the diagnosis of dementia Is made. Clinicians tend to easily apply the diagnosis or Alzheimer's dementia without looking at all of the potential causes of pseudodementia in this population. They often assume that loss of ability is due to dementia because of a study published in 1985 that showed plaques and tangles in the brain tissue of all people with Down syndrome over the age of 35. Wisniewskl & Rabe subsequently wrote that there was a discrepancy between neuropathology and the occurrence of dementia in people with Down syndrome. Just as the population of typically developed older adults the diagnosis of Alzheimer's dementia in people with Down syndrome should be made only after evaluation for causes of pseudodementia.

Smith, K.J., Gupta, S., Fortune, J., Lowton, K., Victor, C., Burke, E., Carew, M.T., Livingstone,E., Creeger, M., Shanahan, P., Walsh, M., & Ryan, J.M.
Aging well with a lifelong disability: A scoping review.
The Gerontologist, 2024 Sept, 64(9), gnae092.
https://doi.org/10.1093/geront/gnae092
Abstract: Existing literature highlights notable health and social inequalities for people aging with a lifelong disability and the need for research to better understand how we can support this group to age well. This scoping review mapped existing literature related to "aging well" in people with lifelong disabilities. Five scientific databases and gray literature sources were searched for studies related to "aging well" and "lifelong disability" (defined as a disability that a person had lived with since birth or early childhood). We identified 81 studies that discussed aging well with a lifelong disability, with most (70%) focusing on intellectual disabilities. Two themes captured existing research on aging well with a lifelong disability: (1) framing aging well with a lifelong disability, which included the ways that people with lifelong disability, their supporters, and existing research frame aging well for this group and (2) supporting people to age well with a lifelong disability, which involves the micro-, meso-, and macro-level factors where research suggests interventions to facilitate aging well could be situated. This synthesis highlights how aging well is currently framed in the literature and where interventions to improve aging well in this group could be situated. Literature highlights the importance of considering multilevel interventions to improve aging well. Evidence gaps include the lack of research conducted with groups other than those with intellectual disabilities and the need for more research examining aging well interventions.

Smith, K.J., Peterson, M.D., Victor, C., & Ryan, J.M.
Risk of dementia in adults with cerebral palsy: a matched cohort study using general practice data.
BMJ Open. 2021 Jan 25, 11(1), e042652. doi: 10.1136/bmjopen-2020-042652.
Abstract: Authors sought to determine the risk of incident dementia in adults with **cerebral palsy** (CP) compared with age, sex and general practice (GP) matched controls by using a retrospective cohort study. The setting was UK GPs linked into the Clinical Practice Research Datalink (CPRD). CPRD data were used to identify adults aged 18 or older with a diagnosis of CP. Each adult with CP was matched to three controls who were matched for age, sex and GP. In total, 1703 adults with CP and 5109 matched controls were included in the analysis. The mean baseline age of participants was 33.30 years (SD: 15.48 years) and 46.8% of the sample were female. New diagnosis of dementia during the follow-up period (earliest date of 1987 to latest date of 2015). During the follow-up, 72 people were identified with a new diagnosis of dementia. The overall proportion of people with and without CP who developed dementia was similar (CP: n=19, 1.1%; matched controls n=54, 10.0%). The unadjusted HR suggested that people with CP had an increased hazard of being diagnosed with dementia when compared with matched controls (HR 2.69, 95% CI 1.44 to 5.00). This association was attenuated when CP comorbidities (sensory impairment, intellectual disability and epilepsy) were accounted for (HR 1.92, 95% CI 0.92 to 4.02). There was no difference in the proportion of people with CP and matched controls who were diagnosed with dementia during the follow-up. Furthermore,

while there was evidence for an increased hazard of dementia among people with CP, the fact that this association was attenuated after controlling for comorbidities indicates that this association may be explained by comorbidities rather than being a direct result of CP. Findings should be interpreted with caution due to the low number of incident cases of dementia.

Snyder HM, Bain LJ, Brickman AM, Carrillo MC, Esbensen AJ, Espinosa JM, Fernandez F, Fortea J, Hartley SL, Head E, Hendrix J, Kishnani PS, Lai F, Lao P, Lemere C, Mobley W, Mufson EJ, Potter H, Zaman SH, Granholm AC, Rosas HD, Strydom A, Whitten MS, Rafii MS.
Further understanding the connection between Alzheimer's disease and Down syndrome.
Alzheimers Dement. 2020 Jul;16(7):1065-1077. doi: 10.1002/alz.12112. Epub 2020 Jun 16
Abstract: Improved medical care of individuals with Down syndrome (DS) has led to an increase in life expectancy to over the age of 60 years. In conjunction, there has been an increase in age-related co-occurring conditions including Alzheimer's disease (AD). Understanding the factors that underlie symptom and age of clinical presentation of dementia in people with DS may provide insights into the mechanisms of sporadic and DS-associated AD (DS-AD). In March 2019, the Alzheimer's Association, Global Down Syndrome Foundation and the LuMind IDSC Foundation partnered to convene a workshop to explore the state of the research on the intersection of AD and DS research; to identify research gaps and unmet needs; and to consider how best to advance the field. This article provides a summary of discussions, including noting areas of emerging science and discovery, considerations for future studies, and identifying open gaps in our understanding for future focus.

Social Care Institute for Excellence
Learning disabilities and dementia
SCIE, Isosceles Head Office, One High Street, Egham TW20 9HJ, (2020). https://www.scie.org.uk/dementia/living-with-dementia/learning-disabilities/
Abstract: Webpage informational matter on dementia and intellectual disability, recognising early changes, diagnostic and assessment issues, environmental design, and health care.

Soliman, A, & Hawkins, D.
The link between Down's syndrome and Alzheimer's disease: 1.
British Journal of Nursing, 1998, Jul 9-22, 7(13), 779-784.
Abstract: This article, the first of two parts, considers the link between Down's syndrome and Alzheimer's disease and how this link has been a significant factor with regards to research into the aetiology of Alzheimer's disease. It describes some of the suggested causes of Alzheimer's disease in people with Down's syndrome. The diagnosis, signs and symptoms of Alzheimer's disease are briefly discussed. The second article concludes with the implications of Alzheimer's disease in people with Down's syndrome for family careers, services and nurses.

Soliman, A., & Hawkins, D.
The link between Down's syndrome and Alzheimer's disease: 2.
British Journal of Nursing, 1998, Jul 23-Aug 12, 7(14), 847-850. DOI: 10.12968/bjon.1998.7.14.5638
Abstract: In this article, the second of two parts, the needs of family and professional carers of people with Down's syndrome and Alzheimer's disease are examined. Substantial numbers of people with Down's syndrome survive to the age of 50 and beyond and so work still needs to be done on finding solutions to the problems faced by this client group and its carers. As well as the difficulties faced by any family carer of a person with dementia, those caring for someone with Down's syndrome and Alzheimer's disease may also have to deal with additional worries and problems. Consideration is given to service provision and the implications for nursing. A case study will illustrate some of the points made.

Soriano, L.D.H., Sandkühler, K., Videla, L., Benejam, B., Carmona-Iragui, M., Wlasich, E., Kustermann, J., Barroeta, I., Vaqué-Alcázar, L., Rodríguez-Baz, I., Bejanin, A., Fernández, S., Arranz, J., Arriola-Infante, J.E., Maure-Blesa, .L, Juan, A.S., Nübling, G., Wagemann, O., Stockbauer, A., Hassenstab, J., Levin, J., & Fortea, J.
Discrepancies in assessing intellectual disability levels in adults with Down syndrome: Implications for dementia diagnosis.
Alzheimer's & Dementia, 2025 Jun, 21(6), e70307. doi: 10.1002/alz.70307.
Abstract: Cut-offs derived from baseline cognitive assessments, stratified by intellectual disability (ID) level, have been proposed to diagnose symptomatic Alzheimer's disease (AD) in Down syndrome (DS). However, discrepancies in ID classification risk misclassification when applying cut-offs across sites. This dual-center cohort study included 673 adults with mild to moderate ID at different AD stages. We assessed ID classification discrepancies across sites and the impact on Cambridge Cognitive Examination for Older Adults with Down's Syndrome (CAMCOG-DS) cut-offs for AD dementia diagnosis derived from receiver operating characteristic analysis. Inter-rater agreement for ID level classification was 95% within sites but 60% between sites. While CAMCOG-DS score distributions in the whole cohort were similar across sites, ID classification discrepancies caused higher cut-offs in Barcelona for mild and moderate ID compared to Munich. Applying site-specific cut-offs to another cohort reduced sensitivity and specificity. Standardizing ID classification is critical for generalizable cut-offs to accurately diagnose AD dementia based on neuropsychological assessments in DS. CAMCOG-DS cut-offs by intellectual disability level classify dementia in Down syndrome. ID classification discrepancies between sites impact CAMCOG-DS diagnostic cut-offs. Applying site-specific cut-offs to other cohorts reduces sensitivity and specificity. Standardized ID classification is essential for generalizable cognitive cut-offs. Use site-specific cut-offs until ID classification is standardized.

Specialist Learning Disability and Forensic Service Hertfordshire Partnership NHS TRUST
HPT and ACS practice guidelines for screening, assessment and management of dementia in persons with a learning disability.
Specialist Learning Disability and Forensic Service, Hertfordshire Partnership NHS TRUST, Trust Head Office, 99 Waverley Road, St Albans AL3 5TL UK (2006). [34pp]. https://www.choiceforum.org > docs > dementiahpt
Abstract: Booklet containing background information on dementia among adults with intellectual disability and guide for assessment, interventions, and care practices.

Speckemeier, C., Niemann, A., Weitzel, M., Abels, C., Höfer, K., Walendzik, A., Wasem, J., & Neusser, S.
Assessment of innovative living and care arrangements for persons with dementia: a systematic review.
BMC Geriatrics, 2023 Aug 1, 23(1), 464. doi: 10.1186/s12877-023-04187-4.
Abstract: Alternative forms of housing for persons with dementia have been developed in recent decades. These concepts offer small groups of residents familiar settings combined with efforts to provide normal daily life. The aim of this systematic review is to collate and analyze these more innovative forms of housing regarding residents' quality of life, behavioral aspects, as well as functional, cognitive and emotional aspects. Searches were conducted in PubMed, EMBASE and PsycInfo in November 2020. Studies comparing traditional and more innovative living environments for persons with dementia were eligible. Concepts are described based on the results of additional searches. Risk of bias of included studies was assessed using checklists from the Joanna Briggs Institute. A total of 21 studies corresponding to 11 different concepts were included, namely Green Houses (USA), Group Living (Sweden), Cantou (France), Group Homes (Japan), Small-scale Group Living (Austria), Special Care Facilities (Canada), Shared-housing Arrangements (Germany), Residential Groups (Germany), Residential Care Centers / Woodside Places (USA/Canada), Small-scale Living (Netherlands/ Belgium), and Green Care Farms (Netherlands). The concepts are broadly similar in terms of care concepts,

but partly differ in group sizes, staff qualifications and responsibilities. Several studies indicate that innovative forms of housing may encourage social behavior, preserve activity performance and/or positively influence emotional status compared to more traditional settings, while other studies fail to demonstrate these effects. Some studies also show increased behavioral and psychological symptoms of dementia (BPSD) in residents who live in more innovative housing concepts. The effect on cognition remains indistinct.
The positive effects may be attributable to the inherent characteristics, including small group sizes, a stimulating design, and altered staff roles and responsibilities. Arguably, some of these characteristics might also be the reason for increased BPSD. Studies had variable methodological quality and results have to be considered with caution. Future research should examine these effects more closely and should investigate populations' preferences with regards to housing in the event of dementia. A

Stanton, L.R., & Coetzee, R.H.
Down's syndrome and dementia
Advances in Psychiatric Treatment, 2004 January, 10(1), 50 - 58.
doi: https://doi.org/10.1192/apt.10.1.50
Abstract: Downs syndrome is the most common genetic disorder seen in clinical practice: about 94% occurs because of non-disjunction of chromosome 21 and 3-5% because of translocation. Individuals increasingly survive to middle and old age, probably because of advances in medical treatment and improved living conditions. People with Downs syndrome have an increased risk of developing Alzheimer's disease in middle age. Within ICD-10 and DSM-IV classifications there is no consensus on the diagnosis of dementia in people with leaming disability. New treatments have been licensed for use in mild to moderate Alzheimer's disease (e.g. acetylcholinesterase inhibitors and memantine). The comorbid picture of Downs syndrome and Alzheimer's disease presents a unique challenge to the clinician in both diagnosis and management.

Starkey, H., Bevns, S., & Bonell, S.
The role of prospective screening in the diagnosis of dementia in people with Down's syndrome
Advances in Mental Health and Intellectual Disabilities, 2014, 8(5), 283-291.
https://doi.org/10.1108/AMHID-12-2013-0067
Abstract: People with Down's syndrome are at increased risk of developing early onset Alzheimer's disease. It has been recommended that all adults with Down's syndrome receive baseline neuropsychological testing for dementia. In certain areas prospective screening of people with Down's syndrome takes place to ensure the early diagnosis of the condition. However, little has been published on the value of this type of screening. The purpose of this paper is to report on a prospective screening programme and asks whether the programme is effective in identifying dementia-related changes in people with Down's syndrome and whether the current screening intervals are appropriate. All adults with Down's syndrome in Plymouth (UK) are identified and offered a comprehensive test battery at baseline at the age of 20 and then have testing biennially from 40 to 50 and annually after 50. All individuals diagnosed with dementia between 2001 and 2013 were identified and their case notes examined. The symptoms at the time of diagnosis were identified and whether these symptoms had been identified through the screening programme or by other routes were recorded. Prevalence data and age at diagnosis were also recorded. In total, 26 people were diagnosed with dementia during the study period. Of these, the diagnosis of dementia followed concerns being identified during the routine screening programme in 54 per cent of cases. In the younger age group (age 40-49) 63 per cent of people were identified through the screening programme. At the time of diagnosis a mean of 5.5 areas of concern were in evidence. This paper adds to the growing evidence base around the value of prospective dementia screening in people with Down's syndrome. It is also one of a few studies exploring the frequency of screening. Additionally, it adds further data about prevalence of dementia in people with Down's syndrome.

Startin, C.M., Hamburg, S., Hithersay, R., Al-Janabi, T., Mok, K.Y., Hardy, J. LonDownS Consortiu, & Strydom, A.
Cognitive markers of preclinical and prodromal Alzheimer's disease in Down syndrome.
Alzheimer's & Dementia, 2019, Feb, 15(2), 245-257.
doi: 10.1016/j.jalz.2018.08.009. Epub 2018 Nov 28.
Abstract: Down syndrome (DS) is associated with an almost universal development of Alzheimer's disease. Individuals with DS are therefore an important population for randomized controlled trials to prevent or delay cognitive decline, though it is essential to understand the time course of early cognitive changes. We conducted the largest cognitive study to date with 312 adults with DS to assess age-related and Alzheimer's disease-related cognitive changes during progression from preclinical to prodromal dementia, and prodromal to clinical dementia. Changes in memory and attention measures were most sensitive to early decline. Resulting sample size calculations for randomized controlled trials to detect significant treatment effects to delay decline were modest. Our findings address uncertainties around the development of randomized controlled trials to delay cognitive decline in DS. Such trials are essential to reduce the high burden of dementia in people with DS and could serve as proof-of-principle trials for some drug targets.

Startin, C.M., Lowe, B., Hamburg, S., Hithersay, R., Strydom, A., & LonDowns Consortium.
Validating the Cognitive Scale for Down Syndrome (CS-DS) to detect longitudinal cognitive decline in adults with Down syndrome.
Frontiers in Psychiatry, 2019, Apr 16,10, 158. doi: 10.3389/fpsyt.2019.00158. eCollection 2019.
Abstract: Down syndrome (DS) is associated with intellectual disability and an ultra-high risk of developing dementia. Informant ratings are invaluable to assess abilities and related changes in adults with DS, particularly for those with more severe intellectual disabilities and/or cognitive decline. We previously developed the informant rated Cognitive Scale for Down Syndrome (CS-DS) to measure everyday cognitive abilities across memory, executive function, and language domains in adults with DS, finding CS-DS scores are a valid measure of general abilities, and are significantly lower for those with noticeable cognitive decline compared to those without decline. To further test the validity of the CS-DS in detecting changes associated with cognitive decline we collected longitudinal data across two time points, approximately 1.5-2 years apart, for 48 adults with DS aged 36 years and over. CS-DS total scores (78.83 ± 23.85 vs. 73.83 ± 25.35, p = 0.042) and executive function scores (46.40 ± 13.59 vs. 43.54 ± 13.60, p = 0.048) significantly decreased between the two time points, with scores in the memory domain trending towards a significant decrease (22.19 ± 8.03 vs. 20.81 ± 8.63, p = 0.064). Adults with noticeable cognitive decline at follow-up showed a trend to significantly greater change in total scores (7.81 ± 16.41 vs. 3.59 ± 16.79, p = 0.067) and significantly greater change in executive function scores (5.13 ± 9.22 vs. 1.72 ± 9.97, p = 0.028) compared to those without decline. Change in total scores showed significant correlations with change in scores from other informant measures of everyday adaptive abilities and symptoms associated with dementia, and participant assessment of general cognitive abilities (all p < 0.005), while change in memory scores (R 2 = 0.28, p = 0.001) better predicted change in participant cognitive assessment scores than change in executive function (R 2 = 0.15, p = 0.016) or language (R 2 = 0.15, p = 0.018) scores. These results suggest informants may better detect changes in the executive function domain, while change in informant rated memory scores best predicts change in assessed cognitive ability. Alternatively, memory domain scores may be sensitive to changes across both early and late cognitive decline, whereas executive function domain scores are more sensitive to changes associated with later noticeable cognitive decline. Our results provide further support for the validity of the CS-DS to assess everyday cognitive abilities and to detect associated longitudinal changes in individuals with DS.

Startin, C.M., Rodger, R., Fodor-Wynne, L., Hamburg, S., & Strydom, A.

Developing an informant questionnaire for cognitive abilities in Down syndrome: The Cognitive Scale for Down Syndrome (CS-DS)
PLOS ONE, Published: May 6, 2016
https://doi.org/10.1371/journal.pone.0154596
Abstract: Down syndrome (DS) is the most common genetic cause of intellectual disability (ID). Abilities relating to executive function, memory and language are particularly affected in DS, although there is a large variability across individuals. People with DS also show an increased risk of developing dementia. While assessment batteries have been developed for adults with DS to assess cognitive abilities, these batteries may not be suitable for those with more severe IDs, dementia, or visual / hearing difficulties. Here we report the development of an informant rated questionnaire, the Cognitive Scale for Down Syndrome (CS-DS), which focuses on everyday abilities relating to executive function, memory and language, and is suitable for assessing these abilities in all adults with DS regardless of cognitive ability. Complete questionnaires were collected about 128 individuals with DS. After final question selection we found high internal consistency scores across the total questionnaire and within the executive function, memory and language domains. CS-DS scores showed a wide range, with minimal floor and ceiling effects. We found high interrater (n = 55) and test retest (n = 36) intraclass correlations. CS-DS scores were significantly lower in those aged 41+ with significant cognitive decline compared to those without decline. Across all adults without cognitive decline, CS-DS scores correlated significantly to measures of general abilities. Exploratory factor analysis suggested five factors within the scale, relating to memory, self-regulation / inhibition, self-direction / initiation, communication, and focussing attention. The CS-DS therefore shows good interrater and test retest reliability, and appears to be a valid and suitable informant rating tool for assessing everyday cognitive abilities in a wide range of individuals with DS. Such a questionnaire may be a useful outcome measure for intervention studies to assess improvements to cognition, in addition to detecting dementia-related cognitive decline. The CS-DS may also be a useful tool for other populations with ID.

Startin, C. M., D'Souza, H., Ball, G., Hamburg, S., Hithersay, R., Hughes, K. M. O., Massand, E., Karmiloff-Smith, A., Thomas, M. S. C., LonDownS Consortium, & Strydom, A.
Health comorbidities and cognitive abilities across the lifespan in Down syndrome.
Journal of Neurodevelopmental Disorders, 2020, 12(1), 4.
https://doi.org/10.1186/s11689-019-9306-9
Abstract: Down syndrome (DS) is associated with variable intellectual disability and multiple health and psychiatric comorbidities. The impact of such comorbidities on cognitive outcomes is unknown. We aimed to describe patterns of physical health and psychiatric comorbidity prevalence, and receptive language ability, in DS across the lifespan, and determine relationships with cognitive outcomes. Detailed medical histories were collected and cognitive abilities measured using standardised tests for 602 individuals with DS from England and Wales (age range 3 months to 73 years). Differences in prevalence rates between age groups and between males and females were determined using chi-squared or Fisher's exact tests. In adults, rates for psychiatric comorbidities were compared to expected population rates using standardised morbidity ratios (SMRs). Adapted ANCOVA functions were constructed to explore age and sex associations with receptive language ability across the lifespan, and regression analyses were performed to determine whether the presence of health comorbidities or physical phenotypes predicted cognitive abilities. Multiple comorbidities showed prevalence differences across the lifespan, though there were few sex differences. In adults, SMRs were increased in males and decreased in females with DS for schizophrenia, bipolar disorder, and anxiety. Further, SMRs were increased in both males and females with DS for dementia, autism, ADHD, and depression, with differences more pronounced in females for dementia and autism, and in males for depression. Across the lifespan, receptive language abilities increasingly deviated from age-typical levels, and males scored poorer than females. Only autism and epilepsy were associated with poorer cognitive ability in those aged 16-35 years, with no relationships for physical health comorbidities, including congenital heart defects. Our results indicate the prevalence of multiple comorbidities varies across the lifespan in DS, and in adults, rates for psychiatric comorbidities show different patterns for males and females relative to expected population rates. Further, most health comorbidities are not associated with poorer cognitive outcomes in DS, apart from autism and epilepsy. It is essential for clinicians to consider such differences to provide appropriate care and treatment for those with DS and to provide prognostic information relating to cognitive outcomes in those with comorbidities

Stephens, M.M., Herge, E., & Wright, C.
Down syndrome and dementia: A patient and care-giver centered approach.
Delaware Journal of Public Health, 2021, Sep 27. 7(4), 128-130. doi: 10.32481/djph.2021.09.016.
https://djph.org/wp-content/uploads/2021/09/djph-74-016.pdf
Abstract: Reviewed are the common characteristics and co-morbidities of individuals with Down syndrome (DS), he epidemiology of AD and clinical presentation of AD in this cohort, and discussed are practical considerations for diagnosis and treatment as well as strategies to maximize support for caregivers. Consistent with the National Plan to Address Alzheimer's Disease, we will focus on the importance of the early detection of AD and what it means to patients, caregivers, and the healthcare system. The most commonly used screening tool for evaluating the early indicators of dementia in DS is the National Task Group - Early Detection Screen for Dementia (NTG-EDSD). The NTG-EDSD scoring scale helps clinicians compare patients with DS to their own baseline as the questions are scored always been the case, always but worse, new symptom in the past year, or does not apply. The framework of analysis reflects transitions from "clinically stable" to concern for MCI-DS to suspicious for AD. Pharmacotherapy and non-pharmacologic approaches to treat co-existing depression and anxiety, and improve quality of life are important considerations in the care of patients with AD and DS. Caring for and supporting patients with DS and MCI-DS and AD is best managed from an interprofessional team approach to achieve the best outcomes. Social workers, case managers, nurses and occupational therapists can evaluate the needs of the patient and caregiver and provide recommendations to support patient participation in activities and routines which can improve patient overall health and well-being as well as reduce unwanted behaviors. The goal of the primary care physician is to recognize and manage the medical and socioeconomic disparity that exists for patients with DS, especially as they age.

Struckmeyer, L.R., & Pickens, N.
Home modifications for people with Alzheimer's disease: A scoping review
American Journal of Occupational Therapy, 2015, 70(1), 1-9. doi:10.5014/ajot.2015.016089Corpus ID: 31243965.
Abstract: The purpose of this review was twofold: (1) to gain insight into what is known from the literature about home modifications for people with Alzheimer's disease (AD) and (2) to identify gaps in the literature that could lead to opportunities for research. A systematic scoping review of peer-reviewed articles published from 1994 through 2014 explored home modifications and AD. Seventeen articles met the inclusion criteria. The three major findings pertain to (1) the caregiver role and caregiver training, (2) a client-centered collaborative approach to assessment and intervention, and (3) modifications for safety and function. Home modifications involved the physical and social environments as well as cognitive strategies at the task level. Opportunities exist for the development of assessment procedures, the exploration of home modifications in the later stages of AD, and the study of home modification needs of people with dementia who live alone.

Strydom, A.
Clinical trials to prevent or delay Alzheimer's disease in individuals with Down syndrome

Journal of Intellectual Disability Research, 2019, 63(8), 640-641. https://onlinelibrary.wiley.com/doi/abs/10.1111/jir.12651
Abstract: Adults with Down syndrome (DS) have neuropathological features identical to individuals with sporadic Alzheimer's disease (AD) and this discovery played an important role in the identification of the amyloid precursor protein gene on chromosome 21. Individuals with DS have a lifetime risk for dementia in excess of 90% and DS is now acknowledged to be the most common genetic cause of AD, but this group is often excluded from AD medication trials. This review will discuss primary and secondary prevention trials for AD in DS and the potential barriers and solutions to such trials. It will include a brief overview of the epidemiology, diagnosis and outcome measurement issues pertinent to prevention trials, as well as important ageing-related co-morbidities that need to be considered in the design of such trials. Ddescribed is the work of the Europe-wide Horizon21 Consortium and other efforts to establish DS clinical trials networks, as well as to consider the methodological issues for trials to prevent or delay AD in DS. It was noted that individuals with DS could benefit from treatments to prevent or delay AD. Improved knowledge of pathogenic processes and their clinical consequences in DS will hopefully lead to new clinical trials.

Strydom, A., & Hassiotis, A.
Diagnostic instruments for dementia in older people with intellectual disability in clinical practice
Aging & Mental Health, 2003, 7(6), 431-437. doi: 10.1080/1360786031000159468 2.
Abstract: There is a need for simple and reliable screening instruments for dementia in the intellectual disability (ID) population that can also be used to follow their progress, particularly if they are being treated with anti-dementia drugs. Commonly used tests for the general population such as the Mini Mental State Examination (MMSE) are not appropriate for many people with ID. This paper is a literature review of alternative instruments that have been used in research or recommended by experts since 1991 and have the potential to be used as screening instruments. Two types of tests have been identified: those administered to informants, and those that rely on direct assessment of the individual. The most promising informant rated screening tool in most adults with ID including Down syndrome (DS) diagnosis is the Dementia Questionnaire for Persons with Mental Retardation (DMR). However, sensitivity in single assessments is variable and cut-off scores need further optimization. In those with DS, the Dementia Scale for Down Syndrome (DSDS) has good specificity but mediocre sensitivity. The Test for Severe Impairment and Severe Impairment Battery are two direct assessment tools that show promise as screening instruments, but need further evaluation.

Strydom, A., Al-Janabi, T., Houston, M., & Ridley, J.
Best practice in caring for adults with dementia and learning disabilities.
Nursing Standard (Royal College of Nursing UK) , 2016, Oct, 31(6), 42-51. doi: 10.7748/ns.2016.e10524.
Abstract: People with intellectual [learning] disabilities, particularly Down's syndrome, are at increased risk of dementia. At present, services and care tailored to people with both dementia and a intellectual disability are unsatisfactory. This article reviews the literature specific to dementia in people with learning disabilities, including: comprehensive screening, diagnosis, management, environmental considerations, end of life care and training issues for nursing staff. Recommendations for best practice and service improvement are made to improve the quality of life for individuals with dementia and learning disabilities, pre and post-diagnosis.

Strydom, A., & Hassiotis, A., Livingston, G., & King, M.
Prevalence of dementia in older adults with intellectual disability without Down syndrome
Journal of Applied Research in Intellectual Disabilities, 2006, 19, 253. https://doi.org/10.1111/j.1468-3148.2006.00322_9.x
Abstract: The aim of this study was to determine the prevalence of dementia in older adults with intellectual disability(ID) without Down syndrome. The authors identified the total population of adults with ID aged 60+ in the five London boroughs served through local social services registers, ID teams and residential services providers and then screened the Ss with a simple object memory task, information about functional status, and the Dementia Questionnaire for Persons with Mental Retardation (DMR). Screen positives on the DMR, or those with unexplained functional decline or memory deficits underwent detailed examination. Full assessment of cognitive and physical function was undertaken and additional information was collected from informants and medical records. All information was summarized to determine dementia status with IDC-10, DSM-IV, and DC-LD criteria. The authors identified 264 adults with ID and 222 (84%) participated in the study. One in four screened positive. The prevalence rate for ICD-10 or DSM-IV was 12%. Prevalence differed between those with mild and severe ID, and between diagnostic criteria. The authors concluded that dementia is common in older adults with ID without DS, but prevalence in severe ID deviated from prediction and the use of diagnostic criteria needs to be reviewed.

Strydom, A., Hassiotis, A., & Livingston, G.
Mental health and social care needs of older people with intellectual disabilities
Journal of Applied Research in Intellectual Disabilities, 2005, 18(3), 229-235. Doi:10.1111/J.1468-3148.2005.00221.X
Abstract: Older people with intellectual disabilities (ID) are a growing population but their age-related needs are rarely considered and community services are still geared towards the younger age group. We aimed to examine the mental health and social care needs of this new service user group. We identified all adults with ID without Down syndrome (DS) aged 65+ living in the London boroughs of Camden and Islington. The Psychiatric Assessment Schedule for Adults with a Developmental Disability (PASADD) checklist was used to detect psychiatric disorder, the Vineland Behavior Scale (maladaptive domain) for problem behaviors and the Dementia Questionnaire for Persons with Mental Retardation (DMR) to screen for dementia. Carers reported health problems and disability. Needs were measured with the Camberwell Assessment of Need for adults with Intellectual Disabilities (CANDID-S). A total of 23 older people with ID (13 had mild ID and nine more severe ID) and their carers participated in the survey. In which, 74% had one or more psychiatric symptoms; 30% were previously known with a diagnosis of mental illness. One-third of the older people screened positive for dementia (range: 17-44%, depending on sensitivity of DMR scores used). Three quarters of the group had physical health problems, 74% had poor sight, 22% had hearing loss and 30% had mobility problems. Carers rated unmet needs for accommodation (22%), day activities, and eyesight and hearing. The people with ID rated unmet needs to be social relationships (44%), information and physical health. Authors concluded that older people with ID without DS have considerable prevalence of health problems and psychiatric disorders, including symptoms of functional decline and dementia. Such symptoms are often not recognized and further research into their needs is a priority.

Strydom, A., Livingston, G., King, M., & Hassiotis. A.
Prevalence of dementia in intellectual disability using different diagnostic criteria.
British Journal of Psychiatry, 2007, 191(2), 150-157. doi: 10.1016/j.ridd.2013.02.021. Epub 2013 Apr 9.
Abstract: Diagnosis of dementia is complex in adults with intellectual disability owing to their pre-existing deficits and different presentation. To describe the clinical features and prevalence of dementia and its subtypes, and to compare the concurrent validity of dementia criteria in older adults with intellectual disability. The Becoming Older with Learning Disability (BOLD) memory study is a two-stage epidemiological survey of adults with intellectual disability without Down syndrome aged 60 years and older, with comprehensive assessment of people who screen positive. Dementia was diagnosed according to ICD–10, DSM–IV and DC–LD criteria. The DSM–IV dementia criteria were more inclusive. Diagnosis using ICD–10 excluded people with even moderate dementia. Clinical subtypes of dementia can be recognized in adults with intellectual disability. Alzheimer's dementia was the most common, with a

prevalence of 8.6% (95% CI 5.2–13.0), almost three times greater than expected. Dementia is common in older adults with intellectual disability, but prevalence differs according to the diagnostic criteria used. This has implications for clinical practice.

Strydom, A., Hassiotis, A., King, M., Livingston, G.
The relationship of dementia prevalence in older adults with intellectual disability (ID) to age and severity of ID.
Psychological Medicine, 2008, 15, 1-9. doi: 10.1017/S0033291708003334.
Abstract: Previous research has shown that adults with intellectual disability (ID) may be more at risk of developing dementia in old age than expected. However, the effect of age and ID severity on dementia prevalence rates has never been reported. We investigated the predictions that older adults with ID should have high prevalence rates of dementia that differ between ID severity groups and that the age-associated risk should be shifted to a younger age relative to the general population. A two-staged epidemiological survey of 281 adults with ID without Down syndrome (DS) aged 60 years; participants who screened positive with a memory task, informant-reported change in function or with the Dementia Questionnaire for Persons with Mental Retardation (DMR) underwent a detailed assessment. Diagnoses were made by psychiatrists according to international criteria. Prevalence rates were compared with UK prevalence and European consensus rates using standardized morbidity ratios (SMRs). Dementia was more common in this population (prevalence of 18.3%, SMR 2.77 in those aged 65 years). Prevalence rates did not differ between mild, moderate and severe ID groups. Age was a strong risk factor and was not influenced by sex or ID severity. As predicted, SMRs were higher for younger age groups compared to older age groups, indicating a relative shift in age-associated risk. Criteria-defined dementia is 2-3 times more common in the ID population, with a shift in risk to younger age groups compared to the general population.

Strydom, A., Shooshtari, S., Lee, L., Raykar, V., Torr, J., Tsiouris, J., Jokinen, N., Courtenay, K., Bass, N., Sinnema, M., & Maaskant, M.
Dementia in older adults with intellectual disabilities—epidemiology, presentation, and diagnosis
Journal of Policy and Practice in Intellectual Disabilities, 2010, 7(2), 96-110. https://doi.org/10.1111/j.1741-1130.2010.00253.x
Abstract: As life expectancy of people with intellectual disabilities (ID) extends into older age, dementia is an increasing cause of morbidity and mortality. To update and summarize current knowledge on dementia in older adults with ID, the authors conducted a comprehensive review of the published literature from 1997–2008 with a specific focus on: (1) epidemiology of dementia in ID in general as well as in specific genetic syndromes; (2) presentation; and (3) diagnostic criteria for dementia. The review drew upon a combination of searches in electronic databases Medline, EMBASE, and PsycINFO for original research papers in English, Dutch, or German. The authors report that varied methodologies and inherent challenges in diagnosis yield a wide range of reported prevalence rates of dementia. Rates of dementia in the population with intellectual disability not because of Down syndrome (DS) are comparable with or higher than the general population. Alzheimer's disease onset in DS appears earlier and the prevalence increases from under 10% in the 40s to more than 30% in the 50s, with varying prevalence reported for those 60 and older. Incidence rates increase with age. Few studies of dementia in other genetic syndromes were identified. Presentation differs in the ID population compared with the general population; those with DS present with prominent behavioral changes believed to be because of frontal lobe deficits. Authors recommend large-scale collaborative studies of high quality to further knowledge on the epidemiology and clinical presentation of dementia in this population.

Strydom, A., Chan, T., King, M., Hassiotis, A., & Livingston, G.
Incidence of dementia in older adults with intellectual disabilities
Research in Developmental Disabilities, 2013, 34(6), 1881-1885. doi:10.1016/j.ridd.2013.02.021. Epub 2013 Apr 9.
Abstract: Dementia may be more common in older adults with intellectual disability (ID) than in the general population. The increased risk for Alzheimer's disease in people with Down syndrome (DS) is well established, but much less is known about dementia in adults with ID who do not have DS. We estimated incidence rates from a longitudinal study of dementia in older adults with ID without DS and compared them to general population rates. 222 participants with ID without DS aged 60 years and older were followed up an average of 2.9 years later to identify those who had declined in functional or cognitive abilities. Those who screened positive had a comprehensive assessment for dementia, diagnosed using ICD 10 and DSM IV criteria. 134 participants who did not have dementia at initial assessment were alive and interviewed at follow up; 21 (15.7%) were diagnosed with dementia. Overall incidence rate for those aged = 60 was 54.6/1000 person years (95% CI 34.1-82.3). The highest incidence rate (97.8/1000 person years) was in the age group 70-74. Standardised incidence ratio for those aged = 65 was 4.98 (95% CI 1.62-11.67). Incidence of dementia in older people with intellectual disabilities are up to five times higher than older adults in the general population. Screening may be useful in this population given the high incident rates, particularly as more effective treatments become available. Studies to explore the underlying aetiological factors for dementia associated with intellectual disability could help to identify novel protective and risk factors.

Strydom, A., Coppus, A., Blesa, R., Danek, A., Fortea, J., Hardy, J., Levin, J., Nuebling G., Rebillat A.S., Ritchie, C., van Duijn, C., Zaman, S., & Zetterberg, H.
Alzheimer's disease in Down syndrome: An overlooked population for prevention trials.
Alzheimers Dementia (N Y), 2018, 13(4), 703-713. doi: 10.1016/j.trci.2018.10.006.
Abstract: The discovery that adults with Down syndrome (DS) have neuropathological features identical to individuals with sporadic Alzheimer's disease (AD) played a key role in the identification of the amyloid precursor protein gene on chromosome 21 and resulted in the amyloid cascade hypothesis. Individuals with DS have a lifetime risk for dementia in excess of 90%, and DS is now acknowledged to be a genetic form of AD similar to rare autosomal-dominant causes. Just as DS put the spotlight on amyloid precursor protein mutations, it is also likely to inform us of the impact of manipulating the amyloid pathway on treatment outcomes in AD. Ironically, however, individuals with DS are usually excluded from AD trials. This review will discuss primary and secondary prevention trials for AD in DS and the potential barriers and solutions to such trials and describe the Europe-wide Horizon21 Consortium to establish a DS-AD prevention clinical trials network.

Stuart, C.
Estimating the number of people with Down's syndrome in Scotland and the cohort at elevated risk of early onset dementia.
Tizard Learning Disability Review, 2017, 22(3), 164-171 https://doi.org/10.1108/TLDR-11-2016-0041
Abstract: The purpose of this study was to ascertain the size of the population of adults with Down syndrome in Scotland and provide a basis for estimating their number and age ranges with dementia. Data were requested from all general practitioners (GP) in Scotland on people with an identified READ code denoting Down syndrome. A statistical weighting model was then applied to account for non-response bias. Findings were that there were 3,261 adults with Down estimated by the application of a statistical weighting model. Of these, 1,118 (34%) were aged between 40 and 59. This age group includes those adults with the highest incidence of early onset dementia. It was not possible to apply a benchmark to the percentage of observed data so as to determine the accuracy of the estimates. Adults with Down have an elevated risk of developing dementia significantly earlier than the general population and require specific age appropriate supports and services to meet their needs both pre- and post-diagnosis. The reality of this is currently not fully realized in either standard practice or national policy concerning the issue.

Sullivan, W.F., Diepstra, H., Heng, J., Ally, S., Bradley, E., Casson, I., Hennen, B., Kelly, M., Korossy, M., McNeil, K., Abells, D., Amaria, K., Boyd, K., Gemmill, M., Grier, E., Kennie-Kaulbach, N., Ketchell, M., Ladouceur, J., Lepp, A., Lunsky, Y., McMillan, S., Niel, U., Sacks, S., Shea, S., Stringer, K., Sue, K., & Witherbee, S.
Primary care of adults with intellectual and developmental disabilities: 2018 Canadian consensus guidelines.
Canadian Family Physician, 2018 Apr, 64(4), 254-279. PMID: 29650602; PMCID: PMC5897068. https://pubmed.ncbi.nlm.nih.gov/29650602/
Abstract: To update the 2011 Canadian guidelines for primary care of adults with intellectual and developmental disabilities (IDD). Family physicians and other health professionals experienced in the care of people with IDD reviewed and synthesized recent empirical, ecosystem, expert, and experiential knowledge. A system was developed to grade the strength of recommendations. Adults with IDD are a heterogeneous group of patients and have health conditions and factors affecting their health that can vary in kind, manifestation, severity, or complexity from those of others in the community. They require approaches to care and interventions that are adapted to their needs. These guidelines provide advice regarding standards of care. References to clinical tools and other practical resources are incorporated. The approaches to care that are outlined here can be applied to other groups of patients that have impairments in cognitive, communicative, or other adaptive functioning. As primary care providers, family physicians play a vital role in promoting the health and well-being of adults with IDD. These guidelines can aid their decision making with patients and caregivers.

Sung, H., Hawkins, B.A., Eklund, S.J., Kim, K.A., Foose, A., May, M.E., Rogers, N.B.
Depression and dementia in aging adults with Down syndrome: a case study approach.
Mental Retardation, 1997, 35(1), 27-38. https://doi: 10.1352/0047-6765(1997)035<0027:DADIAA>2.0.CO;2
Abstract: Patterns of symptoms associated with depression and dementia were examined in 3 aging adults with Down syndrome. A case study approach (Yin, 1994) was employed to identify and link these symptoms. Results of the case analyses provide further insight into distinguishing between depression and dementia in older persons with Down syndrome.

Svanelov, E.
An observation study of power practices and participation in group homes for people with intellectual disability.
Disability and Society, 2020, 35(9), 1419-1440. https://doi.org/10.1080/09687599.2019.1691978
Abstract: This study explored how participation constitutes and is constituted by practices of power in group homes for people with intellectual disability. The study used disciplinary power as theoretical perspective and was based on 50 h of observation in two group homes with a total of 15 residents. The analysis identifies practices of power and their relationship to individual agency and participation. The results show that institutional structures construct practices of power that define codes of conduct for the group home residents and their possibility for participation. This study offers implications for the daily lives of residents in group homes for people with intellectual disability.

te Boekhorst, S., Depla, M.F.I.A., de Lange, J., Pot, A.M. & Eefsting, J.A.
The effects of group living homes for older people with dementia: A comparison with traditional nursing home care.
International Journal of Geriatric Psychiatry, 2009, 24(9), 970-978. doi:10.1002/gps.2205.
Abstract: [Non-ID population] The aim of this study was to investigate the effects of group living homes on quality of life and functioning of people with dementia. The study had a quasi-experimental design with a baseline measurement on admission and an effect measurement six months later. Participants were 67 residents in 19 group living homes and 97 residents in seven traditional nursing homes. DQOL and QUALIDEM measured quality of life, functional status was examined with MMSE, IDDD, RMBPC, NPI-Q and RISE from RAI. Use of psychotropic drugs and physical restraints was also assessed. Linear and logistic regression analyses analyzed the data. After adjustment for differences in baseline characteristics, residents of group living homes needed less help with ADL and were more socially engaged. There were no differences in behavioral problems or cognitive status. Also after adjusting, two of the 12 quality of life subscales differed between the groups. Residents of group living homes had more sense of aesthetics and had more to do. While there were no differences in prescription of psychotropic drugs, residents of group living homes had less physical restraints. Group living homes had some beneficial effects on its residents, but traditional nursing homes performed well as well. Possible study limitations included the baseline differences between the study groups and the use of different informants on T0 and T1. Future nursing home care may very well be a combination of the best group living care and traditional nursing home care have to offer.

te Boekhorst, S., Willemse, B., Depla, F.I.A., Eefsting, J.A., & Pot, A.M.
Working in group living homes for older people with dementia: The effects on job satisfaction and burnpout and the role of job characteristics
International Psychogeriatrics, 2008, 20(5), 927-940. doi:10.1017/S1041610208007291. Epub 2008 May 19.
Abstract: [Non-ID population] Group living homes are a fast-growing form of nursing home care for older people with dementia. This study seeks to determine the differences in job characteristics of nursing staff in group living homes and their influence on well-being. We examined the Job Demand Control Support (JDCS) model in relation to 183 professional caregivers in group living homes and 197 professional caregivers in traditional nursing homes. Multilevel linear regression analysis was used to study the mediator effect of the three job characteristics of the JDCS-model (demands, control and social support) on job satisfaction and three components of burnout (emotional exhaustion, depersonalization and decreased personal accomplishment). Demands were lower in group living homes, while control and social support from co-workers were higher in this setting. Likewise, job satisfaction was higher and burnout was lower in group living homes. Analysis of the mediator effects showed that job satisfaction was fully mediated by all three psychosocial job characteristics, as was emotional exhaustion. Depersonalization was also fully mediated, but only by control and social support. Decreased personal accomplishment was partially mediated, again only by job characteristics, control and support. This study indicates that working in a group living home instead of a traditional nursing home has a beneficial effect on the well-being of nursing staff, largely because of a positive difference in psychosocial job characteristics.

Taggert, L., Truesdale-Kennedy, M., Ryan, A., McConkey, R.
Examining the support needs of ageing family carers in developing future plans for a relative with an intellectual disability
Journal of Intellectual Disabilities, 2012 Sep;16(3):217-234. doi: 10.1177/1744629512456465.
Abstract: Planning for the future care of adults with an intellectual disability after the main family carer ceases their care, continues to be a sensitive and difficult time posing challenges for service providers internationally. Limited research has been undertaken on this topic because until recently, people with intellectual disability usually pre-deceased their parents. This study examined ageing carers' preferences for future care and the support systems required to make such future plans. The study was conducted in one region of the United Kingdom with a high proportion of family carers. A mixed methods design was employed. In Stage 1, a structured questionnaire was used to collate information on the health, caregiving demands and future planning preferences of 112 parent and sibling carers; aged 60-94 years. In Stage 2, 19 in-depth semistructured interviews were undertaken with a sample of carers to explore a range of issues around future planning. Over half of the carers were lone carers, mainly female, with many reporting a wide range of health problems. A third of these carers reported that their caregiving resulted in high levels of anxiety. The main preference of the carers was for the

person to remain in the family home, with either the family and/or paid staff to support them. A minority of parent carers preferred the person to move into the home of a sibling, although some favored the person moving to a residential facility with other people with intellectual disabilities. The majority of carers did not want their relative to move into an older people's residential/nursing facility. In the qualitative data, four main themes were identified around future planning: unremitting apprehension, the extent of planning, obstacles encountered and solutions for future planning. Avoidance, lack of guidance and a lack of appropriate residential provision were cited as obstacles to making future plans compounded by the emotional upset experienced by carers in thinking about the future. Findings of this study clearly identify the emotional, informational and practical supports required by these ageing family carers. These findings have national and international relevance in influencing how governments and service providers support parent and sibling carers to proactively plan for the future, and in the development of both in-home and out-of-home options when a family carer can no longer provide care. This is more urgent than ever given the growing numbers of older persons with intellectual disabilities in future decades.

Takenoshita, S., Terada, S., Kuwano, R., Inoue, T., Kurozumi, T., Choju, A., Suemitsu, S., & Yamada, N.
Validation of the Japanese version of the Dementia Screening Questionnaire for Individuals with Intellectual Disabilities.
Journal of Intellectual Disability Research, 2020 Dec; 64(12), 970-979. doi: 10.1111/jir.12788. Epub 2020 Oct 5.
Abstract: Dementia in people with intellectual disabilities (IDs) is difficult to detect because of preexisting cognitive deficits. An effective screening method is required. The Dementia Screening Questionnaire for Individuals with Intellectual Disabilities (DSQIID) was developed as an observer rating tool to screen dementia in people with ID. The aim of this study was to verify the screening accuracy of the DSQIID for Japanese people with ID. Four-hundred ninety-three subjects with ID participated in this study. Caregivers who had observed the participants for more than 2 years scored the Japanese version of the DSQIID (DSQIID-J) of the participants. Three doctors examined participants directly and diagnosed dementia using the Diagnostic and Statistical Manual of Mental Disorders, Fifth Edition criteria. To identify the key screening items that predict dementia, the specificities of a single and pairs of items with 100% sensitivity were evaluated relative to the dementia diagnosis. Of 493 participants, 34 were people with Down syndrome (DS), and 459 were people without DS. Seventeen participants were diagnosed with dementia. The suitable cut-off score of the DSQIID-J was 10/11 (sensitivity 100% and specificity 96.8%) for screening dementia. The inter-rater reliability, test-retest reliability and internal consistency of the DSQIID-J were excellent. Regarding key items, there was no single item with 100% sensitivity, and the best two-item combination was the pair of 'Cannot dress without help' and 'Walks slower' (sensitivity 100% and specificity 93.5%). We identified several important question items of the DSQIID-J related to the diagnosis of dementia in people with ID. The DSQIID-J is a useful screening tool for dementia in adults with ID.

Takenoshita, S., Terada, S., Kuwano, R., Inoue, T., Cyoju, A., Suemitsu, S., & Yamada, N.
Prevalence of dementia in people with intellectual disabilities: Cross-sectional study
International Journal of Geriatric Psychiatry, 2020 (April), 35(4), 314-422. https://doi.org/10.1002/gps.5258
Abstract: There are only a few studies of the prevalence of dementia in people with intellectual disability (ID) without Down syndrome (DS), and there is a large difference in the prevalences between reported studies. Moreover, the prevalence of mild cognitive impairment (MCI) in ID has not been reported. We aimed to evaluate the prevalence of dementia in adults of all ages and the prevalence of MCI in people with ID. Furthermore, we tried to clarify the differences depending on the various diagnostic criteria. The survey included 493 adults with ID at 28 facilities in Japan. The caregivers answered a questionnaire, and physicians directly examined the participants who were suspected of cognitive decline. Dementia and MCI were diagnosed according to ICD-10, DC-LD, and DSM-5 criteria. The prevalence of dementia was 0.8% for the 45 to 54 years old group, 3.5% for the 55 to 64 years old group, and 13.9% for the 65 to 74 years old group in people with ID without DS. The prevalence of MCI was 3.1% for patients 45 to 54, 3.5% for patients 55 to 64, and 2.8% for patients 65 to 74 with ID without DS. DSM-5 was the most inclusive in diagnosing dementia and MCI in people with ID. People with ID without DS may develop dementia and MCI at an earlier age and higher rate than the general population. Among the diagnostic criteria, DSM-5 was the most useful for diagnosing their cognitive impairment.

Takenoshita, S., Kuwano, R., Inoue, T., Kurozumi, T., Terada, S., Yamada, N., & Suemitsu, S.
Prevalence of dementia in people with intellectual disabilities without Down syndrome in Japan
Journal of Applied Research in Intellectual Disabilities, 2021, 34(5), 1305. doi: 10.1002/gps.5258.
Abstract:There are few large-scale studies of the prevalence of dementia in people with intellectual disabilitywho do not have Down syndrome, although Down syndrome is well known to be related with Alzheimer's disease. We investigated 1831 adults with intellectual disabilitybut without Down syndrome who were residents of a facility for people with intellectual disabilityin Japan. The caregivers answered a questionnaire, and physicians directly examined the participants suspected of cognitive decline. Of the 1831 patients, 118 were diagnosed with dementia and 51 with mild cognitive impairment (MCI). The prevalence of dementia was 5.6% for the 55 to 64 age group, 13.2% for the 65 to 74 age group, 22.2% for the 75 to 84 age group, and 42.3% for the 85 to 94 age group. The prevalence of MCI was 2.1% for the 55 to 64 age group, 7.4% for the 65 to 74 age group, and 11.5% for the 85 to 94 age group. Sixty-five patients (55.1%) had not been diagnosed with dementia before the survey. People with intellectual disability without Down syndrome may develop dementia at an earlier age than those without intellectual disability. Our nationwide survey suggested there are many undiagnosed dementia patients in the community of people with intellectual disability.

Takenoshita, S., Terada, S., Inoue, T., Kurozumi, T., Yamada, N., Kuwano, R., & Suemitsu, S.
Prevalence and modifiable risk factors for dementia in persons with intellectual disabilities.
Alzheimer's Research & Therapy, 2023 Jul 18, 15(1), 125. doi: 10.1186/s13195-023-01270-1.
Abstract: : People with intellectual disability (ID) without Down syndrome (DS) are presumed to be at higher risk of developing dementia due to their lower baseline cognitive reserve. We aimed to determine the prevalence of dementia in people with ID without DS and to identify risk factors of dementia. This was a cross-sectional survey and multicenter study in Japan. Adults with ID without DS residing in the facilities were included. Caregivers of all participants were interviewed by medical specialists, and participants suspected of having cognitive decline were examined directly. ICD-10 criteria for dementia, DC-LD criteria for dementia, and DSM-5 criteria for neurocognitive disorders were used to diagnose dementia. The severity of ID, educational history, and comorbidities were compared by dividing the groups into those with and without dementia. A total of 1831 participants were included; 118/1831 (6.44%) were diagnosed with dementia. The prevalence of dementia for each age group was 8.8%, 60-64 years; 9.0%, 65-69 years; 19.6%, 70-74 years; and 19.4%, 75-79 years. Age, severity of ID, duration of education, hypertension, depression, stroke, and traumatic brain injury were significantly associated with the presence of dementia. Although the prevalence of dementia in people with ID without DS was found to be higher at a younger age than in the general population, the results of this study suggested that adequate education, prevention of head trauma and stroke, and treatments of hypertension and depression may reduce the risk of dementia. These may be potentially important modifiable risk factors for the prevention of dementia in these people.

Tarasova, D., Rösner, P., Deb, S., & Sappok, T.
Validation of the German version of the DSQIID in adults with intellectual disabilities.
Research in Developmental Disabilities, 2024 May, 148, 04721. https://doi.org/10.1016/j.ridd.2024.104721.
Abstract: An observer-rated screening questionnaire for dementia for people with intellectual disabilities (ID), DSQIID, was developed in the UK. So far, the German version has not yet been validated in adults with ID. We validated a German version of DSQIID (DSQIID-G) among adults with ID attending a German clinic. DSQIID-G was completed by the caregivers of 104 adults with ID at baseline (T1), 94 at six months (T2) and 83 at 12 months (T3). A Receiver Operating Curve (ROC) was used to determine the total DSQIID-G cutoff score for the best fit between sensitivity and specificity. Sixteen of the 104 participants at T1 (15%) received a diagnosis of dementia. At T1, the scores among the non-dementia group ranged from 0 to 33 (mean: 6.7; SD: 7.65), and the dementia group ranged from 3 to 43 (mean: 22.12; SD: 11.6). The intergroup difference was statistically significant (W: 158; $p < .001$) (AUC:.89). A total score of 9 provided the best fit between sensitivity (.94) and specificity (.72). DSQIID-G total score can discriminate between dementia and non-dementia cases in adults with ID. A lower cutoff score with a higher sensitivity is desirable for a screening instrument.

Temple, V., & Konstantareas, M.M.
A comparison of the behavioural and emotional characteristics of Alzheimer's dementia in individuals with and without Down syndrome.
Canadian Journal of Aging, 2005, 24(2),179-190. doi: 10.1353/cja.2005.0071.
Abstract: The behavioral and emotional changes associated with Alzheimer's disease (AD) are compared for individuals with Down syndrome and AD and individuals with AD from the general population (AD-only). The primary caregivers of 30 people with Down syndrome and AD and 30 people with AD-only completed the BEHAVE-AD and the Apathy subscale of the CERAD. As well, behavioral observations at adult day programs were undertaken with selected participants (n=26). The Down syndrome group experienced fewer delusions and had lower total scores on the BEHAVE-AD, indicating fewer problem behaviors overall. Day program observations suggested that the AD-only group were more likely to be sedentary and observe the activities of others, while the Down syndrome group were more physically active. Improving our understanding of the similarities and differences between these two groups may help facilitate the integration of individuals with Down syndrome into adult day programs for the general population.

Temple, V., Jozsvai, E., Konstantareas, M.M., & Hewitt, T.A.
Alzheimer dementia in Down's syndrome: the relevance of cognitive ability.
Journal of Intellectual Disability Research, 2001, 45(1), 47-55. doi:10.1046/j.1365-2788.2001.00299.x.
Abstract: More years of education have been found to be associated with a lower rate of Alzheimer disease (AD) in individuals without intellectual disability. It has been proposed that education reflects greater 'synaptic reserve' and that greater synaptic reserve may defer the development of AD. The present study compared individuals with Down's syndrome (DS) who were found to have symptoms of dementia with those who remained symptom-free to determine if the two groups differed in their level of education, employment, recreational activities, years in an institution or overall level of cognitive functioning. Thirty-five adults with DS aged between 29 and 67 years were assessed. The participants were recruited from a community health facility and included individuals with a wide range of ability levels. Neuropsychological testing, caregiver report and the Dementia Scale for Down Syndrome (Gedye 1995) were used to identify decline in participants over periods of 6 months to 3 years. After the effect of age was statistically removed, multiple regression analyses revealed that level of cognitive functioning was significantly associated with decline such that a higher level of cognitive functioning predicted less decline. None of the environmental variables (i.e. educational level, years in an institution and employment) were directly associated with decline; however, a post hoc regression using level of cognitive functioning as the outcome variable revealed that level of cognitive functioning itself was associated with these environmental variables. A higher level of cognitive functioning was associated with fewer cases of dementia in individuals with DS, and level of cognitive functioning appears to be associated with environmental factors such as level of education, years in an institution and employment. The present findings suggest that environmental interventions aimed at improving level of cognitive functioning may also be useful in deferring the onset of dementia.

Thalen, M., Volkers, K.M., van Oorsouw, W.M.W.J.,& Embregts, P.J.C.M.
Psychosocial interventions for older people with intellectual disabilities and the role of support staff: A systematic review.
Journal of Applied Research in Intellectual Disabilities, 2022 Mar, 35(2), 312-337. doi: 10.1111/jar.12953. Epub 2021 Nov 15.
Abstract: The life expectancy of people with intellectual disabilities has increased. The implications of ageing have resulted in changes in their support needs and challenges to support staff. Access to evidence-based strategies for support staff providing care to elderly with intellectual disabilities remains scarce. A systematic review was conducted to provide an overview of available psychosocial interventions. Four databases were searched and assessed according to the PRISMA guidelines. A narrative, integrative method of analysis was conducted to synthesize quantitative and qualitative data. The 36 studies included in the review reported on interventions aimed at either identifying and meeting the needs or perceptions of older individuals or at improving their behavior and skills. Furthermore, the role of support staff in the implementation of interventions was either active, assisting or undefined. This overview of studies could contribute to the existing body of knowledge and help to optimize psychosocial support for a growing population.

Thase, M.E., Tigner, R., Smeltzer, D.J., & Liss. L
Age-related neuropsychological deficits in Down's syndrome
Biological Psychiatry, 1984 Apr; 19(4), 571-585. https://pubmed.ncbi.nlm.nih.gov/6234031/
Abstract: Down's syndrome (DS) has been suggested as a high-risk condition for Dementia Alzheimer's type (DAT). In the present study, neuropsychological variables were assessed in 165 DS subjects and 163 matched controls with intellectual disability. Overall, DS subjects had lower scores for orientation, digit span, visual memory, object-naming, and general knowledge, as well as more "released" reflexes. Impairments were most evident in DS subjects greater than 50 years old. These findings provide further support for an association between aging and DAT in Down's syndrome. Methodological issues and areas for future research are discussed.

The Arc
Developmental disabilities and Alzheimer disease: what you should know.
43 pp.
Silver Spring, Maryland: The Arc of the United States [1010 Wayne Avenue, Suite 650, Silver Spring, MD 20910 – www.TheArc.org] (1995)
Abstract: A booklet covering some of the fundamentals concerning adults with intellectual disabilities and Alzheimer's disease including what is Alzheimer's disease, its course and outcome, diagnostic suggestions, care considerations, and how to obtain assistance. Contains resource list and glossary.

Thompson, S. B.
Assessing dementia in Down's syndrome: A comparison of five tests.
Clinical Gerontologist, 2001, 23(3-4), 3-19. doi: 10.1300/J018v23n03_02
Abstract: Five standardized psychological tools are critically evaluated for their use in assessing dementia in people with Down's syndrome. The difficulties in assessing dementia in people who have Down's syndrome are discussed together with some recommendations for the use of these potential assessment tools.

Thompson, D.J., Ryrie, I., & Wright, S.
People with intellectual disabilities living in generic residential services for older people in the UK
Journal of Applied Research in Intellectual Disabilities, 2004, 17(2), 101-108. https://doi.org/10.1111/j.1360-2322.2004.00187.x
Abstract: As part of a UK program of work focusing on older people with ID, the circumstances of those who reside in generic services for older people were investigated. Some 215 people with ID were identified living in 150 homes. They were significantly younger than other residents and were placed in these homes more because of organizational change or the aging/death of family carers, rather than due to their own needs. Of the residents, 24 adults had Down syndrome, 8 of whom were noted to have dementia. Of the 215, 45 had dementia. Average age of people with DS upon entry was 60 and those remaining at the homes was about 65.

Thorpe, L., Pahwa, P., Bennett, V., Kirk, A., & Nanson, J.
Clinical predictors of mortality in adults with intellectual disabilities with and without Down syndrome
Current Gerontology and Geriatrics Research, 2012, 943890. doi: 10.1155/2012/943890. Epub 2012 May 16.
Abstract: Mood, baseline functioning, and cognitive abilities as well as psychotropic medications may contribute to mortality in adults with and without Down Syndrome (DS). Methods. Population-based (nonclinical), community-dwelling adults with intellectual disabilities (IDs) were recruited between 1995 and 2000, assessed individually for 1-4 times, and then followed by yearly phone calls. Results. 360 participants (116 with DS and 244 without DS) were followed for an average of 12.9 years (range 0-16.1 years as of July 2011). 108 people died during the course of the followup, 65 males (31.9% of all male participants) and 43 females (27.6% of all female participants). Cox proportional hazards modeling showed that baseline practical skills, seizures, anticonvulsant use, depressive symptoms, and cognitive decline over the first six years all significantly contributed to mortality, as did a diagnosis of DS, male gender, and higher age at study entry. Analysis stratified by DS showed interesting differences in mortality predictors. Conclusion. Although adults with DS have had considerable improvements in life expectancy over time, they are still disadvantaged compared to adults with ID without DS. Recognition of potentially modifiable factors such as depression may decrease this risk.

Tom, S.E., Hubbard, R.A., Crane, P.K., Haneuse, S.J., Bowen, J., McCormick, W.C., McCurry, S., & Larson, E.B.
Characterization of dementia and Alzheimer's disease in an older population: updated incidence and life expectancy with and without dementia
American Journal of Public Health, 2015 Feb,105(2), 408-413. doi: 10.2105/AJPH.2014.301935.
Abstract: Authors estimated dementia incidence rates, life expectancies with and without dementia, and percentage of total life expectancy without dementia. Authors studied 3605 members of Group Health (Seattle, WA) aged 65 years or older who did not have dementia at enrollment to the Adult Changes in Thought study between 1994 and 2008. We estimated **incidence rates of Alzheimer's disease and dementia**, as well as **life expectancies** with and without dementia, defined as the average number of years one is expected to live with and without dementia, and percentage of total life expectancy without dementia. Dementia incidence increased through ages 85 to 89 years (74.2 cases per 1000 person-years) and 90 years or older (105 cases per 1000 person-years). Life expectancy without dementia and percentage of total life expectancy without dementia decreased with age. Life expectancy with dementia was longer in women and people with at least a college degree. Percentage of total life expectancy without dementia was greater in younger age groups, men, and those with more education. Efforts to delay onset of dementia, if successful, would likely benefit older adults of all ages.

Tookey, S.A., Greaves, C.V., Rohrer, J.D., & Stott, J.
Specific support needs and experiences of carers of people with frontotemporal dementia: A systematic review.
Dementia (London). 2021 Nov;20(8):3032-3054. doi: 10.1177/14713012211022982. Epub 2021 Jun 11.
Abstract: Frontotemporal dementia (FTD) is one of the most common types of dementia in persons younger than 65 years of age. Diagnosis is often delayed due to slow, gradual decline and misinterpretation of 'non-typical' dementia symptoms. Informal carers of people with FTD experience greater levels of overall burden than carers of people with other forms of dementia. The aim of this systematic review was to describe the subjective experience of being an informal carer of a person with FTD and to identify the specific needs, coping strategies and helpful support resources of this carer population. Four electronic databases were used to search for published literature presenting experiences of carers of people with FTD between January 2003 and July 2019. Search strategy followed PRISMA guidelines. Findings were analyzed using framework analysis, employing five stages of analysis to develop a coding index and thematic framework that included key aspects of the carer experience, which were grouped into themes and presented in a narrative format. Some 1213 articles were identified in total. Twelve studies were included in the final synthesis of the review. Six themes were identified: 'Challenging road to and receipt of diagnosis', 'relationship change and loss', 'challenging experiences in caring', 'positive experiences and resilience', 'coping' and 'support needs'. **Findings highlight an increased need for carers of people with FTD to receive support during the pre-diagnostic stage, including support to manage symptoms**. Further research should explore relationship changes and loss amongst carers to inform approaches for carer support. In conclusion, the lack of knowledge and unique needs of carers highlight the importance of public awareness campaigns and healthcare professional education to support carers with FTD symptom impact.

Torr, J., & Davis, R.
Ageing and mental health problems in people with intellectual disability
Current Opinions in Psychiatry, 2007, 20(5), 467-471. doi: 10.1097/YCO.0b013e328278520d.
Abstract: Increasing numbers of people with intellectual disability are now living well into old age. This paper will review the recent literature pertaining to the mental health of older people with intellectual disability. Overall, the prevalence of mental health problems is high in adults of all ages with intellectual disability. A major epidemiological study did not report sufficient detail to examine the effect of ageing on specific disorders or the differential effects of ageing and early mortality in people with Down's syndrome. At least a third of people with Down's syndrome can expect to develop Alzheimer's disease in middle age whilst for other people with intellectual disability, Alzheimer's disease is probably no more common than in the general population. Diagnosis and management of dementia is complicated by the high rates of comorbid physical and mental health problems. Overall, mental health problems in older people with intellectual disability are similar to younger people with intellectual disability, however there are more cases of dementia and physical health problems. Further research is needed to improve our understanding of the effects of ageing on the mental health and care needs of older people with intellectual disability.

Torr, J., Strydom, A., Patti, P. & Jokinen N.
Aging in Down syndrome: Morbidity and mortality.
Journal of Policy and Practice in Intellectual Disabilities, 2010, *7*(1), 70-81. https://doi.org/10.1111/j.1741-1130.2010.00249.x
Abstract: The life expectancy of adults with Down syndrome has increased dramatically over the last 30 years, leading to increasing numbers of adults with Down syndrome now living into middle and old age. Early-onset dementia of the Alzheimer type is highly prevalent in adults with Down syndrome in the sixth decade, and this has overshadowed other important conditions related to aging among adults with Down syndrome. The authors' aim was to update and summarize current knowledge on these conditions, and examine causes of morbidity and mortality in older people with Down syndrome by conducting a systematic review of the published literature for the period: 1993–2008. They

reviewed English-language literature drawn from searches in the electronic databases Medline, CINAHL, and PsycINFO, as well as supplementary historical papers. The authors conclude that functional decline in older adults with Down syndrome cannot be assumed to be due only to dementia of the Alzheimer type (which is not inevitable in all adults with Down syndrome). Functional decline may be the result from a range of disorders, especially sensory and musculoskeletal impairments. Given the high rates of early-onset age-related disorders among adults with Down syndrome, programmatic screening, monitoring, and preventive interventions are required to limit secondary disabilities and premature mortality. With respect to assessment and treatment, in the absence of specialist disability physicians, geriatricians have a role to play.

Trumble, S.
Dementia in Down syndrome. Untangling the threads.
Australian Family Physician, 1999 Jan, 28(1), :49-53.
https://pubmed.ncbi.nlm.nih.gov/9988915/
Abstract: Despite frequent neuropathological findings, the prevalence of dementia in people with Down syndrome is thought to be no higher than in the general population. The age of onset, however, is approximately two decades earlier. The most important factor in the general practitioner's assessment of a person with this syndrome who appears to be dementing is the exclusion of medical problems that may be masquerading as Alzheimer disease. This article presents an efficient system for the clinical review of people with Down syndrome who are declining in function. The system is also adaptable for use with people who do not have this syndrome.

Tsang, W., Oliver, D., & Triantafyllopoulou, P.
Quality of life measurement tools for people with dementia and intellectual disabilities: A systematic review.
Journal of Applied Research in Intellectual Disabilities, 2023 Jan, 36(1), 28-38.
doi: 10.1111/jar.13050.
Abstract: Adults with intellectual disabilities are an at-risk group of developing dementia. In the absence of a cure for dementia, emphasis on treatment is the promotion of Quality of life (QoL). The aim of this review is to identify and describe QoL tools for people with intellectual disabilities and dementia. A systematic review was carried out using 10 databases and papers from up to March year 2021. Two instruments were identified and examined. The QoL in late-stage dementia, which showed evidence of good levels of internal consistency, intra-rater reliability, test-retest reliability, and convergent validity. The Dementia Quality of Life - proxy was also used; however, its psychometric properties have yet to be studied within the intellectual disabilities population. Due to the degenerative nature of dementia involving cognitive impairment such as memory and language difficulties, there can be significant barriers in capturing the subjective experience of QoL in a person with dementia. Therefore, it may not be possible to capture the patient's perceived QoL from self-report measures. Interpretation of concepts can be problematic, especially when life situation of the respondent is vastly different to that of a researcher. Also, the conceptualisation of QoL can vary in different stages of dementia in the general population, such that in the early stages of dementia enjoyment of daily activities is a relevant domain for QoL, but may no longer seem to be applicable in severe or advanced dementia. It is recommended instruments should be developed and psychometrically tested specifically for adults with intellectual disabilities and dementia to help inform policy makers, measure outcomes of interventions and personal outcomes.

Tse, M.M., Kwan, R.Y, & Lau, J.L.
Ageing in individuals with intellectual disability: issues and concerns in Hong Kong
Hong Kong Medical Journal, 2018, Feb, 24(1), 68-72.
doi: 10.12809/hkmj166302. Epub 2018 Jan 12.
Abstract: The increasing longevity of people with intellectual disability is testimony to the positive developments occurring in medical intervention. Nonetheless, early-onset age-related issues and concerns cause deterioration of their overall wellbeing. This paper aimed to explore the issues and concerns about individuals with intellectual disability as they age. Articles that discussed people older than 30 years with an intellectual disability and those that identified ageing health issues and concerns were included. Only studies reported in English from 1996 to 2016 were included. We searched PubMed, Google Scholar, and Science Direct using the terms 'intellectual disability', 'ageing', 'cognitive impairment', 'health', and 'screening'. Apart from the early onset of age-related health problems, **dementia** is more likely to develop by the age of 40 years in individuals with intellectual disability. Geriatric services to people with intellectual disability, however, are only available for those aged 60 years and older. Cognitive instruments used for the general population are not suitable for people with intellectual disability because of floor effects. In Hong Kong, the Chinese version of the Dementia Screening Questionnaire for Individuals with Intellectual Disabilities is the only validated instrument for people with intellectual disability. The use of appropriate measurement tools to monitor the progression of age-related conditions in individuals with intellectual disability is of great value. Longitudinal assessment of cognition and function in people with intellectual disability is vital to enable early detection of significant deterioration. This allows for therapeutic intervention before substantial damage to the brain occurs such as dementia that hastens cognitive and functional decline.

Tsiouris, J.A., & Patti, P.J.
Drug treatment of depression associated with dementia or presented as "pseudodementia" in older adults with Down syndrome.
Journal of Applied Research in Developmental Disabilities, 1997, *10* (4), 312-322,
https://doi.org/10.1111/j.1468-3148.1997.tb00026.x
Abstract: The response to antidepressant drugs, mainly the selective serotonin reuptake inhibitors (SSRIs), was evaluated in adults with intellectual disability (ID) and Down syndrome (DS) who presented with depression and decline in activities of daily living (ADL) skills. Among other patients with ID referred to a specialised clinic for diagnostic work-up, 37 adults with DS over the age of 40 and a mean age of 51.4 years were evaluated and 34 were followed-up. Depression associated with dementia was diagnosed in 16 cases, and depression presented as functional decline 'pseudodementia' was found in 4 cases. Recommendations for treatment with antidepressants were followed in 10 cases with a marked improvement in functioning compared to a rapid decline in 10 cases where treatment was refused. Treatment with the SSRI antidepressant drugs resulted in improved quality of life, differentiated 'pseudodementia' from dementia, and possibly delayed the dementing process in adults with DS and presentation of depression associated with dementia.

Tsiouris, J.A., Patti, P.J., & Flory, M.J.
Effects of antidepressants on longevity and dementia onset among adults with Down syndrome: a retrospective study
Journal of Clinical Psychiatry, 2014, 75(7), 731-737.
doi:10.4088/JCP.13m08562.
Abstract: To investigate the effects of antidepressants on longevity, age at dementia onset, and survival after onset among adults with Down syndrome, controlling for late-onset seizures, trisomy 21 mosaicism, and cholinesterase inhibitor use. The charts of 357 adults with Down syndrome (mean age at first visit = 46.3 years, SD = 9.0) evaluated in a metropolitan diagnostic and research clinic between 1990 and 2008 were reviewed. Seventeen patients had trisomy 21 mosaicism; 155 patients were diagnosed with depressive disorders using DSM-III-R and IV criteria, 78 of whom received antidepressants for over 90 days. Of 160 patients who developed dementia, **the estimated mean age at onset was 52.8 years**. Fifty-six patients (demented and nondemented) had late-onset seizures. Longevity and age at estimated onset among those receiving and not receiving antidepressants were compared. Cox proportional hazards models examined risks for dementia onset and death. The mean age at dementia onset among those receiving antidepressants before onset was 53.75 years versus 52.44 years among others. Proportional hazards models showed a significant delay of onset among those taking antidepressants (hazard ratio = 0.69; 95% CI,

0.48–0.98; P = .038). Mean age at death or at end of study for those receiving antidepressants was 54.71 years; among others, it was 52.60 years (hazard ratio = 0.63; 95% CI, 0.42–0.94; P = .024). Among the 35 adults with late-onset seizures and dementia who died, mean survival after seizure onset was 4.23 years. The findings in this retrospective study revealed that antidepressant use was associated with delayed dementia onset and increased longevity in adults with Down syndrome; mean survival after late-onset seizures was longer than previously reported. Further studies, however, are needed to confirm these associations, optimally in a clinical trial to confirm causality.

Tsiouris, J.A., Patti, P.J., Tipu, O. & Raguthu, S..
Adverse effects of phenytoin given for late-onset seizures in adults with Down syndrome.
Neurology, 2002, Sept 10, 59(5), 779-780. doi: 10.1212/wnl.59.5.779.
Summary (no abstract provided): A brief report that indicates the adverse effects of therapeutic levels of phenytoin and the improvement observed when phenytoin was replaced with other antiepileptics in 17 adults with DS, Alzheimer disease (AD) and late-onset seizures (LOS). The reported deterioration in the patients' condition was found to be due to the adverse effects from phenytoin and not to AD. It was suggested that practitioners avoid prescribing phenytoin to treat LOS in persons with DS and AD. If phenytoin is already prescribed, it should be replaced with another anticonvulsive agent.

Tsou, A.Y., Bulova, P., Capone, G., Chicoine, B., Gelaro, B., Odell Harville, T., Martin, B.A., McGuire, D.E., McKelvey, K.D. Peterson, M., Tyler, C., Wells, M., Whitten, M.S. & the Global Down Syndrome Foundation Medical Care Guidelines for Adults with Down Syndrome Workgroup
Medical care of adults with Down syndrome: A clinical guideline
JAMA, 2020, 324(15), 1543-1556. doi:10.1001/jama.2020.17024
Abstract: Down syndrome is the most common chromosomal condition, and average life expectancy has increased substantially, from 25 years in 1983 to 60 years in 2020. Despite the unique clinical comorbidities among adults with Down syndrome, there are no clinical guidelines for the care of these patients.
To develop an evidence-based clinical practice guideline for adults with Down syndrome. The Global Down Syndrome Foundation Medical Care Guidelines for Adults with Down Syndrome Workgroup (n=13) developed 10 Population/ Intervention/ Comparison/Outcome (PICO) questions for adults with Down syndrome addressing multiple clinical areas including mental health (2 questions), dementia, screening or treatment of diabetes, cardiovascular disease, obesity, osteoporosis, atlantoaxial instability, thyroid disease, and celiac disease. These questions guided the literature search in MEDLINE, EMBASE, PubMed, PsychINFO, Cochrane Library, and the TRIP Database, searched from January 1, 2000, to February 26, 2018, with an updated search through August 6, 2020. Using the GRADE (Grading of Recommendations, Assessment, Development, and Evaluation) methodology and the Evidence-to-Decision framework, in January 2019, the 13-member Workgroup and 16 additional clinical and scientific experts, nurses, patient representatives, and a methodologist developed clinical recommendations. A statement of good practice was made when there was a high level of certainty that the recommendation would do more good than harm, but there was little direct evidence. From 11295 literature citations associated with 10 PICO questions, 20 relevant studies were identified. An updated search identified 2 additional studies, for a total of 22 included studies (3 systematic reviews, 19 primary studies), which were reviewed and synthesized. Based on this analysis, 14 recommendations and 4 statements of good practice were developed. Overall, the evidence base was limited. Only 1 strong recommendation was formulated: screening for Alzheimer-type dementia starting at age 40 years. Four recommendations (managing risk factors for cardiovascular disease and stroke prevention, screening for obesity, and evaluation for secondary causes of osteoporosis) agreed with existing guidance for individuals without Down syndrome. Two recommendations for diabetes screening recommend earlier initiation of screening and at shorter intervals given the high prevalence and earlier onset in adults with Down syndrome. These evidence-based clinical guidelines provide recommendations to support primary care of adults with Down syndrome. The lack of high-quality evidence limits the strength of the recommendations and highlights the need for additional research.

Tuffrey-Wijne I.
How to Break Bad News to People with Intellectual Disabilities: A Guide for Carers and Professionals.
Jessica Kingsley Publishers, London (2012).
Abstract: How to tell bad news to people with dementia? First assess the person's current framework of knowledge, then give the new chunks of information one by one, then finally check and reassess the person's knowledge. This might be useful for bereaved people with dementia to fill the gap.

Tuffrey-Wijne, I., & Watchman, K.
Breaking bad news to people with learning disabilities and dementia
Learning Disability Practice, 2015 Sept, 18(7), 16-23.
DOI:10.7748/ldp.18.7.16.e1672
Abstract: People with learning disabilities are now enjoying a longer life expectancy than ever before as a result of enhanced medical and social interventions and improved quality of life. Some, particularly individuals with Down's syndrome, are susceptible to dementia at a significantly younger age than the average age of onset in the rest of the population. Currently, there is limited guidance on how to talk to people with learning disabilities about dementia and, until such information is shared, individuals cannot be positioned as an authority on their own condition. The new model presented here suggests a way of supporting staff and families to have enabling conversations about dementia that center on the person's current situation, level of understanding and capacity.

Tuffrey-Wijne, I., & Watchman, K.
Sharing the diagnosis of dementia: Breaking bad news to people with intellectual disabilities.
In: Watchman, Karen, (ed.) Intellectual disability and dementia: research into practice. London : Jessica Kingsley Publishers. pp. 184-203. ISBN 9781849054225
Abstract: Not available - see Tuffrey-Wijne & Watchman 2015

Turky, A., Felce, D., Jones, G., & Kerr, M.
A prospective case control study of psychiatric disorders in adults with epilepsy and intellectual disability
Epilepsia, 2011 July, 52(7), 1223-1230.
https://doi.org/10.1111/j.1528-1167.2011.03044.x
Abstract: No study to date has prospectively investigated the impact of epilepsy on psychiatric disorders among adults with an intellectual disability (ID). This study aimed to determine prospectively the influence of epilepsy on the development of psychiatric disorders in adults with ID. Psychiatric symptoms were measured prospectively over a 1-year period among 45 adults with ID and active epilepsy and 45 adults with ID without epilepsy, matched on level of ID. The 1-year incidence rate (IR) of commonly occurring Axis 1 psychiatric disorders was compared with and without controlling for possible confounding factors. Total psychiatric symptom scores over the period were compared between the two groups using repeated-measures analysis of covariance. Adults with epilepsy and ID had a more than seven times increased risk for developing psychiatric disorders, particularly depression and unspecified disorders of presumed organic origin, including dementia, over a 1-year period compared to those with ID only. Comparison of the psychiatric scores showed the epilepsy group to have significantly higher unspecified disorder and depression symptom scores. The findings point to an increased risk of depression and unspecified disorders, including dementia, among adults with ID and epilepsy. Further exploration of the nature and treatment of these unspecified disorders may help the care of people with epilepsy and ID.

Tyler, C.V., & Shank, J.C.
Dementia and Down syndrome

The Journal of Family Practice, 1996, 42(6), 619-621.
https://cdn.mdedge.com/files/s3fs-public/jfp-archived-issues/1996-volume_42-43/JFP_1996-06_v42_i6_dementia-and-down-syndrome.pdf
Abstract: Case report of a 43-year old woman with Down syndrome and progressive decline over three years that was attributed to dementia of the Alzheimer's type. Authors describe the medical conditions evident during decline, whilst living with her family. Identifies typical features associated with decline for persons with Down syndrome and defines areas for concern during examinations by physicians.

Tyrrell, J.F., Cosgrave, M.P., McLaughlin, M., & Lawlor, B.A.
Dementia in an Irish population of Down's syndrome people.
Irish Journal of Psychological Medicine, 1996, 13(2), 51-54.
doi:10.1017/S0790966700002408
Abstract: To determine the prevalence of dementia in an Irish sample of Down syndrome (DS) patients and to examine the utility of a number of cognitive and functional scales in the assessment of dementia in this population. 76 DS patients diagnosed clinically (range 33-72 years; mean age 47.3 ± 8.8 years) were included in the study. The diagnosis of dementia was made on clinical grounds using DSM-III-R criteria. Cognitive and functional impairment were evaluated using the following scales; Test for Severe Impairment (TSI), Down Syndrome Mental Status Examination (DS-MSE), Daily Living Skills Questionnaire (DLSQ), and the Mental State Performance (MSP). The overall prevalence of dementia was 7.9% (95% C.I = 2.95-16.39). The presence of dementia was associated with late onset epilepsy, anticonvulsant medication, and deafness. Standard cognitive tests such as the MSP showed an early `floor' effect in this population. In contrast the TSI and DLSQ showed a satisfactory range of scores in these patients with moderate to severe intellectual disability. The low prevalence of dementia in this study may be explained by the strict conservative criteria applied in the clinical diagnosis. Prospective assessment of DS patients on a longitudinal basis using decline on scales such as the TSI and DLSQ may allow more accurate diagnosis of dementia at an earlier stage in this at-risk population.

Tyrrell, J., Cosgrave, M., McCarron, M., McPherson, J., Calvert, J., Kelly, A., McLaughlin, M., Gill, M.,& Lawlor, B.A.
Dementia in people with Down's syndrome.
International Journal of Geriatric Psychiatry, 2001, Dec, 16(12), 1168-1174.
Abstract: To determine the prevalence of dementia in an Irish sample of people with Down syndrome (DS) and to examine associated clinical characteristics of dementia in this group. Some 285 people with DS (Age 35-74 years, mean age +/- SD 46.5 +/- 8.2 years) were included in this cross-sectional study. The diagnosis of dementia was made using modified DSMIV criteria. Cognitive tests used were the Down Syndrome Mental Status Examination (DS-MSE), Test for Severe Impairment (TSI) and adaptive function was measured by the Daily Living Skills Questionnaire (DLSQ). The overall prevalence of dementia was 13.3%. The presence of dementia was associated with epilepsy, myoclonus, and head injury. The demented DS group were significantly older (n = 38, mean age 54.7 years SD +/- 7.5) than the non-demented (n = 246, mean age 45.6, SD +/- 7.3). The TSI and DLSQ had a satisfactory spread of scores without 'floor' or 'ceiling' effects in people with moderate and severe learning disability. Median scores in demented versus the non-demented groups were significantly different for each measure of function. Authors conclude that dementia had a prevalence of 13.3% and occurred at **a mean age of 54.7 years**. The combination of DLSQ score, age, and presence of epilepsy were found to predict presence of dementia.

Udell, L.
Supports in small group home settings
In M.P. Janicki & A.J. Dalton (Eds.), Dementia, Aging, and Intellectual Disabilities.
pp. 316-329
Philadelphia: Brunner-Mazel (1999)
Abstract: This book chapter covers what organizations that provide residential supports to adults with an intellectual disability need to consider in terms of planning and implementing program changes. Covered are areas that examine the nature of dementia and its possible impact on service provision, Its particular focus is on how agencies that decide to support people with dementia in small group home settings can accommodate their organizational and operational structure and offers insight ion the perspectives and questions that agencies need to consider. Suggestions are offered on how to address some of the difficulties that organizations will encounter.

University of Maryland School of Medicine
Hi Buddy... The developmentally delayed individual with Alzheimer disease
19 minutes
VideoPress, the University of Maryland School of Medicine [100 North Greene Street, Suite 300, Baltimore, Maryland USA (1 800 328 7450; fax: 1 410 706 8471; www.videopress.org)]
Abstract: Video on the subject of Alzheimer's disease and adults with developmental disabilities.

University of Stirling
Building networks - Conference on learning disabilities and dementia
58 pp.
Dementia Services Development Centre, Department of Applied Social Science, Faculty of Human Sciences, University of Stirling, Stirling, Scotland FH9 4LA (2000).
Abstract: Proceedings of conference on community dementia care and people with intellectual disabilities held in Dunblane, Scotland (November 11, 1999). The report summarizes the main points made by the numerous speakers at the conference. The conference highlighted the need for wider awareness among managers and service personnel of the need for (and for resources and developing expertise on) training staff in residential and home support services on responding to the needs of people with intellectual disabilities who have dementia. The 16 papers range from the theoretical to the practical.

Urv, T.K., Zigman, W.B., & Silverman,W.
Psychiatric symptoms in adults with down syndrome and Alzheimer's disease.
American Journal on Intellectual and Developmental Disabilities, 2010, 115(4), 265-276. doi: 10.1352/1944-7558-115.4.265.
Abstract: Changes in psychiatric symptoms related to specific stages of dementia were investigated in 224 adults 45 years of age or older with Down syndrome. Findings indicate that psychiatric symptoms are a prevalent feature of dementia in the population with Down syndrome and that clinical presentation is qualitatively similar to that seen in Alzheimer's disease within the general population. Psychiatric symptoms related to Alzheimer's disease vary by the type of behavior and stage of dementia, but do not seem to be influenced by sex or level of premorbid intellectual impairment. Some psychiatric symptoms may be early indicators of Alzheimer's disease and may appear prior to substantial changes in daily functioning. Improvements in understanding the progression of dementia in individuals with Down syndrome may lead to improved diagnosis and treatment.

van Hoof, J., & Kort, H.
Supportive living environments: A first concept of a dwelling designed for older adults with dementia.
Dementia, 2009, 8, 293 - 316. doi:10.1177/1471301209103276
Abstract: The vast majority of older adults want to remain living independently at home, with or without a sufficient amount of professional home care, even when overall health is starting to decline. The ageing of society and the increase in the number of very old elders goes together with an increase in the number of people with dementia. About two thirds of the diagnosed people in the Netherlands live at home. Dementia has severe implications to the quality of daily life, in particular

to independent functioning. This sets extra demands to living environments. Older adults with dementia and their partners ask for living environments that support independence, compensate for declining vitality, and lower the burden of family care. For this purpose, a first concept of a design for a dementia dwelling is presented in this paper, which incorporates modifications in terms of architecture, interior design, the indoor environment, and technological solutions. These design features were derived from literature search and focus group sessions. Current design guidelines are frequently based on practical experience only, and therefore, more systematic field research should be carried out to find evidence for the various design modifications. Also, it needs to be studied how the design features of the dementia dwelling can be incorporated into the existing housing stock.

van Hoof, J., Kort, H.S.M., van Waarde, H., & Blom, M.M.
Environmental interventions and the design of homes for older adults with dementia: An overview.
American Journal of Alzheimer's Disease & Other Dementias, 2010, 25(3), 202-232. doi:10.1177/1533317509358885
Abstract: In Western societies, the vast majority of people with dementia live at home and wish to remain doing so for as long as possible. Aging in place can be facilitated through a variety of environmental interventions, including home modifications. This article provides an overview of existing design principles and design goals, and environmental interventions implemented at home, based on literature study and additional focus group sessions. There is a multitude of design principles, design goals, and environmental interventions available to assist with activities of daily living and functions, although few systematic studies have been conducted on the efficacy of these goals and interventions. The own home seems to be a largely ignored territory in research and government policies, which implies that many problems concerning aging in place and environmental interventions for dementia are not adequately dealt with.

van Hoof, J., Kort, H.H., Duijnstee, M.S., Rutten, P.P., & Hensen, J.J.
The indoor environment and the integrated design of homes for older people with dementia.
Building and Environment, 2010, 45, 1244-1261. doi: 10.1016/J.BUILDENV.2009.11.008
Abstract: There are currently about 6 million - mainly older - people with dementia in the European Union. With ageing, a number of sensory changes occur. Dementia syndrome exacerbates the effects of these sensory changes and alters perception of stimuli. People with dementia have an altered sensitivity for indoor environmental conditions, which can induce problematic behavior with burdensome symptoms to both the person with dementia and the family carer. This paper, based on literature review, provides an overview of the indoor environmental parameters, as well as the integrated design and implementation of relevant building systems. The overview is presented in relation to the intrinsic ageing of senses, the responses of older people with dementia and the impact on other relevant stakeholders through the combined use of the International Classification of Functioning, Disability and Health, and the Model of Integrated Building Design. Results are presented as indicators of the basic value, functional value and economic value, as well as a synthesis of building-related solutions. Results can help designers and building services engineers to create optimal environmental conditions inside the living environments for people with dementia, and can be used to raise awareness among health care professionals about of the influence of the indoor environment on behavior of the person with dementia.

Varcin, K.J., Herniman, S.E., Lin, A., Chen, Y., Perry, Y., Pugh, C., Chisholm, K., Whitehouse, A.J.O., & Wood, S.J.
Occurrence of psychosis and bipolar disorder in adults with autism: A systematic review and meta-analysis.
Neuroscience & Biobehavioral Reviews, 2022 Mar;134:104543. doi: 10.1016/j.neubiorev.2022.104543. Epub 2022 Jan 19.
Abstract: Evidence suggests that individuals with autism spectrum disorder have increased rates of co-occurring psychosis and/or bipolar disorder. Considering the peak age of onset for psychosis and bipolar disorder occurs in adulthood, we investigated the co-occurrence of these disorders in adults with autism. We conducted a systematic review and meta-analysis (PROSPERO Registration Number: CRD42018104600) to (1) examine the prevalence of psychosis and bipolar disorder in adults with autism, and (2) review potential risk factors associated with their co-occurrence. Fifty-three studies were included. The pooled prevalence for the co-occurrence of psychosis in adults with autism was 9.4 % (N = 63,657, 95 %CI = 7.52, 11.72). The pooled prevalence for the co-occurrence of bipolar disorders in adults with autism was 7.5 % (N = 31,739, 95 %CI = 5.79, 9.53). Psychosis and bipolar disorder occur at a substantially higher prevalence in adults with autism compared to general population estimates. While there is an overall dearth of research examining risk factors for these disorders in autism, males had increased likelihood of co-occurring psychosis, and females of co-occurring bipolar disorder. These results highlight the need for ongoing assessment and monitoring of these disorders in adults with autism.

Vasconcelos Pais, M., Talin, L.L., & Forlenza, O.V.
Biomarkers of cognitive decline and dementia in Down syndrome.
In: Teixeira, A.L., Rocha, N.P., Berk, M. (eds) *Biomarkers in Neuropsychiatry*. Springer, Cham. https://doi.org/10.1007/978-3-031-43356-6_12
Abstract: Alzheimer's disease (AD) dementia in Down syndrome (DS) is now conceptualized as a form of genetically determined AD. Increased dementia risk in this population is mainly driven by the amyloid precursor protein (APP) overexpression on chromosome 21, resulting in exponential accumulation of amyloid-beta (Aß) in the brain of individuals with DS. The neuropathology hallmarks and some clinical aspects of AD in DS are similar to those of sporadic AD. However, there are specific similarities with autosomal dominant AD. Some of the typical changes in biomarkers seen in both familial and sporadic AD have not been completely confirmed in DS. Cerebrospinal fluid (CSF) Aß1-42 levels, for example, are elevated in early childhood and young adults in this population, with CSF tau levels remaining low. The CSF Aß1-42 levels then decline and CSF tau levels increase over time with progression of AD pathology. Other fluid biomarkers follow the same trend, with various and mixed results reported in the literature. Conversely, as in AD in euploid individuals, plasma p-tau217 has shown to be very accurate in predicting both tau and Aß pathological brain changes in DS. Also, magnetic resonance imaging (MRI) findings in the brain of individuals with DS are more compatible with those observed in AD. Furthermore, other explored biomarkers in DS include inflammatory, biomarkers of oxidative stress, and cerebrovascular biomarkers.

Vaughan, R.M., McGee, C., Guerin, S., Tyrrell, J., & Dodd, P.
The challenges of diagnosis and treatment of dementia in Down's syndrome
Irish Journal of Psychological Medicine, 2016, 33(3), 151-158. doi: 10.1017/ipm.2016.1.
Abstract: This study analysed retrospective data on twenty adults with Down's syndrome (DS), who are clients of a specialist service in Dublin. The aim was to compare the practice of this service on diagnosis and treatment of dementia, with the consensus recommendations of the Royal College of Psychiatry, British Psychological Society and National Institute for Health and Care Excellence. Further aims were to establish average time to make a diagnosis and commence pharmacotherapy, and to describe tolerability to acetylcholinesterase inhibitors. It was found that screening for dementia did not take place before the age of 30yrs with the mean age for first assessment being 48yrs and average age at diagnosis being 51yrs. Average length of time from first identified symptoms to diagnosis was 1.3yrs. Of those diagnosed, 83% were prescribed acetylcholinesterase inhibitors but the authors were concerned at the continued use of the drug even when there appeared to be no benefit to the person. They found that a wide range of assessments were in use and that longitudinal assessment and follow up were not practiced. The authors recommend the streamlining of assessment tools and repeating assessment on a longitudinal basis.

Weden, M.M., Frank, L., Dick A.W., Wang, Z., Peschin, S., Bovenkamp, D.E., Rossi, S.L., Sciullo, D., Hillerstrom, H., & Fisher, R.A.
Investment in Alzheimer's disease research for the next generation of adults with Down syndrome will yield health benefits for future generations
Alzheimer's & Dementia, 2025, 21(9), e70348.
https://doi.org/10.1002/alz.70348.
Abstract: Recent innovations in Alzheimer's disease (AD) treatment highlight critical gaps in knowledge about how to support healthy aging of adults with Down syndrome (DS). RAND researchers updated demographic and epidemiological evidence about the DS population to assess the impact of increased investment in treatment innovations for DS-associated Alzheimer's disease (DS-AD). They estimated life expectancy at birth in 2020 to be 55 years, with ≈ 5 years of DS-AD. They found that the results of investment were dramatic. Between 2020 and 2070, adult years of life are expected to increase by 5 years without any increase in unhealthy years of life with DS-AD. Caregiving hours for individuals with DS-AD are expected to be reduced by 40%, which will generate large annual savings. The new evidence underscores the magnitude of the impact that investment in DS-AD treatments could have for individuals with DS, their families, and caregivers.

Verbeek, H., Zwakhalen, S.M., van Rossum, E., Kempen, G..I, & Hamers, J.P.
Kleinschalig wonen voor ouderen met dementie: de invloed op bewoners, mantelzorgers en medewerkers [Small-scale, homelike care environments for people with dementia: effects on residents, family caregivers and nursing staff].
Tijdschrift voor Gerontologie en Geriatrie, 2013 Dec,. 44(6), 261-271. In Dutch
DOI: 10.1007/s12439-013-0044-2
Abstract: Institutional dementia care is increasingly directed towards small-scale and homelike care environments, in The Netherlands as well as abroad. In these facilities, a small number of residents, usually six to eight, live together, and normal daily household activities and social participation are emphasized. In a quasi-experimental study, we studied the effects of small-scale, homelike care environments on residents (n = 259), family caregivers (n = 206) and nursing staff (n = 305). We compared two types of institutional nursing care during a 1 year period (baseline assessment and follow-up measurements at 6 and 12 months): (28) small-scale, homelike care environments and (21) psychogeriatric wards in traditional nursing homes. A matching procedure was applied to increase comparability of residents at baseline regarding functional status and cognition. This study was unable to demonstrate convincing overall effects of small-scale, homelike care facilities. On our primary outcome measures, such as quality of life and behaviour of residents and job satisfaction and motivation of nursing staff, no differences were found with traditional nursing homes. We conclude that small-scale, homelike care environments are not necessarily a better care environment than regular nursing homes and other types of living arrangements should be considered carefully. This provides opportunities for residents and their family caregivers to make a choice which care facility suits their wishes and beliefs best.

Verbeek, H., van Rossum, E., Zwakhalen, S.M., Ambergen, T., Kempen, G.I.,& Hamers, J.P.
The effects of small-scale, homelike facilities for older people with dementia on residents, family caregivers and staff: Design of a longitudinal, quasi-experimental study.
BMC Geriatrics, 2009 Jan 20, 9(3). doi: 10.1186/1471-2318-9-3.
Abstract: Small-scale and homelike facilities for older people with dementia are rising in current dementia care. In these facilities, a small number of residents live together and form a household with staff. Normal, daily life and social participation are emphasized. It is expected that these facilities improve residents' quality of life. Moreover, it may have a positive influence on staff's job satisfaction and families involvement and satisfaction with care. However, effects of these small-scale and homelike facilities have hardly been investigated. Since the number of people with dementia increases, and institutional long-term care is more and more organized in small-scale and homelike facilities, more research into effects is necessary. This paper presents the design of a study investigating effects of small-scale living facilities in the Netherlands on residents, family caregivers and nursing staff. A longitudinal, quasi-experimental study is carried out, in which 2 dementia care settings are compared: small-scale living facilities and regular psychogeriatric wards in traditional nursing homes. Data is collected from residents, their family caregivers and nursing staff at baseline and after 6 and 12 months of follow-up. Approximately 2 weeks prior to baseline measurement, residents are screened on cognition and activities of daily living (ADL). Based on this screening profile, residents in psychogeriatric wards are matched to residents living in small-scale living facilities. The primary outcome measure for residents is quality of life. In addition, neuropsychiatric symptoms, depressive symptoms and social engagement are assessed. Involvement with care, perceived burden and satisfaction with care provision are primary outcome variables for family caregivers. The primary outcomes for nursing staff are job satisfaction and motivation. Furthermore, job characteristics social support, autonomy and workload are measured. A process evaluation is performed to investigate to what extent small-scale living facilities and psychogeriatric wards are designed as they were intended. In addition, participants' satisfaction and experiences with small-scale living facilities are investigated. A longitudinal, quasi-experimental study is presented to investigate effects of small-scale living facilities. Although some challenges concerning this design exist, it is currently the most feasible method to assess effects of this relatively new dementia care setting.

Verbeek, H., Van Rossum, E., Zwakhalen, S. M., Kempen, G. I., & Hamers, J. P.
Small, homelike care environments for older people with dementia: A literature review.
International Psychogeriatrics, 2009, 21, 252-264.
https://doi.org/10.1017/S104161020800820X
Abstract: There is large cross-national variation in the characteristics of small, domestic-style care settings which emphasize normalized living. However, a systematic overview of existing types is lacking. This study provides an international comparison of the care concepts which have adopted a homelike philosophy in a small-scale context. Insight into their characteristics is vital for theory, planning and implementation of such dementia care settings. A literature search was performed using various electronic databases, including PubMed, Medline, CINAHL and PsycINFO. In addition, "gray" literature was identified on the internet. Concepts were analyzed according to five main characteristics: physical setting, number of residents, residents' characteristics, domestic characteristics and care concept. Authors included 75 papers covering 11 different concept types in various countries. Similarities among concepts reflected a focus on meaningful activities centered around the daily household. Staff have integrated tasks and are part of the household, and archetypical home-style features, such as kitchens, are incorporated in the buildings. Differences among concepts were found mainly in the physical settings, numbers of residents and residents' characteristics. Some concepts have become regular dementia care settings, while others are smaller initiatives. The care concepts are implemented in various ways with a changing staff role. However, many aspects of these small, homelike facilities remain unclear. Future research is needed, focusing on residents' characteristics, family, staff and costs.

Verbeek H, Zwakhalen SM, van Rossum E, Ambergen T, Kempen GI, Hamers JP.
Dementia care redesigned: Effects of small-scale living facilities on residents, their family caregivers, and staff.
Journal of the American Medical Directors Association, 2010 Nov, 11(9), 662-670. doi: 10.1016/j.jamda.2010.08.001.
Abstract: The purpose of this study was to evaluate the effects of small-scale living facilities in dementia care on residents, family caregivers, and staff. This was a quasi-experimental study including 2 types of institutional nursing care: small-scale living facilities (experimental group), and regular psychogeriatric nursing home wards (control group). Three measures were conducted: at

baseline and follow-ups after 6 and 12 months. Twenty-eight houses in small-scale living facilities and 21 regular psychogeriatric nursing home wards. In total, 259 residents were included in the study: 124 in small-scale living facilities and 135 controls, matched on cognitive and functional status. Furthermore, 229 family caregivers were included and 305 staff members. For residents, main outcome measures were quality of life, neuropsychiatric symptoms, and agitation. Main outcome measures for family caregivers included perceived burden, satisfaction, and involvement with care. Main outcome measures for staff were job satisfaction and motivation. No effects were found for residents' total quality of life, neuropsychiatric symptoms, and agitation. Family caregivers in small-scale living reported significantly less burden (adjusted mean difference 0.8, 95% CI 0.1-1.5) and were more satisfied with nursing staff (0.3, 0.2-0.5) than family caregivers in regular wards. No differences were found in their involvement with care. Overall, no significant differences were found for staff's job satisfaction and motivation, although subgroup analyses using contrast groups (regarding typical small-scale living and regular wards) revealed more job satisfaction (2.0, 0.5-3.5) and motivation (0.6, 0.0-1.3) in small-scale living compared with regular wards. This study was unable to demonstrate convincing overall effects of small-scale living facilities. Because governmental policies and, in some countries, financial support, are increasingly aimed at providing small-scale, homelike care, it is suggested that this may not be a final solution to accomplish high-quality dementia care and that other options should be considered.

Verbeek, H., Zwakhalen, S.M., van Rossum, E., Kempen, G.I., & Hamers, J.P.
Small-scale, homelike facilities in dementia care: a process evaluation into the experiences of family caregivers and nursing staff.
International Journal of Nursing Studies, 2012 Jan, 49(1), 21-9. doi: 10.1016/j.ijnurstu.2011.07.008. Epub 2011 Aug 5
Abstract: Current developments in institutional dementia care aim at the downsizing of facilities and increasing their homelike appearance. Small-scale living facilities are an example of this movement, in which a small group of residents (usually six to eight) live together in a homelike environment. Residents are encouraged to participate in normal daily activities and nursing staff is part of the household with integrated tasks. Despite the increase of these facilities, little is known about experiences of family caregivers of residents and nursing staff. To gain an in-depth insight into the experiences of family caregivers and nursing staff with small-scale living facilities. A process evaluation was conducted alongside the final measurement of an effectiveness study, using a cross-sectional, descriptive design. Two types of institutional dementia care in the Netherlands: small-scale living facilities and regular wards in nursing homes. In total, 130 family caregivers and 309 nursing staff workers in both care settings participated in a survey questionnaire. Additional in-depth interviews were conducted with a random selection of 24 participants in small-scale living facilities: 13 family caregivers and 11 nursing staff workers. Survey questions for family caregivers focused on care service delivery; questions for nursing staff were related to skills. The interviews especially related to positive and negative aspects of small-scale living facilities and skills for nursing staff. Both family caregivers and staff mainly reported positive experiences with small-scale living facilities, especially the personal attention that nursing staff provides to residents, their involvement with residents and the emphasis on autonomy in daily life. Barriers mainly related to nursing staff working alone during a large part of the day. Family caregivers in small-scale living facilities were more satisfied with the care facility and nursing staff than those in regular wards. The findings of the study revealed several positive aspects of small-scale living facilities related to physical, social and organizational aspects that could be used as tools to implement changes in institutional dementia care settings.

Verghese, J., Ayers, E., Barzilai, N., Bennett, D.A., Buchman, A.S., Holtzer, R., Katz, M.J., Lipton, R.B., & Wang, C.
Motoric cognitive risk syndrome: Multicenter incidence study.
Neurology, 2014 Dec 9, 83(24), 2278–2284. doi: 10.1212/WNL.0000000000001084
Abstract: Authors report on the incidence and risk factors for motoric cognitive risk syndrome (MCR), a newly described predementia syndrome characterized by slow gait and cognitive complaints. They examined incidence rates of MCR in 3,128 adults aged 60 years and older, MCR- and dementia-free at baseline, participating in 4 US-based cohort studies. Hazard ratios (HRs) with 95% confidence intervals (CIs) for the association of modifiable risk factors with risk of MCR were computed using Cox models. Over a median follow-up time of 3.2 years, 823 of the 3,128 participants met MCR criteria. The overall age- and sex-adjusted incidence of MCR was 65.2/1,000 person-years (95% CI: 53.3-77.1), and ranged from 50.8/1,000 person-years to 79.6/1,000 person-years in the individual cohorts. MCR incidence was higher with older age but there were no sex differences. In the pooled sample adjusted for age, sex, education, and cohort source, strokes (HR 1.42, 95% CI: 1.14-1.77), Parkinson disease (HR 2.52, 95% CI: 1.68-3.76), depressive symptoms (HR 1.65, 95% CI: 1.28-2.13), sedentariness (HR 1.76, 95% CI: 1.44-2.17), and obesity (HR 1.39, 95% CI: 1.17-1.65) predicted risk of incident MCR. Authors found that the incidence of MCR is high in older adults. Identification of modifiable risk factors for MCR will improve identification of high-risk individuals and help develop interventions to prevent cognitive decline in aging.

Verlinden, V.J.A., van der Geest, J.N., de Bruijn, R.F.A.G., Hofman, A., Koudstaal, P.J., & Ikram, M.A.
Trajectories of decline in cognition and daily functionng in preclinical dementia
Alzheimer's & Dementia, 2016, Feb, 12(2), 144-153.
doi: 10.1016/j.jalz.2015.08.001. Epub 2015 Sep 9.
Abstract: [*Note: this article refers to adults in the general population]* Although preclinical dementia is characterized by decline in cognition and daily functioning, little is known on their temporal sequence. We investigated trajectories of cognition and daily functioning in preclinical dementia, during 18 years of follow-up. In 856 dementia cases and 1712 controls, we repetitively assessed cognition and daily functioning with memory complaints, mini-mental state examination (MMSE), instrumental activities of daily living (IADL), and basic activities of daily living (BADL). Dementia cases first reported memory complaints 16 years before diagnosis, followed by decline in MMSE, IADL, and finally BADL. Vascular dementia related to earlier decline in daily functioning but later in cognition, compared with Alzheimer's disease. Higher education related to larger preclinical cognitive decline, whereas apolipoprotein E (APOE) e4 carriers declined less in daily functioning. These results emphasize the long hierarchical preclinical trajectory of functional decline in dementia. Furthermore, they show that various pathologic, environmental, and genetic factors may influence these trajectories of decline.

Videla, L., Benejam, B., Pegueroles, J., Carmona-Iragui, M., Padilla, C., Fernández, S., Barroeta, I., Altuna, M., Valldeneu, S., Garzón, D., Ribas, L., Montal, V., Arranz Martínez, J., Rozalem Aranha, M., Alcolea, D., Bejanin, A., Iulita, M.F., Videla Cés, S., Blesa, R., Lleó, A., & Fortea, J.
Longitudinal clinical and cognitive changes along the Alzheimer disease continuum in Down syndrome.
JAMA Netw Open. 2022 Aug 1;5(8):e2225573. doi: 10.1001/jamanetworkopen.2022.25573.
Abstract: Alzheimer disease (AD) is the main medical problem in adults with Down syndrome (DS). However, the associations of age, intellectual disability (ID), and clinical status with progression and longitudinal cognitive decline have not been established. To examine clinical progression along the AD continuum and its related cognitive decline and to explore the presence of practice effects and floor effects with repeated assessments, authors used a single-center cohort study of adults (aged >18 years) with DS with different ID levels and at least 6 months of follow-up between November 2012 and December 2021. The data are from a population-based health plan designed to screen for AD in adults with DS in Catalonia, Spain. Individuals were classified as being asymptomatic, having prodromal AD, or having AD dementia. The main outcome was clinical change

along the AD continuum. Cognitive decline was measured by the Cambridge Cognitive Examination for Older Adults With Down Syndrome and the modified Cued Recall Test. Authors assessed a total of 632 adults with DS (mean [SD] age, 42.6 [11.4] years; 292 women [46.2%]) with 2847 evaluations (mean [SD] follow-up, 28.8 [18.7] months). At baseline, there were 436 asymptomatic individuals, 69 patients with prodromal AD, and 127 with AD dementia. After 5 years of follow-up, 17.1% (95% CI, 12.5%-21.5%) of asymptomatic individuals progressed to symptomatic AD in an age-dependent manner (0.6% [95% CI, 0%-1.8%] for age <40 years; 21.1% [95% CI, 8.0%-32.5%] for age 40-44 years; 41.4% [95% CI, 23.1%-55.3%] for age 45-49 years; 57.5% [95% CI, 38.2%-70.8%] for age ?50 years; P < .001), and 94.1% (95% CI, 84.6%-98.0%) of patients with prodromal AD progressed to dementia with no age dependency. Cognitive decline in the older individuals was most common among those who progressed to symptomatic AD and symptomatic individuals themselves. Importantly, individuals with mild and moderate ID had no differences in longitudinal cognitive decline despite having different performance at baseline. This study also found practice and floor effects, which obscured the assessment of longitudinal cognitive decline. This study found an association between the development of symptomatic AD and a high risk of progressive cognitive decline among patients with DS. These results support the need for population health plans to screen for AD-related cognitive decline from the fourth decade of life and provide important longitudinal data to inform clinical trials in adults with DS to prevent AD.

Videla L, Benejam B, Carmona-Iragui M, Barroeta I, Fernández S, Arranz J, Azzahchi SE, Altuna M, Padilla C, Valldeneu S, Pegueroles J, Montal V, Aranha MR, Vaqué-Alcázar L, Iulita MF, Alcolea D, Bejanin A, Videla S, Blesa R, Lleó A, Fortea J.
Cross-sectional versus longitudinal cognitive assessments for the diagnosis of symptomatic Alzheimer's disease in adults with Down syndrome.
Alzheimers Dement. 2023 Sep;19(9):3916-3925. doi: 10.1002/alz.13073. Epub 2023 Apr 11. PMID: 37038748.
Abstract: Down syndrome (DS) is a genetic form of Alzheimer's disease (AD). However, clinical diagnosis is difficult, and experts emphasize the need for detecting intra-individual cognitive decline. We compared the performance of baseline and longitudinal neuropsychological assessments for the diagnosis of symptomatic AD in DS. Longitudinal cohort study of adults with DS. Individuals were classified as asymptomatic, prodromal AD, or AD dementia. We performed receiver operating characteristic curve analyses to compare baseline and longitudinal changes of CAMCOG-DS and mCRT. We included 562 adults with DS. Baseline assessments showed good to excellent diagnostic performance for AD dementia (AUCs between 0.82 and 0.99) and prodromal AD, higher than the 1-year intra-individual cognitive decline (area under the ROC curve between 0.59 and 0.79 for AD dementia, lower for prodromal AD). Longer follow-ups increased the diagnostic performance of the intra-individual cognitive decline. Baseline cognitive assessment outperforms the 1-year intra-individual cognitive decline in adults with DS.

Visootsak, J., & Sherman, S.
Neuropsychiatric and behavioral aspects of trisomy 21
Current Psychiatry Reports, 2007, 9(2), 135-140.
doi: 10.1007/s11920-007-0083-x.
Abstract: Down syndrome (DS), or trisomy 21, is the most common identifiable genetic cause of mental retardation. The syndrome is unique with respect to its cognitive, behavioral, and psychiatric profiles. The well-known cheerful and friendly demeanor often creates a personality stereotype, with parents and observers commenting on the positive attributes. Despite these strengths, approximately 20% to 40% of children with DS have recognized behavioral problems. Such problems persist through adulthood, with a decrease in externalizing symptoms of aggressiveness and attention problems and the emergence of internalizing symptoms of depression and loneliness. In adulthood, the presence of early-onset dementia of the Alzheimer type and cognitive decline may pose a challenge in recognizing these internalizing symptoms. Understanding the age-related changes in cognitive functioning and behavioral profiles in individuals with DS provides insight into clinical and treatment implications.

Visser, F.E., Aldenkamp, A.P., van Huggelen, A.C., Kuilman, M., Overweg, J., & van Wijk, J.
Prospective study of the prevalence of Alzheimer-type dementia in institutionalized individuals with Down syndrome
*American Journal on Mental Retardation,*1997, Jan, 101(4), 400-412.
https://pubmed.ncbi.nlm.nih.gov/9017086/
Abstract: Institutionalized patients with Down syndrome (n = 307) were monitored for 5 to 10 years prospectively to determine prevalence of Alzheimer-type dementia. Clinical signs, cognitive functioning, and EEGs were assessed. When possible, postmortem neuropathological examinations were conducted. Progressive mental and physical deterioration was found for 56 of the residents. **Mean age at onset of dementia was 56 years**. Prevalence increased from 11% between ages 40 and 49 to 77% between 60 and 69. All patients 70 and over had dementia. Neuropathological findings were consistent with clinical diagnosis. Use of a dementia checklist, cognitive skills inventory, and EEG reliably detected Alzheimer-type dementia at an early stage.

Vivanti, G., Tao, S., Lyall, K., Robins, D.L., & Shea, L.L.
The prevalence and incidence of early-onset dementia among adults with autism spectrum disorder.
Autism Research, 2021 Oct,14(10), 2189-2199. doi: 10.1002/aur.2590.
Abstract: The prevalence and incidence of early-onset dementia among adults with autism spectrum disorder (ASD) is currently unknown. In this case-control study, the prevalence and incidence of early-onset dementia in individuals with ASD was examined during 2008-2012 using Medicaid Analytic eXtract files. Participants were 30-64 year-old adults who were Medicaid beneficiaries and had either a diagnosis of ASD only (n = 12,648), a diagnosis of ASD with co-occurring intellectual disability (ID) (n = 26,168), a diagnosis of ID without ASD (n = 406,570), or no ASD nor ID diagnoses (n = 798,828). The 5-year prevalence of dementia was 4.04% among adults with ASD only, and 5.22% for those with ASD and co-occurring ID. This prevalence was higher compared to the prevalence of dementia in individuals with no ASD and no ID (0.97%), but lower compared to individuals with ID only (7.10%). Risk factors associated with the increased prevalence in the general population were similarly associated with the increased risk of dementia in individuals with ASD. Even after adjusting for these risk factors, compared to the general population, dementia was found to occur more frequently in individuals with ASD only (adjusted hazard ratio, 1.96; 95% CI, 1.69-2.28), as well as individuals with ASD and co-occurring ID (adjusted hazard ratio, 2.89; 95% CI, 2.62-3.17). In conclusion, adults with ASD under the age of 65 were approximately 2.6 times more likely to be diagnosed with dementia compared to the general population in our study. LAY SUMMARY: It is unclear whether adults diagnosed with autism spectrum disorder (ASD) are at higher risk of being diagnosed with early-onset dementia compared to those who are not on the autism spectrum. In this study we examined for the first time the nationwide prevalence and incidence of Alzheimer's Disease and other types of dementia in ASD in a sample of adults with ASD aged 30-64 years who were enrolled in Medicaid, the largest insurer of behavioral health services in the US. Medicaid claims data, which include information on the diagnoses that beneficiaries receive, suggested that the adults with ASD were approximately 2.6 times more likely to be diagnosed with early-onset Alzheimer's disease and related dementias compared to the general population.

Vivanti, G., Lee, W., Ventimiglia, J., Tao, S., Lyall, K., & Shea, L.L.
Prevalence of dementia among US adults with autism spectrum disorder.
JAMA Network Open, 2025, 8(1), e2453691.
doi:10.1001/jamanetworkopen.2024.53691
Abstract: This retrospective cohort study reports the nationwide prevalence of identified dementia diagnoses in individuals with assigned ASD diagnoses in

linked Medicare and Medicaid data. Participants were individually linked Medicaid and fee-for-service Medicare beneficiaries aged 30 years and older enrolled January 1, 2014, to December 31, 2016, whose diagnosis of ASD was detected via International Classification of Diseases, Ninth Revision or Tenth Revision diagnostic codes. Race and ethnicity data were drawn from Medicare and Medicaid enrollments. Racial and ethnic differences were assessed to track disparities from previous reports and across diagnostic groups. Given the known association between intellectual disabilities (ID) and dementia, we created an ASD-only group including 46 877 individuals and an ASD plus ID group, including 67 705 individuals. Individuals with Down syndrome were excluded because of their genetic risk for dementia. Prevalence of dementia diagnoses in each group was examined as a period prevalence estimate over the 3 years of enrollment and annually. Denominators were calculated using the total enrolled for each group. Additionally examined was whether common risk factors for dementia increased the odds of assigned dementia diagnoses in the sample. Data were analyzed from June 2023 to June 2024. Of the 114 582 individuals included in the study, most (63 194 [55.2%]) were aged 40 or older, 80 104 (69.9%) were male, 18 627 (16.3%) were Black, 8048 (7.02%) were Hispanic or Latino, and 80 680 (70.4%) were White. An identified dementia diagnosis was present in 8.03% of the ASD-only group and 8.88% of the ASD plus ID group. The odds of a dementia diagnosis increased with age, with a prevalence of 35.12% in the ASD-only group and 31.22% in the ASD plus ID group for individuals older than 64. The odds were higher in individuals with cardiovascular risk factors and depression or other psychiatric conditions, after controlling for residence state. Authors note that linked Medicaid and Medicare records suggest a markedly elevated prevalence of identified dementia diagnoses in individuals with an ASD diagnosis. Findings were consistent with our previous research, and prevalence estimates were higher than those for the general population of Medicaid and Medicare beneficiaries reported in the literature.

Walaszek, A., Schroeder, M., Krainer, J., Pritchett, G., Wilcenski, M., Endicott, S. , Albrecht, T., Carlsson, C.M., & Mahoney, J.
Effectively training professional caregivers to screen and refer persons with dementia and intellectual/developmental disability
AAIC 2020 Conference, Poster presentation, July 30. 2020. *Alzheimer's & Dementia*, 16(S8), First published: 07 December 2020. https://doi.org/10.1002/alz.037966
Abstract: By age 40, almost all people with Down syndrome, the most common cause of intellectual/developmental disability (I/DD), have neuropathological changes consistent with Alzheimer's disease; by age 60, about half have dementia. Detecting dementia in persons with I/DD can be challenging because baseline cognitive impairment can be severe and because persons with I/DD may have difficulty reporting symptoms. The National Task Group Early Detection Screen for Dementia (NTG-EDSD) was developed to aid detection of cognitive impairment in adults with I/DD. We implemented an educational curriculum to increase the ability of professional caregivers to screen for dementia in persons with I/DD using the NTG-EDSD. In November 2018 to April 2019, we held five training sessions for professional caregivers of persons with I/DD, partnering with various managed care organizations (MCO), aging and disability resource centers, adult day programs, and adult family homes. We assessed knowledge and attitudes at baseline, immediately after training, and one week, one month and six months after training. Participants (N=154) included direct care workers, case managers, healthcare providers, and other social services staff. Participants reported a marked increase in confidence in their ability to detect changes associated with mild cognitive impairment or dementia ($p<0.001$), decline in activities of daily living ($p=0.02$), and changes in behavior and affect ($p<0.001$). Satisfaction with the training was very high, and 94.0% of participants agreed or strongly agreed they could use the NTG-EDSD tool with their clients. Following the training, one MCO we partnered with, serving 62 of 72 counties in Wisconsin, made the NTG-EDSD a standard part of the assessment of adults with Down syndrome starting at age 40. Authors note that a wide variety of social services and healthcare professionals can be effectively trained to screen for dementia in persons with I/DD using a standardized screening tool, the NTG-EDSD. Satisfaction with the training was high, and use of the NTG-EDSD was thought to be feasible. This educational intervention led to change in practice at a systems level within an MCO. Next steps could include assessing impact of such training on the quality of life and healthcare outcomes of persons with I/DD.

Walaszek, A., Albrecht, T., LeCaire, T., Sayavedra, N., Schroeder, M., Krainer, J., Prichett, G., Wilcenski, M., Endicott, S., Russmann, S., Carlsson, C.M., & Mahoney, J. Training professional caregivers to screen for report of cognitive changes in persons with intellectual disability.
Alzheimer's & Dementia: Translational Research & Clinical Interventions, 2022 Aug, 8(1), 1-10, e12345 https://doi.org/10.1002/trc2.12345
Abstract: By age 60, 60% of adults with Down syndrome (DS) have dementia. Detecting dementia in persons with intellectual disability (ID) can be challenging because their underlying cognitive impairment can confound presentation of dementia symptoms and because adults with ID may have difficulty reporting symptoms. The National Task Group Early Detection Screen for Dementia (NTG-EDSD) was developed to aid detection of report of cognitive impairment in adults with ID. We implemented an educational curriculum using the NTG-EDSD and evaluated the impact of the intervention on professional caregivers' self-assessed capacity to identify persons with ID and dementia. We held five in-person training sessions for professional caregivers of persons with ID, partnering with various managed care organizations and social services agencies. We assessed knowledge and attitudes at baseline; immediately after training; and 1 week, 1 month, and 6 months after training. A total of 154 direct care workers, case managers, health-care providers, and other social services staff attended the trainings. Satisfaction with the NTG-EDSD training was high; 94% of attendees agreed or strongly agreed that they could use the NTG-EDSD with their clients. After training, attendees reported a marked increase in confidence in their ability to track various health circumstances and detect functional decline in their clients, although some gains were not sustained over time. As a result of the training, one managed care organization made the NTG-EDSD a standard part of its assessment of adults with DS starting at age 40. Social services and health-care professionals can learn to document signs of cognitive decline in adults with ID using the NTG-EDSD. Attendees were highly satisfied with the training, experienced an increase in confidence in their care of persons with ID, and found the NTG- EDSD feasible to use. Because not all gains were sustained over time, booster trainings may be necessary.

Walker, B., MacBryer, S., Jones, A., & Law, J.
Interinformant agreement of the dementia questionnaire for people with learning disabilities
British Journal of Learning Disabilities, 2015, Sept, 43(3), 227-233.. https://doi.org/10.1111/bld.12102
Abstract: Because of difficulties with neuropsychological assessments for dementia in people with intellectual disabilities, professionals in clinical practice have relied heavily on carer interviews, one of the most widely used being the Dementia Questionnaire for People with Learning Disabilities (DLD – Evenhuis et al. 2006 Dementia questionnaire for people with intellectual disabilities manual (second edition). Amsterdam, Netherlands, Harcourt Test Publishers). Because dementia is indicated by the magnitude of changes in scores between longitudinal assessments, interinformant agreement is paramount. We carried out the DLD interview independently with two carers for each of 26 people with Down syndrome. Only 15% of pairs of carers achieved 'good' agreement. Levels of agreement varied widely across the DLD subscales and individual questions. Interinformant agreement was better for less able people with Down syndrome than for more able individuals.

Walker, C.A., & Walker, A.
Uncertain Futures: people with learning difficulties and their ageing family carers 54 pp.

Brighton, UK: Pavilion Publishing (1998)
Abstract: This monograph provides an overview of research, policy and practice relating to service responses to adults with learning difficulties living at home with older family carers in the UK. The authors' premise is that as life expectancy increases, a growing proportion of people with learning difficulties continues to live with family members, most frequently parents, whose caring role is being extended into their own advanced old age. Highlighted are some of the issues raised by service users, carers and service providers, including care for someone with diminishing abilities. The text argues that there is urgent need for the paid service sector to work with families to provide the necessary support and planning to take the uncertainty out of the future.

Wallace, E., Harp, J., Van Pelt, K.I., Koehl, L., Caban-Holt, .A.M., Anderson-Mooney, A.J., Robertson, W., Lightner, D., Jicha, G.A., Head, E., & A Schmitt, F.A.
Validity of the Severe Impairment Battery, Brief Praxis Test, and Dementia Questionnaire for Persons with Intellectual Disabilities in differentiating dementia status in individuals with Down syndrome
AAIC2020, Poster presentation, July 29, 2020. *Alzheimer's & Dementia*, 16 (S6), First published: 07 December 2020. https://doi.org/10.1002/alz.044227
Abstract: Individuals with Down syndrome (DS) are at high risk for dementia, specifically Alzheimer's disease (AD). However, many measures regularly used for the detection of AD in the general population are not suitable for individuals with DS. Some measures, including the Severe Impairment Battery (SIB), Brief Praxis Test (BPT), and Dementia Questionnaire for Persons with Intellectual Disabilities (DMR), have been used in clinical trials and other research with this population. Validity research is limited, however, particularly regarding identification of predementia symptoms in the DS population. The current project presents baseline cross-sectional SIB, BPT, and DMR performance in order to characterize their ability to discriminate normal cognition, possible AD, and probable AD in DS. Baseline SIB, BPT, and DMR performances from 117 individuals were analyzed as part of a large longitudinal cohort of aging individuals with DS. Receiver operating characteristic (ROC) curves were calculated to investigate accuracy in differentiating levels of dementia status. In comparing no/possible AD vs. probable AD, the SIB and BPT exhibited fair discrimination ability (AUC = .78 and .79, respectively). In comparing no/possible AD vs. probable AD, the DMR exhibited good discrimination ability (AUC = .89), with qualitatively similar performance of the DMR-Cognitive and DMR-Social subscales (AUC = .89 and .83, respectively). In comparing no AD vs. possible AD, the SIB and BPT failed to differentiate these groups (AUC = .53 and .55, respectively), whereas the DMR exhibited good differentiation (AUC = .80). Au thors note that the results suggest that the SIB, BPT, and DMR are able to discriminate between levels of dementia status in individuals with DS, supporting their continued use in the clinical assessment of dementia in DS. Specifically, the DMR, based on informant ratings of social and cognitive behaviors of daily living, outperformed the SIB and BPT, tests of cognitive performance, in discriminating no/possible AD vs. probable AD as well as no AD vs. possible AD. Such findings suggest that the DMR is better equipped to identify symptoms of overt dementia as well as predementia in this population. Findings reinforce the importance of including informant behavior ratings in assessment of this population.

Wallace, E.R., Harp, J.P., Van Pelt, K.L., Koehl, L.M., Caban-Holt, A.M., Anderson-Mooney, A.J., Jicha, G.A., Lightner, D.D., Robertson, W.C., Head, E. & Schmitt, F.A.
Identifying dementia in Down syndrome with the Severe Impairment Battery, Brief Praxis Test and Dementia Scale for People with Learning Disabilities.
Journal of Intellectual Disability Research, 2021 Dec, 65(12),1085-1096. doi: 10.1111/jir.12901. Epub 2021 Nov 16.
Abstract: Individuals with Down syndrome (DS) are at high risk for dementia, specifically Alzheimer's disease. However, many measures regularly used for the detection of dementia in the general population are not suitable for individuals with DS due in part to floor effects. Some measures, including the Severe Impairment Battery (SIB), Brief Praxis Test (BPT) and Dementia Scale for People with Learning Disabilities (DLD), have been used in clinical trials and other research with this population. Validity research is limited, particularly regarding the use of such tools for detection of prodromal dementia in the DS population. The current project presents baseline cross-sectional SIB, BPT and DLD performance in order to characterise their predictive utility in discriminating normal cognition, possible dementia and probable dementia in adult DS. Baseline SIB, BPT and DLD performances from 100 individuals (no dementia = 68, possible dementia = 16 & probable dementia = 16) were examined from a longitudinal cohort of aging individuals with DS. Receiver operating characteristic curves investigated the accuracy of these measures in relation to consensus dementia diagnoses, diagnoses which demonstrated high percent agreement with the examining neurologist's independent diagnostic impression. The SIB and BPT exhibited fair discrimination ability for differentiating no/possible versus probable dementia [area under the curve (AUC) = 0.61 and 0.66, respectively]. The DLD exhibited good discrimination ability for differentiating no versus possible/probable dementia (AUC = 0.75) and further demonstrated better performance of the DLD Cognitive subscale compared with the DLD Social subscale (AUC = 0.77 and 0.67, respectively). Results suggest that the SIB, BPT and DLD are able to reasonably discriminate consensus dementia diagnoses in individuals with DS, supporting their continued use in the clinical assessment of dementia in DS. The general performance of these measures suggests that further work in the area of test development is needed to improve on the AUCs for dementia status discrimination in this unique population. At present, however, the current findings suggest that the DLD may be the best option for reliable identification of prodromal dementia in this population, reinforcing the importance of including informant behaviour ratings in assessment of cognition for adults with DS.

Walsh, D.M., Doran, E., Silverman, W., Tournay, A., Movsesyan, N., & Lott, I.T.
Rapid assessment of cognitive function in down syndrome across intellectual level and dementia status
Journal of Intellectual Disability Research, 2015, Nov, 59(11), 1071-1079. doi:10.1111/jir.12200. Epub 2015 May 29.
Abstract: Adults with Down syndrome (DS) are at risk of developing dementia and cognitive assessment is a fundamental part of the diagnostic process. Previously, we developed a Rapid Assessment for Developmental Disabilities (RADD), a brief, broadly focused direct test of cognition. In the current report, we assess whether the RADD is sensitive to dementia in DS and the degree to which it compares with other cognitive measures of dementia in this population.
In a sample of 114 individuals with DS, with dementia diagnosed in 62%, the RADD was compared with the Dementia Questionnaire for Mentally Retarded Persons (DMR), the Bristol Activities of Daily Living Scale, Severe Impairment Battery (SIB), and the Brief Praxis Test (BPT). The RADD showed predicted effects across intellectual disability (ID) levels and dementia status ($p < 0.001$). Six-month test-retest reliability for the subset of individuals without dementia was high ($r(41) = 0.95$, $p < 0.001$). Criterion-referenced validity was demonstrated by correlations between RADD scores and ID levels based upon prior intelligence testing and clinical diagnoses ($rs (114) = 0.67$, $p = 0.001$) and with other measures of cognitive skills, such as the BPT, SIB, and DMR-Sum of Cognitive scores (range 0.84 through 0.92). Using receiver operating characteristic curves for groups varying in pre-morbid severity of ID, the RADD exhibited high sensitivity (0.87) and specificity (0.81) in discriminating among individuals with and without dementia, although sensitivity was somewhat lower (0.73) for the subsample of dementia cases diagnosed no more than 2 years prior to their RADD assessment. Taken together, findings indicated that the RADD, a relatively brief, easy-to-administer test for cognitive function assessment across ID levels and dementia status, would be a useful component of cognitive assessments for adults with DS, including assessments explicitly focused on dementia.

Waninge, A., Wissing, M., Hobbelen, H., Fokkens, A., Dekker, A., & De Deyn, P.

Dementia in people with severe/profound intellectual disabilities
Journal of Applied Research in Intellectual Disabilities, 2021, 34(5), 1214-1215. https://doi.org/10.1111/jar.12917
Abstract: In people with severe or profound intellectual disabilities, it is difficult to diagnose dementia. As timely identification and diagnosis of dementia allows for a timely response to changing client wishes and needs, this study examined symptoms, and diagnosis of dementia in practice. Family members and professionals were invited to fill out a survey about symptoms and diagnosis of dementia in people with severe or profound intellectual disabilities. Results of the survey were further explored within semi-structured interviews with professionals having experience with signaling and diagnosing dementia in these people. Symptoms found in the survey and transcripts of the interviews were qualitatively analyzed, using thematic analyses based on a developed symptom-matrix. The survey was filled out completely by 14 family members and 90 professionals with different backgrounds. Results showed that behavioral changes were recognized more frequently than cognitive decline. Compared to those without dementia, epilepsy and motor decline were more present in case of dementia. Fifteen interviews (until saturation) with professionals provided an in-depth view into the symptoms, and how to identify them, again stressing behavioral alterations and to a lesser extent cognitive symptoms.

Waninge, A., Wissing, M., Hobbelen, J., Fokkens, A., Dekker, A., & De Deyn, P.
Dementia symptoms in persons with severe/profound intellectual disability: expertise of practice. (2021). pp. 1214-1215. Abstract from 6th IASSIDD virtual Europe Congress, Amsterdam, Netherlands.
Abstract: Life expectancy of people with severe or profound intellectual disability (SPID) increases, which contributes to the risk of developing dementia. However, early detection and diagnosing dementia is complex, because of their low-level baseline functioning. Therefore, the aim is to identify observable dementia symptoms in adults with SPID in available literature. A systematic literature search, in line with PRISMA guidelines, was conducted in PubMed, PsycINFO and Web of Science using a combination of search terms for SPID, dementia/aging and aged population. In total, fifteen studies met inclusion criteria. Cognitive, behavioral and psychological symptoms (BPSD) and a decline in the ability to perform activities of daily living as well as neurological and physical changes were found. This presentation gives an overview of reported symptoms of (possible) dementia-related symptoms in SPID. Despite growing attention for dementia in people with ID in literature, only very few studies have studied dementia symptoms in SPID. Given the complexity of signaling and diagnosing dementia in SPID, dedicated studies are required to unravel the natural history of dementia in SPID, specifically focusing on observable symptoms for caregivers of (early) dementia in this population.

Ward, S., Opie, J., O'Connor, D.W.
Family carer's responses to behavioural and psychological symptoms of dementia
International Journal of Geriatric Psychiatry, 2003, Nov, 18(11), 1007-1012. doi: 10.1002/gps.1005.
Abstract: The author sought to describe the responses of family carers to the behavioural and psychological symptoms associated with dementia. Thirty family carers of people with dementia were identified in a survey of mental disorder in general practice. Another 20 were referred by local aged mental health services. Carers were interviewed using the Manchester and Oxford University Scale for the Psychopathological Assessment of Dementia (MOUSEPAD) which rates behavioural and psychological disturbances. Carers' customary responses to current symptoms were recorded verbatim and categorised using a structured typology. Symptom frequency increased in line with dementia severity. Disturbances were generally well tolerated. Most were ignored where possible, except for wandering from home. Other common responses included avoiding triggers, providing reassurance, reality orientation, diversion, and collusion with false beliefs. Restrictive or punitive responses were uncommon. Few carers articulated clear strategies to deal with behavioural and psychological symptoms. For most, tolerance proved more effective and less distressing than arguments and reprimands. Carers' responses are likely to be influenced by social and cultural factors and may differ in other settings.

Wark, S., Hussain, R., & Parmenter, T.
Down syndrome and dementia: Is depression a confounder for accurate diagnosis and treatment?
Journal of Intellectual Disabilities, 2014, 18(4), 305-314. doi: 10.1177/1744629514552152.
Abstract: The past century has seen a dramatic improvement in the life expectancy of people with Down syndrome. However, research has shown that individuals with Down syndrome now have an increased likelihood of early onset dementia. They are more likely than their mainstream peers to experience other significant co-morbidities including mental health issues such as depression. This case study reports a phenomenon in which three individuals with Down syndrome and dementia are described as experiencing a rebound in their functioning after a clear and sustained period of decline. It is hypothesized that this phenomenon is not actually a reversal of the expected dementia trajectory but is an undiagnosed depression exaggerating the true level of functional decline associated with the dementia. The proactive identification and treatment of depressive symptoms may therefore increase the quality of life of some people with Down syndrome and dementia.

Washington, S.E., Cler., Lowery, C., & Stark, S.L.
Down syndrome and Alzheimer's disease: A scoping review of functional performance and fall risk.
Alzheimers Dement (N Y). 2023 May 22;9(2):e12393. doi: 10.1002/trc2.12393. eCollection 2023 Apr-Jun.
Abstract: Alzheimer's disease (AD) occurs in aging adults with Down syndrome (DS) at a higher prevalence and an earlier age than in typical aging adults. As with the general aging adult population, there is an urgent need to understand the preclinical and early phases of AD progression in the adult population with DS. The aim of this scoping review was to synthesize the current state of the evidence and identify gaps in the literature regarding **functional activity performance and falls and their significance to disease staging** (i.e., mild, moderate, and severe defined staging criteria) in relation to Alzheimer's disease and related dementias (ADRD) in adults with DS. This scoping review included six electronic databases (e.g., PsycInfo, Academic Search Complete, CINAHL, COCHRANE Library, MEDLINE, and PubMed). Eligible studies included participants with DS ≥25 years of age, studies with functional measures and/or outcomes (e.g., activities of daily living, balance, gait, motor control, speech, behavior, and cognition; falls; and fall risks), and studies that investigated AD pathology and implications. Fourteen eligible studies were included and categorized through a thematic analysis into the following themes: (1) physical activity and motor coordination (PAMC), (2) cognition, (3) behavior, and (4) sleep. The studies indicated how functional activity performance and engagement may contribute to early identification of those at risk of cognitive decline and AD development and/or progression. There is a need to expand the research regarding ADRD pathology relative to functional outcomes in adults with DS. **Functional measures related to disease staging and cognitive impairment are essential to understanding how AD progression** is characterized within real-world settings. This scoping review identified the need for additional mixed-methods research to examine the use of assessment and intervention related to function and its detection of cognitive decline and AD progression.

Watchman, K., Kerr, D., & Wilkinson, H.
Supporting Derek: A new resource for staff working with people who have a learning difficulty and dementia.
58 pp.
York, United Kingdom: Joseph Rountree Foundation (2010)

Access: http://www.jrf.org.uk/publications/supporting-derek
Abstract: This resource pack published by the Joseph Rowntree Foundation in partnership with the University of Edinburgh, is aimed at staff supporting people with intellectual disability who develop dementia. Its focus in on helping care staff and training officers from intellectual disability and dementia care settings, as well as community, housing and health care staff. The pack is composed of 10 topic area (chapters), including basics on dementia, understanding behavior, development care environments, pain, communication, meaningful activities, friends with dementia, nutrition and hydration, night-time care, and palliative care. The pack includes a DVD and training materials which cover many of the key issues related to diagnosing and responding to dementia in people with intellectual disabilities. A short drama included on the DVD (acted by people with an intellectual disability) provides an insight into the reality of dementia and how it might feel to the individual affected.

Watchman, K.
Critical issues for service planners and providers of care for people with Down's syndrome and dementia.
British Journal of Learning Disabilities, 2003, 31(2), 81-84.
https://doi.org/10.1046/j.1468-3156.2003.00228.x
Abstract: This discussion paper raises critical issues that need to be addressed along with suggestions as to how they may be met with. Author notes that the role of service planners and providers of care is one that cannot be understated while considering the future needs of people with Down's syndrome and dementia. Discussed are appropriateness of accommodations, care management, diagnosis, and training.

Watchman, K.
Why wait for dementia?
Journal of Learning Disabilities, 2003, 7, 221-230.
https://doi.org/10.1177/14690047030073
Abstract: Adults with Down syndrome living in supported accommodation, who develop dementia, may also experience other preventable difficulties caused by the environment in which they live. This can result in their enforced move to a different accommodation. Yet it is known that it is beneficial for people with intellectual disabilities and dementia to remain in familiar surroundings for as long as possible. This article puts forward a new set of guidelines suggesting the modification of the living environment of adults with Down syndrome before they develop dementia. The guidelines are discussed along with possible barriers to their implementation.

Watchman, K.
Practitioner-raised issues and end-of-life care for adults with Down syndrome and dementia
Journal of Policy and Practice in Intellectual Disabilities, 2005, 2(2), 156-162.
https://doi.org/10.1111/j.1741-1130.2005.00026.x
Abstract: The author interviewed a small group of practitioners working in intellectual disability and palliative care settings about their perceptions of a number of end-of-life issues related to people with Down syndrome who were affected by dementia. The study, which took place in Scotland, identifed a number of issues and perceptions expressed by the subjects as well as gaps in services and practice. Key among the findings were the need for people with Down syndrome to be more involved in planning for their own end-of-life care; a lack of communication between those persons working in palliative care and intellectual disability settings; identification of a “care culture clash;” deficits in training programs for staff involving dying, death, and bereavement; and that end-of-life care for people with Down syndrome and dementia is a neglected area of research. The author highlights the lack of uniform practice when working with people with Down syndrome in the end stages of dementia and provides some recommendations for further discourse and research.

Watchman, K.
Changes in accommodation experienced by people with Down syndrome and dementia in the first five years after diagnosis
Journal of Policy and Practice in Intellectual Disabilities, 2008, 5(1), 65-68.
https://onlinelibrary.wiley.com/doi/abs/10.1111/j.1741-1130.2007.00140.x
Abstract: Research that has tracked living situation changes is lacking for people with Down syndrome post-diagnosis of dementia. Extant studies have not considered reasons for a move, the stage at which it happened, and how involved in the decision the person with Down syndrome was. To study this, a postal questionnaire was used with 35 carers of persons with Down syndrome who had been diagnosed with dementia during the previous five years. Results showed that there are fewer accommodation changes in the early stages of dementia among people with Down syndrome than have previously been suspected and that confusion exists over the interpretation of existing care models. Findings also revealed that adults with Down syndrome were often denied the opportunity to take part in discussions about their future accommodations and there was a lack of forward planning on the part of carers.

Watchman, K.
People with a learning disability and dementia: reducing marginalisation
Journal of Dementia Care [Research Review], 2012, 20(5), 34- 39.
http://www.learningdisabilityanddementia.org/uploads/1/1/5/8/11581920/journal_of_dementia_care.pdf
Abstract: The awareness that people with a learning disability, particularly Down’s syndrome, are at risk of dementia at a younger age brings an associated need for clarity over service planning and delivery. In order to record changes and developments in approaches, research literature documents the changing history of people with a learning disability and, separately, people with dementia. We not have the same knowledge about the most appropriate ways of supporting individuals who have both a learning disability and dementia. People will already experience social exclusion due to society’s interpretation of their learning disability and this review identifies from the literature factors that have contributed to the further marginalization of this group. The review highlights the need for accurate data and statistics, an individualized approach to sharing information about the diagnosis, general and specialist training, an increased use of adapting methods of communication as dementia progresses and a consistent staff approach across care settings.

📖 **Watchman, K**.
Intellectual Disability and Dementia: Research into Practice.
336 pp.
London/Philadelphia: Jessica Kingsley Publishers (2014).
Abstract: In 16 chapters, this edited text offers a balanced appraisal of the evidence base on people with intellectual disabilities who develop dementia. It includes a range of resources, and is split into three sections that address the following: (1) The association between intellectual disabilities and dementia: what do we know? (2) Experiences of dementia in people with intellectual disabilities: how do we know?, and (3) Service planning: what are we going to do? Section one explores issues such as defining and diagnosing dementia in people with intellectual disabilities, prevalence and incidence and treatment options. The authors explain the differing theories about why people with Down’s syndrome are more likely to experience dementia, which provides a useful foundation for discussions about the use of medication. Section two explores the perspectives of people with learning disabilities and their families and the experiences of families via case studies. This section also explores some checklists for use with family members to help plan for the future. Section three focuses on service planning by describing a framework that can be used by practitioners for discussing diagnosis and prognosis of dementia. This section also considers the issues related to ageing in place and dementia-specific services and suggests that training is important for staff supporting those with learning disabilities and dementia.

Watchman, K.

Stand by Me - Perspectives of older couples with a learning disability when one partner has dementia: identifying support needs of carer dyads. (Version 1). University of Stirling, Faculty of Health Sciences and Sport. 2023, Dataset. http://hdl.handle.net/11667/212
Abstract: The risk of dementia at an earlier age among people with a learning disability, especially people with Down's syndrome, is known. Experiences of carer dyads in dementia care is typically limited to couples who do not have a learning disability, with no previous evidence base informing how each partner with a learning disability may best be supported and how relationships may be sustained. Prior to this research, it was not known what coping mechanisms were developed or how couples wished to be supported. Evidence to inform practice was needed to support not just couples with a learning disability, but also staff and family members who find themselves at the intersection between learning disability and dementia. This study used life story interviews to identify the perspectives and support needs of couples with a learning disability where one partner has/had dementia. Social care support staff and family members of one or both partners were also interviewed with participants recruited from Scotland and England. Thematic analysis was conducted highlighting struggles for some couples to have their relationship accepted and past experiences of group living or institutional care which influenced perceptions of longer-term support. Factors that supported sustainability in relationships included access to consistent support networks, support to understand the diagnosis, recognition of identities - firstly as wife and husband or partners, later a shift to carer, and counselling for people with a learning disability. Anonymised interview transcripts from 5 participants with a learning disability, four family members and nine learning disability support staff, plus NVivo analysis and accessible study information are provided within this dataset.

Watchman, K., Wilkinson, H., & Hare, P.
Supporting People with Learning Disabilities and Dementia: Practice Development Guide (A training pack for support staff based on the Supporting Derek film and guide). (2018).
33pp
Pavilion Publishing and Media Ltd, Rayford House, School Road, Hove BN3 5HX UK

Abstract: This practice development guide has been developed to support participants attending the training course Supporting People with Learning Disabilities and Dementia. (There is a separate and more detailed self-study guide for support staff and carers available that can be purchased at www.pavpub.com/supporting-people-with-learning-disabilities-and-dementia/.) The guide provides information on ten topics: Learning disability and dementia, Understanding behaviour, developing supportive and meaningful environments, responding to pain, effective communication, meaningful activities, when a friend has dementia, eating and drinking, night-time care, and advanced dementia.

Watchman, K., Janicki, M.P., and the members of the International Summit on Intellectual Disability and Dementia
The intersection of intellectual disability and dementia: Report of the international summit on intellectual disability and dementia
Gerontologist, 2019, 59(3), 411-419. doi: 10.1093/geront/gnx160.
Abstract: An International Summit on Intellectual Disability and Dementia, held in Glasgow, Scotland (October 13-14, 2016) drew individuals and representatives of numerous international and national organizations and universities with a stake in issues affecting adults with intellectual disability (ID) affected by dementia. A discussion-based consensus process was used to examine and produce a series of topical reports examining three main conceptual areas: (1) human rights and personal resources (applications of the Convention for Rights of People with Disabilities and human rights to societal inclusion, and perspectives of persons with ID), (2) individualized services and clinical supports (advancing and advanced dementia, post-diagnostic supports, community supports and services, dementia-capable care practice, and end-of-life care practices), and (3) advocacy, public impact, family caregiver issues (nomenclature/ terminology, inclusion of persons with ID in national plans, and family caregiver issues). Outcomes included recommendations incorporated into a series of publications and topical summary bulletins designed to be international resources, practice guidelines, and the impetus for planning and advocacy with, and on behalf of, people with ID affected by dementia, as well as their families. The general themes of the conceptual areas are discussed and the main recommendations are associated with three primary concerns.

Watchman, K., Janicki, M.P., Splaine, M., Larsen, F.K., Gomiero, T., & Lucchino, R.
International summit consensus statement: Intellectual disability inclusion in national dementia plans.
American Journal of Alzheimer's Disease & Other Dementias, 2017, Jun, 32(4), 230-237 https://doi.org/10.1177/1533317517704082
Abstract: The World Health Organization (WHO) has called for the development and adoption of national plans or strategies to guide public policy and set goals for services, supports, and research related to dementia. It called for distinct populations to be included within national plans, including adults with intellectual disability (ID). Inclusion of this group is important as having Down's syndrome is a significant risk factor for early-onset dementia. Adults with other ID may have specific needs for dementia-related care that, if unmet, can lead to diminished quality of old age. An International Summit on Intellectual Disability and Dementia, held in Scotland, reviewed the inclusion of ID in national plans and recommended that inclusion goes beyond just description and relevance of ID. Reviews of national plans and reports on dementia show minimal consideration of ID and the challenges that carers face. The Summit recommended that persons with ID, as well as family carers, should be included in consultation processes, and greater advocacy is required from national organizations on behalf of families, with need for an infrastructure in health and social care that supports quality care for dementia.

Watchman, K., Janicki, M.P., Udell, L., Hogan, M., Quinn, S., Beránková, A.
Consensus statement of the International Summit on Intellectual Disability and Dementia on valuing the perspectives of persons with intellectual disability.
Journal of Intellectual Disabilities,2019 Jun;23(2):266-280. doi: 10.1177/1744629517751817. Epub 2018 Jan 17.
Abstract: The International Summit on Intellectual Disability and Dementia covered a range of issues related to dementia and intellectual disability, including the dearth of personal reflections of persons with intellectual disability affected by dementia. This article reflects on this deficiency and explores some of the personal perspectives gleaned from the literature, from the Summit attendees and from the experiences of persons with intellectual disability recorded or scribed in advance of the two-day Summit meeting. Systemic recommendations included reinforcing the value of the involvement of persons with intellectual disability in (a) research alongside removing barriers to inclusion posed by institutional/ethics review boards, (b) planning groups that establish supports for dementia and (c) peer support. Practice recommendations included (a) valuing personal perspectives in decision-making, (b) enabling peer-to-peer support models.

Watchman, K., Mattheys, K., & McKernon, M.
Effects of the implementation of non-drug Interventions on behaviour and psychosocial symptoms of dementia in people who have an Intellectual disability
Journal of Intellectual Disability Research, 2019, 63(7), 647.
https://onlinelibrary.wiley.com/doi/epdf/10.1111/jir.12651
Abstract: This three-year study investigates if non-drug interventions result in positive changes in behaviour associated with dementia in people with intellectual disability. People with intellectual disability are involved as advisors (n = 1) and co-researchers (n = 4) in both cycles. Cycle 1 (concluded) included 7 participants with intellectual disability in the early stage of dementia (4 with Down syndrome) and 12 support staff. Cycle 2 (ongoing) includes participants who have a more profound intellectual disability, and/or are experiencing advanced dementia. In both cycles, a goal-setting tool firstly helped to identify individualised

non-drug interventions. In Cycle 1, a pre- and post-behaviour change tool (NPI-Q), was completed alongside semi-structured interviews, a bespoke tool to measure 'in the moment' changes, intervention diaries, and photovoice. Cycle 1 interventions included reminiscence, life story, music playlists, cookery, aromatherapy, environmental design change, exercise and cognitive games. Of 239 separate intervention over a 6-month period in Cycle 1, 193 resulted in positive behaviour change with 75% of goals being achieved or exceeded. The study offers insight into the support of people with intellectual disability and dementia. Use of non-drug supports in response to distress has led to cultural change within participating organizations with less reliance on medication as a first response.

Watchman, K., Mattheys, K., McKernon, M., Strachan, H., Andreis, F., & Murdoch, J.
A person-centred approach to implementation of psychosocial interventions with people who have an intellectual dis☻ability and dementia-A participatory action study.
Journal of Applied Research in Intellectual Disabilities, 2021, 34(1), 164-177. https://doi.org/10.1111/jar.12795
Abstract: Numbers of people with an intellectual disability and dementia present a global health and social challenge with associated need to reduce stress or agitation and improve quality of life in affected individuals. This study aimed to identify effectiveness of psychosocial interventions in social care settings and, uniquely, explore use of photovoice methodology to develop dialogue about dementia. This mixed-method participatory action study used individualized goal-setting theory with 16 participants with intellectual disability and dementia, and 22 social care staff across 11 sites. Five co-researchers with intellectual disability were part of an inclusive research team collecting data using existing and bespoke tools including photovoice. Analysis used descriptive and inferential statistics and framework analysis. Seventy four percentage of individual goals met or exceeded expectations with reduction in some "as required" medication. Qualitative findings include themes of enabling care and interventions as tools for practice. Photovoice provided insight into previously unreported fears about dementia. Individualized psychosocial interventions have potential to reduce distress or agitation.

Watchman, K., McKernon, M., Boustead, L., & Doyle, A.
Intellectual disability and dementia: Understanding the effectiveness of psychosocial interventions
Journal of Applied Research in Intellectual Disabilities, 2021, 34(5),1273. https://onlinelibrary.wiley.com/doi/epdf/10.1111/jar.12763
Abstract: The study aim was to identify effectiveness of psychosocial interventions with people who have an intellectual disability and dementia. This mixed-method participatory action study used goal-setting theory with 16 participants with intellectual disability and dementia, and 22 social care staff across 11 sites. Five core researchers with intellectual disability were part of an inclusive research team collecting data using existing and bespoke tools, including photovoice. Psychosocial interventions included: music playlists, reminiscence, animal therapy, robotic animals, and design changes. Analysis used descriptive and inferential statistics and framework analysis. Found that 74% of individual goals met or exceeded expectations with reduction in some "as required" medication. Qualitative findings include themes of enabling care and interventions as tools for practice. Photovoice provided insight into previously unreported fears about dementia. This poster combines an easy-read and pictorial summary of the study. Recommendations are made to maximise wellbeing and ensure the perspectives of people with dementia are heard: medication review, design changes to the home of a person with dementia, staff training, and talking about dementia more with people who have an intellectual disability. Individualized psychosocial interventions have potential to reduce distress or agitation in persons with intellectual disability and dementia, and to increase quality of life.

📖 **Warner, M.L.**
The complete guide to Alzheimer's-proofing your home.
470 pp.
West Lafayette, Indiana: Purdue University Press (1998)
Abstract: General text on adapting homes and living environments for persons with dementia; applicable to home and other residential situations for adults with intellectual disabilities and dementia.

Webber, R., Bowers, B., & Bigby, C
Confidence of group home staff in supporting the health needs of older residents with intellectual disability
Journal of Intellectual and Developmental Disabilities, 2016, 41(2), 107-114. doi.org/10.3109/13668250.2015.1130218
Abstract: Increased life expectancy for people with intellectual disability is accompanied by increased age-related health concerns. People ageing with intellectual disability experience more health conditions and are relocated to aged care earlier than their age peers. Group home staff were surveyed about their (a) training and confidence in 11 health conditions and 7 health procedures, and (b) attitude to relocating residents with health needs to aged care. Staff training in each of 10 health conditions and 7 health procedures was positively associated with increased confidence in supporting residents with those health issues. Higher staff confidence in caring for residents with 9 conditions and requiring 4 procedures was negatively associated with a likelihood of recommending that a person with those health needs should relocate to aged care. Targeted training of staff in age-related health issues may contribute to better health care and delay residents relocating to aged care.

Webber, R., Bowers, B. & McKenzie-Green, B.
Staff responses to age-related health changes in people with an intellectual disability in group homes.
Disability and Society, 2010, 25(6), 657-671. https://doi.org/10.1080/09687599.2010.505736
Abstract: The purpose of this study was to explore how supervisors in group homes caring for people with intellectual disability responded to the development of age-related health changes in their residents. Ten group home supervisors working in the disability sector were interviewed once. Data were analyzed using Dimensional Analysis. The study identified several factors related to whether a resident could stay 'at home' or would need to be moved to residential aged care (nursing home) including: nature and extent of group home resources, group home staff comfort with residents' health changes, staff skill at navigating the intersection between the disability and ageing sectors, and the supervisor's philosophy of care. The ability of older people with an intellectual disability to 'age in place' is affected by staff knowledge about and comfort with age-related illnesses, staff skills at navigating formal services, staffing flexibility, and the philosophy of group home supervisors. Despite the growing international concern for the rights of people with disability, particularly in relation to decision making, questions about the older person's choice of residence and participation in decision making about what was best for them, were almost nonexistent. Rather, decisions were made based on what was considered to be in 'the best interest.

Wegiel, J., Flory, M., Kuchna, I. Nowicki, K., Wegiel, J., Ma, S.Y., Zhong, N., Bobrowicz, T.W., de Leon, M., Lai, F., Silverman, W.P.,& Wisniewski, T.
Developmental deficits and staging of dynamics of age associated Alzheimer's disease neurodegeneration and neuronal loss in subjects with Down syndrome.
Acta Neuropathologica Communications, 2022, 10(1), 2. https://doi.org/10.1186/s40478-021-01300-9
Abstract: The increased life expectancy of individuals with Down syndrome (DS) is associated with increased prevalence of trisomy 21-linked early-onset Alzheimer's disease (EOAD) and dementia. The aims of this study of 14 brain regions including the entorhinal cortex, hippocampus, basal ganglia, and cerebellum in 33 adults with DS 26-72 years of age were to identify the magnitude of brain region-specific developmental neuronal deficits contributing to

intellectual deficits, to apply this baseline to identification of the topography and magnitude of neurodegeneration and neuronal and volume losses caused by EOAD, and to establish age-based staging of the pattern of genetically driven neuropathology in DS. **Both DS subject age and stage of dementia, themselves very strongly correlated, were strong predictors of an AD-associated decrease of the number of neurons, considered a major contributor to dementia**. The DS cohort was subclassified by age as pre-AD stage, with 26-41-year-old subjects with a full spectrum of developmental deficit but with very limited incipient AD pathology, and 43-49, 51-59, and 61-72-year-old groups with predominant prevalence of mild, moderately severe, and severe dementia respectively. This multiregional study revealed a 28.1% developmental neuronal deficit in DS subjects 26-41 years of age and 11.9% AD-associated neuronal loss in DS subjects 43-49 years of age; a 28.0% maximum neuronal loss at 51-59 years of age; and a 11.0% minimum neuronal loss at 61-72 years of age. A total developmental neuronal deficit of 40.8 million neurons and AD-associated neuronal loss of 41.6 million neurons reflect a comparable magnitude of developmental neuronal deficit contributing to intellectual deficits, and AD-associated neuronal loss contributing to dementia. This highly predictable pattern of pathology indicates that successful treatment of DS subjects in the fourth decade of life may prevent AD pathology and functional decline.

Whitaker, S., & Read, S.
The prevalence of psychiatric disorders among people with intellectual disabilities: An analysis of the literature
Journal of Applied Research in Intellectual Disabilities, 2006, 19(4), 330-345. Doi: https://doi.org/10.1111/j.1468-3148.2006.00293.x
Abstract: It has often been stated that the prevalence of psychiatric disorders in people with intellectual disabilities is greater than it is in the population as a whole. The epidemiological studies on psychiatric disorders in people with intellectual disabilities were reviewed. There is evidence that the prevalence of psychiatric disorder is greater in children with intellectual disabilities, compared with children with normal IQs, that it is higher in both adults and children with severe intellectual disabilities compared with people with mild or no intellectual disability and that the rate of problematic behavior is higher in both adults and children having intellectual disabilities, than in their non-disabled peers. There is no sound evidence that the prevalence of psychiatric disorders in adults with mild intellectual disability is greater than in the population as a whole.

Whitehouse, R., Chamberlain, P., & Tunna, K.
Dementia in people with learning disability: a preliminary study into care staff knowledge and attributions
British Journal of Learning Disabilities, 2000, 28(4) 148- 153. https://doi.org/10.1046/j.1468-3156.2000.00057.x
Abstract: This paper describes the findings of a pilot study funded by the NHS Executive Primary and Community Care Research Initiative Small Projects Scheme that investigated the knowledge and attributions of dementia held by care staff who work with older adults with learning disability. Meetings took place with 21 members of care staff identified as “keyworkers” to older adults with learning disability living in residential houses provided by Solihull Healthcare NHS Trust, Solihull, UK. The results suggest that staff have knowledge of ageing at a similar level to that of college students. Forgetfulness was the sign that they would most expect to see if they thought someone was suffering from dementia. When a change in behavior was attributed to dementia, it was most likely to be viewed as 'stable, uncontrollable' with staff feeling pessimistic about being able to change the behavior.

Whittick, J.E.
Dementia and mental handicap: attitudes, emotional distress and caregiving
British Journal of Medical Psychology, 1989, 62, 181-189. doi: 10.1111/j.2044-8341.1989.tb02825.x.
Abstract: Against the current climate of hospital closure programs and community care, attitudes to caregiving were examined in three groups of carers, namely mothers caring for a mentally handicapped child, mothers caring for a mentally handicapped adult and daughters caring for a parent with dementia. An 'attitude questionnaire' was developed by the author and administered, postally, to the three groups. Daughters were found to be more likely than the mothers to see their caring role in a negative way and were more inclined to favor institutional care. Possible reasons for this are discussed. The relationship between attitudes and emotional distress (as measured by the GHQ-30) were also examined for the sample as a whole. Negative and pro-institutional attitudes towards the caregiving situation were associated with elevated levels of emotional distress. Implications at both a local and a national level for all those involved with carers are discussed in the light of these findings.

Whitwham, S., McBrien, J., & Broom, W.
Should we refer for a dementia assessment? A checklist to help know when to be concerned about dementia in adults with Down syndrome and other intellectual disabilities.
British Journal of Learning Disabilities, 2011, 39(1), 17-21. https://doi.org/10.1111/j.1468-3156.2009.00606.x
Abstract: The aim of this research was to develop a simple screening checklist to help carers and professionals know when to make a referral for a dementia assessment. A checklist was completed for all new referrals to a dementia service for people with intellectual disabilities. The obtained scores were compared to the diagnostic outcome of a comprehensive dementia assessment. The data (n = 159) indicate a higher score on the checklist correlates significantly with a subsequent diagnosis of dementia. Cut-off scores are explored. The checklist appears to be a useful tool to prompt referrals for a full dementia assessment. By helping the referrer to know when to be concerned about dementia, it may reduce the number of people referred late or not at all.

Wiener, J.M., & Pazzaglia, F.
Ageing- and dementia-friendly design: theory and evidence from cognitive psychology, neuropsychology and environmental psychology can contribute to design guidelines that minimise spatial disorientation.
Cognitive Processing, 2021, 22, 715-730 (2021). https://doi.org/10.1007/s10339-021-01031-8
Abstract: Many older people, both with and without dementia, eventually move from their familiar home environments into unfamiliar surroundings, such as sheltered housing or care homes. Age-related declines in wayfinding skills can make it difficult to learn to navigate in these new, unfamiliar environments. To facilitate the transition to their new accommodation, it is therefore important to develop retirement complexes and care homes specifically designed to reduce the wayfinding difficulties of older people and those with Alzheimer's disease (AD). Residential complexes that are designed to support spatial orientation and that compensate for impaired navigation abilities would make it easier for people with dementia to adapt to their new living environment. This would improve the independence, quality of life and well-being of residents, and reduce the caregivers' workload. Based on these premises, this opinion paper considers how evidence from cognitive psychology, neuropsychology and environmental psychology can contribute to ageing- and dementia-friendly design with a view to minimising spatial disorientation. After an introduction of the cognitive mechanisms and processes involved in spatial navigation, and the changes that occur in typical and atypical ageing, research from the field of environmental psychology is considered, highlighting design factors likely to facilitate (or impair) indoor wayfinding in complex buildings. Finally, psychological theories and design knowledge are combined to suggest ageing- and dementia-friendly design guidelines that aim to minimize spatial disorientation by focusing on residual navigation skills.

Wiese, M., Stancliffe, R.J., Dew, A., Balandin, S., & Howarth, G.
What is talked about? Community living staff experiences of talking with older people with intellectual disability about dying and death
Journal of Intellectual Disability Research, 2014 Jul, 58(7), 679-690. doi: 10.1111/jir.12065

Abstract: This study explored what community living staff talked about and did with people with intellectual disability (ID) to assistthem to understand dying and death. Guided by grounded theory methodology,focus groups and one-to-one interviews were conducted with 22 staff who had talked about any topic relating to dying and death with their clients. There was little evidence that staff talked with, or did things with clients to assist understanding of the end of life, both prior to and after a death. Prior to death staff assisted clients in a limited way to understand about determining wishes in preparation for death, and what dying looks like by observance of its passage. Following a death staff offered limited assistance to clients to understand the immutability of death, and how thedead can be honored with ritual, and remembered.
The findings have implications for why people with ID have only partial understanding of the end of life, the staff skills required to support clients' understanding, and when conversations about the end of life should occur

Wilkinson, H., Janicki, M.P., & Edinburgh Working Group on Dementia Care Practices (EWGDCP).
The Edinburgh Principles with accompanying guidelines and recommendations.
Journal of Intellectual Disability Research, 2002, 46, 279-284. doi: 10.1046/j.1365-2788.2002.00393.x.
Abstract: A panel of experts attending a 3-day meeting held in Edinburgh, UK, in February 2001 was charged with producing a set of principles outlining the rights and needs of people with intellectual disability (ID) and dementia, and defining service practices which would enhance the supports available to them. The Edinburgh Principles, seven statements identifying a foundation for the design and support of services to people with ID affected by dementia, and their carers, were the outcome of this meeting. The accompanying guidelines and recommendations document provides an elaboration of the key points associated with the Principles and is structured toward a four-point approach: (1) adopting a workable philosophy of care; (2) adapting practices at the point of service delivery; (3) working out the coordination of diverse systems; and (4) promoting relevant research. It is expected that the Principles will be adopted by service organizations world-wide, and that the accompanying document will provide a useful and detailed baseline from which further discussions, research efforts and practice development can progress.

Wilkinson, H., Kerr, D., & Rae, C.
People with a learning disability: their concerns about dementia
Journal of Dementia Care, 2003, 11(1), 27-29
https://www.researchgate.net/publication/294334582_People_with_a_learning_disability_Their_concerns_about_dementia
Abstract: With people with a learning disability live longer, more of them are developing dementia. In planning the services they need, an important first step is to ask them what they think. Authors report information from surveying a group of older adults with intellectual disabilities.

Wilkinson, H., Kerr, D., & Cunningham, C.
Equipping staff to support people with an intellectual disability and dementia in care home settings.
Dementia -The International Journal of Social Research and Practice, 2005, 4(3), 387-400. https://doi.org/10.1177/147130120505
Abstract: The knowledge, experiences and skills of direct care staff working in care home settings are essential in ensuring a good quality of life and care for a person with an intellectual disability (ID) who develops dementia. Drawing on the findings of a wider study, the issues of training, support and the wider needs of staff when trying to support a resident who develops dementia are explored, specifically as relating to the role played by staff and the need to determine their experiences and related training needs. Following an introduction to the policy and practice context for working with people with an ID and dementia, and a brief description of the research method, the authors discuss the attitudes and practices of staff; supportive changes at an organizational level; and the knowledge and training needs of staff and specific gaps in knowledge. The authors argue that, within the policy and practice context of aiming to support residents to 'age in place', support for staff is a crucial aspect of ensuring that such an approach is effective and provides a coordinated approach to planning, resourcing and support.

Wilkinson H., & Watchman, K.
Down's Syndrome and dementia: A framework for practice to support people with Down's Syndrome and dementia living in generic care homes
In *Supportive Care for the Person with Dementia, Julian Hughes, Mari Lloyd-Williams,& Greg Sachs (Editors).* Oxford University Press, Jan 2011. DOI:10.1093/acprof:oso/9780199554133.003.0009
Abstract: Despite the recognized longevity of people with Down's syndrome, the age-related long-term care needs of this population have yet to be fully understood and addressed. Owing to an increase in medical and social interventions, people with Down's syndrome are living longer. This has led to the current awareness that there are a number of illnesses in later life that people with Down's syndrome are susceptible to, one of which is dementia. This chapter focuses on generic care provision and, in particular, the use by people with Down's syndrome and dementia of mainstream care home provision for older people when they are moved from other services, or need a placement when home or family care structures break down. It provides an overview of the demographics and policy imperative in developing stronger policy and practice responses to the needs of people growing older with Down's syndrome and dementia. It then turns to the proposed framework using a case study to illustrate the urgent requirement for more detailed research evidence and policy responses to this underserved area of practice.

Wicki, M.T.,& Riese, F.
Prevalence of dementia and organization of dementia care in Swiss disability care homes.
Disability and Health Journal, 2016 Oct, 9(4), 719-723. doi: 10.1016/j.dhjo.2016.05.008.
Abstract: With higher life expectancy an increasing number of people with intellectual disability (PWID) are at risk for developing dementia. Since PWID are an often neglected patient population, the objective of this study was to investigate the prevalence of dementia in residential disability group homes in Switzerland and to describe how residential homes organize dementia care. All residential group homes for adults with disabilities in Switzerland (N = 437) were invited to participate in a cross-sectional survey. A subset of questions covered the number of residents with diagnosed and suspected dementia and the organization of dementia care. The response rate to the dementia-related questions was 32% (n = 140 care homes with 10403 residents). In residential homes specialized in PWID, 5.8% of the residents were reported to have a diagnosed or suspected dementia. In 140 deaths of PWID, 26% (n = 37) died with a diagnosed or suspected dementia. Residential homes for PWID mostly rely on internal resources (67.7%), general practitioners (61.3%) or psychiatrists (45.2%) for the care of residents with dementia, while specialized dementia nurses are rarely involved (16.1%). This is the first study in Switzerland to assess the prevalence of dementia in PWID. The study indicates a diagnostic gap. Dementia care is provided in a heterogeneous way across Swiss residential group homes for people with disability. Since the number of PWID requiring such care will likely increase in the future, best-practice examples and guidelines are needed.

Wilson, B., Jones, K.B., Weedon, D., & Bilder, D.
Care of adults with intellectual and developmental disabilities: Down syndrome.
FP Essentials, 2015 Dec, ;439, 20-25.
https://pubmed.ncbi.nlm.nih.gov/26669211/ PMID: 26669211.
Abstract: Down syndrome (DS) is a genetic disorder involving excess genetic material from chromosome 21. The incidence of DS is increasing, and the life expectancy for individuals with DS has increased to a median age of 55 years. Adults with DS are at increased risk of several conditions, including significant neurologic, cardiovascular, pulmonary, gastrointestinal, musculoskeletal, endocrine, psychiatric, hematologic, and social comorbidities, and additional

screening or monitoring may be needed. Additional preventive measures for patients with DS include regular screening for thyroid dysfunction, hearing loss, eye disorders, heart disease, osteoporosis, and dementia, and one-time vaccination with the polyvalent pneumococcal polysaccharide vaccine (PPV23). Quality of life should be the main focus of treatment, with patients being involved in medical decisions as much as possible.

Wisch, J.K., McKay, N.S., Boerwinkle, A.H., Kennedy, J., Flores, S., Handen, B.L., Christian, B.T., Head, E., Mapstone, M., Rafii, M.S., O'Bryant, S.E., Price, J.C., Laymon, C.M., Krinsky-McHale, S.J., Lai, F., Rosas, H.D., Hartley, S.L., Zaman, S., Lott, I.T., Tudorascu, D., Zammit, M., Brickman, A.M., Lee, J.H., Bird, T.D., Cohen, A., Chrem, P., Daniels, A., Chhatwal, J.P., Cruchaga, C., Ibanez, L., Jucker, M., Karch, C.M., Day, G.S., Lee, J.H., Levin, J., Llibre-Guerra, J., Li, Y., Lopera, F., Roh, J.H., Ringman, J.M., Supnet-Bell, C., van Dyck, C.H., Xiong, C., Wang, G., Morris, J.C., McDade, E., Bateman, R.J., Benzinger, T.L.S., Gordon, B.A., Ances, B.M.; Alzheimer's Biomarker Consortium-Down syndrome; Dominantly Inherited Alzheimer Network.
Comparison of tau spread in people with Down syndrome versus autosomal-dominant Alzheimer's disease: a cross-sectional study.
Lancet Neurology, 2024 May; 23(5), 500-510. doi: 10.1016/S1474-4422(24)00084-X. PMID: 38631766.
Abstract: In people with genetic forms of Alzheimer's disease, such as in Down syndrome and autosomal-dominant Alzheimer's disease, pathological changes specific to Alzheimer's disease (ie, accumulation of amyloid and tau) occur in the brain at a young age, when comorbidities related to ageing are not present. Studies including these cohorts could, therefore, improve our understanding of the early pathogenesis of Alzheimer's disease and be useful when designing preventive interventions targeted at disease pathology or when planning clinical trials. We compared the magnitude, spatial extent, and temporal ordering of tau spread in people with Down syndrome and autosomal-dominant Alzheimer's disease. In this cross-sectional observational study, we included participants (aged ?25 years) from two cohort studies. First, we collected data from the Dominantly Inherited Alzheimer's Network studies (DIAN-OBS and DIAN-TU), which include carriers of autosomal-dominant Alzheimer's disease genetic mutations and non-carrier familial controls recruited in Australia, Europe, and the USA between 2008 and 2022. Second, we collected data from the Alzheimer Biomarkers Consortium-Down Syndrome study, which includes people with Down syndrome and sibling controls recruited from the UK and USA between 2015 and 2021. Controls from the two studies were combined into a single group of familial controls. All participants had completed structural MRI and tau PET (18F-flortaucipir) imaging. We applied Gaussian mixture modelling to identify regions of high tau PET burden and regions with the earliest changes in tau binding for each cohort separately. We estimated regional tau PET burden as a function of cortical amyloid burden for both cohorts. Finally, we compared the temporal pattern of tau PET burden relative to that of amyloid. We included 137 people with Down syndrome (mean age 38•5 years [SD 8•2], 74 [54%] male, and 63 [46%] female), 49 individuals with autosomal-dominant Alzheimer's disease (mean age 43•9 years [11•2], 22 [45%] male, and 27 [55%] female), and 85 familial controls, pooled from across both studies (mean age 41•5 years [12•1], 28 [33%] male, and 57 [67%] female), who satisfied the PET quality-control procedure for tau-PET imaging processing. 134 (98%) people with Down syndrome, 44 (90%) with autosomal-dominant Alzheimer's disease, and 77 (91%) controls also completed an amyloid PET scan within 3 years of tau PET imaging. Spatially, tau PET burden was observed most frequently in subcortical and medial temporal regions in people with Down syndrome, and within the medial temporal lobe in people with autosomal-dominant Alzheimer's disease. Across the brain, people with Down syndrome had greater concentrations of tau for a given level of amyloid compared with people with autosomal-dominant Alzheimer's disease. Temporally, increases in tau were more strongly associated with increases in amyloid for people with Down syndrome compared with autosomal-dominant Alzheimer's disease. Although the general progression of amyloid followed by tau is similar for people Down syndrome and people with autosomal-dominant Alzheimer's disease, we found subtle differences in the spatial distribution, timing, and magnitude of the tau burden between these two cohorts. These differences might have important implications; differences in the temporal pattern of tau accumulation might influence the timing of drug administration in clinical trials, whereas differences in the spatial pattern and magnitude of tau burden might affect disease progression.

Wisniewski, K., Howe, J., Williams, D. G., & Wisniewski, H. M.
Precocious aging and dementia in patients with Down's syndrome.
Biological Psychiatry, 1978,13, 619–627. PMID: 153156
Abstract: Studied 50 unselected institutionalized patients with Down's syndrome to determine the clinical course of precocious aging and mental and neurological deterioration. Significant differences were established in neurological and psychiatric abnormalities and mental deterioration in patients below and above age 35, indicating progressive changes in the CNS. Demonstrated were higher incidence of recent memory loss, impairment of short-term visual retention, frontal release signs, hypertonia, hyperreflexia, long-tract signs, and psychiatric problems. Also noted was the presence of external features of precocious aging. Down's syndrome appears to be a human chromosomal abnormality in which genetically determined biochemical defects leading to precocious aging and dementia can be studied.

Wisniewski, K.E., Wisniewski, H.M., & Wen, G.Y.
Occurrence of neuropathological changes and dementia of Alzheimer's disease in Down's syndrome.
Annals of Neurology,1985, Mar;17(3), 278-282. doi: 10.1002/ana.410170310.
Abstract: One hundred brains of patients with Downs syndrome (DS) who died in institutions for chronic care were examined for clinicopathological correlation of Alzheimer's disease. Fifty-one were below and 49 were above age 30 years at death. Tissues from the right, prefrontal, and hippocampal cortices were processed for microscopy using H&E and Bodian-periodic acid-Schiff impregnation. Morphometric evaluations of plaques and tangles were carried out. Plaques or plaques and tangles were found in the brains of 56 patients with DS, 7 below age 30 and 49 above that age. A history of dementia was evident in the medical records of 15 of these patients; of these only 2 were below the age of 30. The brains of the patients with DS who also had clinical dementia had more than twenty plaques or plaques and tangles per 1.5 X 10(6) micron 2 of cortex. The numbers of plaques and tangles found in the brains of the patients with DS above the age of 30 greatly increased with age but varied from brain to brain. These observations suggest a correlation among dementia, the density of plaques and tangles, and age. All 100 brains studied showed early arrest of brain growth and brain atrophy, a condition that may have been due to prenatal arrest of neurogenesis mainly in the granular cell layers, prenatal and postnatal arrest of synaptogenesis, and early aging. Plaques and tangles developed twenty to thirty years earlier and dementia was clinically detected at least three times more frequently (20 to 30%) in DS than it is known to occur in the non-DS population.

Wissing, M. B. G., Ulgiati, A. M., Hobbelen, J. S. M., De Deyn, P. P., Waninge, A., & Dekker, A. D.
The neglected puzzle of dementia in people with severe/profound intellectual disabilities: A systematic literature review of observable symptoms.
Journal of Applied Research in Intellectual Disabilities, 2022, 35, 24-45. https://doi.org/10.1111/jar.12920
Abstract: Dementia is increasingly prevalent in people with severe/profound intellectual disabilities. However, early detection and diagnosis of dementia is complex in this population. This study aimed to identify observable dementia symptoms in adults with severe/profound intellectual disabilities in available literature. A systematic literature search was conducted in PubMed, PsycINFO and Web of Science with an exhaustive search string using a combination of search terms for severe/profound intellectual disabilities and dementia/ageing. Eleven studies met inclusion criteria. Despite the few small-sized studies, a range of dementia symptoms were identified, subdivided in cognitive decline (e.g., memory loss, for getfulness, deterioration in speech, losses of social skills),

decline in activities of daily living (e.g., self-cares skills, everyday functioning/ skills), BPSD (e.g., apathy, aggression, irritability, altered eating/ drinking behaviour) as well as neurologic and other physical symptoms (e.g., incontinence, (late-onset) epilepsy, hypotonia, gait deterioration). Cognitive decline, behavioral and psychological alterations, decline in activities of daily living as well as neurological and physical changes were found. Only a very limited number of studies reported symptoms ascribed to dementia in adults with severe/profound intellectual disabilities. Given the complexity of signaling and diagnosing dementia, dedicated studies are required to unravel the natural history of dementia in this population.

Wissing, M.B.G., Dijkstra, R., van der Wal, I.A., Grootendorst, E.S., Hobbelen, J.S.M., van der Putten, A.A.J., De Deyn, P.P., Waninge, A., Dekker, A.D.
Dementia in people with severe/profound intellectual (and multiple) disabilities: applicability of items in dementia screening instruments for people with intellectual disabilities
Journal of Mental Health Research in Intellectual Disabilities, 2022, 15(4), 322-363. https://doi.org/10.1080/19315864.2022.2111737
Abstract: Diagnosing dementia in people with severe/profound intellectual (and multiple) disabilities (SPI(M)D) is complex. Whereas existing dementia screening instruments as a whole are unsuitable for this population, a number of individual items may apply. Therefore, this study aimed to identify applicable items in existing dementia screening instruments. Informant interviews about 40 people with SPI(M)D were conducted to identify applicable items in the Dementia Scale for Down Syndrome, Behavioral and Psychological Symptoms of Dementia in Down Syndrome II scale, Dementia Questionnaire for persons with Mental Retardation and Social competence Rating scale for people with Intellectual Disabilities. Among 193 items, 101 items were found applicable, categorized in β5 domains: behavioral and psychological functioning (60 items), cognitive functioning (25), motor functioning (6), activities of daily living (5) and medical comorbidities (5). Identifying applicable items for people with SPI(M)D is an essential step in developing a dedicated dementia screening instrument for this population.

Wissing, M.B.G., Fokkens, A.S., Dijkstra, R., Hobbelen, J.S. M., van der Putten, A.A.J., De Deyn, P.P., Waninge, A., & Dekker, A.D.
Dementia in people with severe/profound intellectual (and multiple) disabilities: practice-based observations of symptoms
Journal of Mental Health Research in Intellectual Disabilities, 2022, 15(4), 364-393. https://doi.org/10.1080/19315864.2022.2061092
Abstract: Observable dementia symptoms are hardly studied in people with severe/profound intellectual (and multiple) disabilities (SPI(M)D). Insight in symptomatology is needed for timely signaling/diagnosis. This study aimed to identify practice-based observations of dementia symptoms in this population. Care professionals and family members were invited to complete a survey about symptoms. Quantitatively analyzed survey data were further deepened through semi-structured interviews with care professionals having vast experience in signaling/diagnosing dementia in this population. Symptoms were categorized using a symptom matrix. Survey respondents and interviewees frequently observed a decline in activities of daily living (ADL) functioning and behavioral and psychological changes, like increased irritability, anxiety, apathy and decreased eating/drinking behavior. Cognitive symptoms were particularly recognized in persons with verbal communication and/or walking skills. To lesser extent motor changes and medical comorbidities were reported. Increased insight in dementia symptoms contributes to developing a dedicated screening instrument for dementia in people with SPI(M)D.

Woods, R.T., Moniz-Cook, E., Lliffe, S., Campion, P., Vernooij-Dassen, M., Zanetti, O., & Franco, M.
Dementia: Issues in early recognition and intervention in primary care.
Journal of the Royal Society of Medicine, 2003, 96, 320-324. doi: 10.1258/jrsm.96.7.320
Abstract: Generic article about the need for quality and accurate screening and assessment of adults suspected of showing signs of Alzheimer's disease and the need for psychosocial interventions and family carer supports. Authors note need for better training of medical practitioners who may be screening for dementia, indicating that there is a need for timely detection and diagnosis that will prevent crises, facilitate adjustment and provide access to treatments and supports.

Yang, Q., Rasmussen, S. A., & Friedman, J. M.
Mortality associated with Down's syndrome in the USA from 1983 to 1997: A population-based study.
The Lancet, 2002, 359(9311), 1019-1025.
https://doi.org/10.1016/s0140-6736(02)08092-3
Abstract: : Down syndrome is the most frequently identified cause of mental retardation, but information about mortality and comorbidity in people with Down syndrome is limited. We used data from US death certificates from 1983 to 1997 to calculate median age at death and standardized mortality odds ratios (SMORs) for common medical disorders in people with Down syndrome. Of 17897 people reported to have Down syndrome, median age at death increased from 25 years in 1983 to 49 years in 1997, an average increase of 1.7 years per year studied ($p<0.0001$). Median age at death was significantly lower in black people and people of other races than in white people with Down syndrome. As expected, death certificates with a diagnosis of Down syndrome were more likely to list congenital heart defects (SMOR 29.1, 95% CI 27.8-30.4), **dementia** (21.2, 19.6-22.7), hypothyroidism (20.3, 18.5-22.3), or leukemia (1.6, 1.4-1.8) than were those that did not report Down syndrome. By contrast, malignant neoplasms other than leukemia were listed on death certificates of people with Down's syndrome less than one-tenth as often as expected (0.07, 0.06-0.08). A strikingly low SMOR for malignancy was associated with Down syndrome at all ages, in both sexes, and for all common tumor types except leukemia and testicular cancer. Identification of factors responsible for the racial differences recorded could facilitate further improvement in survival of people with Down syndrome. Reduced exposure to environmental factors that contribute to cancer risk, tumor-suppressor genes on chromosome 21, or a slower rate of replication or higher likelihood of apoptosis in Down syndrome cells, could be possible reasons for paucity of cancer in people with Down syndrome.

Zaman, S., & Fortea, J.
The crucial history of Down syndrome.
Lancet Neurology, 2022 Mar, 21(3), 222. doi: 10.1016/S1474-4422(22)00047-3.
Abstract (Full article): In 1866, John Langdon Down was the first clinician to provide a detailed account of Down syndrome. A decade later, Fraser and Mitchell highlighted the early onset of senility in this population. The observations of neuropathological features of Alzheimer's disease in Down syndrome were published in 1929 by Struwe. In 1948, Jervis noted, "[with the] exception of the age of onset, the clinical and pathological manifestations are those of senile dementia". In 1984, using samples taken from the brain of people with Down syndrome, Glenner and Wong biochemically characterized amyloid β (Aβ), predicted that the gene for Alzheimer's disease would be on chromosome 21, and proposed a pathogenic role for Aβ. Using the Aβ peptide sequence, the locus of the APP gene was discovered on chromosome 21. Genetic linkage studies later identified the first mutations in APP, PSEN1, and PSEN2. Furthermore, Down syndrome studies have shown the importance of endosomal misprocessing of APP in Alzheimer's disease. Down syndrome is now regarded as a genetically determined form of Alzheimer's disease, similar to autosomal dominant Alzheimer's disease. Clinical and biomarker changes show a parallel natural history in these two forms. These genetic types of Alzheimer's disease provide the strongest evidence supporting the amyloid cascade hypothesis and give researchers a unique opportunity to study pathophysiology and its temporality, and to undertake preventive trials.

Zammit, M.D., Betthauser, T.J., McVea, A.K., Laymon, C.M., Tudorascu, D.L., Johnson, S.C., Hartley, S.L., Converse, A.K., Minhas, D.S., Zaman, S.H.,

Ances, B.M., Stone, C.K., Mathis, C.A., Cohen, A.D., Klunk, W.E., Handen, B.L., Christian, B.T.; Alzheimer's Biomarker Consortium - Down Syndrome.
Characterizing the emergence of amyloid and tau burden in Down syndrome.
Alzheimers & Dementia, 2024 Jan, 20(1), 388-398. doi: 10.1002/alz.13444.
Abstract: Almost all individuals with Down syndrome (DS) will develop neuropathological features of Alzheimer's disease (AD). Understanding AD biomarker trajectories is necessary for DS-specific clinical interventions and interpretation of drug-related changes in the disease trajectory. A total of 177 adults with DS from the Alzheimer's Biomarker Consortium-Down Syndrome (ABC-DS) underwent positron emission tomography (PET) and MR imaging. Amyloid-beta (Aß) trajectories were modeled to provide individual-level estimates of Aß-positive (A+) chronicity, which were compared against longitudinal tau change. Elevated tau was observed in all NFT regions following A+ and longitudinal tau increased with respect to A+ chronicity. Tau increases in NFT regions I-III was observed 0-2.5 years following A+. Nearly all A+ individuals had tau increases in the medial temporal lobe. These findings highlight the rapid accumulation of amyloid and early onset of tau relative to amyloid in DS and provide a strategy for temporally characterizing AD neuropathology progression that is specific to the DS population and independent of chronological age. Longitudinal amyloid trajectories reveal rapid Aß accumulation in Down syndrome NFT stage tau was strongly associated with A+ chronicity Early longitudinal tau increases were observed 2.5-5 years after reaching A.

Zeilinger, E.L., Gärtner, C, Janicki, M.P., Esralew, L., Weber, G.
Practical applications of the NTG-EDSD for screening adults with intellectual disability for dementia: A German-language version feasibility study.
Journal of Intellectual and Developmental Disability, 2016, 41(1), 42-49. https://doi.org/10.3109/13668250.2015.1113238
Abstract: Authors evaluated the feasibility of using the German-language version of a recently developed screening tool for dementia for persons with intellectual disability (ID): the National Task Group – Early Detection Screen for Dementia (NTG-EDSD). Some 221 paid carers of ageing persons with ID were asked to use the NTG-EDSD and report back on its utility and on 4 feasibility dimensions, and to provide detailed feedback on aspects deemed critical or missing. All feasibility dimensions were rated good to very good, and 80% of respondents found the NTG-EDSD useful or very useful for the early detection of dementia. This highlights a high acceptability of this instrument by the main target group. The positive feasibility evaluation of the NTG-EDSD indicates the usability and adequacy of this instrument for application of early detection of dementia in persons with ID.

Zeilinger, E.L., Komenda, S., Zrnic, I., Franken, F., & Woditschka, K.
Informant-based assessment instruments for dementia and their measurement properties in persons with intellectual disability: systematic review protocol
BMJ Open, 2020, 10(12), 1136. https://bmjopen.bmj.com/content/10/12/e040920
Abstract: Persons with intellectual disability (ID) are at a higher risk of developing dementia than persons without ID, with an expected earlier onset. Assessment methods for the general population cannot be applied for persons with ID due to their pre-existing intellectual and functional impairments. As there is no agreed-upon measure to assess dementia in persons with ID, multiple instruments for this purpose have been developed and adapted in the past decades. This review aimed to identify all available informant-based instruments for the assessment of dementia in persons with ID, to evaluate and compare them according to their measurement properties, and to provide a recommendation for the most suitable instruments. Additionally, an overview of the amount and quality of research on these instruments was provided.

Zeilinger, E.L, Novakovic, I.Z., Komenda, S., Franken, F., Sobisch, M.., Mayer, A-M., Neumann, L.C., Loosli, S.V., Hoare, S., & Pietschnig, J.
Informant-based assessment instruments for dementia in people with intellectual disability: A systematic review and standardised evaluation,
Research in Developmental Disabilities, 2022, 121(Feb),104148, https://doi.org/10.1016/j.ridd.2021.104148.
Abstract: Dementia in people with intellectual disability (ID) is frequent but hard to recognize. Evidence-based recommendations for suitable instruments are lacking. The present study set out to evaluate informant-based dementia assessment instruments and to provide evidence-based recommendations for instruments most suitable in clinical practice and research. A systematic review was conducted across ten international electronic databases. The COnsensus-based Standards for the selection of health Measurement INstruments (COSMIN) guidelines, including a risk of bias assessment, was applied to extract information and to evaluate measurement properties and the quality of available evidence. In total, 42 studies evaluating 18 informant-based assessment instruments were analysed. For screening purposes, we recommend the Behavioral and Psychological Symptoms of Dementia in Down Syndrome Scale (BPSD-DS), the Cognitive Scale for Down Syndrome (CS-DS), and the Dementia Screening Questionnaire for Individuals with Intellectual Disabilities (DSQIID). For a more thorough dementia assessment, we recommend the Cambridge Examination for Mental Disorders of Older People with Down Syndrome and Others with Intellectual Disabilities (CAMDEX-DS).
Our study informs clinicians and researchers about adequate, well-evaluated dementia assessment instruments for people with ID, and highlights the need for high quality studies, especially regarding content validity.

Zeilinger, E.L., Stiehl, K.A.M., & Weber, G.
A systematic review on assessment instruments for dementia in persons with intellectual disabilities.
Research in Developmental Disabilities, 2013, 34, 3962–3977. doi:10.1016/j.ridd.2013.08.013
Abstract: This work describes an extensive systematic literature review on assessment instruments for dementia in persons with intellectual disability (ID). Existing instruments for the detection of dementia in persons with ID were collected and described systematically. This allows a direct and quick overview of available tools. Additionally, it contributes to the availability and usability of information about these instruments, thus enhancing further developments in this field. A systematic literature search in five databases (CINAHL, PsycInfo, PubMed, Scopus, and Web of Science) was conducted. In order to include gray literature an invisible college approach was used. Relevant studies were identified and selected using defined inclusion and exclusion criteria. After the selection process all instruments were coded and classified. It was determined which concepts they assess, whether they were especially developed or adapted for persons with ID, and whether they were designed to assess dementia. The selection of relevant papers, as well as the coding of instruments was done independently by two researchers. In total, 97 records met the search criteria. Out of these, 114 different instruments were extracted. There were 79 instruments to be completed by the person with ID, and 35 informant-based instruments. Additionally, four test batteries were found. Some of these instruments were neither designed for the assessment of dementia, nor for persons with ID. There are a variety of different tools used for the assessment of dementia in ID. Nevertheless, an agreed-upon approach or instrument is missing. Establishing this would improve the quality of assessment in clinical practice, and benefit research. Data collected would become comparable and combinable, and allow research to have more informative value.

Zigman, W.B
Atypical aging in Down syndrome
Developmental Disabilities Research Review, 2013, 18(1), 51-67. doi:10.1002/ddrr.1128.
Abstract: At present, there may be over 210,000 people with Down syndrome (DS) over the age of 55 in the United States (US) who have significant needs for augmented services due to circumstances related to ordinary and/or pathological aging. From 1979 through 2003, the birth prevalence of DS rose from 9.0 to 11.8 (31.1%) per 10,000 live births in 10 representative US regions. This increase, largely due to women conceiving after age 35, portends an ever-growing

population of people with DS who may be subject to pathogenic aging. Whereas Trisomy 21 is one of the most widespread genetic causes of intellectual disability (ID), it still is one of the least understood of all genetic ID syndromes. While longevity in people with DS has improved appreciably in as modest a period as 30 years, age-specific risk for mortality still is considerably increased compared both with other people with ID or with the typically developing population. The penetrance of the phenotype is widely distributed, even though a consistent genotype is assumed in 95% of the cases. Some, but not all body systems, exhibit signs of premature or accelerated aging. This may be due to both genetic and epigenetic inheritance. We now know that the long-term outcome for people with DS is not as ominous as once contemplated; a number of people with DS are living into their late 60s and 70s with few if any major signs of pathogenic aging. Alzheimer's disease (AD), a devastating disease that robs a person of their memory, abilities and personality, is particularly common in elder adults with DS, but is not a certainty as originally thought, some 20% to 30% of elder adults with DS might never show any, or at most mild signs of AD. DS has been called a mature well-understood syndrome, not in need of further research or science funding. We are only beginning to understand how epigenetics affects the phenotype and it may be feasible in the future to alter the phenotype through epigenetic interventions. This chapter is divided into two sections. The first section will review typical and atypical aging patterns in somatic issues in elder adults with DS; the second section will review the multifaceted relationship between AD and DS.

Zigman, W.B,, & Lott. I.T.
Alzheimer's disease in Down syndrome: Neurobiology and risk.
Mental Retardation and Developmental Disabilities Research Reviews, 2007, 13, 237–246. https://doi.org/10.1002/mrdd.20163
Abstract: Down syndrome (DS) is characterized by increased mortality rates, both during early and later stages of life, and age-specific mortality risk remains higher in adults with DS compared with the overall population of people with mental retardation and with typically developing populations. Causes of increased mortality rates early in life are primarily due to the increased incidence of congenital heart disease and leukemia, while causes of higher mortality rates later in life may be due to a number of factors, two of which are an increased risk for Alzheimer's disease (AD) and an apparent tendency toward premature aging. In this article, we describe the increase in lifespan for people with DS that has occurred over the past 100 years, as well as advances in the understanding of the occurrence of AD in adults with DS. Aspects of the neurobiology of AD, including the role of amyloid, oxidative stress, Cu/ZN dismutase (SOD-1), as well as advances in neuroimaging are presented. The function of risk factors in the observed heterogeneity in the expression of AD dementia in adults with DS, as well as the need for sensitive and specific biomarkers of the clinical and pathological progressing of AD in adults with DS is considered.

Zigman, W.B., .Devenny, D.A., Krinsky-McHale, S.J., Jenkins, E.C., Urv, T.K., Wegiel, J., Schupf, N., & Silverman, W.
Alzheimer's disease in adults with Down syndrome.
International Review of Research in Mental Retardation, 2008, 36, 103-145. doi: 10.1016/S0074-7750(08)00004-9
Abstract: Down syndrome is associated with increased mortality rates due to congenital cardiac defects and leukemia early in life, and with Alzheimer's disease and a tendency toward premature aging later in life. Alzheimer's disease was once considered an inexorable result of growing old with Down syndrome, but recent data indicate that risk does not reach 100%. Although some individuals exhibit signs and symptoms of Alzheimer's disease in their 40s, other individuals have reached the age of 70 without developing dementia. This chapter presents a wealth of data from a longstanding longitudinal study with the overall objective of understanding and recounting the mechanisms responsible for these substantial individual differences.

Zigman, W.B., Schupf, N., Devenny, D., Miezejeski, C., Ryan, R., Urv, T.K., Schubert, R., & Silverman, W.
Incidence and prevalence of dementia in elderly adults with mental retardation without Down syndrome.
American Journal on Mental Retardation, 2004, 109, 126–141. DOI: 10.1352/0895-8017(2004)109<126:IAPODI>2.0.CO;2
Abstract: Rates of dementia in adults with mental retardation without Down syndrome were equivalent to or lower than would be expected compared to general population rates, whereas prevalence rates of other chronic health concerns varied as a function of condition. Given that individual differences in vulnerability to Alzheimer's disease have been hypothesized to be due to variation in cognitive reserve, adults with mental retardation, who have long-standing intellectual and cognitive impairments, should be at increased risk. This suggests that factors determining intelligence may have little or no direct relationship to risk for dementia and that dementia risk for individuals with mental retardation will be comparable to that of adults without mental retardation unless predisposing risk factors for dementia are also present.

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Denotes report
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