

NTG Bibliography of Published Work on Identifying Symptoms of Dementia in Adults with Intellectual Disability

v.12/15/22

This section is in development and new information will be added.

Study authors	Subjects	Method	Key factors/Comments
Burt et al., 2005	78 adults with DS	Review of instruments against clinical determinations	"... assessment for psychiatric disorders, delayed memory decline, adaptive behavior decline, and the presence of seizures would be useful for the diagnosis of dementia..."
Cooper, S-A., & Prasher, V. 1998	DS N=19, other ID N=26	Assessed maladaptive behaviors and psychiatric symptoms	"...with Down syndrome had a higher prevalence of low mood, restlessness/excessive overactivity, disturbed sleep, being excessively uncooperative and auditory hallucinations." Aggression more frequent in other ID.
Deb et al., 2007	DS adults N=24	Interview with caregivers	"...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..." "...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..."
Dekker et al., 2018	DS w/o dementia N=149 DS w/possible dementia (Q) N=65 Ds w/dementia N=67	Interviews with informants	"...increases in frequency and severity in items of anxiety, sleep disturbances, agitation & stereotypical behavior, aggression, apathy, depressive symptoms, and eating/drinking behavior..." "...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."
Dekker et al., 2015		Review of studies	"...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..."
Holland et al., 2000	Adults, age 30+ with DS	Cambridge Examination for Mental Disorders of the Elderly informant interview	"... looked for changes in memory, personality, general mental functioning, and daily living skills..." Found "...the initial changes reported were predominately in behavior and personality..."
Mascarenhas Fonseca, Prado Mattar et al., 2020	Adults with DS N=92 (N=13 with AD; N=17 with possible AD; N=62 w/no symptoms	Cambridge Examination for Mental Disorders of Older People with Down's Syndrome and Others with Intellectual	"...Hallucination, agitation, apathy, aberrant motor behavior and night-time behavior disturbance showed significant difference among the DS groups (p<0.05), with higher prevalence in the DS group with AD..."

		Disabilities (CAMDEX-DS).	
Mascarehas Fonseca, Padilla et al., 2020	Adults with DS N=162	Cambridge Examination for Mental Disorders of Older People with Down's Syndrome and Others with Intellectual Disabilities (CAMDEX-DS).	"...Memory impairments were more marked in the prodromal dementia than the stable cognition group, as was executive dysfunction. Disinhibition was greater in the AD than in the prodromal dementia group. Apathy was more pronounced in the AD than in the stable cognition group. Premorbid amnesic and non-amnesic symptoms as reported by informants increase with the progression to AD..."
Wissing et al., 2022	Adults with severe/profound ID	Review of 11 studies	Predominantly "...Cognitive decline, behavioral and psychological alterations, decline in activities of daily living as well as neurological and physical changes were found..."

Source articles

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.



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.



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.

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]



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 about 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 behaviors 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.



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.



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 an easy observable sign, however, general slowness has been reported and apathy itself might cause reduced activity. Agitation appears to be more prevalent in adults with dementia than in DS adults absent dementia, 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., Sacco, S., Carfi, A., Benejam, B., Vermeiren, Y., Beugelsdijk, G., Schippers, M., Hassefras, L., Eleveld, J., Grefelman, S., Fopma, R., Bommer-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 Alzheimer's 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.



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 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., 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. 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.



Mascarenhas Fonseca, L., 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 behavior and night-time behavior 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 to reduce the odds of generating a false positive.



Mascarehas Fonseca, L., 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.
 Amnesic and non-amnesic 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-amnesic informant reported symptoms (disinhibition, apathy, and executive dysfunction) and amnesic 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. Amnesic and non-amnesic 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 amnesic and non-amnesic 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 considered. An understanding of the unique clinical presentation of DS in AD should inform treatment options.



Wissing, M. B. G., Ulgiati, A. M., Hobbelen, J. S. M., De Deyn, P. P., Waning, 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/aging. 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, forgetfulness, 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 behavior) 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., 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 a 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.

