Research in Developmental Disabilities 121 (2022) 104148

Contents lists available at ScienceDirect

Research in Developmental Disabilities

journal homepage: www.elsevier.com/locate/redevdis

Informant-based assessment instruments for dementia in people with intellectual disability: A systematic review and standardised evaluation

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ARTICLE INFO

No. of reviews completed is 2

Keywords:
Alzheimer disease
Dementia
Down syndrome
Intellectual disability
Mental status and dementia tests
Psychometrics
Systematic review

ABSTRACT

Background: Dementia in people with intellectual disability (ID) is frequent but hard to recognise. Evidence-based recommendations for suitable instruments are lacking.
Aims: 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.
Method and procedures: 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.
Outcomes and results: 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's Syndrome and Others with Intellectual Disabilities* (CAMDEX-DS).
Conclusions and implications: 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.

What this paper adds

The present review is the first to systematically focus not only on descriptive characteristics of dementia assessment instruments,

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https://doi.org/10.1016/j.ridd.2021.104148
Received 18 August 2021; Received in revised form 4 November 2021; Accepted 5 December 2021
Available online 27 December 2021

but also on measurement properties and on the amount and quality of available research for each instrument and each measurement property. We apply a very comprehensive state-of-the-art methodology for reviews on psychometric properties, the COSMIN approach, and complement it with aspects derived by the CAPs-IDD, an evaluation tool specifically designed for assessment instruments for people with ID. We provide transparent, evidence-based recommendations for the most suitable and best-evaluated informant-based dementia assessment instruments, which will support clinicians and researchers in choosing the most adequate instruments for their respective purpose. Our work thus facilitates the detection of dementia in people with ID and supports the demands raised in the UN-CRPD regarding health care standards without discrimination on the basis of disability.

1. Introduction

People with an intellectual disability (ID) are at a higher risk to be affected by mental health disorders, including dementia, than people without ID (Strydom, Chan, King, Hassiotis, & Livingston, 2013; Takenoshita, Terada, Kuwano, Inoue, Cyoju et al., 2020). People with Down syndrome (DS) are especially prone to develop dementia, particularly Alzheimer's disease (Lott & Head, 2019; Nieuwenhuis-Mark, 2009). Failure to recognise mental disorders in people with ID can not only have a negative impact on their health, but also promote social disintegration (Dias et al., 2020).

Recognising dementia in people with ID is challenging. Assessment instruments designed for people without ID cannot be reliably applied to people with ID (Burt, 2018; Zeilinger, Stiehl, & Weber, 2013), as pre-existing cognitive deficits and limitations in adaptive behaviour make it difficult to distinguish early signs of dementia from disability symptoms (Jopp & Keys, 2001, p. 2001; Mason & Scior, 2004). Furthermore, dementia can manifest itself differently in people with ID compared to people without ID, because behavioural symptoms and personality changes appear more frequently and presumably earlier throughout the course of the illness, especially in people with DS (Hartley et al., 2015; Lautarescu, Holland, & Zaman, 2017). People with DS are especially prone to develop Alzheimer's disease, which can present differently than other types of dementia (Sheehan et al., 2015). Therefore, it is important to consider if an assessment tool was developed specifically for people with DS and/or for the assessment of Alzheimer's disease.

Early recognition of dementia is essential for the provision of adequate health care and for the quality of life of people with ID, their family members and caregivers. It facilitates adequate future planning as well as development of early interventions and tailored support (Heller, Scott, & Janicki, 2018; Robinson, Tang, & Taylor, 2015; Summers & MacDonald, 2020). Not being able to reliably recognise dementia in people with ID constitutes a disadvantage for this population, hinders the possibility to lead a dignified life with dementia, and contradicts the *Convention on the Rights of Persons with Disabilities by the United Nations* (UN-CRPD, United Nations., 2006). Article 25 and 26 of the UN-CRPD demand the "highest attainable standard of health without discrimination on the basis of disability."

The present review is the first to systematically focus not only on descriptive characteristics of instruments, but also on measurement properties and on the amount and quality of available research for each instrument and each measurement property. We apply a comprehensive state-of-the-art methodology for reviews on psychometric properties, the *COnsensus-based Standards for the selection of health Measurement INstruments* (Mokkink, Prinsen et al., 2018; Prinsen et al., 2018; Terwee et al., 2018), and the *Characteristics of Assessment Instruments for Psychiatric Disorders in Persons with Intellectual Developmental Disorders* (CAPs-IDD, Zeilinger, Nader, Brehmer-Rinderer, Koller, & Weber, 2013).

We focus on informant-based scales, as they are most frequently used to screen for dementia in people with ID, and provide evidence-based recommendations for the most suitable and best-evaluated instruments. This will support clinicians and researchers in choosing the most adequate instruments for their respective purpose. Our work thus facilitates the detection of dementia in people with ID, which consequently can improve the lives of people with ID affected by dementia.

The objectives of this systematic review are to (1) identify informant-based dementia assessment instruments for people with ID, to (2) provide an overview of the characteristics of instruments, to (3) systematically describe and evaluate measurement properties for each instrument, including the amount and quality of available research, and to (4) draw recommendations for the most suitable instruments based on the collected information.

2. Method

The review was registered prospectively with the *International Prospective Register of Systematic Reviews* (PROSPERO; No. CRD42020181773), and reported in accordance with the *Preferred Reporting Items for Systematic Reviews and Meta-Analyses* (PRISMA) guidelines (Moher, Liberati, Tetzlaff, Altman, & The PRISMA Group, 2009; Page et al., 2021). The study protocol was published in a peer-reviewed journal (Zeilinger, Komenda, Zrnic, Franken, & Woditschka, 2020), and the study was conducted according to the protocol without any amendments. For collecting and evaluating information on each instrument, we applied an internationally agreed standard for evaluating outcome measures, the comprehensive COSMIN methodology (Prinsen et al., 2018), which ensures a transparent and replicable evaluation of assessment instruments. In addition, we used the CAPs-IDD to adequately address assessment criteria particularly relevant for people with ID (Zeilinger, Nader et al., 2013), such as level of ID or respondent requirements.

2.1. Search strategy and selection criteria

Two systematic literature searches were applied consecutively. The first search resulted in an inventory of informant-based assessment instruments for dementia in people with ID. The second search located evaluation studies for each instrument found in

the first search. To include grey and unpublished literature in both searches, we applied an invisible college approach, contacting authors in the field for information or manuscripts on this topic, and we followed up on meeting abstracts. Full texts of reviews on assessment instruments as well as book chapters and manuals identified in the course of the two searches were screened for possible further studies. References of papers meeting the inclusion criteria were hand-searched. Both searches were updated before the final analyses to include the most recent publications.

2.1.1. First search

Search strings for the first search are provided in (Appendix A in Supplementary Information). The search was carried out in ten international electronic databases: ASSIA, CINAHL, Cochrane Library, ERIC, MEDLINE, PsycINFO, Scopus, Web of Science, OpenGrey, and ProQuest Dissertations & Theses Global. For a study to be eligible, it had to meet the following inclusion criteria: (1) focusing on assessing dementia in people with ID, (2) describing the development or evaluation of an informant-based instrument for the assessment of dementia, and (3) the instrument had to be specifically developed or adapted for people with ID. Studies were excluded if they were about scales including dementia, but focusing on a broader spectrum of disorders for screening purposes or differential diagnosis.

We did not include publications prior to the year 2012. Instruments published up to the year 2012 were acquired from a previous systematic review that used a comprehensive search strategy and listed 114 instruments for assessing dementia in people with ID (Zeilinger, Stiehl et al., 2013). We examined these instruments applying our inclusion criteria. No language restrictions were imposed.

2.1.2. Second search

For the second search we conducted a cited reference search using the initial publication(s) of each instrument as a reference point. We based this strategy on the assumption that a paper evaluating an instrument would cite the initial publication of the respective instrument. If more than one publication was considered initial (e.g., a manual and a published paper), we used all possible initial publications as reference points for the second search. The papers and manuals used as a reference point were also included in the further appraisal of the literature. For published studies, we used five international databases that allowed cited reference searches: ERIC, PsycInfo, MEDLINE, Scopus, and Web of Science. For published manuals not listed in one of the five databases we searched Google Scholar.

To be included, studies had to describe an evaluation of the respective instrument in people with ID. Studies were excluded when: (1) the respective instrument was used primarily for other investigations, not related to an evaluation of the instrument, or (2) the study was a review of assessment instruments, not providing novel information.

2.2. Screening

To detect duplicates, automation tools were used. These were then checked and excluded by one reviewer. Duplicates were defined as records being exactly the same, i.e. studies reporting the same data but published in different journals were not considered duplicates. Title and abstract screening, as well as full-text evaluation were done by two team members independently, i.e. blinded to each other's decisions (FF, SK, AMM, ELZ). Team members included in the screening process were psychologists or clinical psychologists with knowledge and previous experience in the fields of intellectual disability, dementia, and psychometrics. In cases of disagreement, discrepancies were discussed until agreement was reached. If no agreement was reached, a third team member served as arbiter. Average percentage agreement between reviewers in the screening phase was 94.1 %.

2.3. Data extraction & quality assessment

In the first search, the names of the instruments and information on their initial publication(s) were extracted. In the second search, we extracted psychometric evaluation data of instruments, i.e. nine measurement properties as listed in the COSMIN checklist: content validity, structural validity, internal consistency, cross-cultural validity, reliability, measurement error, criterion validity, construct validity, and responsiveness. The exact definition of each measurement property can be found in the COSMIN guidelines (Mokkink, Prinsen et al., 2018; Prinsen et al., 2018). We further coded interpretability and feasibility aspects (length of instrument, completion time, ease of scoring, costs, and availability) as specified by the COSMIN guidelines, and extracted descriptive data according to the CAPs-IDD – Part I (e.g. level of ID, respondent requirements, item content; see Appendix B in Supplementary Information).

First, we evaluated the methodological quality of each measurement property per study as very good, adequate, doubtful, or inadequate using the COSMIN Risk of Bias Checklist (Mokkink, de Vet et al., 2018). The checklist consists of up-to-date standards regarding design requirements and statistical methods for studies on measurement properties. We then applied criteria for good measurement properties to rate the results of each single study as adequate (+), inadequate (-) or indeterminate (?). These evidence-based criteria (Prinsen et al., 2018) specify how to rate each of the nine measurement properties (e.g., reliability is considered adequate when intraclass correlation coefficient or weighted Kappa is greater than or equal 0.70).

Data extraction was done independently by two team members (FF, SH, SK, SVL, LCN, MS, IZN) for each study. We used the template data collection forms provided by the COSMIN initiative (https://www.cosmin.nl/tools/guideline-conducting-systematic-review-outcome-measures). Before coding of the studies began, all team members took part in a COSMIN guidelines training including a practical coding trial of four sample studies. Again, assessment discrepancies were discussed until consensus was reached, with the help of a third reviewer if necessary. To ensure that the rating procedure was consistent for all studies, emerging issues were discussed on a regular basis. Average percentage agreement between the initial coding of reviewers was 81.8 %.

2.4. Data analysis

The overall quality rating of each instrument was based on the quality ratings of single studies. The final evaluation was done independently by two reviewers (IZN, ELZ), both psychologists with knowledge and experience in intellectual disability, dementia, psychometrics, and evaluation research. Discrepancies were resolved in consensus meetings. Following COSMIN guidelines (Prinsen et al., 2018; Terwee et al., 2018), we determined whether the psychometric properties per instrument were sufficient (+), insufficient (-), inconsistent (±), or indeterminate (?). The COSMIN guidelines specify a very clear, transparent procedure for each measurement property to arrive at a final rating (Prinsen et al., 2018).

Thereafter, we applied the modified *Grading of Recommendations, Assessment, Development and Evaluation* (GRADE) approach as suggested by the COSMIN guidelines to evaluate the overall quality of the body of evidence for each psychometric property per instrument. In the GRADE approach the quality of evidence is assessed. Larger risk of bias, inconsistencies, and imprecision lead to lower ratings. Publication bias is not included in the modified GRADE approach, because publication bias detection methods are typically non-applicable to measurement property studies (Mokkink, Prinsen et al., 2018). The quality of evidence is summarized to be either high, moderate, low, or very low. For each of these four quality levels, COSMIN guidelines provide definitions and step-by-step instructions in order to arrive at a final rating (Prinsen et al., 2018). For example, the risk of bias is considered as extremely serious when there is only one study of inadequate quality, leading to a "very low" final rating.

2.4.1. Providing recommendations

To provide evidence-based, transparent recommendations we followed COSMIN guidelines and categorized instruments threefold: *Category A* includes instruments that can be recommended. These have to fulfil the following two criteria: (1) sufficient content validity as demonstrated by studies that have any level of evidence, and (2) sufficient internal consistency from at least low quality evidence. *Category B* includes instruments that have the potential to be recommended, but need further evaluation. All instruments not classified as *A* or *C* are assigned to category *B*. *Category C* includes instruments that have been shown to possess insufficient measurement properties in high quality accounts and can therefore not be recommended. Furthermore, recommendations are based on interpretability and feasibility aspects (Prinsen et al., 2018). Interpretability is the extent to which one can assign qualitative meaning to quantitative scores, e.g. by providing cut-off scores. Feasibility includes the length of an instrument, completion time, ease of scoring, costs, and availability aspects. Since this review evaluates instruments that are associated with a clinical diagnosis, we determined criterion validity (sensitivity/specificity) as another important measurement property for our recommendations.

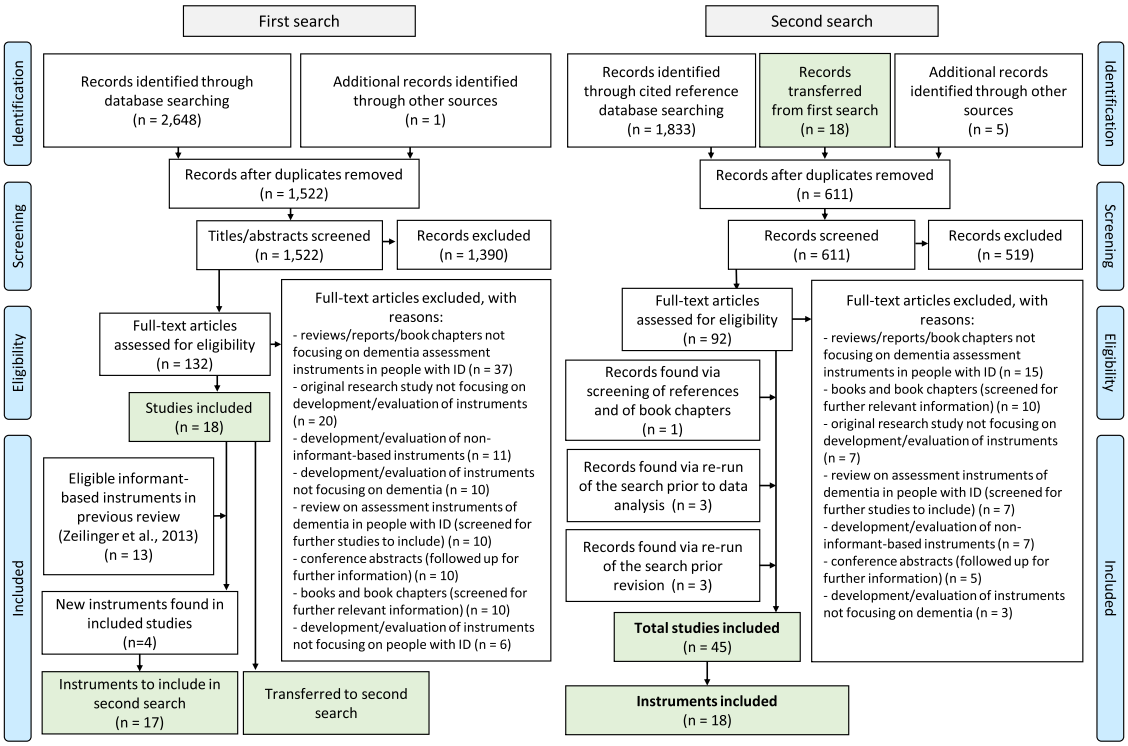

Fig. 1. PRISMA flow chart.

Table 1
Characteristics of included studies.

Study	Language of paper	Language of study	Country	Instruments used	Sample size (female)	Sample characteristics			
						Level of ID	Age-range	ID etiology	DS target population met[a]
Ball et al., 2004	English	English	UK	CAMDEX-DS	74 (31)	Mild-profound	> 30	DS	–
Beresford-Webb et al., 2021	English	English	UK	CAMDEX-DS	85 (42)	Mild-moderate	19–65	DS	–
Burt et al., 1999	English	English	USA	DLD	138 (74)	Mild-profound	29–82	ID/DS	–
(Burt et al., 2005a) (Comparing dementia diagnostic methods)	English	English	USA	MPC	168 (n.r.)	Mild-profound	30–69	ID/DS	–
(Burt et al., 2005b) (Tests and medical conditions)	English	English	USA	DSDS, DLD	78 (n.r.)	Mild-profound	30–61	DS	y
(Dalton, Fedor, Patti, Tsiouris, & Mehta, 2002)	English	English	USA	MOSES-DS	336 (151)	No ID, Mild-profound	17–88	No ID, ID/DS	n
(De Vreese, Mantesso, DeBastiani, Marangoni, & Gomiero, 2007)	Italian	Italian	Italy	DLD	60 (29)	Mild-severe	38-63	ID/DS	–
De Vreese et al., 2011[b]	English	Italian	Italy	AADS	63 (35)	Mild-profound	21–64	ID/DS	–
(De Vreese et al., 2015)	English	Italian	Italy	AFAST	61 (47)	Mild-profound	39–64	ID/DS	–
(Deb & Braganza, 1999)	English	English	UK	DSDS, DLD	62 (28)	Mild-severe	35–72	DS	y
(Deb, Hare, Prior, & Bhaumik, 2007)	English	English	UK	DSQIID	193 (95)	Mild-profound	23–77	DS	–
(Dekker et al., 2018)	English	English	Europe: Netherlands, France, UK	BPSD-DS	281 (141)	Mild-severe	31–74	DS	y
Dekker et al., 2021	English	English	Europe: Netherlands, Belgium, France, Italy, Spain	BPSD-DS	524 (246)	Mild-severe	30–74	DS	y
(Esteba-Castillo et al., 2013)	Spanish	Spanish	Spain	CAMDEX-DS	146 (51)	Mild-moderate	30–75	ID/DS	–
(Evenhuis, 1992)	English	Dutch	The Netherlands	DLD	139 (79)	Mild-profound	43–93	Only ID; no DS	–
(Evenhuis, 1996)	English	Dutch	The Netherlands	DLD	78 (n.r.)	Mild-profound	35–98	ID/DS	–
(Fonseca et al., 2019)	English	Spanish	Brazil	CAMDEX-DS	92 (33)	Mild-severe	n.r.	DS	–
(Friedman & Brown, 2001)	English	English	Canada	Friedman interview	50 (15)	n.r.	20–56	DS	y
Gomiero et al., 2008[b]	Italian	Italian	Italy	AADS	63 (35)	Mild-profound	21–64	ID/DS	–
(Gomiero et al., 2017)	English	Italian	Italy	DSQIID	200 (82)	Mild-profound	40–80	ID/DS	–
(Hoekman & Maaskant, 2002)	English	Dutch	The Netherlands	CLD /ESDC, DLD	329 (n.r.)	Mild-profound	40–91	ID/DS	–
(Huxley, Prasher, & Haque, 2000)	English	English	UK	DSDS	40 (11)	Mild-severe	26–66	DS	y
(Johansson & Terenius, 2002)	English	Swedish	Sweden	Johansson interview	9 (3)	Mild-severe	26–56	ID/DS	n
(Kirk, Hick, & Laraway, 2006)	English	English	UK	DLD	88 (33)	n.r.	41–86	ID/DS	–
Kuske & Mueller, 2017[c]	German	German	Germany	DSQIID	102 (51)	Mild-profound	41–96	ID/DS	–
Kuske et al., 2017[c]	English	German	Germany	DSQIID	102 (51)	Mild-profound	41–96	ID/DS	–
(Lessov-Schlaggar et al., 2019)	English	English	USA	CDR	34 (15)	n.r.	18–55	DS	y
(Li et al., 2015)	English	Chinese	Hong Kong	DSQIID	200 (86)		40–73	ID/DS	–

(*continued on next page*)

Table 1 (*continued*)

Study	Language of paper	Language of study	Country	Instruments used	Sample size (female)	Sample characteristics			
						Level of ID	Age-range	ID etiology	DS target population met[a]
(Nübling et al., 2020)	German	German	Germany	CAMDEX-DS	11 (n.r.)	Mild-profound n.r.	19–58	DS	–
(Oliver, Kalsy, McQuillan, & Hall, 2011)	English	English	UK	AADS	36 (19)	n.r.	30–64	DS	–
(Prasher, Farooq, & Holder, 2004)	English	English	UK	ABDQ	150 (67)	Mild-severe	16–76	DS	y
(Prasher, 1997)	English	English	UK	DLD	100 (44)	Mild-severe	19–78	DS	–
(Rebillat, Hiance-Delahaye, Falquero, Radice, & Sacco, 2021)	English	French	France	DSQIID	151 (74)	Mild-profound	40–74	DS	–
(Rösner et al., 2021)	English	German	Germany	DLD	71 (37)	Mild-profound	n.r.	ID/DS	–
(Sano, Aisen, Dalton, Andrews, & Tsai, 2005)	English	English	USA	BFT	108 (47)	Mild-profound	49–71	DS	y
(Shultz et al., 2004)	English	English	USA	DSDS, DLD	38 (17)	Mild-severe	45–74	ID/DS	n
(Silverman et al., 2004)	English	English	USA	DLD	273 (n.r.)	Mild-severe	45–70+	ID/DS	–
Silverman et al., 2021	English	English	USA	NTD-EDSD	185 (77)	Mild-severe	40–82	DS	–
(Startin, Rodger, Fodor-Wynne, Hamburg, & Strydom, 2016)	English	English	UK	CS-DS	128 (60)	Mild-severe	16–66	DS	y
(Startin et al., 2019)	English	English	UK	CS-DS	48 (22)	Mild-severe	36–64	DS	y
(Sturmey, Tsiouris, & Patti, 2003)	English	English	USA	MOSES-DS	163 (69)	Mild-profound	30–84	ID/DS	n
Takenoshita et al., 2020	English	Japanese	Japan	DSQIID	493 (182)	Mild-severe	20–83	ID/DS	–
(Walker, MacBryer, Jones, & Law, 2015)	English	English	UK	DLD	26 (11)	Mild-profound	40–69	DS	–
(Whitwham, McBrien, & Broom, 2011)	English	English	UK	Plymouth DSC	159 (69)	n.r.	18–89	ID/DS	–
(Zeilinger, Gärtner, Janicki, Esralew, & Weber, 2016)	English	German	Austria and Germany	NTG-EDSD	221 (172)	n.r.	19–65	ID/DS	–

Note: n.r. = not reported. [a]Some instruments were developed to be used with people with DS, only. Information in this column describes whether or not this target population specification was met by the respective study. [b,c]These papers describe the same study in two different languages.

3. Results

3.1. Results of the literature search

The first search yielded 2,649 records, 18 of which were eligible for inclusion. In these 18 records, 17 instruments were found to be included in the second search. The second search yielded 1,856 records, 39 of which were eligible. We found one additional instrument via screening of book chapters, leading to a total of 18 instruments which we evaluated further. Fig. 1 depicts the detailed search results. We updated the search twice: once prior to the final analysis on June 22, 2021, and once prior to the submission of a revised version of the paper on October 20, 2021. In each update, we found three additional papers, leading to a total of 45 papers included in this review. Appendix C in Supplementary Information lists the references of all included papers.

3.2. Characteristics of studies and instruments

The 45 papers included in this review describe 43 individual studies. Two studies each were duplicate publications in two different languages (De Vreese, Mantesso, De Bastiani, Marangoni, & Gomiero, 2011; Gomiero, Mantesso, De Bastiani, & De Vreese, 2008; Kuske & Mueller, 2017; Kuske, Wolff, Goevert, & Mueller, 2017). In these two cases, we kept both papers but considered them as a single

Table 2
Descriptive characteristics, feasibility, and availability of instruments.

Acronym	Name of instrument	Purpose of the measurement	Developed for DS[a]	Content of items	No. of items	Completion time[b]	Interpretability	Availability	No. of individual studies[c]
AADS	Assessment for Adults with Developmental Disabilities	Screening, research	n	Behavioural excesses and deficits	28	20 min.	No cut-offs	Free	2
ABDQ	Adaptive Behavior Dementia Questionnaire	Screening, research	y	Adaptive behaviour	15	10 min.	Cut-offs provided	Free	1
AFAST	Alzheimer's Functional Assessment Tool	Screening, diagnosis, monitoring of progress	n	Adaptive behaviour, ADLs, functionality, autonomy	46	20 min.	n.i.	Free	1
BFT	Behavior and Function Test	Screening, diagnosis research	y	Behaviour, function	58	no info	n.i.	n.i.	1
BPSD-DS	The Behavioral and Psychological Symptoms of Dementia in Down Syndrome Scale	Screening, research	y	Behavioural and psychological symptoms	52	30 min.	Preliminary cut-offs provided (by one study)	Not yet determined; probably free for research; for other purposes the intention is to keep it free or cheap (probably some kind of cost for online use)	2
CAMDEX-DS: informant interview	The Cambridge Examination for Mental Disorders of Older People with Down's Syndrome and Others with Intellectual Disabilities: informant interview	Diagnosis, monitoring of progress, research	y	Adaptive behaviour and cognitive abilities, general mental and intellectual functioning, memory, relevant previous medical and family history	157	50 min.	Preliminary cut-offs provided (by one study)	Can be purchased via publisher	5
CDR-DS	Clinical Dementia Rating Scale for adults with Down syndrome	Screening, diagnosis	y	Adaptive behaviour, ADLs, memory, orientation, judgment	41	30 min.	No cut-offs	License can be purchased; costs dependent on the purpose	1
CS-DS	Cognitive Scale for Down Syndrome	Screening, research	y	Memory, executive functions, language	61	20 min.	No cut-offs	Free	2
DLD / DMR	Dementia Questionnaire for Learning Disabilities / Dementia Questionnaire for Mentally Retarded Person	Screening	n	Adaptive and problem behaviour, memory, orientation, speech, practical skills, mood, activity	50	20 min.	Cut-offs provided for single completion and for change scores	Can be purchased via publisher	13 [c]
DSDS	Dementia Scale for Down Syndrome	Screening, diagnosis	y	Behaviour, ADLs, cognitive abilities	60	30 min.	Cut-offs provided	Only psychologists and psychometrists	4 [c]

(*continued on next page*)

Table 2 (*continued*)

Acronym	Name of instrument	Purpose of the measurement	Developed for DS[a]	Content of items	No. of items	Completion time[b]	Interpretability	Availability	No. of individual studies[c]
								can purchase the instrument	
DSQIID	Dementia Screening Questionnaire for Individuals with Intellectual Disabilities	Screening, research	n	Adaptive and problem behaviour, ADLs, cognitive abilities, psychological symptoms	53	15 min.	Cut-offs provided	Free	6
ESDC / CLD	Early Signs of Dementia Checklist / Checklist with Symptoms of Dementia	Screening	n	Adaptive behaviour, ADLs, cognitive and intellectual abilities, personality changes, physical and psychological symptoms	64	20 min.	Cut-offs provided	n.i.	1 [c]
Friedman Assessment Protocol	Friedman Assessment Protocol for Dementia in Down Syndrome	Diagnosis, research	y	Adaptive behaviour, cognitive abilities, memory, emotion regulation, motivation	no info	no info	n.i.	n.i.	1
Johansson Interview	Johansson Interview for Early Detection of Dementia in Persons with Down Syndrome	Screening, diagnosis	y	Adaptive and problem behaviour, cognitive abilities, emotions	92	40 min.	n.i.	Can be purchased via publisher	1
MOSES - DS	Multidimensional Observation Scale for Elderly Subjects - adapted for persons with Down syndrome	Screening, research	y	Adaptive and problem behaviour, ADL, cognitive abilities, physical and psychological symptoms, social withdrawal	40	20 min.	Cut-offs from MOSES, not extra provided	Probably free for research; n.i. for other purpose	2
MPC	Memory Problems Checklist	Screening, research	n	Everyday memory	10	10 min.	n.i.	Free	1
NTG-EDSD	NTG-Early Detection Screen for Dementia	Aiding health checks, documenting changes	n	Adaptive and problem behaviour, ADLs, cognitive abilities, psychological and physical symptoms	103	30 min.	No cut-offs	Free	2
Plymouth DSC	Plymouth Dementia Screening Checklist	Screening	n	Behaviour, memory, mood	3	3min.	Cut-offs provided	Free	1

Note: [a]DS = Down syndrome. [b]If no information on completion time was provided in the papers, the review team estimated this time based on inspection of the instrument. [c]Four papers reported the evaluation of two instruments in one study: Three papers on the DSDS and the DLD, and one paper on the ESDC and the DLD.

Table 3
Overall ratings of measurement properties and gradings of the quality of evidence across all instruments and studies.

Measurement properties		AADS	ABDQ[a]	AFAST	BFT[a]	BPSD-DS[a]	CAMDEX-DS	CDR-QDS[a]	CS-DS[a]	DLD/DMR
Content validity	Overall rating	+	±	+	? [b]	+	+	+	+	+
	Quality of evidence	low	low	very low (RoR)	very low	moderate	low	low	low	very low (RoR)
Structural validity	Overall rating	?	?	?	?	?	?	?	+	?
	Quality of evidence	n.i.	n.i.	very low	n.i.	very low	n.i.	n.i.	very low	n.i.
Internal consistency	Overall rating	+	?	+	?	+	+	?	+	+
	Quality of evidence	moderate	n.i.	very low	n.i.	high	high	n.i.	high	moderate
Reliability	Overall rating	intrarater: +interrater: -	?	+	?	+	+	+	+	+
	Quality of evidence	low	low	low	n.i.	high	moderate	low	moderate	high
Measurement error	Overall rating	?	?	?	?	?	?	?	?	?
	Quality of evidence	n.i.	n.i.	n.i.	n.i.	n.i.	n.i.	n.i.	n.i.	n.i.
Criterion validity	Overall rating	?	?	+	?	+	+	?	?	+
	Quality of evidence	n.i.	very low	moderate	n.i.	high	high	very low	n.i.	moderate
Construct validity: convergent/ discriminative[c]	Overall rating	+ / +	? / ?	+ / ?	? / -	? / +	+ / +	+ / ?	+ / +	- / -
	Quality of evidence	low / low	n.i. / n.i.	moderate / n.i.	n.i. / high	n.i. / high	high / high	very low / n.i.	high / high	moderate / low
Responsiveness	Overall rating	?	?	?	?	?	+	?	+	sensitivity: - specificity: +
	Quality of evidence	n.i.	n.i.	n.i.	n.i.	n.i.	moderate	n.i.	low	moderate
Category[d]		A	B	B	C	A	A	B	A	A

Measurement properties		DSDS[a]	DSQIID	ESDC/CLD	Friedman[a]	Johansson[a]	MOSES-DS[a]	MPC	NTG-EDSD	Plymouth
Content validity	Overall rating	+	+	+	? [b]	+	+	±	+	+
	Quality of evidence	very low (RoR)	very low	very low (RoR)	low	low	very low (RoR)	very low	moderate	low
Structural validity	Overall rating	?	?	?	?	?	?	?	?	?
	Quality of evidence	n.i.	moderate	n.i.	n.i.	n.i.	very low	n.i.	n.i.	n.i.
Internal consistency	Overall rating	?	+	?	?	?	+	?	?	?
	Quality of evidence	n.i.	low	n.i.	n.i.	n.i.	low	n.i.	n.i.	n.i.
Reliability	Overall rating	?	+	?	?	?	?	?	?	?
	Quality of evidence	n.i.	high	n.i.	n.i.	n.i.	very low	n.i.	n.i.	n.i.
Measurement error	Overall rating	?	+	?	?	?	?	?	?	?
	Quality of evidence	n.i.	high	n.i.	n.i.	n.i.	n.i.	n.i.	n.i.	n.i.
Criterion validity	Overall rating	+	+	sensitivity: - specificity: +	?	?	?	?	+	-
	Quality of evidence	sensitivity: moderate specificity: high	high	low	n.i.	n.i.	n.i.	n.i.	high	low
Construct validity: convergent/ discriminative[c]	Overall rating	+ / ?	+ / +	- / -	? / +	? / ?	? / ?	?	? / +	?
	Quality of evidence	moderate / n.i.	moderate / high	moderate / moderate	n.i. / low	n.i. / very low	n.i. / very low	n.i.	n.i. / high	n.i.
Responsiveness	Overall rating	+	?	?	?	?	?	?	?	?
	Quality of evidence	sensitivity: low specificity: moderate	n.i.	n.i.	n.i.	n.i.	n.i.	n.i.	n.i.	n.i.
Category[d]		B	A	B	B	B	A	B	B	B

Note: RoR = Based solely on the ratings of reviewers. n.i. = not investigated. *Overall rating of psychometric property*: + (sufficient), - (insufficient), ± (inconsistent),? (indeterminate).
Overall quality of the evidence: high, moderate, low, very low. *Cross-cultural validity/measurement invariance* is not included, as this property was not investigated in either of the studies.
[a]Instrument specifically for people with Down syndrome. [b]Content validity was not assessed by reviewers, as no access to instrument could be gained. [c]For construct validity, convergent and discriminative validity are reported separately: the first rating relates to convergent, the second to discriminative validity. [d]A: instruments that can be recommended. B: instruments that may have the potential to be recommended, but further evaluation studies are needed. C: instruments that cannot be recommended.

evaluation study. The majority of studies were based on data from high-income, English-speaking countries. Sample sizes ranged from $n = 9$ to $n = 524$ ($Mdn = 101$); percentages of women within samples varied between 27.5 % and 77.83 %. Study characteristics are presented in Table 1.

Out of the 18 identified instruments, 9 were evaluated by a single study only. Most studies ($n = 13$) were available for the *Dementia Questionnaire for Learning Disabilities* (DLD), followed by six for the *Dementia Screening Questionnaire for Individuals with Intellectual Disabilities* (DSQIID). Nine instruments were designed for people with DS, only. Most instruments include or consist mainly of behavioural aspects, which is in great contrast to dementia assessment instruments for people without ID. Characteristics of instruments are shown in Table 2 and in Table D.1 in In (Appendix D in Supplementary Information). References of evaluation papers for each instrument are also listed in Table D.1.

3.3. Psychometric properties, feasibility, and availability of instruments

Feasibility and availability aspects of all instruments are detailed in Table 2. Ratings of measurement properties and quality of the evidence of all instruments are depicted in Table 3. Table 4 lists detailed psychometric properties of recommended instruments. We describe results in relation to the COSMIN guidelines for providing recommendations (see Section 2.4.1. for more details).

3.3.1. Instruments for people with all ID aetiologies

Four instruments that are usable for people with all ID aetiologies (including people with DS) were classified as category A instruments. Of these, three had a sufficient rating for criterion validity: the CAMDEX-DS, the DLD, and the DSQIID. However, for content validity, arguably the most important measurement property, quality of evidence was low to very low for all four instruments. The only instrument with a superior quality of evidence for content validity was the *NTG-Early Detection Screen for Dementia* (NTG-EDSD), with high evidence for sufficient relevance and comprehensiveness, and moderate evidence for sufficient comprehensibility. However, internal consistency has not been evaluated for the NTG-EDSD. Therefore, it is not classified as category A instrument.

The CAMDEX-DS is the only instrument in category A not designed as a screening instrument, but as a more thorough assessment of possible dementia, aiming at establishing a diagnosis. The CAMDEX-DS is one of the best-evaluated instruments with a number of sufficient ratings (including criterion validity) supported by high quality evidence. Completion time is about 50 min. Scoring includes a differential diagnosis and is therefore sophisticated. Preliminary cut-offs have been established via one study (Beresford-Webb et al., 2021). The CAMDEX-DS has been translated into many different languages. It is a published instrument and has to be purchased.

The DLD is one of the most frequently used instruments for dementia assessment in persons with ID. However, ratings of measurement properties varied from sufficient (including criterion validity) to insufficient, and quality of the evidence was mostly moderate. The DLD is one of three instruments reviewed in this study with moderate or high quality evidence on insufficient ratings. For the DLD, this implies potential problems for construct validity and responsiveness. Completion of the DLD takes about 20 min, scoring is easy, and cut-offs for interpretability are provided. The DLD is a published instrument and has to be purchased.

The DSQIID received a number of sufficient ratings mostly with high quality evidence, including criterion validity. No measurement property was rated as insufficient. Completion of the DSQIID takes about 15 min, scoring is easy, and cut-offs for interpretation are provided. The DSQIID is freely available and has been translated and evaluated in many different languages, including Chinese, English, German, Italian, and Japanese.

3.3.2. Instruments specifically for people with Down syndrome

In addition to instruments recommended for all ID aetiologies, three instruments specifically developed for DS were assigned to *category A*, two of which had sufficient ratings on a variety of measurement properties: the *Behavioral and Psychological Symptoms of Dementia in Down Syndrome Scale* (BPSD-DS) and the *Cognitive Scale for Down Syndrome* (CS-DS).

The BPSD-DS had the best (i.e., moderate) quality of evidence for content validity. Criterion validity was sufficient with high quality evidence and preliminary cut-offs were established in one study (Dekker, Ulgiati, Groen, & Boxelaar, 2021). Administration time is 30 min, and scoring is straightforward. Costs for the BPSD-DS have not yet been determined. According to the authors, it will be cost-free for research purposes. For other purposes there may be a fee, especially for online use of the questionnaire. The BPSD-DS is available in many languages including Dutch, English, French, Italian, Norwegian, and Spanish.

For the CS-DS criterion validity has not yet been investigated, but it was the only instrument in this review with a sufficient rating for structural validity. Administration time is 20 min, scoring is easy, but currently no cut-offs are available. The CS-DS is free of charge and can be obtained from the authors of the instrument.

4. Discussion

The present paper describes the systematic and comprehensive review and evaluation of informant-based assessment instruments for dementia in people with ID, based on the analysis of 45 evaluation papers on a total of 18 assessment instruments. Among instruments usable for all ID aetiologies, including DS, we found two screening instruments, the DLD and the DSQIID, and one comprehensive assessment instrument, the CAMDEX-DS, that can generally be recommended. Based on our evaluation results, we recommend the DSQIID as the most appropriate screening instrument. The DSQIID is well evaluated in relation to measurement properties with high quality evidence for a number of positive ratings and has excellent feasibility, including interpretation, ease of administration, and scoring. It is available free of charge in many languages, which is especially valuable for international comparisons. Furthermore, the authors of the DSQIID constantly include new research results in their recommendations for use. Recent studies

Table 4
Measurement properties of recommended instruments.

Measurement property	Instruments for all ID etiologies		Instruments for people with DS only	
	CAMDEX-DS	DSQIID	BPSD-DS	CS-DS
Content validity[a]	Moderate evidence for sufficient comprehensibility and very low evidence for sufficient relevance and comprehensiveness of CAMDEX provided	Low evidence for sufficient comprehensibility, and very low evidence for sufficient relevance and comprehensiveness of DSQIID provided	Moderate evidence for sufficient relevance, comprehensiveness and comprehensibility of BPSD provided	Moderate evidence for sufficient comprehensibility and low evidence for sufficient relevance and comprehensiveness of CS-DS provided
Structural validity	n.i.	Two studies found a 4-factor-solution explaining between 45 % and 57 % of variance One study found a 3-factor-solution explaining 40 % of variance	11 clinically defined sections; satisfactory items' discriminative ability	5 factors (RMSEA = 0.05)
Internal consistency	Cronbach's α = 0.93	Cronbach's α: 0.91 – 0.95 for total scale	Cronbach's α: Frequency change: 0.85−0.90 Severity change: 0.80−0.88	Cronbach's α: Memory domain: 0.92 Executive function domain: 0.93 Language domain: 0.86
Reliability	*Interrater reliability:* Kappa = 0.60 – 0.91 for all items *Test-retest reliability:* Kappa = 0.92 for all items	*Interrater reliability:* ICC: 0.88–1.00 *Test-retest reliability:* ICC: 0.84 – 0.98	*Interrater reliability* Percentage agreement for individual items: 92%–100% *Test-retest reliability:* Percentage agreement for individual items: 62%–97%	*Interrater reliability* ICC: 0.84 *Test-retest reliability* ICC: 0.95
Criterion validity	*Sensitivity:* 0.80 – 0.98 *Specificity:* 0.81–1.00 AUC of 0.99 for the total score	*Sensitivity:* 0.92–1.00 *Specificity:* 0.96 – 0.99 AUC of .98 for the total score	*Sensitivity:* 69.80–76.70 *Specificity:* 72.60–83.20	n.i.
Construct validity: convergent/ discriminative	*Convergent validity:* CAMDEX-based diagnosis of AD shown to be consistent with objectively observed cognitive decline CAMDEX-DS versus DSM-IV: Kappa = 0.95; $p < 0.001$ CAMDEX-DS versus ICD-10: Kappa = 0.97; $p < 0.001$ *Discriminative validity*: Good discriminative ability with epsilon squared suggesting strong effects (.67 for total score)	*Convergent validity:* DMR vs. DSQIID: Pearson correlation coefficient rho = .64 (total score) DMR vs DSQIID: Spearman's rho = .24 (SCS); rho = .28 (SOS) *Discriminative validity:* Consistently higher DSQIID scores in groups with dementia/cognitive decline than those without dementia/cognitive decline Subgroup analyses (male/female; moderate/ severe ID) revealed comparable cut-off scores and psychometric properties to those in total study population	Total scale scores significantly higher in the group with dementia as compared to those without dementia	*Convergent validity:* CS-DS scores significantly correlated to measures of general abilities. *Discriminative validity:* Significantly lower scores for adults with cognitive decline than those without
Responsiveness	CAMDEX-based diagnosis of dementia shown to be a good predictor of future diagnosis Predictive value of precision: 85 % Positive predictive value: 0.50 – 0.58 Negative predictive value: 0.93 – 0.94	n.i.	n.i.	Changes in CS-DS scores found to be a valid measure to detect longitudinal changes in everyday cognitive abilities in adults with DS

Note: Percentages are reported as whole numbers. Other values are rounded to two decimal places. Included are only measurement properties that were evaluated for the respective instruments. DSM-IV = Diagnostic and Statistical Manual of Mental Disorders, Fourth Edition; ICD-10 = International Classification of Diseases, Tenth Revision; ICC = Intraclass correlation coefficient; AUC = Area under the receiver operating curve. n.i. = not investigated. [a]According to COSMIN, comprehensiveness, relevance, and comprehensibility are three relevant aspects of content validity.

(Silverman et al., 2021; Takenoshita, Terada, Kuwano, Inoue, Cyoju et al., 2020) suggest a lower cut off than the one initially proposed by the development study. In light of these findings, the authors advised that although a lower cut off may identify a number of false positives, it will miss a smaller number of false negatives, and adjusted the instructions of the DSQIID accordingly. The DSQIID was also chosen and successfully tested as the proxy rating of a recently developed German-language dementia assessment kit for people with ID (Mueller & Kuske, 2020), which further highlights its usefulness for clinical practice. The quality of the evidence for the DLD, an instrument that is frequently described as "well-evaluated" was unexpectedly low considering the large amount of evaluation studies available for this instrument. This further underlines the need for and relevance of high quality studies, as the quality of evaluation studies should be prioritized over quantity.

For people with DS only, we recommend two further instruments: the BPSD-DS and the CS-DS. Both instruments are well-evaluated with excellent feasibility aspects. The BPSD-DS comprises mainly behavioural and psychological symptoms, whereas the CS-DS focuses on cognitive abilities. Remarkably, the CS-DS is the only instrument in this review that had a rating for its structural validity, and it is available free of charge. The BPSD-DS is available in many languages and may therefore be especially valuable for international comparisons and research purposes.

Our results indicate the need for further evaluation studies in various domains, most importantly for content validity. For most instruments, the quality of evidence for content validity was low to very low. Only two instruments had a sufficient content validity rating with moderate quality evidence: the BPSD-DS and the NTG-EDSD. The BPSD-DS is among our recommendations for screening instruments for people with DS. The NTG-EDSD was developed by a large panel of experts to complement health screening requirements under the National Plan to Address Alzheimer's Disease in the United States (U.S. Department of Health & Human Services, 2012). Its main purpose is to document changes, communicate those changes to physicians, and thus aid physical health checks. It is mainly based on the DSQIID, one of the best-evaluated instruments in this review, and is complemented with health-related aspects like medication and chronic health conditions. Due to the superior rating for content validity, we provisionally recommend this instrument for the specific purpose of aiding health checks.

4.1. Strengths and limitations

Our review contributes to a more reliable detection of dementia in people with ID by providing an evidence-based evaluation of and recommendations for well-evaluated informant-based instruments, using the internationally agreed and comprehensive COSMIN guidelines. Thus, it supports the demands raised in the UN-CRPD regarding health care standards without discrimination on the basis of disability (United Nations., 2006). However, some limitations have to be mentioned. First, despite applying a comprehensive search strategy, there may be evaluation studies we have missed. Second, only studies focusing on evaluation of instruments were included. However, internal consistency is also frequently reported in studies focusing on other aspects. This may have contributed to the lack of information on this measurement property. Finally, we did not apply any restrictions related to language of studies. However, databases searched index mainly English-language journals. Thus, there may be studies in other languages not found via our search strategy.

4.2. Conclusion

People with ID should receive the same quality of mental health care as people without ID. For people at risk of developing dementia this includes an adequate and reliable screening and assessment procedure. Applying well-evaluated assessment instruments can considerably facilitate the detection of dementia in people with ID. Based on the information of evaluation studies we recommend the BPSD-DS, the CAMDEX-DS, the CS-DS, the DSQIID, and provisionally the NTG-EDSD. Our recommendations can be used by clinicians as well as researchers. Further evaluation studies, especially regarding content validity and structural validity are needed to strengthen the evidence-base of those instruments.

Data availability

The data that support the findings of this study are available from the corresponding author upon reasonable request.

CRediT authorship contribution statement

Elisabeth L. Zeilinger: Conceptualization, Data curation, Formal analysis, Investigation, Methodology, Project administration, Supervision, Validation, Writing - original draft, Writing - review & editing. **Irina Zrnic Novakovic:** Conceptualization, Data curation, Formal analysis, Investigation, Methodology, Visualization, Writing - original draft, Writing - review & editing. **Sophie Komenda:** Conceptualization, Data curation, Formal analysis, Investigation, Methodology, Writing - original draft, Writing - review & editing. **Fabian Franken:** Conceptualization, Data curation, Formal analysis, Investigation, Methodology, Writing - original draft, Writing - review & editing. **Marc Sobisch:** Formal analysis, Investigation, Visualization, Writing - review & editing. **Anna-Maria Mayer:** Formal analysis, Investigation, Visualization, Writing - review & editing. **Lennart C. Neumann:** Formal analysis, Investigation, Visualization, Writing - review & editing. **Sandra V. Loosli:** Formal analysis, Investigation, Visualization, Writing - review & editing. **Sarah Hoare:** Formal analysis, Investigation, Visualization, Writing - review & editing. **Jakob Pietschnig:** Conceptualization, Investigation, Supervision, Validation, Writing - review & editing.

Appendix F. Supplementary data

Supplementary material related to this article can be found, in the online version, at doi:https://doi.org/10.1016/j.ridd.2021.104148.

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