Received: 29 January 2025 | Revised: 25 April 2025 | Accepted: 15 May 2025

DOI: 10.1002/alz.70356

Alzheimer's & Dementia®
THE JOURNAL OF THE ALZHEIMER'S ASSOCIATION

SHORT REPORT

Specialists' perceptions of clinical instruments, practices, and staging of DS-AD: Results from an international survey

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Funding information
LuMind IDSC Foundation; National Center for Chronic Disease Prevention and Health Promotion; Healthy Brain Initiative, Grant/Award Number: 1NU58DP006782-01-00; Eli Lilly and Company; Rainwater Charitable Foundation

Abstract

INTRODUCTION: Diagnosing and staging Down syndrome–associated Alzheimer's disease (DS-AD) is hindered by the lack of standardized criteria, complicating clinical decision making, trial participation, and access to advanced therapies. This study aimed to explore perceptions of these issues.

METHOD: An international survey of 42 clinicians and researchers specializing in DS-AD gathered perspectives on instruments, symptomatic staging, clinical practices, and research priorities.

RESULTS: Respondents noted that key domains of impairment in mild cognitive impairment in Down syndrome and DS-AD dementia included memory, executive functioning, personality, social behavior, attention, mood, and language. Among the 10 assessment tools evaluated, informant-based interviews were noted as critical for individuals with severe intellectual disability (ID), while direct assessments were noted as useful for those with mild to moderate ID. Common diagnostic confounders like hypothyroidism and sleep disorders were identified.

DISCUSSION: Behavioral assessments provide a valuable function; however, future efforts should integrate behavioral assessments with biomarkers and develop standardized staging frameworks to improve diagnostic reliability, care planning, and treatment strategies for DS-AD.

KEYWORDS
assessment, dementia, diagnosis, Down syndrome, instruments, staging

Highlights

- Personality, social behavior, language, mood/affect, memory, executive functioning, and attention are recognized as key domains of impairment in both mild cognitive impairment in Down syndrome (MCI-DS) and Down syndrome–associated Alzheimer's disease (DS-AD).

- Ten prominent informant and direct assessment tools were noted as appropriate for individuals with DS and mild to moderate intellectual disability (ID) for identifying both MCI-DS and DS-AD; however, for individuals with severe/profound ID, there was less assurance of applicability.
- Harmonizing recommended tools in a standardized list was identified as a strategy to promote consistency across clinical and research contexts.

The rising need for standardized recognition and assessment of early dementia symptoms in adults with Down syndrome (DS) reflects broader shifts in dementia care and policy. Accurate assessment tools are increasingly critical for determining eligibility for federal dementia assistance programs,[1] prescribing newly approved Alzheimer's disease (AD)-modifying therapies,[2] and enrolling participants in clinical trials.[3] Traditionally, clinical guidelines and instrument reviews offered a range of assessment instruments for use with adults with DS and other intellectual disability (ID), outlining their strengths and limitations,[4–10,11] leaving selection to the discretion of individual practitioners. Although adaptations tailored to individuals with DS have been developed, global consensus on their equivalence to tools used for the general population remains absent, posing challenges for program access and treatment authorization.

The anticipated growth of the dementia-affected population heightens the urgency to expand the use of reliable instruments for early AD detection.[12] While biomarkers are becoming central to diagnosing mild cognitive impairment (MCI) and AD dementia,[13] cognitive, behavioral, and functional assessments remain indispensable for complementing biomarker diagnostics, staging disease progression, and informing care strategies. This dual need applies universally but is especially pertinent for adults with DS, who face a 90% lifetime risk of developing AD and a 75% probability of dementia-related mortality.[14] Recent efforts aim to define biomarker thresholds and identify behavioral assessment instruments that can be standardized for diagnostic use in this population.

The Alzheimer's Association's 2024 Revised Criteria for Alzheimer's Disease Diagnosis and Staging[13] provides the conceptual foundation for this work. These guidelines classify individuals with DS as having Stage 0 AD even prior to becoming biomarker positive due to the deterministic link between DS and early-onset AD, resulting from triplication of the amyloid precursor protein (*APP*) gene. DS-associated AD (DS-AD) presents unique clinical characteristics, shaped by the cognitive and functional profile of DS itself. Within this framework, Clinical Stage 1 represents a transitional phase during which individuals test positive for AD biomarkers but do not yet exhibit measurable cognitive or functional impairments. However, this stage was not addressed within the scope of our work. Clinical Stages 2 and 3, characterized by mild cognitive changes with minimal to early functional impairment, are collectively categorized in this paper as MCI-DS to represent the prodromal stage of DS-AD. Clinical Stages 4 through 6, encompassing mild to severe dementia, are categorized as DS-AD dementia.

Our study investigated the clinical assessment processes and tools for detecting DS-AD, focusing on key diagnostic markers, staging domains, exclusion of comorbidities, and the influence of ID severity on dementia screening outcomes. In addition, we envision an alignment of these findings with emerging biomarker frameworks specific to DS-AD. This report presents insights from an international survey of DS-AD clinicians and researchers regarding their use of assessment instruments, levels of comfort and familiarity with these tools, and perceptions of their applicability to different ID levels in adults with DS. We also highlight common diagnostic challenges raised by survey respondents, including behavioral and medical conditions that can mimic dementia, and propose areas for further research and resource development to better support the clinical community in diagnosing and managing dementia in this population.

1 | METHODOLOGY

1.1 | Survey design

Data were collected using a purpose-built survey designed to explore key aspects of diagnosing MCI and dementia in adults with DS. The survey content covered symptomatic clinical stages, clinical practice implications, research priorities, and relevance for clinical trials. The survey was refined through expert clinician review, resulting in a final version with four sections and 38 questions: (1) Clinical Presentation of MCI-DS/DS-AD Dementia (11 questions), (2) Clinical Practice Implications of Staging DS-AD (19 questions), (3) Research Implications (6 questions), and (4) Societal Implications (2 questions). Questions used Likert scales, dropdown menus, and open-text responses. Demographic data, including geographic location, profession, years of experience with adults with DS, affiliations, and primary focus (clinical or research), were also collected (see supporting information for a full survey).

Assessment tools evaluated in the survey were identified from a prior review of commonly used instruments. Informant-based tools included the Cambridge Examination for Mental Disorders of Older People with Down Syndrome (CAMDEX-DS) Informant Interview,[15] CAMDEX-DS-II Informant Interview,[16] Dementia Questionnaire for

People with Learning Disabilities (DLD),[17] Dementia Screening Questionnaire for Individuals with Intellectual Disabilities (DSQIID),[18] and NTG-Early Detection Screen for Dementia (NTG-EDSD).[19] Direct assessments included the Cambridge Examination Modified for Use in People with Down Syndrome (CAMCOG-DS),[15] CAMCOG-DS-II,[18] CANTAB Paired Associates Learning (CANTAB-PAL) test,[20] Down Syndrome Mental Status Examination (DS-MSE),[21] and modified Cued Recall Test (mCRT).[22] Domains of potential impairment were also presented for respondent feedback, selected from widely cited sources in the literature.

1.2 | Data collection

We used a snowball sampling method, initially targeting key respondents identified from publications and the DS-AD research/clinical network ($N = 78$). Additionally, we requested distribution through various organizations and other prominent clinicians/researchers. The initial invitations, sent via e-mail in August 2024, outlined the study's objectives, and provided a link to the survey. Another wave was distributed to various international DS organizations, requesting dissemination to clinicians. Although this wave was distributed worldwide, we only provided an English-language version. Recipients were encouraged to share the survey with colleagues. Subsequent waves of invitations continued through October 2024. Due to the survey's anonymity and lack of required identifiers, we could not determine the exact number of recipients. However, we estimate that an additional 25 to 50 individuals might have seen the survey request, although we cannot confirm if they were clinicians experienced in assessing adults with DS.

1.3 | Data analysis

Descriptive statistics were calculated as frequencies and percentages of survey responses. Results were presented to a multidisciplinary work group during a virtual meeting in December 2024, at which findings were reviewed, and additional insights were gathered.

2 | RESULTS

2.1 | Respondents

A total of 51 responses were received from Africa (2%), Asia (10%), Europe (29%), North America (53%), and Oceania (6%). Of these, 42 (82%) were complete and suitable for analysis, while the remainder were excluded due to insufficient familiarity with specialized assessment tools for older adults (14%) or incomplete responses (4%). Of the 42 analyzed responses, 16 respondents identified as clinicians who see adults with DS in their practice, 17 identified as clinicians who see adults with DS in their practice and conduct DS research, and 9 identified as non-clinicians who conduct DS research. Among the 33 clinicians, 9 respondents (27.3%) identified as psychiatrists, 8 (24.2%) as neurologists, 5 (15.2%) as primary care providers, 4 (12.1%) as pediatricians, and 7 (21.2%) as in other specialties. Among the nine non-clinicians, six respondents (66.7%) identified as psychologists, two (22.2%) as psychiatrists, and one (11.1%) as specializing in neuroimaging.

2.2 | Dementia features

Some 75% of respondents identified personality, social behavior, language, mood/affect, memory, executive functioning, and attention as

RESEARCH IN CONTEXT

1. **Systematic review**: Diagnosing and staging Down syndrome–associated Alzheimer's disease (DS-AD) remains a significant challenge due to the absence of standardized criteria. This complicates clinical decision making, enrollment in clinical trials, and access to advanced therapies. An international survey identified core domains of impairment in mild cognitive impairment in Down syndrome (MCI-DS) and DS-AD dementia and evaluated 10 assessment (informant-based interviews and direct assessments) tools with application to adults by varied levels of intellectual disability (ID) and stage of dementia.
2. **Interpretation**: The survey findings underscored the complexity of diagnosing DS-AD due to overlapping symptoms, co-occurring conditions, and variability in ID levels. Informant-based interviews were seen as critical for capturing nuanced impairments in individuals with severe ID, while direct assessments complemented these approaches in less severe cases. The identified diagnostic confounders emphasize the need for comprehensive clinical evaluations to avoid misdiagnoses. Additionally, the variation in preferred assessment tools signaled the need for greater consensus on diagnostic criteria and methodologies to ensure reliability and comparability across clinical and research settings.
3. **Future directions**: To advance the field, future research should focus on correlating behavioral assessments with biomarkers to enhance diagnostic precision. Developing standardized, consensus-driven staging frameworks will be essential for improving diagnostic reliability, care planning, and treatment alignment. Furthermore, addressing the gaps in assessment tools tailored to different levels of ID can optimize the evaluation process for individuals with DS-AD. Collaborative efforts among clinicians, researchers, and advocacy groups will be critical in addressing these priorities and ultimately improving outcomes for individuals with DS-AD.

key domains of impairment in both MCI-DS and DS-AD. Orientation, praxis, and gait were reported as substantively more relevant in DS-AD dementia compared to MCI-DS. Higher importance for personality, social behavior, mood/affect, and language was also noted in DS-AD dementia compared to MCI-DS. Conversely, memory, executive functioning, and attention were reported as similarly important across both stages, reflecting a broad consensus on their centrality to the diagnostic process.

2.3 | Assessment tools

Among the five informant interview tools surveyed, at least 75% of respondents rated them as moderately to highly useful for identifying MCI-DS, with > 90% agreement for their utility in diagnosing DS-AD dementia (see Figure 1). Similarly, four of the five direct assessment tools were rated moderately to highly useful by > 80% of respondents for MCI-DS, with ratings for all five > 90% for DS-AD dementia.

Familiarity and usage of assessment tools varied by region. The NTG-EDSD (83.3% North America, 94.1% Europe/Oceania) and DLD (72.7% North America, 76.5% Europe/Oceania) were the most recognized tools across North America and Europe/Oceania. The DSQIID was less recognized in North America than in Europe/Oceania (56.5% vs. 88.2%), while the DS-MSE was slightly more recognized in North America than in Europe/Oceania (65.2% vs. 47.1%). The CAMDEX-DS (17.4% vs. 70.6%), CAMDEX-DS-II (17.4% vs. 82.4%), CAMCOG-DS (21.7% vs. 64.7%), and CAMCOG-DS-II (21.7% vs. 76.5%) were all much less familiar to respondents in North America than in Europe/Oceania.

Greater than 70% of respondents considered all 10 informant and direct assessment tools appropriate for individuals with DS and mild to moderate ID for identifying both MCI-DS and DS-AD (see Figure 2). For individuals with severe ID, ≈ 60% of respondents rated the five informant interviews as suitable for MCI-DS, with a similar to slightly higher proportion endorsing these tools for DS-AD dementia. Among the five direct assessments, only the DS-MSE was rated appropriate for indicating both MCI-DS and DS-AD dementia for individuals with severe ID by > 40% of respondents, while the remaining tools received < 20% approval for both stages. For individuals with profound ID, between 30% and 40% of respondents rated all five informant interviews as being appropriate for indicating both MCI-DS and DS-AD dementia, whereas < 10% of respondents rated any of the direct assessments as being appropriate for either stage.

Respondents highlighted specific advantages and limitations of individual tools. The CAMDEX-DS-II was valued for its detailed structure and capacity to track changes over time, making it well suited for ongoing clinical management. Tools incorporating caregiver reports, such as the NTG-EDSD, were noted for ease of use in detecting significant changes but were perceived as more appropriate for dementia screening than early-stage MCI-DS detection.

Feasibility considerations influenced tool preferences. Respondents noted that lengthy assessments requiring multiple hours were less practical for general clinical use, particularly in resource-limited settings. In multidisciplinary environments, dividing assessments among specialized professionals could enhance efficiency, but replicating this approach in smaller practices posed challenges. Cost, including staff time and licensing fees, was noted as a barrier to the widespread adoption of certain tools.

Harmonizing recommended tools in a standardized list was identified as a strategy to promote consistency across clinical and research contexts. Such standardization could guide clinicians with limited expertise in DS-AD assessments and facilitate international comparisons. However, respondents cautioned that familiarity biases often drive tool selection, complicating global harmonization efforts. To address this, any standardized framework should offer multiple equivalent tools for each clinical purpose, ensuring both flexibility and broader acceptance.

2.4 | Excluding co-occurring conditions

Respondents identified at least 17 non-AD causes that could confound the diagnosis of DS-AD and should be ruled out before confirming a diagnosis. Common considerations for both DS-AD and MCI-DS included hypothyroidism, depression, obstructive sleep apnea (OSA) and other sleep disorders, general medical issues, life events, and vision or hearing impairments. Pseudodementia caused by psychiatric conditions was frequently cited as a diagnosis of exclusion.

A key distinction emphasized by respondents was between historical medical conditions that are well managed (e.g., treated hypothyroidism or repleted B_{12} deficiency) and acute, untreated conditions that could cause or exacerbate cognitive, behavioral, or functional changes (e.g., unmanaged OSA). Failure to evaluate the current treatment status of such conditions may lead to diagnostic errors by attributing symptoms to DS-AD rather than reversible causes.

Respondents supported the development of a standardized checklist to improve consistency and thoroughness in clinical evaluations. A minimum recommended checklist would include common reversible or treatable conditions such as hypothyroidism, vitamin B_{12} deficiency, sleep apnea, mood disorders, and seizures. This approach could reduce diagnostic variability by providing structured guidance, especially for clinicians with limited experience in DS or DS-AD.

The proposed checklist would prioritize addressing treatable conditions first, allowing clinicians to reassess residual cognitive or behavioral symptoms once these factors are managed. Respondents highlighted the importance of flexibility in adapting checklists to different health-care systems and resource availability, ensuring feasibility and broad adoption in diverse clinical settings.

2.5 | Staging DS-AD transitions

Respondents provided mixed evaluations regarding the precision of defining transitions between stages of DS-AD. More than 60% rated the transition from Asymptomatic DS-AD to MCI-DS (Stage 0–1 to Stage 2–3) as having low to somewhat low precision. The transition

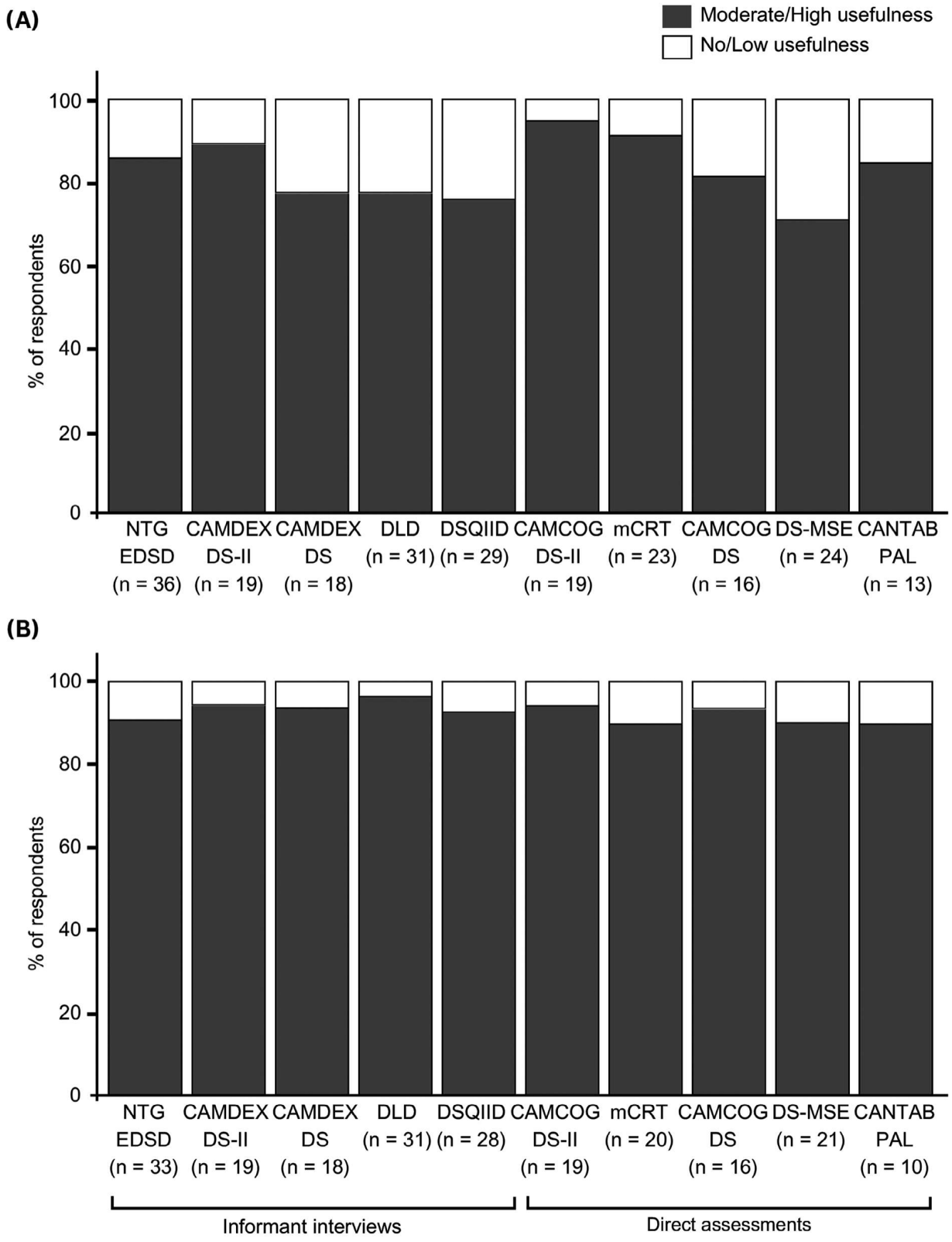

FIGURE 1 Perceived usefulness of 10 assessment tools for adults with Down syndrome in indicating (A) the presence of MCI-DS, and (B) the presence of DS-AD dementia. Respondents rated each assessment tool on a four-option scale: not useful, low usefulness, moderate usefulness, or high usefulness. The number of responses varies for each tool because respondents only rated the assessments with which they were familiar. CAMCOG-DS, Cambridge Examination Modified for Use in People with Down Syndrome; CAMDEX-DS, Cambridge Examination for Mental Disorders of Older People with Down Syndrome; CANTAB PAL, CANTAB Paired Associates Learning; DLD, Dementia Questionnaire for People with Learning Disabilities; DS-AD, Down syndrome–associated Alzheimer's disease; DS-MSE, Down Syndrome Mental Status Examination; DSQIID, Dementia Screening Questionnaire for Individuals with Intellectual Disabilities; MCI-DS, mild cognitive impairment in Down syndrome; mCRT, modified Cued Recall Test; NTG-EDSD, NTG-Early Detection Screen for Dementia

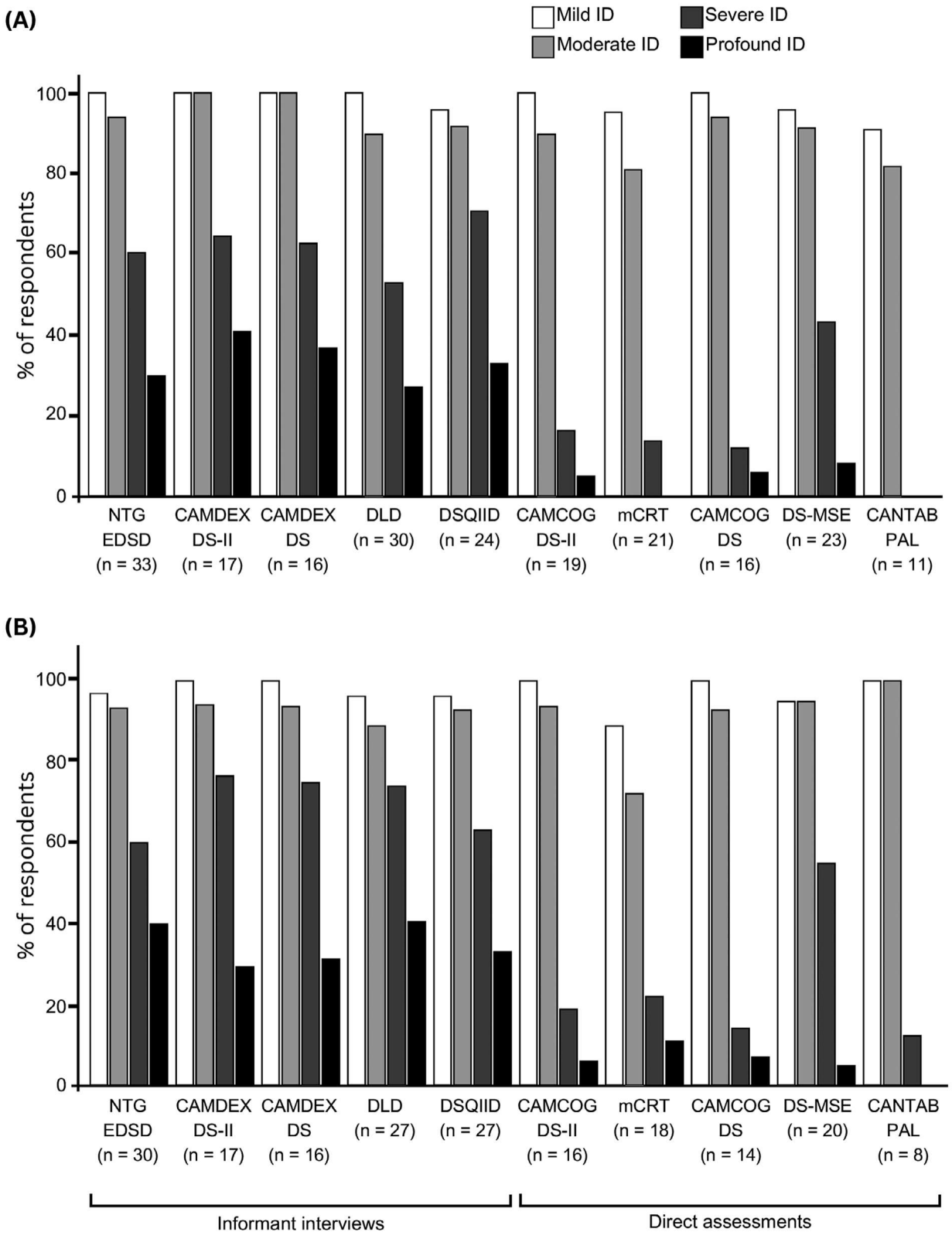

FIGURE 2 Perceived appropriateness of 10 assessment tools for use with adults with Down syndrome at different levels of ID in indicating (A) the presence of MCI-DS, and (B) the presence of DS-AD dementia. Respondents rated only the tools they were familiar with for perceived appropriateness. Tools not rated were assumed to reflect a lack of endorsement. CAMCOG-DS, Cambridge Examination Modified for Use in People with Down Syndrome; CAMDEX-DS, Cambridge Examination for Mental Disorders of Older People with Down Syndrome; CANTAB PAL, CANTAB Paired Associates Learning; DLD, Dementia Questionnaire for People with Learning Disabilities; DS-AD, Down syndrome–associated Alzheimer's disease; DS-MSE, Down Syndrome Mental Status Examination; DSQIID, Dementia Screening Questionnaire for Individuals with Intellectual Disabilities; ID, intellectual disability; MCI-DS, mild cognitive impairment in Down syndrome; mCRT, modified Cued Recall Test; NTG-EDSD, NTG-Early Detection Screen for Dementia

from MCI-DS to DS-AD Dementia (Stage 2–3 to Stage 4) was viewed as having greater precision, with 40% rating it as moderate and 25% as somewhat high in precision. The precision of the shift from Mild to Moderate DS-AD Dementia (Stages 4 to 5) received lesser ratings, with 35% noting somewhat low and 35% moderate precision. Opinions on the transition from Moderate to Severe DS-AD Dementia (Stages 5 to 6) showed no consensus, with responses ranging from somewhat low to somewhat high precision.

Despite variability in precision ratings, respondents agreed that defining stage transitions is clinically meaningful when it affects care or treatment decisions. Although most individuals with DS will eventually develop dementia if they live long enough, clearly delineating DS-AD stages can inform timely treatment initiation and care planning. Respondents noted the importance of diagnosing stage transitions as soon as dementia criteria are met and recommended the integration of biomarker evidence to enhance diagnostic accuracy and consistency in clinical practice.

3 | DISCUSSION

This survey's results underscore a broad consensus within the international DS-AD clinical and research community regarding the utility of behavioral/clinical indicators for detecting dementia associated with amyloid-based neuropathology in adults with DS. Despite the diversity of tools available, many assessment instruments align on core cognitive, behavioral, and functional domains, suggesting a degree of interchangeability. However, practical considerations, such as administration time, depth of inquiry, adaptability to varying levels of ID, and diagnostic output, ultimately will guide tool selection. To enhance clinical practice the field would benefit from an international consensus list of clinical assessments.

The survey findings suggest that although most tools are effective for identifying DS-AD in its early stages, their sensitivity may be limited for detecting nuanced staging differences as dementia progresses (i.e., Stages 5 to 6) or when evaluating individuals with more severe ID. This underscores the potential need for developing or refining specialized tools to enhance diagnostic precision. Additionally, the availability of translated versions was not assessed in this study, but language accessibility must be prioritized when recommending tools for use in non–English-speaking regions to ensure broader applicability and equitable access.

Overall, the results point to critical priorities for harmonizing diagnostic frameworks for DS-AD staging, generating a consensus list of clinical assessments, standardizing checklists to exclude non-AD conditions, and advancing collaborative research to validate tools and integrate biomarker findings. Standardized, globally relevant frameworks are essential for improving diagnostic precision, enabling personalized care planning, and ensuring equitable access to emerging disease-modifying therapies, particularly in low- and middle-income countries.[23] The need for robust staging criteria, adaptable across ID levels, and clarity in distinguishing dementia from other cognitive and behavioral changes remains pivotal. Combining clinical and biomarker-based assessments holds promise for further enhancing diagnostic reliability and staging accuracy.

This study faced several limitations. First, reaching a diverse international audience of clinicians specializing in DS-AD was challenging, particularly those using specific diagnostic instruments for dementia assessment. Second, to maintain a high response rate, we limited the number of survey questions. While this approach encouraged participation, it also restricted the depth of data collected. Third, as an exploratory survey, responses varied significantly in detail—some were highly informative, while others were brief—creating challenges in data analysis. Fourth, participation was restricted to clinicians proficient in English, potentially limiting input from non–English-speaking experts. Additionally, we cannot confirm how representative our sample is of the broader clinical community. However, given our targeted recruitment of recognized experts, we believe the findings reflect key practices among leading DS-AD practitioners. Follow-up work with a more diverse and systematically recruited sample could enhance the generalizability and applicability of these insights.

Future research should prioritize studies correlating biomarkers and imaging with behavioral assessment stages to improve diagnostic granularity and reliability. Investigations exploring the use of biomarkers for staging DS-AD in individuals with severe to profound ID are especially warranted, as these approaches could mitigate challenges in tracking cognitive decline in this population. Finally, research should aim to refine tools suited for different clinical purposes—whether to generate detailed cognitive profiles or to provide a straightforward dementia diagnosis—ensuring flexibility for varied clinical and research settings.

ACKNOWLEDGMENTS

The funding for the project was provided by LuMind IDSC Foundation, the Rainwater Charitable Foundation, and Eli Lilly & Company. Partial support for M.P.J. was provided by a grant from the Centers for Disease Control and Prevention (CDC), National Center for Chronic Disease Prevention and Health Promotion, and the Healthy Brain Initiative Award No. 1NU58DP006782-01-00, to the University of Illinois Chicago.

CONFLICT OF INTEREST STATEMENT

Hampus Hillerstrom is the co-founder and a board member of Anaka Pharmaceuticals, Inc. Amit Das has nothing to report. Matthew P. Janicki received honoraria from the LuMind IDSC Foundation to aid in the work on the study reported in this article. He serves in a non-paid capacity as the co-president of the board for the National Task Group in Intellectual Disabilities and Dementia Practices, and the executive committee of the ISTAART DS-AD PIA. Natalia S. Rozas has nothing to report. Stephanie Santoro has received research funding from LuMind IDSC Down Syndrome Foundation to conduct clinical trials for people with DS within the past 2 years. She serves in a non-paid capacity on the Medical and Scientific Advisory Council of the Massachusetts Down Syndrome Congress, the board of directors of the Down Syndrome Medical Interest Group (DSMIG-USA), and the executive committee of the American Academy of Pediatrics

Council on Genetics. Author disclosures are available in the supporting information.

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SUPPORTING INFORMATION

Additional supporting information can be found online in the Supporting Information section at the end of this article.

How to cite this article: Hillerstrom H, Das A, Janicki MP, Rozas NS, Santoro SL. Specialists' perceptions of clinical instruments, practices, and staging of DS-AD: Results from an international survey. *Alzheimer's Dement*. 2025;21:e70356. https://doi.org/10.1002/alz.70356