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A critical literature review of the effectiveness of various instruments in the diagnosis of dementia in adults with intellectual disabilities

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Abstract

Introduction: Currently, there is no consensus on dementia diagnostics in adults with intellectual disabilities (ID). There are three types of assessments available: direct cognitive tests, test batteries, and informant reports.

Methods: A systematic literature search was conducted in four databases yielding 9840 records. Relevant studies were identified and selected using predefined inclusion and exclusion criteria and then coded and classified according to assessment type. This was completed by two independent researchers, with a third consulted when discrepancies arose. The review collates diagnostic instruments and presents strengths and weaknesses.

Results: Overall 47 studies met the search criteria, and 43 instruments were extracted from the selected studies. Of which, 10 instruments were classified as test batteries, 23 were classified as direct cognitive tests, and the remaining 10 were informant reports.

Discussion: This review can recommend that cognitive test batteries can offer the most practical and efficient method for dementia diagnosis in individuals with ID.

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Keywords: Dementia; Diagnosis; Intellectual disability; Cognitive testing; Test battery; Informant reports; Assessment tools

1. Introduction

An intellectual disability (ID), similar to the UK specific term learning disability, onsets during the developmental period and is characterized by impairments of general mental abilities that impact adaptive functioning in three main domains: conceptual, social, and practical (American Psychological Association, 2013). [1] Various studies discussed throughout this review refer specifically to Down syndrome (DS). This is the most common genetic ID disorder seen in clinical practice. DS is caused 94% of the time by nondisjunction of chromosome 21 and 3%–5% of the time by translocation. The IQ of people with DS falls within the mild to moderately severe ID spectrum [2].

The life expectancy of individuals with ID is increasing due to improvements in medicine and living circumstances [3,4]. Individuals with mild ID are even experiencing life spans equal to those of the general population [5]. Adults with ID are subsequently in a position where age-related illnesses are becoming a greater concern. The most notable of these illnesses is dementia, for which an individual's age is the strongest risk factor (e.g., Daviglus et al [6]). Dementia of the Alzheimer's type is a cognitive impairment that gradually onsets, is progressive, and leads to interference with social and occupational functioning [7]. Dementia can be caused by a variety of underlying pathology. For example, Alzheimer's disease (AD) [8] consists of amyloid plaques and neurofibrillary tangles in the cerebral cortex, temporal lobe cortex, and hippocampus, among other brain areas [9].

Furthermore, individuals with ID often experience onset of aging characteristics earlier than in the general population [10], and this is reflected in age of dementia diagnosis. Onset of dementia usually occurs among older adults over the age of 65 years; however, in individuals with DS, onset is usually around the early 50s [11].

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The literature has shown substantial conflict in prevalence estimates of dementia in ID populations with and without DS when compared to the general population. Dementia has been shown to be common in older adults with ID but prevalence rates reported differ according to the diagnostic criteria applied [12]. It was found that diagnoses of dementia are substantially higher than in the general population, for people who have ID but do not have DS [13]. 21.6% of participants were diagnosed with dementia, compared to 5.7% that was expected in a non-ID group with this age structure. This was further supported by Strydom et al. [14] who highlighted the incidence rate of dementia in ID to be five times higher than that of older adults in the general population. Other studies have shown prevalence rates to only be comparable or higher than in the general population (e.g., Strydom et al. [15]). Opposing studies have shown risk of dementia to be equivalent to or lower than in the general population (e.g., Zigman et al. [16]). The variety in prevalence estimates further highlights the divergence in the understanding and application of dementia diagnostics for individuals with ID.

Stronger evidence has been established regarding dementia rates in individuals with ID and DS. Incidence of early-onset dementia of the Alzheimer's type has been shown to be higher than in the general population (e.g., Bush et al. [17]). Genetic findings have suggested that owing to the complex etiology of DS and the triplication of the amyloid precursor protein gene on chromosome 21, DS could be considered a model of early-onset dementia [18]. Almost all adults with DS over the age of 35–40 years show neuropathologic changes characteristic of AD [19], including senile plaques and neurofibrillary tangles. Although this does not necessarily reflect a clinical diagnosis, genetic evidence has merely begun to highlight similarities between the neuropathology of the two conditions. Unsurprisingly however, individuals with DS in many cases have been shown to be at higher risk of developing Alzheimer's disease than the general population (e.g., Nieuwenhuis-Mark [20]).

There is a need for further clarification of the difference in prevalence rates between the three populations, individuals from the general population with no pre-existing impairment, individuals with ID but without DS and individuals with ID and DS. Regardless of comparisons to the general population, evidence does show that the prevalence rates of dementia in ID increase dramatically between the ages of 40 and 60 years [21]. Therefore, dementia diagnostic assessments should be targeted at this age group or before.

Diagnosing dementia can be an incredibly difficult and complicated process. This is remarkably more complex in individuals with ID, as dementia and related pathology is manifested in areas of functioning that are, more than likely, already impaired by the intellectual disability [22]; thus leading to inherent difficulties in assessing cognitive functioning to aid with dementia diagnostics in people with ID [21]. There is no agreement in the literature or in practice on how dementia diagnosis should be informed in ID populations (e.g., Moran et al. [23]).

Assessments within the general population that build up a picture to aid the clinician when diagnosing dementia involves direct cognitive tests that indicate progressive cognitive decline in areas such as short-term and long-term memory, orientation, communication and mood, among others. But these tests are frequently not appropriate for individuals with ID as they often require abilities that individuals with ID may find more difficult due to their pre-existing impairment. The tests are not often developed for use in ID populations, and therefore, they do not reliably screen for dementia in this group [24]. Moreover, there are no normed data for this population, and thus, results cannot be interpreted meaningfully [23]. Consequently, floor effects are often observed on the chosen test and problems of accuracy in diagnosis ensue. There are three potential assessment methods that practitioners can apply to help inform diagnosis. These include a single test that directly assesses the individual's cognitive functioning, a test battery which comprises of multiple tests that assess a range of cognitive functions and lastly, informant reports which are completed by a carer or close relative who can report on the individual's functioning.

This review aims to collate existing instruments used in the diagnosis of dementia in individuals with ID. The instruments will then be coded according to whether they are (1) a direct cognitive test, (2) informant report, or (3) a test battery. The review will then discuss the benefits of each type of test. This review shall build on previous reviews by presenting an up to date overview of the instruments available, as well as discussing instruments that have been proposed for diagnostics in adults with ID but have yet to be established as such. This could include instruments that are designed for use in the general population, in the intellectually disabled populations or in people who have already been diagnosed with dementia. This review will therefore help clinicians to extend their knowledge of the potential cognitive assessments available as well as discuss non-cognitive assessments being used and give recommendations based on previous literature.

2. Methods

2.1. Literature search

A systematic literature search was conducted in four databases: PubMed, Science Direct, Google Scholar and PsycInfo. These databases were selected due to the depth and breadth that they offer in literature searching and their relevance to the reviewed topic. The search string included various terms for (1) the measure of interest (e.g., Alzheimer's disease, Dementia, Dementia of Alzheimer's type) and (2) the output of interest (e.g., diagnosis, assessment, instrument, screening tool). The search was performed once for the (3) specified population (e.g., intellectual disability, learning disability, mental retardation) and again for (4) Down syndrome, due to the well documented increased risk of dementia of Alzheimer's type in

this subgroup of individuals with ID. Table 1 shows the logic of the search strategy. References of included studies were also hand-searched, to include further relevant studies. Both English and non-English publications were sought after. However due to searches being conducted in English only, publications that had been originally written in English or translated into English were able to be included.

Relevant studies were identified and selected using the following inclusion criteria. Identified studies should be suitable dementia assessments for individuals with ID; this included informant reports, independent direct cognitive tests, or test batteries. Test batteries were included with both cognitive assessment and noncognitive assessment reported by an informant. Direct cognitive tests that are not yet used for dementia assessment but test a specific aspect of cognitive functioning like memory, intelligence, or orientation in an intellectually disabled population were included. Participants in selected studies included participants with ID that were classified as mild, moderate, severe, with or without the presence of DS. Included studies compared individuals with ID to individuals with ID who had already been diagnosed with dementia. Reviews, guidance documents, and dissertation projects were included when they pertained to the topic to consider and build on previous findings. These publications included discussion of dementia diagnostics in ID, instruments used for cognitive assessment, informant reporting for the purpose of dementia assessment, and guidance documents regarding dementia diagnostics or assessment in individuals with ID.

Studies were excluded if the instruments presented were not suitable for use in ID or DS populations. The instrument did not need to have been used for the purpose of diagnosis as of yet, but if it had been shown to be tolerated well by participants with ID and had been suggested for use in dementia assessment, then it was considered in this review. Diagnostic checklists and criteria were excluded as this review aimed to assess instruments that assess an individual with ID's functioning, either via an informant or directly, to aid the practitioner to complete checklists and criteria for dementia diagnosis. Checklists, although helpful when making the final decision regarding diagnosis, require heavy input from trained clinicians. This review sought to identify assessment methods that can be completed before input from the clinician as this will give the opportunity for diagnosis to be made more efficiently. Likewise, medical tests or studies focusing on biological or genetic markers were also excluded, due to their differential emphasis in the diagnostic process. Studies looking at interventions and treatments were also excluded due to lack of relevance to the diagnostic process.

2.2. Extraction of information and coding of instruments

Instruments were extracted from included studies and coded according to whether they were (1) an independent direct cognitive test completed by the individual, (2) an informant report completed by a carer or consultee on behalf of the individual or (3) a test battery consisting of multiple tests. Instruments were then put into the table that corresponds with the given code. Therefore if an instrument was coded as a direct cognitive test then it was placed into Table 2. Informant reports were added to Table 3, and finally Test batteries were placed into Table 4. Test batteries contain many different independent direct cognitive tests and informant reports, if the instrument was included in a battery it was described in Table 4, although it may be applicable to Table 2 or 3, this is to avoid repetition, so when considering which instrument to use do bear in mind that individual tests contained within test batteries are also available (see Table 4 for references). Instruments were further coded to highlight the level of ID and whether DS was present during the specified study. Tables therefore are displayed with non-DS participants denoted first, starting with mild ID, then moderate, and finally severe. After this, studies that compared ID participants without DS to participants with equivalent level of ID and DS as well. Finally, the tables displayed studies conducted with participants who have ID and DS. Selection and coding of studies were completed by two independent researchers (JEK, SS), with a third independent researcher consulted when discrepancies arose (RD).

3. Results

3.1. Results of the literature search

The literature searches conducted in all four databases yielded a total of 9840 studies. After excluding duplicates, screening titles and abstracts, 74 studies remained. These were assessed in full text, a further 34 studies were excluded at this point for not meeting the inclusion criteria. Thirty-six studies remained, and their references were hand searched

Table 1
Search string logic

Search Terms	Output	Measure	Population
Synonyms	Informant report, direct test, test battery, diagnosis, diagnostic, screening, assessment, tool, questionnaire, Scale	Dementia, Alzheimer's disease, Dementia of Alzheimer's type	Intellectual Disability, Learning Disability, Mental Retardation, Developmental Disability, Down Syndrome, Downs Syndrome.
Combined and Truncated	Inform* OR Informant Report* OR diagnos* OR screen OR screening* OR instrument* OR tool* OR Assess* OR questionnaire OR Scale*	Dement* OR Alzheimer*	((Intellectual* OR mental* OR learning OR developmental*) AND (disab* OR retard*)) OR (Down* AND syndrom*)

manually, identifying 12 additional relevant studies. An overview of the whole search and the results is shown in Fig. 1. In total, 48 studies met the search criteria.

A total of 44 instruments were found in the 47 included studies. There were 33 instruments to be completed by the individual and 11 to be completed by the carer or consultee. Of the 33 tests completed by the individual, 10 test batteries were identified, and 23 independent direct tests were identified. In the following sections, the instruments extracted are described in further detail.

3.2. Direct cognitive tests

During the literature search, 23 instruments coded as direct cognitive test batteries were identified; these are listed in Table 2. They each assess an aspect of cognitive functioning hypothesized to be associated with dementia, and therefore useful to assess when considering making a diagnosis. Various aspects of memory were the most frequently tested cognitive function. Memory domains included visual recognition, visual spatial, explicit, recall, and cued recall. Numerous tests sought to take a snapshot of overall cognitive functioning and mental status. Alternatively individual cognitive domains assessed included learning, various aspects of language, object recognition, executive function, and intelligence, among others. Many tests still incurred floor effects when participants were classed as having severe ID, reducing the tests potential for practical usage (e.g., PCFT [42]; MMSE [48]; CAMCOG [50]). The comments column in Table 2 denotes when a test has encountered this problem.

3.3. Informant reports

The informant reports highlighted by the studies in this review are detailed in Table 3. A total of 11 informant reports were reviewed. The informant reports nearly all assessed either behavior, dementia status, or daily functioning. These are noncognitive symptoms of dementia that indirectly indicate changes in cognitive functioning. When the participant is classed as having severe ID, these methods are more favored, as they do not require the individual to complete any tests that they may potentially find distressing. All informant reports in Table 3 were shown to be effective during the dementia diagnostic procedure, except for the Activities of Daily Living Questionnaire (ADL [54]), which was shown to not be effective in this population [55]. The Dementia Questionnaire for Mentally Retarded people (DMR [56]), which has been renamed at the Dementia Questionnaire for people with Learning Disabilities (DLD [57]) and Adaptive Behaviour Scale (ABS [43]) was highlighted to be most effective when used together, as they can cover a wide range of factors affected by dementia. This suggests that both adaptive behavior and general cognitive functioning that are assessed with these two scales are implicated in dementia diagnosis.

3.4. Test batteries

There were 10 test batteries identified in the literature search; which are listed in Table 4. Of these 10 batteries, four were designed for individuals with ID and five for individuals with Down syndrome. The remaining battery was designed for individuals in the general population who are already severely demented, rather than for use as an assessment battery. Eight of the batteries contained sections for informant reports as well, whereas two of the batteries chose to focus on just the participant's cognitive abilities. The test batteries varied in length from 20 minutes (severe impairment battery), up to 4 hours (Das Naglieri Cognitive Assessment System).

3.5. Other studies assessed

Table 5 shows the other studies reviewed. Although these studies did not present specific instruments, they discussed instruments available and presented their recommendations for diagnosis of dementia for people with ID and Down syndrome. Overall seven other studies were assessed, of which five were classed as literature reviews and two were classed as recommendation or advise documents, issued with the intention of informing practitioners of which route to take when diagnosing dementia in people with ID. All the documents agreed that a consensus should be reached to further research and benefit clinical practice. Many of the studies also highlighted how crucial longitudinal use of the chosen screening instrument is to accurately inform diagnosis [113] further argues for individually tailored assessment techniques to be employed, as many of the studies noted that the variability in cognition of individuals with ID makes it almost impossible to recommend one set instrument. Therefore selecting appropriate tests for the individual is key. Some studies also note the use of a multidisciplinary approach to most successfully inform diagnosis. This requires obtaining knowledge of the patient's history and making observations of not only their cognitive functioning, but also emotional, motivational and daily functioning.

4. Discussion

4.1. Summary

In this review, instruments that are used in the assessment of dementia in individuals with intellectual disabilities (IDs) were systematically collected and described. This review also presents information regarding the available instruments in a more accessible and condensed form for clinicians to use to inform decisions regarding dementia diagnostics for individuals with ID. Furthermore, strengths and weaknesses of each type of instrument were discussed.

The three categories of diagnostic instruments presented are direct cognitive tests, informant reports, and test batteries. Previous reviews agree that consensus needs to be reached to advance assessment of dementia in ID (e.g., Zeilinger et al.

Table 2
Direct cognitive tests

Author (Year)	Country and setting (clinical or applied)	Test name	Ability tested	Ppts	Type of ID	Groups	Outcome (what was sig?) ><	Comments
McDaniel (2000) [25]	United States—Applied setting (quiet room in their unit)	**Dementia Rating Scale (DRS)** [26]	General cognitive ability	84 ppts Aged: 14–60 years	Mild ID (n = 32) Moderate ID (n = 42) Severe ID (n = 10)	1 = Mild 2 = Moderate 3 = Severe	1 > 2 (sig) on Total Score and all subtests except Construction 2 > 3 (sig) on all measures.	DRS can provide info about the cognitive strengths and weaknesses of individuals with ID. DRS can be administered to a wide range of individuals with ID.
Pyo et al. (2010) [27]	United States—Applied Setting (separate room with a family or staff member present to make ppts feel more comfortable)	The revised Picture Recognition Memory Test (r-PRMT) [28]	Visual recognition memory	59 ppts (26 cases, 33 controls) Age: 40+	Moderate to severe	1 = DAT cases with DS (n = 15) 2 = DAT cases without DS (n = 11) 3 = Controls with DS (n = 9) 4 = Controls without DS (n = 24)	Controls > Cases on r-PRMT Controls with non-DS etiologies scored much lower with a wider score spread, resulting in significant overlap with the score distribution of DAT cases. Effect sizes indicated that ppts with DS were 5.35 for r-PRMT immediate and 4.44 for r-PRMT delayed which were significantly larger compared to non-DS ppts who showed effect sizes of 0.73 and 1.02, respectively.	r-PRMT may be effective at identifying DAT among moderate to severe from DS, however high false positive rate.
		The Modified Objective Memory Test (OMT)	Recall memory				Cases = controls on OMT (no sig difference)	
		Test for severe impairment (TSI) [29]	Mental status as a whole, including immediate memory recall and delayed recall.				Cases = controls on TSI (no sig difference)	
		The Neuropsychology (NEPSY) Comprehension of Instructions [31]	Language comprehension				Cases = controls on The NEPSY (no sig difference)	
Shultz et al. (2004) [30]	United States—Applied setting (designated rooms at ppts' group homes or workshops)	The Shultz Mental Status Exam Paired Associate Learning Task (modified from [32])	Overall mental Status Visual Spatial Explicit Memory	38 ppts aged: 45–74 years	ID without DS (32%) and ID with DS (68%)	Cases = Dementia Controls = Non dementia	Both performance tasks discriminated between groups. The performance tasks were related to dementia and IQ, but not age or sex.	Both the Shultz Mental Status Exam and the paired associate learning task were able to detect cases versus controls and therefore could be informative when diagnosing dementia in ID.

Krinsky-McHale et al. (2002) [33]	United States—Potentially a clinical setting but this is not specified.	Selective Reminding Test (SRT) [34] Modified for use in this population [35]	Explicit Memory	155 ppts	Down Syndrome vs individuals with ID but no DS. Equivalent level of ID between groups.	Cases 1 = DS with DAT Controls 1 = S without DAT Cases 2 = ID without DS with DAT Controls 2 = ID without DS without DAT	Cases 1 < controls 1 and cases 2 < controls 2 on long-term storage and retrieval processing abilities	These declines preceded other DAT symptoms, in most cases by more than 1 year & sometimes up to 3 years. Results confirm SRT can detect affected memory processes during early dementia in adults with DS.
Das et al. (1995) [36]	Canada and United States—Applied setting (quiet rooms located in a workshop, group or independent living setting)	**Dementia Rating Scale (DRS)** [26] Peabody Picture Vocab Test—revised [37] Matrix—Analysis Test—expanded form [38]	General cognitive ability Receptive vocabulary Nonverbal measure of intelligence	63 ppts Age: 40–49 years or 50–62 years	Down syndrome vs individuals with ID but no DS. Equivalent level of ID between groups.	Younger cases = DS aged 40–49 years Younger controls = non-DS aged 40–49 years Older cases = DS aged 50–62 years Older controls = non-DS aged 50–62 years	Older cases < younger cases, younger controls, older controls	Older DS individuals performed most poorly on the tasks involving planning and attention. DRS indicates good clinical utility. PPVT-r also discriminated effectively. Matrix was found to be too difficult for individuals with moderate to severe ID to complete.
Nelson et al. (2007) [39]	United States—Clinical Setting	Simple visual discrimination	Visual discrimination learning	19 ppts Age: 24–55 years	Down syndrome			Results demonstrated good reliability and validity of select tests.
		Reversal learning	Executive function	Mean = 40			Reversal and landmark 0: Sensitivity 71.43 and specificity 72.73	Sensitivity and specificity not given for tests individually.
		Delayed non-match to sample	Object recognition				Delayed non-match to sample and landmark 4: sensitivity, 72.73; specificity 27.27	
		Landmark stimulus—response task	Spatial learning and memory				Landmark 4: sensitivity 75; specificity 60	
McCarron et al (2014) [40]	Ireland & United States—Clinical Setting (Memory clinic)	Downs Syndrome Mental Status Exam (DMSE) [41]	Overall mental status	77 ppts Aged: 35+ years	Down syndrome	Cases = dementia Controls = nondementia	Average age of diagnosis = 55.41 (SD = 7.14) Median survival = 7 years after diagnosis Cases sig older than controls	DMSE was effective at picking up changes in functioning 1 year before diagnosis.

(*Continued*)

Table 2
Direct cognitive tests (*Continued*)

Author (Year)	Country and setting (clinical or applied)	Test name	Ability tested	Ppts	Type of ID	Groups	Outcome (what was sig?) ><	Comments
Kay et al. (2003) [42]	UK—Clinical Setting	Prudhoe cognitive functioning test (PCFT)	Overall mental status, including: orientation, recall, language, praxis, and calculation.	87 ppts Aged: 20+ years	Down syndrome	No dementia cases participated, the sample was made up of individuals with DS only.	PCRT sig. correlated with Adaptive Behaviour Scale (ABS—[43]) given to carers. PCRT sig. correlated with degree of ID More subjects with high levels (i.e., profound to untestable) of ID obtained very low or zero scores on PCFT.	PCFT = reliable quantitative measure of cognitive function in DS. Floor effects suggest that PCRT is limited to detecting cognitive decline to those who are less disabled.
Devenny et al. (2002) [44]	United States—Applied Setting (Quiet rooms in ppts' day program or at their residence)	Cued Recall Test (CRT) [45,46]	Cued memory recall	160 ppts	Down syndrome	Cases = with DS and early stage DAT Controls = DS with no DAT Controls 2 = ID no DS and no DAT	Cut-off value ≤23 on the TS = sensitivity: 94.7%, specificity: 93.9%, positive predictive value: 81.9% when cases compared to controls 2.	Usefulness of CRT needs to be confirmed with longitudinal data. Memory declines can occur several years before DAT identification.
Tyrrell et al. (2001) [47]	Ireland—Potentially a clinical setting but not clearly stated.	**Downs Syndrome Mental Status Exam (DMSE)** [41]	Overall mental status.	285 ppts Aged: 35–74 mean age ±SD = 46.5 ± 8.2 years	Down Syndrome	Cases = DS with dementia Controls = DS without dementia	Sig different Median scores in Cases vs Controls for DMSE.	
		Test for Severe Impairment (TSI) [29]	Mental Status as a whole, including immediate memory recall and delayed recall.				Sig different Median scores in Cases vs Controls for TSI.	No floor or ceiling effects in individuals with moderate and severe ID.
Deb et al. (1999) [48]	UK—Setting not clearly stated.	**The Mini Mental Status Exam (MMSE)** [49]	Overall mental status	62 ppts Aged: 35+ years	Down syndrome	Cases = DS with Dementia (n = 26) Controls = DS without dementia (n = 36)	MMSE could only be completed by 34 (55%) ppts with DS. 30 ppts got MMSE score less than 24 (the usual cut-off for the diagnosis of possible dementia), 23 ppts (77%; of the 30) did not have a diagnosis of dementia.	MMSE not able to be administered to all ppts with DS. And did not accurately identify cases or controls.
Hon et al. (1999) [50]	UK—Applied Setting (Ppts' home or day center)	Cambridge Cognitive Examination (CAMCOG)	Overall cognitive functioning	74 ppts Aged: 30+ years	Down syndrome	1 = Younger DS 2 = Older DS	CAMCOG scores = well distributed. 8 ppts (11%) scored 0. 1 > 2 (sig) on total CAMCOG score 1 > 2 (sig) on 6 out of 7 subscales.	CAMCOG useful unless ID is severe. May need some modifications to make it more accessible. Better than MMSE as well.

Pennington et al. (2003) [51]	United States—Applied Setting	Cambridge Neuropsychological Test Automated Battery, Paired Associates Learning (CANTAB-PAL—Robbins [52])	Visual-spatial explicit memory	56 ppts	Down Syndrome	1 = Children without DS 2 = Children with DS	Study was not assessing dementia but does show that the test is well tolerated in DS populations.	CANTAB-PAL was designed for use for assessing dementia in general population. But this study indicates that CANTAB-PAL may be able to be used in assessment of dementia in ID.
Boanda et al. (2008) [53]	Spain—Clinical Setting	The Mini Mental Status Exam (MMSE) [49]	Overall mental status	45 ppts Age: 40+ years	Down syndrome	Cases = Alzheimer's disease (AD) Cases 2 = Potential AD Control = Absence of AD	MMSE performance sig. correlated with total and cognitive DMR scores as well as SIB scores.	MMSE = useful for assessing cognition.

Abbreviations: ID, intellectual disabilities; DS, Down syndrome; DAT, Dementia Alzheimer's type; ppts, participants.
NOTE. Tests highlighted in bold indicate repeated use within studies. Age is denoted in years.

[24]). Clinicians currently lean toward using instruments that they are previously familiar or comfortable with; however, this has resulted in disparity in the instruments being used across clinical settings. By reaching a consensus, benefits will not only be seen in assessment efficiency and communication between health professionals but also in treatment. Earlier treatment has been suggested to maintain the highest possible level of cognitive functioning, whereas dementia is mild [114].

Many studies agreed that memory impairment is crucial to dementia diagnosis and therefore included assessments of various aspects of memory in their recommendations of instruments. Some studies chose to assess other cognitive domains either in conjunction with memory assessments or instead of, for example, tests of orientation, language, intelligence, executive functioning, to name a few. Although the study by Crayton et al. [97] observed a similar clinical progression in the participants with ID and dementia that is often seen in individuals with dementia but no pre-existing ID, the numerous different cognitive domains tested in the included studies highlight how onset, course, and progression of dementia can notably differ from person to person.

With this in mind, it is vital to consider the level of intellectual disability that the instrument will be most suitable for assessing. It is important to also note that instruments often differ in their applicability to clinical or applied settings. All instruments discussed can be administered in both settings; however, some instruments are better suited to one setting or the other. In any case, level of distraction, how comfortable the participant is, length of the test, and accuracy of information gathered always need to be considered when deciding where to administer various instruments.

4.2. Direct cognitive tests

Evaluation of the direct cognitive tests showed many instruments that are appropriate for application with people who have ID. Studies assessed a variety of levels of ID, DS only, as well as comparing participants with DS to individuals with equivalent level of ID but no DS. Therefore, the instruments assessed present a range of levels of ID and could be applicable across the population, if administered correctly.

Multiple studies indicated good clinical utility for the Dementia Rating Scale (DRS [26]) and the Downs Syndrome Mental Status Exam (DMSE [41]). Furthermore, the study by McCarron et al. [40] commented that the DMSE was particularly useful in detecting cognitive changes 1 year before dementia diagnosis and therefore could prove useful in early detection. Studies looking at the DRS only included a total of 147 participants [25,36], whereas studies looking at the DMSE included 362 participants [40,47]. Therefore, to further support the findings of these studies, more research will need to be completed using the DRS, particularly in a larger sample size. No studies evaluated showed the DRS or the DMSE to be unsuitable for dementia diagnostics in ID. Both instruments were used in applied and clinical settings in the reported studies, indicating their flexibility in

Table 3
Instruments based on informant reports

Author (Year)	Country and setting (clinical or applied)	Test name	Ability tested	Ppts and age	Type of ID	Groups	Outcome (what was sig?) ><	Comments
Zeilinger et al. (2015) [58]	United States—applied (in large residential care homes)	The National Task Group—early detection screen for dementia [59]	Dementia Status	221 carers	ID. All participants are cared for.	Groups	Four feasibility dimensions of use of the NTG-EDSD were reported on by carers. However, data from the NTG-EDSD were not assessed directly. All feasibility dimensions were rated good to very good and 80% of the carers found the NTG-EDSD useful or very useful in the early detection of dementia.	Reliability and validity of the instrument for clinical use in aiding dementia diagnostic assessment was not assessed. Therefore, further research is needed before use of this instrument.
Lin et al. (2014) [55]	Taiwan—Setting is not clearly stated but potentially an applied setting.	**Dementia Screening Questionnaire for Individuals with Intellectual Disabilities (DSQIID** [60]**)**	Dementia status	459 ppts Aged: 45+ years	ID of varying degree	Cases = Dementia Controls = Nondementia	Was used to identify cases and controls in this study. 16.3% of ppts in this study were identified as being demented based on the DSQIID.	Although originally designed for use in DS is an effective tool for diagnosing dementia in ID.
		Activities of Daily living Questionnaire (ADL [54])	Daily functioning				Disability level and comorbidity can explain 10% of the ADL score variation. Dementia conditions can only explain 3% of the ADL score variation in the study.	ADL would not be an effective tool for diagnosing dementia in ID
de Vreese et al. (2011) [61]	Italy—Applied setting	Assessment for adults with Developmental Disabilities Scale (AADS-I [61])	Behavior	63 ppts	All ID included		Good reliability and validity found.	Useful for detecting dementia if used longitudinally.
Kirk et al. (2006) [62]	UK—Setting is not clearly stated.	**Dementia questionnaire for mentally retarded people (DMR)** [56]	Dementia status Behavior	88 ppts Aged: 40+	Varying ID (n = 76) And DS (n = 12)	All ppts completed both tests	DMR significantly related to ABS	Would need to use both to assess an individual for dementia diagnosis as neither covers the full range of factors effected by dementia.
		The Adaptive Behaviour Scale (ABS-RC2) [43]					ABS significantly related to DMR	2 questionnaires showed significant relationships.
Shultz et al. (2004) [30]	United States—Applied setting (rooms at group homes or workshops)	The Dementia Scale for Down Syndrome (DSDS)	Dementia status	38 ppts Aged: 45–74	ID without DS (32%) and ID with DS (68%)	Cases = Dementia Controls = Non dementia	Both dementia scales discriminated between groups.	All informant reports used were able to detect cases vs controls and therefore could be informative to clinicians looking to make a decision regarding dementia diagnostics for people with ID.
		Dementia questionnaire for mentally retarded people (DMR) [56]	Dementia status				The dementia scales were not related to premorbid IQ, age, or sex.	
		Reiss screen for maladaptive behavior [63]	Adaptive behaviour				Various Reiss screen subscales also discriminated between groups.	

Prasher et al. (2004) [64]	UK—Setting is not clearly stated.	Adaptive Behaviour Dementia Questionnaire (ABDQ [64])	Behavior	150 ppts (83 male 67 females) Mean age: 44 years	Down syndrome	Cases = Diagnosed DAT during 5 year study Controls = remained non dementia throughout.	The scale has good reliability and validity. Overall accuracy = 92%.	First tool designed specifically for detecting DAT in DS.
Lin et al. (2014) [65]	Taiwan—Setting is not clearly stated but potentially an applied setting.	**Dementia Screening Questionnaire for Individuals with Intellectual Disabilities (DSQIID—**[60]**)**	Dementia Status	196 ppts Aged: 15–48 years	Down syndrome	Younger = adolescent ppt Older = adult ppts	Older > younger on DSQIID scores. Older age ($P = .001$) and comorbid conditions ($P = .003$) were significantly associated with DSQIID scores. Age ($P < .01$), severe disability level ($P < .05$), and comorbid condition ($P < .01$) significantly explained 13% of variation in DSQIID scores after adjusting for sex, education level, and multiple disabilities.	DSQIID used well to diagnose dementia here in DS but need to consider other demographic factors that play a large influence on dementia status.
Ball et al. (2004) [66]	UK—Setting is not clearly stated.	Modified version of Cambridge examination for mental disorders of the elderly (CAMDEX)	General cognitive functioning	74 ppts at first visit and 56 ppts at repeat 6 years later Aged: 30+ years	Down syndrome		CAMDEX-based diagnosis of AD shown to be consistent with objectively observed cognitive decline (good concurrent validity) and to be a good predictor of future diagnosis. Inter-rater reliability was good with Kappa >0.8 for 91% of items and >0.6 for all items.	Modified CAMDEX informant interview useful when diagnosing dementia in ID and DS.
McCarron et al. (2014) [40]	Ireland & United States—Clinical Setting (Memory clinic)	Daily Living Skills Questionnaire (DLSQ) [67]	Daily Functioning	77 ppts Aged: 35+ years	Down syndrome	Cases = dementia Controls = non dementia	Over 14 year follow-up average age of diagnosis = 55.41 years (SD = 7.14). Median survival of 7 years after diagnosis. Cases older than controls (sig) Decline in DLSQ score was shown 3-4 years prior to diagnosis. Presence of dementia also associated with epilepsy and sensory impairments.	Changes in DLSQ indicated diagnosis 3–4 years apriori. More effective than direct tests used (DMSE and TSI) Also informative about variables that are associated with dementia diagnosis.
		Dementia Questionnaire for Mentally retarded people (DMR) [56]					Among instruments used DMR most sensitive to tracking change in symptoms over time before diagnosis, reporting changes 5 years prior to diagnosis. Direct tests used only reported changes 1 year prior to diagnosis.	DMR most effective at reporting changes in functioning.

(*Continued*)

Table 3
Instruments based on informant reports (*Continued*)

Author (Year)	Country and setting (clinical or applied)	Test name	Ability tested	Ppts and age	Type of ID	Groups	Outcome (what was sig?) ><	Comments
Deb (2007) [60]	UK—Setting is not clearly stated.	**Dementia Screening Questionnaire for Individuals with Intellectual Disabilities (DSQIID—**[60]**)**	Dementia Status	193 ppts Aged: 23–77 years Mean age = 55 years	Down syndrome		Sensitivity = 0.92 and specificity = 0.97 On DSQIID score of 20. Internal consistency (a1/4 0.91) for all its 53 items, and good test–retest and inter-rater reliability. Good construct validity was established by dividing the items into 4 factors.	Valid and reliable screening method for dementia in DS.
Kay (2003) [42]	UK—Clinical setting	Adaptive Behaviour Scale (ABS)	Behavior	87 ppts Aged: 20+ years	Down syndrome	No dementia cases participated, the sample was made up of individuals with DS only.	Significantly correlated with direct test Prudhoe cognitive functioning test (PCFT—see Table 1) ABS correlated significantly with the degree of ID.	Was able to obtain scores for all levels of ID including profound, whereas the direct test was not able to.
Deb et al. (1999) [48]	UK—Setting is not clearly stated.	**Dementia Questionnaire for persons with Mentally Retardation (DMR)** [56] **Dementia Scale for Down Syndrome (DSDS** [68]**)**	Dementia Status Dementia Status	62 ppts Aged: 35+ with DS.	Down syndrome	Cases = Dementia (n = 26) Controls = non dementia (n = 36)	DMR and DSDS showed good positive correlation. A similar positive correlation was found between the overall DSDS score and the scores in the main subcategories of the DMR. Direct test used (MMSE) could not be completed by all ppts.	Informant scales, rather than the direct tests, were more useful for the diagnosis of dementia in people with an intellectual disability.

Abbreviations: ID, intellectual disabilities; DS, Down syndrome; DAT, dementia Alzheimer's type; ppts, participants.
NOTE. Tests highlighted in bold indicate repeated use within studies. Age is denoted in years.

Table 4
Test batteries

Author	Battery name—designed for…	Informant reports contained in battery	Ability tested	Direct tests contained in battery	Ability tested	Ppts, age, and group	Type of ID	Outcome and comments
Burt et al. (2000) [69]—United States	Working Groups Battery—designed for dementia diagnosis in ID.	- DMR [56]	- Dementia status	- Test for Severe Impairment modified (TSI [29,70])	- Memory and Other Cognitive decline	None reported in first study – reliability studies did follow…		**1–1.5 hours to administer.** Longitudinal administration is crucial to observing clinical change.
		- The Dementia Scale for Downs Syndrome (DSDS—[68])	- Dementia status	- Stanford Binet sentences [71]	- Immediate recall	- Pyo et al. [28] showed good reliability on the Autobiographical memory and Orientation tests.		
		- Reiss Screen for maladaptive behavior [63]	- Emotional/motivational changes	- Flud modified [72]	- Immediate and delayed recall	- Pyo et al. [73] only looks at orientation and shows significant differences between DAT group and controls but Orientation tests alone are not reliable for diagnosis.		
		- Scales of Independent Behaviour—revised (SIB-R) [74]	- Behavior	- Spatial Recognition Span [75]	- Immediate spatial recognition	- Pyo et al. [76] showed autobiographical memory tests to be reliable.		
		- AAMR Adaptive Behaviour Scale: Residential and Community [43]	- Behavior	- Autobiographical memory	- Autobiographical memory			
		- Stress Index	- Differential diagnostics (stress)	- Orientation [77]	- Orientation to time and place			
				- Boston Naming Test [78]	- Expressive vocabulary			
				- McCarthy Verbal Fluency [79]	- Language fluency			
				- Simple commands (modified from Haxby [41])	- Receptive language			
				- Purdue Pegboard modified [80]	- Fine motor speed			
				- Developmental Test of Visual Motor Integration [81]	- Perceptual motor skills			

(*Continued*)

Table 4
Test batteries (*Continued*)

Author	Battery name—designed for…	Informant reports contained in battery	Ability tested	Direct tests contained in battery	Ability tested	Ppts, age, and group	Type of ID	Outcome and comments
Palmer (2006) [82] - USA	Not given—designed for dementia assessment in individuals with Mental Retardation.	- The early signs of dementia check-list [83]	- Dementia status	- The Color Trials Test [84]	- Visual Attention and Concentration	22 ppts Aged: 33–66 years Groups: Cases = Dementia Controls = matched for IQ, age, presence of DS and sex but no dementia present.	Mild or Moderate ID.	**2–2.5 hours to administer.** Cases < Controls in all areas assessed.
				- The Boston Naming Test [78]	- Expressive Vocabulary (Agnosia)			
				- The Controlled Oral Word Association Test (COWAT—Spreen et al. [85])	- Language: sematic fluency			
				- The Fuld Object Memory Evaluation [86]	- Memory and Learning			
Van der Wardt et al. (2011) [87]—UK, applied setting	Cognitive computerized test battery for individual's with intellectual disabilities (CCIID)—designed to assess IQ in individuals with ID.	N/A	N/A	- Corsi Block Tapping Test [88]	- Visual/spatial working memory	Reliability and validity studies were conducted in various ID populations and showed the CCIID to be a valid and reliable instrument for testing IQ.	ID all levels	**30 minutes to administer.** Originally designed as an IQ test for verifying eligibility for Paralympic sporting events, but has been suggested for use in dementia assessment – not yet tested however for this purpose.
				- Series	- Inductive Reasoning			
				- Odd one out	- Inductive Reasoning			
				- Jigsaw	- Visual/Spatial Abilities			
Silverman et al. (2004) [89]—United States, applied setting (ppts' residence or day program facility)		- Informant interviews conducted based on a clinical record review of the participants medical history.	- Medical history	- IBR evaluation of mental status [90]	- Orientation	273 ppts Aged: 45+ years After testing grouped into: 1) Not demented 2) Questionable 3) Possible dementia	All levels of ID.	2 hours to administer. 18 month longitudinal analysis presented. Findings suggest that by conducting a full assessment of cognitive abilities like presented here, diagnosis of dementia can be
		- The Dementia Questionnaire for Mentally Retarded persons [56].	- Cognitive abilities and social skills	- Downs syndrome Mental Status Examination [41]—including expanded memory section.	- Overall Cognitive Functioning - Overall cognitive functioning			

		- Part 1 of the American Association on Mental Deficiency Adaptive Behaviour Scale (ABS [91]) - Reiss Screen for Maladaptive Behaviour [63]	- Description of functional abilities - Screening for possible depression, psychosis, and behavior problems.	- Test of severe impairment (TSI—Albert et al. [29]) - The Peabody Picture Vocabulary Test – Revised (PPVT—Dunn et al. [37]) - Verbal Fluency Test [79] - The Beery Visual Motor Integration Test, long form [92] - Block design subtest of WISC-R [93] - Selective Reminding Test [34]	- Receptive vocabulary - Verbal fluency - Construction abilities - Visual spatial memory - Episodic memory	4) Definite dementia 5) Declines with complications		made a lot more rapid and accurate.
Das et al. (1995) [36]—United States and Canada, applied setting (quiet rooms in workshops, group or independent living setting)	Das Naglieri Cognitive Assesment System—designed to assess cognitive decline due to aging among individuals with Downs Syndrome.	N/A	N/A	- Planned search [94] - Matching numbers [95] - Number finding [96] - Expressive Attention [96] - Receptive Attention [95] - Simultaneous Verbal - Figure Memory - Word Series - Color ordering - Speech rate	- Visual search and planning - Planning - Attention, Vigilance - Expressive attention - Receptive Attention - Language - Simultaneous processing memory - Recall Memory - Spatial Memory - Speech rate (Verbal Fluency)	63 ppts Aged: 50–62 Groups 1) Young DS (n = 16) 2) Old DS (n = 16) 3) Young Non-DS (n = 16) 4) Old Non-DS (n = 15)	ID with DS or ID without DS with equivalent level of ID.	**1.5–4 hours to administer** – a lot of variation in time taken. 2 < than all other groups on all tasks. Seen most on tasks requiring planning and attention.

(*Continued*)

Table 4
Test batteries (*Continued*)

Author	Battery name—designed for…	Informant reports contained in battery	Ability tested	Direct tests contained in battery	Ability tested	Ppts, age, and group	Type of ID	Outcome and comments
Crayton et al. (1997) [97]—UK	Neuropsychological Assesment of dementia in adults with intellectual disability—designed for dementia assessment in Downs syndrome.	Cognitive test battery was compared to… - Vineland Adaptive Behaviour Scale (VABS—Sparrow et al. [98])	- Adaptive behaviour	- British Picture Vocabulary Scale (BPVS—Dunn, Dunn, Whetton & Pentilie, 1982) [99] - Orientation (taken from Cambridge Mental Disorders of the Elderly Examination—CAMDEX [100]: - Picture Naming (taken from BPVS) - Picture identification (taken from BPVS) - Acting on request - Card Sorting task Computerized tests - Visual memory - Pattern recognition - Spatial recognition - Matching-to-sample - Delayed response - Conditioned associative learning	- Receptive Language - Orientation - Aphasia - Agnosia - Receptive Language - Executive Functions - Visual Memory - Recognition - Spatial Abilities - Object recognition - Delayed response - Conditioned associative learning	70 ppts Aged: 28+ Mean Age: 42.8 Groups 1) under 40 years old 2) between 40 and 49 years 11 months old 3) aged 3 50+ years	DS	**1.5 hours to administer.** VABS and all neuropsychological tests negatively correlated (sig) – preexisting global cog impairment shown on these tests No difference between age groups (1, 2, and 3) on neuropsychological deficits. – because of screening method used before study. 2 & 3 < 1 performance on memory tests (sig) Results suggest sensitive tests that were used could be useful in dementia diagnostic process.
Oliver et al. (1998) [101] – UK	Different test batteries were collated, including the CANTAB and CAMCOG, plus extra tests added for the purpose of this study. (Please see across) – designed to detect age-related cognitive change in DS.	- Vineland Adaptive Behaviour Scale (VABS [98])	- Adaptive behaviour	- Visual memory battery (part of Cambridge Neuropsychological Automated Test Battery—CANTAB, see [103]), only 2 sections analyzed in this study, delayed response and conditioned associative learning tasks. - Orientation section of CAMCOG (part of the Cambridge Assessment for Mental Disorder in the Elderly—CAMDEX) Asked to name 14 pictures of everyday objects and	- Learning and memory - Aphasia and Agnosia	57 ppts Aged: 30+ Groups 1) No cognitive deterioration 2) Cognitive deterioration 3) Moderate cognitive deterioration 4) Severe cognitive deterioration	DS	**Does not state how long the battery took to administer.** 28.3% of ppts showed severe cognitive deterioration, like apraxia or agnosia. A higher prevalence of these impairments was associated with older age. Rate of cognitive deterioration also ↑ w/age & degree of pre-existing cognitive impairment.

				identification following a verbal instruction. Also asked to carry out simple actions on a verbal cue (e.g., clap your hands). - The British Picture Vocabulary Scale (BPVS) (Dunn, Dunn, Whetton & Pentilie, 1982) - Extra verbal memory test added to batteries used. (adapted from the Memory for Sentences Test – [102]) - Extra procedure added at the start of the memory for objects test, involving naming, identification, immediate recall and delayed recall.	- Apraxia - Receptive language - Verbal memory - Memory			Deterioration in memory, learning and orientation preceded the acquisition of aphasia, agnosia and apraxia. Pattern of cognitive deterioration seen with individuals who have DS in this study is comparable to the pattern reported in individuals who have Alzheimer's disease but do not have DS.
Jozsvai et al. (2002) [104]—UK, Clinical Setting	Not given—designed to assess cognitive decline in DS.	- The Dementia Scale for Downs syndrome (DSDS [68])	- Dementia status	- The Peabody Picture Vocabulary test revised (PPVTr – [37]) Battery included: - Information and Orientation Questions (IO) - Block Design Test (BD – from WISC-R: [93]) - Fuld Object Memory Evaluation (FULD—[86,105]) - Grocery list (GL) - Boston Naming Task (BNT—[78]) - Test of Apraxia (PX)	- Receptive vocabulary, verbal intelligence - Orientation - Visuo-constructional praxis - Immediate and delayed memory - Category fluency - Expressive vocabulary - Apraxia	35 ppts Aged: 28+ years Groups: Cases = diagnosed DAT using DSDS (n = 12) Controls = without DAT (n = 23)	DS	- Does not include more severe ID in sample	**Doesn't state how long the battery took to administer.** FLUD and IO shown to be most useful tests in battery—must be wary of practice effects though. BNT and BD, most effected by aging & had least diagnostic ability.

(*Continued*)

Table 4
Test batteries (*Continued*)

Author	Battery name—designed for…	Informant reports contained in battery	Ability tested	Direct tests contained in battery	Ability tested	Ppts, age, and group	Type of ID	Outcome and comments
Johansson et al. (2002) [106]—Sweden	Not given—designed to assess dementia in DS.	Informants were interviewed with questions regarding the ppts abilities in the following aspects and any changes observed in these abilities: 1) Change (global changes in ppt) 2) Support 3) Learning Adaptability		- Spatial tests of memory - Verbal tests of memory including: objects to be remembered, auditive learning, visual learning, supported learning, sensing items inside a bag, Where did I put it? - Other cognitive tests included: - Understanding pictures - Simplified Arithmetic - Telling the time on a clock - Ability to estimate time taken - Understanding of cause and effect - Drawing ability - Proper prepositions - Copying pictures with and without the original - Agnosia: what did you draw? - Word Fluency - Routine Decisions - Understanding reverse order - Arranging a coffee break - Naming (aphasia, agnosia, and apraxia) - Long-term memory questions included: - Biographical questions - Memory of the dys preceding the interview - Past and present friends and staff at residential and occupational settings - Semantic memory - Prospective memory		9 ppts Aged: 26–56 Groups: 1) No Decline 2) Possible Decline 3) Decline	DS	**Ppt section took 1.5–2 hours to administer.** Advocates a combination of testing and interviewing in order to gain a full clinical picture.
Witts (1998) [107]—UK, applied setting (adult training centers.	Severe Impairment Battery (SIB [108])—designed to assess cognitive functioning of the severely demented client.	- Vineland Adaptive Behaviour Scale (VABS [98])	-Adaptive behavior	Battery tests focus on: - Attention - Orientation - Language - Memory - Visuo-perception - Construction - Praxis - Social interaction Carers provide most of the info for the battery.		33 ppts Mean age = 36 years	DS	**20 minutes to administer.** Good reliability and validity found. No floor effects encountered. Should be used longitudinally.

Abbreviations: ID, intellectual disabilities; DS, Down syndrome; DAT, dementia Alzheimer's type; ↑, increases; ppts, participants.
NOTE. Tests highlighted in bold indicate repeated use within studies. Age is denoted in years.

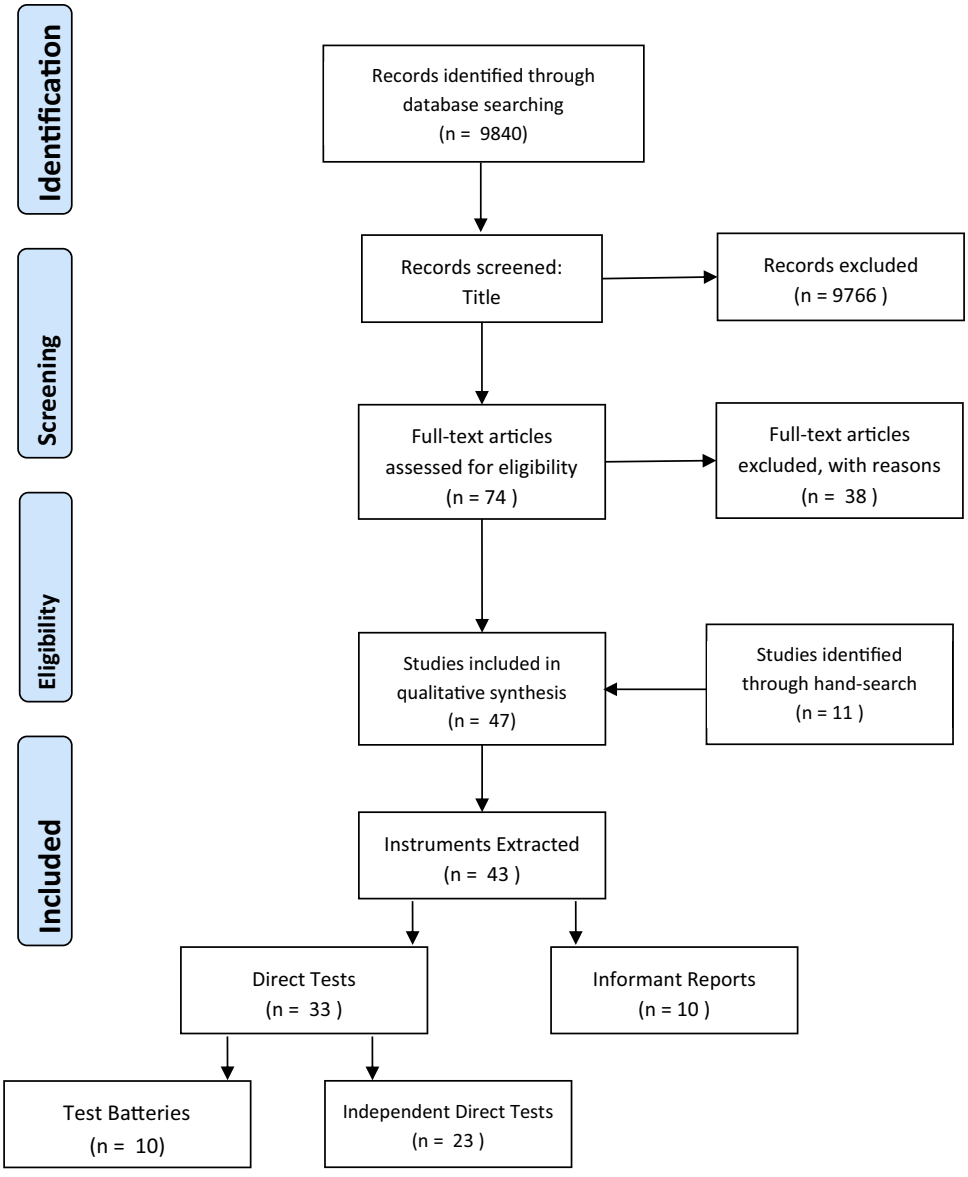


Fig. 1. A PRISMA flow diagram detailing the search strategy and results.

application and therefore good potential for use in informing dementia diagnostics.

Additionally, the modified version of the Selective Reminding Test (SRT [35]) was shown to have good utility in early detection, picking up cognitive changes between 1 and 3 years before dementia diagnosis [33]. Although there were no studies opposing this conclusion, this was only shown in one study of 155 participants; therefore, further research is required to support the clinical utility of the SRT.

Discrepancies in the effectiveness of the mini mental status examination (MMSE [49]) and the test for severe impairment (TSI [29]) arose. For example, the study by Boada et al. [53] was able to show the MMSE discriminated effectively between people with ID and people with ID and dementia. Similarly, Tyrrell et al. [46] replicated findings using the TSI. However, studies are inconsistent as Deb et al. [48] found the MMSE to show no significant difference between people with ID with and without dementia. Pyo et al. [27] also found no significant difference with the TSI.

The direct cognitive assessments shown to be most effective in this literature review included the DRS, DMSE, and SRT; however, each requires further assessment in larger sample sizes. Several studies noted the importance of tests being administered longitudinally, as there are no normative data for individuals with an ID as of yet. If used longitudinally clinicians can observe any cognitive decline, which could be very informative and necessary for making a decision regarding dementia diagnosis. Having said that,

Table 5
Other studies reviewed

Author (year)	Country	Document	Summary of key points
Moran et al. (2013) [23]	United States	Advise document	- A patient's history is key. - Gives a list of cognitive assessments and states that one should be used. - Regardless of the clinician's choice of instrument, the focus should be on recognizing change and decline in relation to a premorbid baseline.
Zelinger et al. (2013) [24]	Austria	Literature review	- No consensus in the literature and practice about what instrument should be used to diagnose dementia in ID. - Establishing consensus would improve the quality of assessment in clinical practice & benefit research.
Nieuwenhuis-Mark (2009) [20]	The Netherlands	Literature review	- No consensus on how dementia should be diagnosed in Down Syndrome. - Longitudinal studies using multiple tests accessing cognitive, emotional, motivational and daily functioning in individuals are recommended as is focus on change in functioning, 'bounce patterns' and multidisciplinary diagnosis
Krinksky-McHale et al. (2013) [109]	United States	Literature review	- While the intellectual and developmental disabilities field has for some time recognized the need to develop best-practices for the diagnosis of MCI and dementia, there remains a pressing need for empirically based assessment methods and classification.
Nagdee (2011) [110]	South Africa	Literature review	- In patients with ID, standard clinical methods need to be supplemented by careful, longitudinal behavioral observations and individually tailored assessment techniques. Co-morbidity, multiple biological, psychological, and socio-environmental factors, and complex interactions among events, are the reality for many aging people with ID. Determining the various influences is often a formidable clinical task but should be systematically carried out using medical, cognitive, behavioral, neuropsychiatric, and psycho-social frameworks.
Strydom et al. (2003) [111]	UK	Literature review	- The most promising informant-rated screening tool in most adults with ID including Down syndrome (DS) diagnosis is the Dementia Questionnaire for Persons with Mental Retardation (DMR). However, sensitivity in single assessments is variable, and cut-off scores need further optimization. In those with DS, the Dementia Scale for Down Syndrome (DSDS) has good specificity but mediocre sensitivity. The Test for Severe Impairment and Severe Impairment Battery are two direct assessment tools that show promise as screening instruments but need further evaluation.
Suh (2013) [112]	Hong Kong	Advise document	- Discusses a method for viewing statistical analyses of diagnostic screening tool results. - Values clinical judgment and advises that we adhere to 2 standard deviations away from population norm as cutoffs rather than a set score. As this will reduce false biases and false negatives and allow common sense to override whether pathology is present there.

Margallo-Lana [115] highlighted findings that longitudinal follow-up is not useful in people with severe ID. So, test selection needs to be carefully tailored to the level of functioning of the individual and the setting in which the testing can take place.

4.3. Informant reports

Instruments classed as informant reports evaluated noncognitive concepts, such as activities of daily living and functioning, as individuals with dementia find many activities of daily living difficult due to decline in episodic memory [116]. Informants are often in a good position to observe these changes. Furthermore, informants reporting on everyday functioning, prospectively or retrospectively, are much more effective than reporting on changes in memory [117]. These noncognitive concepts have also been shown to hold greater significance to individuals with ID and their carers than evaluation of cognitive changes [118]. Although the effectiveness of informant reports often varies from study to study (e.g., Jozsvai et al. [119]), in the studies reviewed here, informant reports were shown overall to be an effective way of aiding in dementia diagnostics. As informant reports are not completed by the participant, they are exceedingly suitable for individuals who have severe ID. A variety of both clinical and applied settings were used in the reviewed studies and no studies commented on the setting being inappropriate for the assessment, but again level of distraction and accuracy of data do always need to be considered when deciding where to administer instruments.

In all the studies that compared informant reports to direct cognitive tests, informant reports were shown to be more effective than cognitive assessments [40,42,48]. The Daily Living Skills Questionnaire (DLSQ [67]) was noted to be effective in early detection, showing changes indicative of dementia 3–4 years before diagnosis. The Dementia Screening Questionnaire for Individuals with Intellectual Disabilities (DSQIID [60]) was administered to 848 participants across numerous reviewed studies and each found the questionnaire to be informative. However, the study by Lin et al. [55,65] did note that other demographic factors that

influence dementia status do need to be considered alongside DSQIID administration.

Results on the Activities of Daily Living Questionnaire (ADL [54]) were better explained by disability level and comorbidity than dementia status, and therefore, this was the only informant report reviewed to be shown to be unsuitable for use in dementia diagnostics for people with ID.

4.4. Test batteries

Test batteries reviewed contained a variety of instruments including both direct cognitive tests and informant reports. All batteries reviewed were effective in discriminating between ID dementia cases and ID controls, and none described floor effects, indicating promise for clinical utility. However, Jozsvai et al. [104] found the Boston Naming Task (BNT [78]) and the Block Design Test (BD from WISC-R [93]) contained in their test battery to be most affected by aging. Thus, these two tests were shown to have least diagnostic utility out of the battery. So, if a practitioner was to select this test battery, it is advised that these tests be removed.

The Cognitive Computerized Test Battery for Individual's with Intellectual Disabilities (CCIID [87]) is yet to be studied for the purpose of dementia diagnostics in individuals with ID. However, the CCIID has been validated in adults with ID. Moving forward, this battery should be assessed in a demented ID sample before clinical utility. Similarly, the Das Naglieri Cognitive Assessment System is yet to be assessed comparing ID dementia cases to ID controls. Das et al. [36] assessed cognitive decline as a result of aging that occurs among adults with DS with this test battery and showed the battery to be effective at detecting age-related cognitive decline. However, research has not yet assessed its utility in discriminating between dementia cases and controls in an ID or DS sample. Therefore, further research would need to be carried out to determine this battery's usefulness in aiding with dementia diagnosis.

Test batteries assess a range of cognitive abilities without relying on informants. Consequently, to best inform dementia diagnostics administering a test battery longitudinally can highlight any decline and track cognitive functioning in the years before dementia onset to best aid a clinician in making a diagnostic decision. Test batteries, however, do have numerous practical implications that need to be considered. Many require touch screen laptops, which are costly if the technology is not already available to the clinician. The laptops would also need to be near an available plug socket to administer tests without interruption, which might not be practical in an applied setting, which limits their potential utility. Paper and pen forms of certain cognitive tests are available, so if it is not feasible to have technology then the same concept of assessing a range of cognitive functions can be applied.

The direct tests and informant reports recommended above and described in the tables can help in deciding which tests to administer. However, comparing dementia cases to controls before clinical utility is advised where limited evidence is available on use for people with ID, which is often the case for numerous instruments presented throughout this review. Likewise, it must be noted that all test batteries presented require further testing to validate their clinical utility in an appropriate sample, particularly those with concerns noted above.

4.5. Combining methods

Previous reviews argue that a combination of methods can best inform dementia diagnosis in individuals with ID (e.g., Burt et al. [120]). Johansson et al. [106] describe how cognitive testing and informant interviewing could be the most effective way to combine methods and gain a full clinical picture. Combining methods for diagnosis, although effective, may be time consuming, and therefore the combination of methods chosen need to be carefully considered. This further supports the recommendation of the use of a test battery to aid diagnosis, as a number of batteries presented contain informant reports as well as cognitive assessments.

4.6. Limitations of this review

This review has some limitations. Most notably, instruments that compiled the test batteries were not evaluated individually as direct cognitive tests. To improve this research, instruments used within the batteries could be assessed individually as well as part of the battery. However, owing to the benefits highlighted in this review of test batteries, it was felt that test batteries would be of more benefit to clinicians to be presented as a whole.

4.7. Conclusion

In summary, it can be recommended that when diagnosing dementia in individuals with ID test, batteries can offer the most informative assessment of cognition. This could be alongside informant reports or a battery that contains informant reports to provide valuable information on the daily functioning of the individual as well as an overall assessment of cognitive functioning. Tables provided highlight previous validation of test batteries, and before selecting a battery clinicians should review literature presented. Particularly considering the length of the test battery, the level of ID of the individual being assessed and the setting in which the instrument will be administered. It may be advised to complete a shorter instrument when the ID is more severe. In this case, the CCIID or the SIB each takes 30 minutes or less to administer. Nonetheless, breaks should be offered to participants throughout testing, and it is always possible to split testing sessions into multiple shorter sessions.

Completing a test battery that covers both informant reports of daily functioning and assesses a full range of cognitive abilities is advised. This can enable clinicians to gain a more in-depth account of a participant's functioning and symptoms, hence can best inform a decision regarding dementia diagnosis.

RESEARCH IN CONTEXT

1. Systematic review: A systematic literature search was conducted in four databases (PubMed, Science Direct, Google Scholar, and PsycInfo). Studies were identified and selected using predefined inclusion and exclusion criteria; coded and classified according to assessment type by two independent researchers, with a third consulted when discrepancies arose.
2. Interpretation: The review collates diagnostic instruments and presents strengths and weaknesses of each type of assessment and each individual test. Findings indicated that test batteries to offer the most practical and efficient method of assessment. The variation in areas of functioning affected by dementia in individuals' with ID can be assessed with a battery to best inform a clinician's decision.
3. Future directions: The review proposes specific test batteries that could be most beneficial; however, small sample sizes in cited studies suggest that further studies need to investigate the use of these batteries in larger samples of individuals with ID to better highlight diagnostic potential for each test.

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