Assessment of Cognition in People with Intellectual Disabilities Using a Novel Set of Neuropsychological Tests

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Abstract

Background*:* Improving the diagnosis of dementia in people with an intellectual disability is vital for early intervention and effective treatment and improving the quality of life for individuals, their families, or carers. Dementia is a risk factor for people with an intellectual disability, and neuropsychological assessment is a valuable diagnosis component. However, there has been difficulty in establishing tests for use with this population. A literature review was undertaken to gather research on neuropsychological tests for assessing cognitive impairment in adults with an intellectual disability. This was followed by an empirical study to pilot a novel test set for assessing dementia in people with an intellectual disability and derive preliminary data. ***Methods****:* This exploratory acceptability and feasibility study adopted a cross-sectional design to develop an understanding of the usefulness and appropriateness of the novel measure by piloting it with a sample of seven participants with an intellectual disability. Quantitative data is reported using the test scores, and qualitative data is reported using verbal feedback on the participants' experience completing the measure. ***Results****:* A total of 36 studies were included in the literature review, reporting on 81 directly administered single-domain instruments used in testing of cognitive deterioration in people with an intellectual disability. The majority of the samples included people with Down syndrome. The results were variable, with some instruments developed specifically for this population. The novel test set proved acceptable and feasible for people with an intellectual disability but requires some modifications. Particular challenges were found for the tests of executive function, confirming the literature review findings. ***Conclusions****:* The results will inform modification of the measure for future piloting with larger and different samples of people with an intellectual disability.

Keywords: intellectual disability, learning disability, dementia, cognitive decline, neuropsychology, cognitive assessment, cognitive screening

CHAPTER 1: INTRODUCTION

1.1. Overview

This research project aimed to review what is currently known about dementia in people with an intellectual disability and pilot a novel cognitive test set with a group of participants recruited through London National Health Service (NHS) learning disability services.

In this chapter, I define the terminology used throughout this thesis and give an overview of cognitive testing, intellectual disability, dementia, dementia in intellectual disabilities and the factors that make assessing dementia problematic in this population. I then provide a literature review exploring what single-domain cognitive tests and tasks have been used to contribute to the assessment of dementia in people with an intellectual disability and their utility. I then identify the gaps this thesis seeks to address through the development of the novel test set designed for this population. Finally, I set out the rationale and aims of the major research project before moving on to the methods, results, and discussion chapters.

1.2. Terminology

The term ‘intellectual disability’ will be used throughout this thesis, as this is the most widely used term in the international and academic communities (Schalock et al., 2007). The term ‘learning disability’ was used in the participant recruitment materials as a less stigmatising term. The term ‘intellectual disability’ is used to avoid confusion since ‘learning disability’ and ‘learning difficulty’ are often used internationally to mean specific difficulties, such as dyslexia and dyscalculia. The term ‘people with an intellectual disability’ is used to promote person-first language, and abbreviations of ‘LD’ or ‘ID’ have been avoided (except for where brevity is required in tables). Person-first language is preferred by people with intellectual disabilities and recommended by The Journal of Applied Research in Intellectual Disabilities (The JARID, n.d.). The terms ‘typically developing’ or ‘typical development’ have been used to refer to people without an intellectual disability.

It is essential to acknowledge that the terminology has changed over time, and current use varies between countries. Outdated terms are only included within the search string of the literature review to include historical research within the specified date range. These terms are not used elsewhere in this thesis and are considered discriminatory.

1.3. Cognitive Assessment

1.3.1. Definitions and Purposes

Cognitive assessment is a “performance-based method to assess cognitive functioning” (Harvey, 2019, p. 91). The neuropsychologist can flexibly combine single-domain measures in a hypothesis-testing approach to assessment. The results identify a person’s pattern of cognitive strengths and difficulties (profile). Researchers design neuropsychological tests for specific populations and provide normative data for each population they are intended for so clinicians can reliably compare an individual to the average score within (a sample of) that population.

The test must be testing what it claims to test (validity) and be shown to do so reliably. There should be no floor or ceiling effects (indicating a test is too hard or too easy, respectively) for that population. There are implications for the cultural validity of most neuropsychological tests, given that they are predominantly developed and tested with (educated) populations in North America and Europe and may not be in a person’s primary language.

Cognitive assessment can be used in various circumstances, including intelligent quotient (IQ) testing and diagnosis of intellectual disability, to inform an educational plan, mapping a person’s profile following an acquired brain injury and to inform a neurorehabilitation plan, or the differential diagnosis of cognitive impairment and dementia.

1.3.2. Cognitive Functions and Assessment

Cognitive functions occur hierarchically, with basic sensory processes and attention being the least complex, and reasoning and problem-solving being the most complex (Stuss & Benson, 1984). Although these cognitive domains and functions are generally associated with specific brain regions, contemporary neuroscience suggests functions arise from distributed neural networks of processes involving multiple brain regions. Cognitive tests often cover a range of domains, outlined below.

Sensorimotor

Sensorimotor abilities refer to the process of receiving information through our senses (e.g., vision, hearing, proprioception) and the motor (movement or verbal) response (Li & Lindenberger, 2002). It is associated with integrating the sensory and motor areas and overlaps with verbal and visuospatial skills. Sensorimotor examination can include motor and tactile skills, basic visual processes, olfaction, and auditory skills.

Attention

There is some debate on conceptualising attention, but generally, it refers to the processes involved in receiving stimuli and processing information. It is difficult to locate this domain in a specific brain region and to isolate it from other cognitive domains during testing, given its involvement in all other functions (see above). Forms of attention generally recognised include sustained attention (the ability to concentrate for an extended period) and selective attention (to direct our attention to a particular item, suppressing competing distractions; Lezak et al., 2012).

Executive Function

Executive functions refer to a range of higher-order cognitive processes involved in planning, self-monitoring, and purposive action (Lezak et al., 2012) that guide, direct and manage cognitive, emotional, and behavioural functioning. There is no agreed model of executive functioning.

However, there is a well-accepted three-factor structure of executive function that includes updating (working memory and monitoring), shifting (self-generative behaviour and set-shifting), and behavioural inhibition (Collette et al., 2005; Miyake et al., 2000). Working memory is the ability to hold something in mind briefly in short-term memory whilst manipulating it somehow (e.g., mathematical, or sequencing operations). The brain region mainly associated with executive functioning is the frontal lobes. Since executive functioning is not a unitary component, multiple tests are required to assess the full range of functions.

Learning and Memory

Learning and memory functions concern the processes of encoding, storing, and retrieving information (Lezak et al., 2012). Encoding means processing information to be stored in memory, which is strengthened through rehearsal. Memory processes generally involve the temporal lobes, though other areas will also be involved, depending on the nature of the sensory information encoded (Dickerson & Eichenbaum, 2010). Memory is often divided into verbal and non-verbal memory: short-term memory (immediate recall) and long-term memory (delayed recall). Memory is also categorised as explicit (conscious recall of facts) and implicit memory (learned procedures needed to complete tasks). Episodic memory (personally experienced events) can be separated into anterograde (newly encountered information) and retrograde (past events). Instruments often used to assess a person's ability to learn new information, recall that information after a delay and recognise material presented along with distractors are usually divided into visual and verbal tests.

Visuospatial Functions

Visuospatial abilities are the cognitive processes of discriminating and recognising objects (form, colour, distinction) and spatial relationships between objects and the environment. Visuospatial abilities are related to the brain's primary visual cortex and the ventral ('what') and dorsal ('where') systems (Mishkin et al., 1983). The ventral system extends to the temporal lobe, and the dorsal stream extends to the parietal lobe.

Construction and praxis are voluntary movements, and so have a motor component. Due to the necessary organisation skills, they may also have an executive function component (Harvey, 2019). Typical tests of visuospatial skills include construction-style tasks, visuospatial perception tasks, and drawing tasks.

Verbal Functions

Verbal functions involve understanding and producing auditory information, speech, and written language (Mesulam et al., 2019). Semantic memory refers to general knowledge and word meaning. Typical language tests include naming tests or verbal comprehension tests. Typically, tests of verbal reasoning (and non-verbal reasoning) are included in general intelligence batteries, such as the Wechsler Adult Intelligence Scale - 4th Edition (WAIS-IV; Wechsler, 2012).

Estimating Optimal Ability

Results of an assessment are compared either to 1) a person's previous assessment scores, 2) an estimation of optimal or premorbid ability based on their educational and occupational attainment, or 3) by using a test such as The Test of Premorbid Functioning (TOPF; Holdnack et al., 2013) or British Picture Vocabulary Scale (BPVS; Dunn et al., 1982). The BPVS is designed for children to assess vocabulary, which is often used as a proxy measure of intelligence. The TOPF requires reading a list of words that have atypical grapheme-to-phoneme translations. It, therefore, uses over-learned vocabulary, which is thought to be unaffected by cognitive decline in many presentations.

1.4. Intellectual Disability

The notion of 'intellectual disability' as intrinsic to an individual is problematic since it may be more suitable to view it as a social construction that has been developed and reinforced through language and societal practices over time. It is used to characterise people outside normative societal expectations of self-sufficiency and social responsibilities (Rapley, 2004).

Similarly, the social model of disability states that people are disabled by their environment and society, as opposed to by their impairments. This is evidenced by how intellectual disability is conceptualised differently across countries and cultures, where societal demands on individuals vary. This lack of universality has implications for diagnosing intellectual disability using IQ and adaptive behaviour assessment.

Furthermore, the construct of IQ as a single factor is highly contested. Although evidence suggests IQ correlates with educational attainment, it has not been found to correlate well with everyday functioning (Whitaker, 2013). Further, using IQ as the basis of diagnosis depends on the accuracy of the tools used to measure it, requiring good validity and reliability. Whitaker (2013) suggests that although modern IQ tests do appear to measure intelligence and are valid in the low range, they do not do so reliably, with variability between test instruments, test examiners, and across time.

Focusing on ‘limitations,’ ‘impairments’ or ‘deficits’ continues to problematise individuals and maintains that low IQ needs eliminating or improving, thus pathologizing difference (Goodley, 2001). However, the social model of disability is also criticised for too firmly rejecting the notion of impairments and the medicalisation of low IQ, risking implying that intrinsic impairments are not a problem for individuals (Shakespeare, 2010). It may be preferable to conceive that the interaction of individual bodies and social environments produces disability. Subsequently, many people with intellectual disabilities face additional medical challenges due to underlying physical pathology, such as a higher risk of dementia, which warrant special consideration and assessment to provide equity of care.

1.4.1. Definitions of Intellectual Disability and Diagnostic Criteria

Various diagnostic criteria exist internationally to categorise intellectual disabilities, which include the three central classification systems: The International Statistical Classification of Diseases and Related Health Problems (ICD-11; WHO, 2021), The Diagnostic and Statistical Manual of Mental Disorders (DSM-5; American Psychiatric Association [APA], 2013), and the American Association on Intellectual and Developmental Disabilities (AAIDD-11, 2010).

However, consistent criteria include:

1) A significant impairment of intellectual functioning based on intelligence quotient (IQ) testing.
2) A significant impairment of adaptive behaviour (social functioning).
3) Both of these impairments starting before adulthood.

As mentioned earlier, this criterion is subject to controversy. The British Psychological Society's (The BPS, 2015a) Guidance on the Assessment and Diagnosis of Intellectual Disabilities in Adulthood now acknowledges that diagnosis can harm how people with intellectual disabilities are perceived and treated within society. Therefore, focusing on a person's needs and providing appropriate support to benefit the person's adaptive functioning and quality of life is preferred over IQ as the defining feature. Although the validity of IQ testing in intellectual disabilities is somewhat disputed, the BPS view the use of an IQ assessment as an essential part of the overall assessment of intellectual disability.

Intellectual disabilities can be categorised by their aetiology (e.g., Down syndrome) and severity. Until recently, a four-level classification system (mild, moderate, severe, profound) was used internationally to categorise severity. However, there is a recent shift to using a two-level classification system since instruments become less accurate the further the scores are from the mean. Therefore, an IQ score below 70 (two standard deviations below the mean of 100 in the general population) indicates an intellectual disability, with more than three standard deviations (IQ score below 55) indicating 'severe' impairments (The BPS, 2015). People with a more severe intellectual disability will have more severe difficulties, likely requiring a higher level of support with activities of daily living (WHO, 2021).

1.4.2. Incidence and Prevalence

Based on Public Health England (PHE) and the Office for National Statistics data, Mencap (2013) estimates that around 1.5 million people in the United Kingdom (UK) have an intellectual disability, with a prevalence of approximately 2.16% of the adult population.

Estimated global prevalence rates range from 0.33 – to 2.42% (Nair et al., 2022) between high- and low-socioeconomic status countries. This wide range in prevalence rates is likely due to differences in adequate healthcare across the lifespan, differences in mortality rates, and proactive assessment and diagnosis of intellectual disability between countries. Furthermore, there is a general lack of worldwide data on the prevalence of intellectual disability.

1.4.3. Causes and Subtypes of Intellectual Disability

Most intellectual disabilities occur before birth, including heritable conditions such as Fragile X syndrome or genetic disorders such as Down syndrome. Intellectual disability, regardless of aetiology, has been associated with differences in frontal lobe functioning compared to people with typical development (Ball et al., 2008; Cornish et al., 2009; Mervis & John, 2010). Although IQ has been shown to not necessarily correlate with everyday functioning, a positive correlation has been found between executive functioning and adaptive functioning in individuals with intellectual disabilities (Shapoval et al., 2022) and in people with dementia who have shown typical development (Razani et al., 2007). However, differing cognitive profiles (patterns of strengths and weaknesses) accompany the different conditions, discussed below.

Down Syndrome

Down syndrome is the most common cause of intellectual disability and occurs in about 1 in 1,000 babies born yearly (Weijerman & de Winter, 2010). Down syndrome occurs due to an extra copy of chromosome 21 caused by a random error in cell division (WHO, 2021), hence the alternative name of trisomy 21. In addition to intellectual disability (mainly mild to moderate), people with Down syndrome have distinctive facial characteristics and physical conditions, such as heart malformations and hearing or vision impairments (Bergström et al., 2016). People with Down syndrome can have low muscle tone of the tongue, small mouth, and high palate, resulting in difficulties with pronunciation (Kelly, 2018).

People with Down syndrome also experience an ‘accelerated ageing’ process, whereby conditions usually seen in older adults occur in those with Down syndrome in their 40s and 50s (Horvath et al., 2015; Patterson & Cabelof, 2012; Zigman, 2013). Chromosome 21 is crucial in developing amyloid precursor protein (APP), which is linked to the development of dementia (Hampel et al., 2021).

The cognitive phenotype of Down syndrome includes strengths in visuospatial short-term memory, implicit (unconscious) long-term memory, and associative (stimulus-response) learning (Lott & Dierssen, 2010). Difficulties are evident in expressive and receptive language, verbal working memory, and explicit (conscious) long-term memory (Lott & Dierssen, 2010; Næss et al., 2011; Silverman, 2007). Research suggests this cognitive profile occurs due to a lack of development of typical automatic processing for speech production and perception (Silverman, 2007). People with Down syndrome are thought to have pre-existing abnormalities in the frontal lobes from birth due to hypoplasia, possibly making them susceptible to early decline in executive function during neurodegeneration (Crome & Stern, 1972; Holland et al., 1998; 2000).

Other Aetiologies

Other common aetiologies include Fragile X syndrome, Williams syndrome and Prader-Willi syndrome. All of which have differing cognitive and behavioural phenotypes. For example, a summary by Huddleston et al. (2014) found that the cognitive phenotype for Fragile X syndrome includes strengths in verbal reasoning and simultaneous processing tasks with difficulties in short-term memory, executive function, visual memory, non-verbal reasoning, and visual-motor coordination. This profile can be contrasted with Williams syndrome, where an uneven cognitive profile is often noted, with strengths in language and face recognition tasks but difficulties in visuospatial and arithmetic tasks (Van Herwegen, 2015).

Intellectual disabilities can also arise from pregnancy (e.g., foetal alcohol syndrome) and birth complications (e.g., hypoxic-ischemic encephalopathy), be due to neglect or can be idiopathic (Huang et al., 2016).

People may acquire an intellectual disability due to a severe head injury, stroke, or severe infections, such as meningitis, during childhood. Some are aetiologically undetermined. In the case of an acquired intellectual disability, children tend to show an uneven pattern of cognitive strengths and weaknesses due to having had a period of typical development prior to the injury; the resultant effects depend on the age at the time of injury, severity, and location of injury (Slomine & Locascio, 2009).

1.4.4. Health Inequalities

As mentioned above, people with an intellectual disability are known globally to experience health inequalities (Doherty et al., 2020). They often have more long-term conditions and a shorter life expectancy than people without an intellectual disability. However, healthcare access rates are considerably lower in people with an intellectual disability, giving rise to unmet needs with low diagnosis and treatment rates (Schützwohl et al., 2016). People with an intellectual disability also experience diagnostic overshadowing, which Emerson and Baines (2010) describe as "symptoms of physical ill health that are mistakenly attributed to either a mental health/ behavioural problem or as being inherent in the person's learning disabilities" (Emerson & Baines, 2010, p. 9); meaning treatable illnesses are often missed. Improving the diagnosis of dementia in people with an intellectual disability is vital for early intervention and effective treatment and improving the quality of life for individuals, their families, and carers (Zeilinger et al., 2013).

1.5. Dementia

Dementia is an umbrella term to describe a specific cluster of clinical features that occur as part of a progressive decline in cognitive and functional ability. Several pathological conditions cause dementia, each subtype reflecting a different disease process.

1.5.1. Incidence and Prevalence

Current estimates by the London School of Economics (LSE) suggest that almost 885,000 people are living with dementia in the UK (Wittenberg et al., 2019). Projections suggest this will rise to 1,183,000 by 2040 (Wittenberg et al., 2020).

Estimates suggest that around 57 million people are living with dementia globally, with estimates suggesting this could rise to around 152 million by 2050 (Nichols et al., 2022). Every year, there are around 10 million new cases globally.

1.5.2. Causes and Subtypes of Dementia

The most common subtypes are Alzheimer's disease (AD), vascular dementia (VaD), Lewy body dementia (LBD), and frontotemporal dementia (FTD) variants. Often, there are Alzheimer's tauopathy and vascular features, referred to as 'mixed' dementia (Dening & Sandilyan, 2015). Each dementia subtype shows a different cognitive profile and trajectory, reflecting the differential pathology outlined below. These differences indicate the need for comprehensive test instruments that cover a range of cognitive domains.

Alzheimer's Disease

AD is the most common, affecting around 50-75% of people diagnosed with dementia (Cunningham et al., 2015). It is closely associated with age, with most cases being in those over 65 years of age and prevalence increasing exponentially in those over 90 in the typically developing population. AD is caused by amyloid-beta (Aβ) plaque deposition and neurofibrillary tangles (Braak & Braak, 1991). This build-up affects the brain's structure and function, particularly areas associated with learning and memory. These pathological processes can occur as early as ten to fifteen years before symptoms start and are associated with a progressive decline (Tarawneh & Holtzman, 2012). AD is characterised by prominent progressive amnesia, followed by emerging executive dysfunction, poor semantic fluency, and problems with naming. Word-finding difficulties are early prominent features with limited insight and social indifference. Later, there is a global deterioration with apraxia, agnosia, and executive function deficits.

Vascular Dementia

This profile can be contrasted with vascular dementia, which affects around 15-20% of those with dementia (Lobo et al., 2000) and is less strictly associated with age.

It is related to a restricted blood supply to the brain due to cerebrovascular disease (CVD) and ischemic or haemorrhagic brain injury (i.e., stroke; Wolters & Ikram, 2019). Cognitive functioning either abruptly deteriorates following a cerebrovascular event, or the course is fluctuating or stepwise, leading to a cumulative cognitive decline (Verdelho et al., 2021). VaD is characterised by a 'patchy' cognitive profile, depending on the location of the pathology, with a general decline in information processing speed, attention, and executive functioning, whilst memory is less affected (WHO, 2021). People may have difficulties with initiation and cognitive flexibility, low and fluctuating mood, or pseudobulbar palsy, amongst other effects (Verdelho et al., 2021).

Lewy Body Dementia

LBD occurs in around 10-15% of people with dementia (McKeith et al., 2017) and is associated with a build-up of Lewy bodies in the brain, which causes symptoms related to those seen in Parkinson's disease. LBD is characterised by fluctuations in attention and alertness and visual hallucinations that are present early in the disease course. AD and LBD pathology modifies the clinical presentation and accelerates cognitive decline (Malek-Ahmadi et al., 2019). In the early stages, memory is less affected due to the relative preservation of hippocampal and medial temporal lobes (Elder et al., 2017). Due to this relative memory sparing, early MCI diagnosis is often missed on assessment (McKeith et al., 2017). More prominent are deficits in visuoperceptual, visuospatial and visuo-constructive abilities, poor attention, psychomotor speed, and executive function (Ferman et al., 2004; Guidi et al., 2006; Stavitsky et al., 2006).

Frontotemporal Dementias

FTD is an umbrella term used to describe several rare dementias affecting the frontal lobes, including the most common behavioural variant (bvFTD), as well as the primary progressive aphasias, including semantic dementia and progressive non-fluent aphasia (Pickering-Brown, 2007). It is caused by changes at a cellular level with white matter tract pathology (Mahoney et al., 2014; Tovar-Moll et al., 2014).

It is more common in people under sixty-five, and the familial variant has a strong genetic association (Lok & Kwok, 2021). In bvFTD, pathological examination shows atrophy of the frontal and anterior temporal lobes and the striatum with changes in behaviour (e.g., apathy, disinhibition, perseveration, lack of insight, decreased speech output) and a prominent dysexecutive syndrome (Snowden et al., 2002). Visuospatial abilities, memory, language, and motor function are relatively spared (Boxer & Miller, 2005; Grossman, 2002; Neary et al., 2005).

1.5.3. Diagnostic Criteria

The ICD-11 (WHO, 2021) defines dementia as a 'marked' impairment in *two* or more cognitive domains, representing a decline from their usual level of functioning and that expected of their age. The DSM-5 (APA, 2013) has renamed dementia 'neurocognitive disorders' (major and minor). The DSM-5 defines major neurocognitive disorder as a 'significant' decline from their previous level of functioning in *one* or more cognitive domains. It requires reporting from either the person themselves or an informant and significant impairment on neuropsychological testing. The cognitive deficits must interfere with the independence of activities of daily living and not be better accounted for by another mental disorder.

Dementia is classified by level of severity: mild, moderate, and severe, which is typically based on functional ability and level of independence (WHO, 2021). The National Institute for Health and Care Excellence (NICE, 2018) states that if the level of cognitive impairment does not meet the clinical threshold for dementia, a person may be diagnosed as having mild cognitive impairment (MCI), otherwise called 'mild neurocognitive disorder' in ICD-11 (WHO, 2021) or 'minor neurocognitive disorder' in DSM-V (APA, 2013). 5–15% of people with MCI develop dementia each year. However, around 50% remain stable at five years; for some, their symptoms may improve over time.

1.5.4. Diagnostic Processes

NICE guidance suggests that diagnosis should utilise a combination of clinical assessment, neurological examination, cognitive screening, and blood tests to rule out reversible causes (NICE, 2018). Brief cognitive screening measures include the Mini-Mental State Examination (MMSE; Folstein et al., 1975) or the Addenbrooke's Cognitive Examination (ACE; Mathuranath et al., 2000). Clinicians should take a history from the person and an informant, such as a family member. This history can include an informant-rated questionnaire, such as the Informant Questionnaire on Cognitive Decline in the Elderly (IQCODE; Jorm & Jacomb, 1989).

If dementia is suspected, the person may be referred to a specialist memory clinic. Validated criteria such as the National Institute of Neurological and Communicative Diseases and Stroke-Alzheimer's Disease and Related Disorders Association (NINCDS-ADRDA; McKhann et al., 1984) or international consensus criteria for dementia with Lewy bodies (McKeith et al., 2017) may be used for differential diagnosis. Structural imaging may be requested to confirm the diagnosis and rule out reversible causes. Neuropathological confirmation is considered by many to be the 'gold standard' for dementia diagnosis (Scheltens & Rockwood, 2011).

1.5.5. Cognitive Assessment of Dementia

A more comprehensive neuropsychological assessment is often sought to confirm a diagnosis or provide a differential diagnosis, with a decline of two standard deviations below the person's cognitive baseline indicating a decline sufficient to reach a diagnosis of dementia (Hugo & Ganguli, 2014). Various cognitive tests can be used as part of a dementia assessment. These should cover a range of cognitive domains according to the cognitive and behavioural profiles of the different dementias described earlier. These domains include orientation, attention, learning and memory (short and long-term), language, executive function, and praxis (National Collaborating Centre for Mental Health, 2007).

Some neuropsychological batteries are explicitly designed to address all these key domains and to be used stand-alone for assessing cognitive decline and dementia, such as the Cambridge Cognition Examination (CAMCOG; Roth et al., 1986) and the Addenbrooke's Cognitive Examination III (ACE-III; Mathuranath et al., 2000).

1.6. Dementia in People with an Intellectual Disability

1.6.1. Causes and Subtypes

As mentioned earlier, people with an intellectual disability have a higher risk of developing dementia compared to those without an intellectual disability (22% *versus* 3-7% of people aged over 65 years; The BPS, 2015b). People with Down syndrome have a particularly significant risk (three times greater) of developing AD compared to the typically developing population (Hill et al., 2003). This increased risk is likely due to people with Down syndrome having three copies of the APP gene, resulting in extensive amyloid-beta (Aβ) plaque deposition by age 35 (Zigman et al., 2008). A review by Head et al. (2012) found that prevalence rates in people with Down syndrome aged 40-49 years varied between 6-55%, 4-55% between ages 50-59 years, and 15-77% over the age of 60.

For people with an intellectual disability from other causes, the picture may be variable. However, there is limited research regarding dementias in intellectual disabilities due to other causes and across the range of severity, which makes the link between dementia and intellectual disability unclear, likely due to greater heterogeneity in presentations and aetiologies (The BPS, 2015b).

1.6.2. Presentation of Dementia in People with an Intellectual Disability

The differing baseline cortical pathology and cognitive profiles outlined earlier mean that dementia may present and progress differently across the intellectual disability aetiologies (Strydom et al., 2010). A review by Lautarescu et al. (2017) found that early signs of AD in Down syndrome were more likely to be executive dysfunction and behavioural changes rather than memory loss.

This finding may reflect the vulnerability of people with Down syndrome to both AD and frontal pathology, as outlined previously. Whereas other studies report memory decline as an early sign of dementia (Esteba-Castillo et al., 2022; García-Alba et al., 2019; Krinsky-McHale et al., 2002; Oliver et al., 1998; Sano et al., 2005). The heterogeneity in these findings may reflect differences in the use of diagnostic criteria to classify dementia status, the confounding effects of severity of intellectual disability and increasing age but also the type of cognitive tests used. For example, criterion-based tests with a cut-off score are likely to be less sensitive than normative tests used longitudinally.

There is evidence that a decline in olfactory ability may be a precursor to MCI or AD in people who have shown typical development (Murphy et al., 1990; Wilson et al., 2007). This decline may be earlier and greater during normal ageing in people with Down syndrome (Manan & Yahya, 2021) with impaired odour detection, identification, and recognition memory (Murphy & Jinich, 1996). People with Down syndrome also score more poorly when compared to age and IQ-matched participants with an intellectual disability from other causes (Nijjar & Murphy, 2002). However, it is still unclear if this is associated with pathological neurodegeneration. Despite these findings, there are no alternative diagnostic criteria for detecting dementia in people with an intellectual disability.

1.6.3. Factors Impeding Cognitive Assessment

As mentioned previously, there is pronounced variability within the intellectual disability population and across the levels of severity, which hinders the accurate assessment of cognitive decline. In addition, various other factors impact assessment, discussed below.

Normative Data

This pronounced variability across aetiologies means it is difficult to provide a normative sample (Moran et al., 2013). For example, in people with autism, low IQ scores are not necessarily associated with low IQ but with differences in information processing.

These differences have widespread effects on the overall functioning of the individual (Anderson, 2008; Scheuffgen et al., 2000). Therefore, using a single area of functioning to define dementia (e.g., memory or adaptive behaviour), or information collected at only one time point, is unsuitable for people with intellectual disabilities (Aylward et al., 1997; Burt et al., 1998).

Floor and Ceiling Effects

In addition, this variability means that floor and ceiling effects are common (Lautarescu et al., 2017), especially if designed for the typically developing population (assuming the premorbid functioning level to be in the average range), making them insensitive to detecting decline in people with an intellectual disability.

Communication Difficulties

Difficulties with verbal expression and comprehension are common in this population. People may need help forming sentences, understanding crucial and abstract concepts, and extra time to process and retrieve information (Hassiotis et al., 2012). Therefore, they cannot respond effectively to complex verbal materials that depend on speech and language skills used in most test batteries. This mainly affects those with a more severe level of intellectual disability (Dalton & Wisniewski, 1990) but risks over-diagnosing dementia in this population.

Difficulties Estimating Premorbid Ability

Using a proxy measurement of premorbid functioning, such as the British Picture Vocabulary Scale (BPVS; Dunn et al., 1982), in intellectual disability may not be suitable because they will have a lower premorbid level of vocabulary than people who have shown typical development. Furthermore, people with an intellectual disability often will not have informants who know the person's history or have experienced poor record-keeping for those in care homes or institutions. Therefore, deterioration in functioning may not have been monitored or described accurately (Holland et al., 2000). Using a current estimate derived from a reading test, such as the Wechsler Test of Adult Reading (WTAR; Wechsler, 2001) or the TOPF (Holdnack et al., 2013) will be inappropriate for a person with low literacy.

Co-morbidities

The presence of conditions such as depression and thyroid problems that often co-occur in people with an intellectual disability compound this situation further because they can mimic symptoms of dementia (Oliver, 1999). Again, this risks over-diagnosing dementia in this population. Also, epilepsy, sensory deficits, and medication use, including antipsychotics often used to manage behaviour that challenges others (The BPS, 2015b), may influence the presentation and trajectory of dementia.

Difficulties in Executive Functioning

Given that dysexecutive symptoms are likely to be early signs of dementia in people with an intellectual disability, it may be essential to prioritise the assessment of these to aid earlier detection (Rowe et al., 2006). However, there has yet to be an explicit agreement on which tests should be adopted (The BPS, 2015b). Executive test batteries developed for people who have shown typical development tend to be too difficult for many people with an intellectual disability. Even if a person with an intellectual disability can complete the tests, the normative samples do not include people with an IQ < 70, meaning the interpretation of results can be difficult.

Furthermore, not all people with an IQ < 70 will be known to services, as they will not meet diagnostic criteria for an intellectual disability unless they also have difficulties in adaptive functioning. Given that executive functions are associated with adaptive behaviour, Willner et al. (2010) suggest a person with a *diagnosis* of an intellectual disability will likely struggle more on tests of executive function than others with an IQ < 70 not known to services. This difference is important to consider regarding the choice of instrument and interpretation of results.

Therefore, several batteries have been developed to assess executive function in intellectual disabilities. These include the Behavioural Assessment of Dysexecutive Functioning – Intellectual Disabilities Adaptation (BADS-ID; Webb et al., 2020), an adaptation of the BADS (Wilson et al., 1996), and the Cambridge Executive Functioning Assessment (CEFA; Sandberg, 2011).

1.6.4. Assessment of Dementia in People with an Intellectual Disability

A diagnosis of dementia requires a significant decline in cognition from baseline functioning. However, as I have shown, distinguishing the subtle, insidious, and progressive signs of dementia from pre-existing divergences in behaviour and verbal and communication skills is challenging. Furthermore, the screening tools developed for the typically developing population are unsuitable.

Current UK guidelines by the National Institute for Health and Care Excellence (NICE, 2018) suggest complementing an assessment of dementia in intellectual disability with:

- A measure of symptoms, such as the Dementia Questionnaire for People with Learning Disabilities (DLD; Evenhuis, 2018), the Down Syndrome Dementia Scale (DSDS; Gedye, 1995), or the Dementia Screening Questionnaire for Individuals with Intellectual Disabilities (DSQID; Deb et al., 2007).
- A measure of cognitive functioning to monitor changes over time, such as the Test for Severe Impairment (TSI; Albert & Cohen, 1992).
- Measures of adaptive functioning, e.g., Adaptive Behavior Assessment System, Third Edition (ABAS-III; Harrison & Oakland, 2018).

However, there is still no agreed-up ‘gold standard’ approach to assessment. A systematic review by Zeilinger et al. (2013), aimed at identifying existing instruments used to assess dementia in people with an intellectual disability, found inconsistency in the assessment methods and instruments. They identified 79 directly administered instruments, 35 informant-report instruments, and four batteries. They noted that some of the instruments were not designed for use with people with an Intellectual disability or to detect dementia. A further review by Paiva et al. (2020), focusing on batteries and scales to inform clinical practice, found 39 instruments and 13 batteries for assessing cognitive and behavioural changes in people with an intellectual disability, with no instrument used in more than one study. Paiva et al. (2020) found that most studies focused on people with Down syndrome and lacked descriptive data to inform the quality assessment but recommended the CAMCOG-DS and DLD.

However, given the need for more consistency across studies, both reviews noted a continued need for a unified, standardised instrument to allow results from research to be combined and to provide greater consistency of diagnosis in clinical practice.

1.6.5. Recommended Cognitive Tests and Batteries in the UK

In terms of directly administered cognitive instruments, The BPS (2015b) recommends using an instrument validated for the assessment of dementia in people with an intellectual disability. They state it should include assessment of new learning and prospective memory, short- and long-term memory (visual and verbal), executive functioning, orientation, language expression and comprehension, and any additional tests needed (e.g., praxis). They also note that many services have developed their own battery but list the most used tests in services in the UK as follows:

1. The Neuropsychological Assessment of Dementia in Individuals with Intellectual Disabilities (NAID; Crayton et al., 1998).
2. Cambridge Cognitive Examination (CAMCOG; Roth et al., 1986) and Cambridge Cognitive Examination adapted for individuals with Down Syndrome (CAMCOG-DS; Ball et al., 2004).
3. Severe Impairment Battery (SIB; Saxton & Swihart, 1989).
4. Test for Severe Impairment (TSI; Albert & Cohen, 1992).

Neuropsychological Assessment of Dementia in Intellectual Disabilities (NAID)

The NAID is a battery used for intellectual disability, previously called the Crayton and Oliver Dementia Battery (CODB). It includes subtests from the CAMCOG and CANTAB (Sahakian et al., 1988). The instructions are in Crayton et al. (1998), which take around 45 minutes to administer. It includes seven subscales: picture and object naming and identification (language), action on request (praxis), object and picture memory (working memory), and memory for sentences (verbal short-term memory). It relies heavily on language and visual perception, and it lacks a non-verbal measure of executive function, and measures of abstraction and concept formation (e.g., a matrix reasoning task). The NAID-R (Gleave et al., 2023) has recently been released with adaptations to the administration and scoring.

The Paiva et al. (2020) review did not report the NAID. However, it was shown to have good split-half reliability (Spearman-Brown formula, 0.74-0.95) and internal consistency (Cronbach's alpha, 0.82-0.96) in a study by Oliver et al. (2021). However, several studies have noted floor and ceiling effects (Adams & Oliver, 2010; Ball et al., 2008; Ball et al., 2010; Bevins & Hurse, 2014; Carr & Collins, 2018; Crayton et al., 1998; Oliver et al., 1998; Oliver et al., 2021; Sinai et al., 2016). These findings suggest that assessment for potential cognitive deterioration at a single time point is possible for some adults with Down syndrome using the NAID but not those with a more severe intellectual disability or advanced dementia.

Cambridge Cognitive Examination (CAMCOG) and the Cambridge Cognitive Examination Adapted for Individuals with Down Syndrome (CAMCOG-DS)

The CAMCOG is the neuropsychological assessment component of the CAMDEX designed to assess for dementia in the typically developing population. The CAMCOG-DS is the adapted version for use with people with Down syndrome. It was based on the original CAMCOG and the SIB. The CAMCOG-DS includes tests of orientation, language (e.g., performing actions on request, picture naming, verbal fluency – naming animals), memory (e.g., recall and recognition of pictures, remote and recent information retrieval), attention (e.g., counting to 20, digit span), praxis (e.g., “Show me how you would…”, clock drawing, picture copying), abstract thinking (similarities), and perception (naming of people, and naming pictures from unusual angles). It provides individual test scores and a total score and appears to rely heavily on verbal ability and general knowledge, which is culturally situated. Beresford-Webb and Zaman (2021) have recently released the CAMCOG-DS-II as part of the CAMDEX-DS-II, emphasising establishing change from the individual’s baseline functioning.

It has been shown to have good inter-rater reliability for most items (Kappa >0.8 for 91% of items and >0.6 for all items) and to be a good predictor of a future diagnosis of dementia in people with an intellectual disability (Ball et al., 2004).

Furthermore, it was shown to correlate highly (Spearman rank correlation of 0.96) with the MMSE (Folstein et al., 1975) in a study by Hon et al. (1999), even when shared items were removed. McPaul et al. (2017) found the recall subtest to show moderate internal consistency (Cronbach's alpha coefficient of 0.72), but this was lower for the recognition subtest (Cronbach's alpha coefficient of 0.56). However, several studies report floor effects (Ball et al., 2004; Hon et al., 1999; McPaul et al., 2017), so it may not be an adequate test set for those with severe intellectual disability or advanced dementia. In addition, Paiva et al. (2020) note that it may have limited diagnostic value when used at a single time-point due to it being a criterion-based test. Therefore, it has limited sensitivity to change over time.

Severe Impairment Battery (SIB)

The SIB assesses cognitive functions in people with severe dementia (age range 51–91). It includes forty items, and administration takes approximately 20-30 minutes. It comprises simple commands that are presented along with gestural cues (e.g., "Please write your name here"). It is divided into six subscales appropriate for the range expected of a severely impaired individual: attention and orientation, language, learning and memory, visuospatial ability, and construction. There are also short evaluations of social interaction and praxis. Cut-offs for severe impairment are provided. A short version was later developed by Saxton et al. (2005) that takes around 10-15 minutes to administer and is better suited to people with more severe cognitive impairment. Furthermore, a brief eight-item version was also developed that takes around three minutes to administer and has been shown to correspond well with the original SIB (Schmitt et al., 2009, 2013).

Paiva et al. (2020) found it effective in following cognitive decline longitudinally in people with Down syndrome but noted a limited description of its effectiveness. Although the SIB was not created for people with an intellectual disability, it has been shown to have good test-retest reliability (Spearman's rho = 0.89) and moderate criterion validity (Spearman's rho = 0.68, $p < 0.001$, referencing the Vineland ABS) in people with an intellectual disability without dementia (Witts & Elders, 1998).

The battery showed moderate concurrent validity when compared to the DLD (Spearman's Rho = -0.73, p = <0.001) in a group of people with Down syndrome without dementia (Hutchinson & Oakes, 2011). Hutchinson and Oakes (2011) and Witts and Elders (1998) also noted minimal floor effects, but ceiling effects have been found in people with Down syndrome and no dementia. However, Head et al. (2011) found no association between scores and the presence of APP, which calls into question its clinical sensitivity and effectiveness. Therefore, the SIB may help detect cognitive deterioration in people with a more severe intellectual disability if used longitudinally but needs further longitudinal data to confirm this.

Test for Severe Impairment (TSI)

The TSI is a 24-item test developed for use with people with severe dementia. It has subsections on language comprehension and production, memory (immediate and delayed), motor performance, and some non-verbal reasoning. Although it was not designed for use in people with an intellectual disability, Paiva et al. (2020) note that most people with an intellectual disability can score on this test, and only those with advanced dementia cannot. For example, Cosgrave et al. (1998) found it to be reliable in tracking the development of dementia in people with severe intellectual disability. They also noted good convergent validity (r = 0.94) for all samples and interrater reliability (r = 0.97) and test-retest reliability (r = 0.98) over a two-year follow-up period, with good internal consistency (Cronbach's alpha of 0.89). McCarron et al. (2014) found it could detect deterioration one year before a diagnosis of dementia in those with Down syndrome using longitudinal assessment. However, a study by Pyo et al. (2010) found no difference in TSI total score or immediate and delayed memory subtest scores between those with AD and those without in a sample of people with a moderate to severe intellectual disability. Furthermore, Krisnky-McHale et al., (2020) noted floor effects. This indicates it may not be sensitive enough to detect early AD and may still be too difficult for those with a more severe intellectual disability.

1.7. Literature Review

1.7.1. Aims

Given the large number of instruments found in the Paiva et al. (2020) and Zeilinger et al. (2013) reviews and the variable findings for the current recommended measures, there remains a need for a single sensitive 'gold standard' measure, co-normed for use across the range of severity of intellectual disability. This review aims to find all single-domain tests and subtests of batteries used to assess dementia in people with an intellectual disability and assess the utility of the available instruments to inform the development of a novel test set.

The aims of the review are to:

1) Compile a list of cognitive single-domain tests and subtests of batteries that are used to assess cognitive decline in the intellectual disability community.
2) Report the findings (including any available psychometric and acceptability data) to assess test utility and inform the development of a novel measure.
3) Find any novel/ adapted tests (if any) produced since the last review on this topic (i.e., Paiva et al., 2020).

1.7.2. Methods

1.7.2.1. Protocol.

No ethical approval for this review was needed. Although this was not a systematic review, the Preferred Reporting Items for Systematic Reviews and Meta-analyses (PRISMA) Statement (Page et al., 2021) checklist and flowchart (Figure 1) were used as a guideline for reporting the findings. Findings are included as a narrative synthesis under established cognitive domains (e.g., attention, memory). Where possible, cognitive measures were categorised by cognitive domain per the original studies. Some measures were used differently across the studies and were included as such under the relevant domains in Appendix C.

1.7.2.2. Search Strategy.

A review was conducted on the 4th of July 2022 through EBSCO to include the following electronic databases: CINAHL, APA PsycArticles and APA PsycInfo. A search of PubMed was also conducted. References of included studies and relevant reviews were searched manually for additional studies. Search terms were adapted from the previous reviews by Zeilinger et al. (2013) and Paiva et al. (2020). See Table 1 for a summary of the search criteria used. Filters were applied to exclude papers published before 1980 and those published in languages other than English. Papers written before 1980 were not included, as the tests and constructs have since been updated. The titles and abstracts were initially screened for relevance using the eligibility criteria, following which full-text manuscripts were accessed and screened for inclusion against the eligibility criteria. Any duplicates were removed. Whilst attempts were made to include all relevant tests, given the constraints of a doctoral thesis (e.g., a second rater was not available), an exhaustive systematic approach and detailed quality assessment were not undertaken.

Table 1

Search Terms Used

	Population	**Measure**	**Output**	**Output Type**	**Exclusions**
	Adult intellectual disability	Dementia/ cognitive change	Instruments	Cognitive assessment	Non-adult
Synonyms	Intellectual disability; learning disability; mental retardation; mental handicap; mental deficiency; developmental disability	Dementia; Alzheimer's; cognitive impairment	Instrument, assessment; battery; screening; interview; measurement; questionnaire; tool; psychometrics; scale; diagnostic; diagnosis; test	Cognitive; cognition; neuropsychological	Child; adolescent; youth
Search Terms	(Adult* or older adult*) AND (intellectual* disabilit* or mental* retar* or general learn* disabilit*)	AND (dementia or Alzheimer* or cognit* impair* or cognit* decline*)	AND (instrument* OR assess* OR battery OR screen OR screening OR interview* OR measure* OR questionnaire* OR tool* OR psychometr* OR scale* OR questionnaire* OR diagnosti* OR test*)	AND (cognit* or neuropsych*)	NOT (Child* or adolesc* or youth*)

1.7.2.3. Eligibility Criteria.

Studies were included if they met the following criteria:

1) Peer-reviewed cohort (cross-sectional or longitudinal) studies and written in English;
2) Population: Adults aged over 18 years diagnosed with an intellectual disability;
3) Intervention: Single-domain cognitive tests and batteries used to assess for cognitive decline in adults with an intellectual disability;
4) Comparison: Longitudinal comparison, between groups comparison of dementia status, comparison between measures, reporting on psychometric properties of tests;
5) Outcome: Cognitive decline in adults with an intellectual disability.

Studies were excluded if they were:

1) Non-peer-reviewed studies, case studies or case reports, meeting abstracts/ conference presentations, protocols, book chapters, reviews/ commentaries, and unpublished dissertations and theses;
2) Adults without an intellectual disability;
3) Papers not written in English;
4) Included participants under 18 years of age in the sample;
5) No cognitive assessment included or not directly administered cognitive single-domain tests included or not measuring cognitive changes;
6) Reported on batteries but did not provide data by subtest;
7) Used an inappropriate comparator group (e.g., young vs. old groups only).

1.7.2.4. Data Collection Process.

Data were extracted to a pre-defined and piloted Microsoft Excel spreadsheet for all included studies. Studies were classified by type following published criteria (Mann, 2003). For example, ‘cross-sectional’ was used if participants were assessed at a single time point and ‘longitudinal’ if participants were assessed at more than a single time point. Data were extracted regarding relevant outcomes (results of the cognitive assessment and psychometric properties), study characteristics (e.g., follow-up period, tests included), and sample characteristics (e.g., dementia subtype, sample size, age, sex, and ethnicity).

Information on acceptability and tolerability was sought. For example, the number of people who could not complete the tests due to language ability or suggestions that some participants found the tests too demanding or stressful.

Where possible, psychometric data was extracted for reliability (inter-rater, test-retest, internal consistency), validity (content, criterion, construct), diagnostic utility (sensitivity and specificity, sensitivity to change), and appropriate scaling (floor and ceiling effects). Reliability coefficients range from 0.0 (no reliability) to 1.0 (perfect reliability). Following general guidelines (Portney & Watkins, 2015), below .50 was taken to indicate poor reliability, from .50 to .75 to indicate moderate reliability, and over .75 to indicate good reliability. However, over .90 is preferred for diagnostic instruments to ensure a valid interpretation of findings. The level of agreement for inter-rater reliability is otherwise measured using a kappa statistic, with a kappa over 0.75 described as excellent, 0.40 to 0.75 as fair to good, and below 0.40 as poor (Fleiss, 1981). However, these guidelines are somewhat arbitrary and not without criticism. Effect sizes (Cohen's d) were referred to as small (d = 0.2), medium (d = 0.5), and large (d = 0.8) based on guidelines suggested by Cohen (1988).

1.7.3. Results

1.7.3.1. Study Selection.

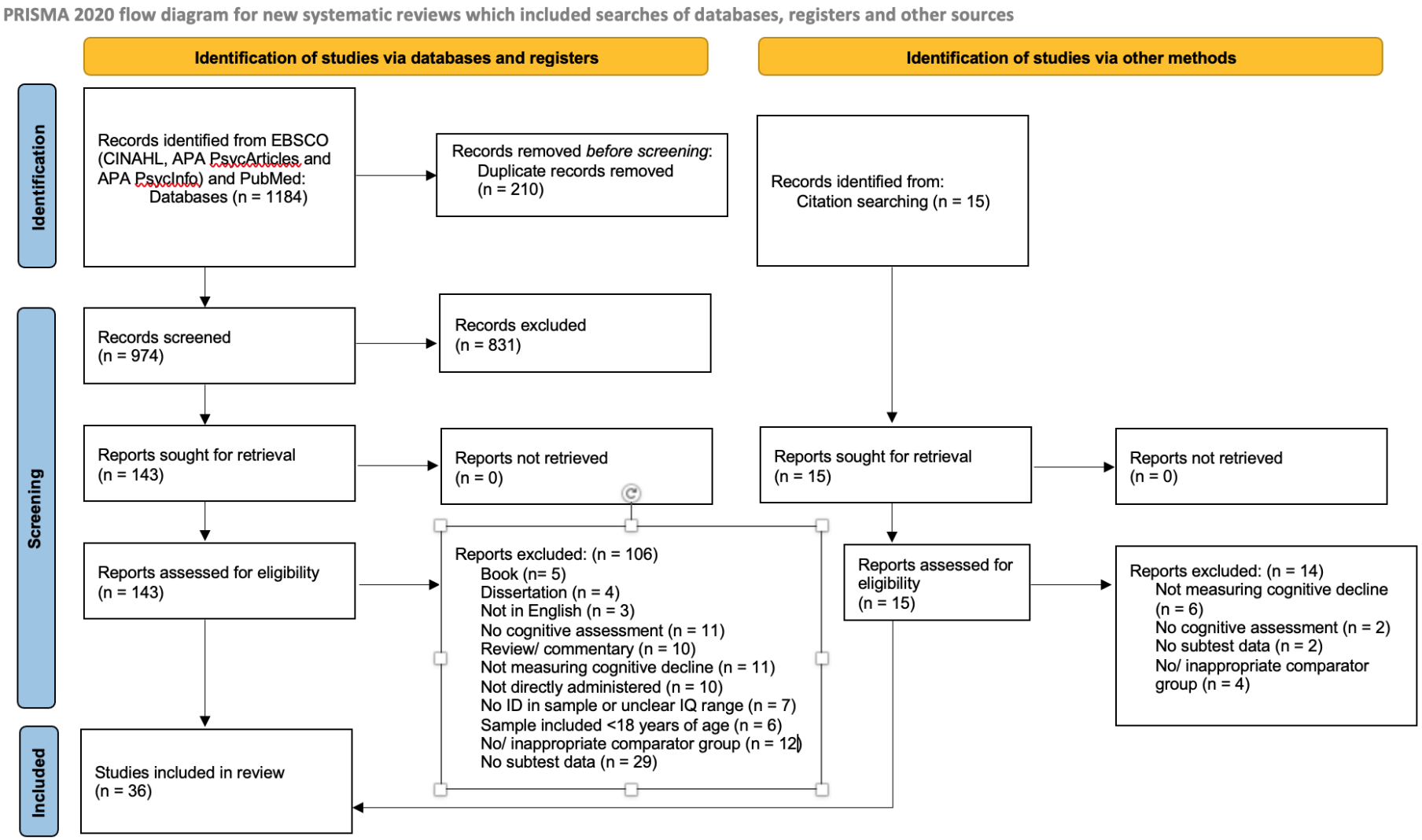
The database search identified a total of 1184 records, including 210 duplicates. A total of 974 papers were screened by the author, with 143 retained for full-text review, and an additional 15 articles were identified from manual reference searching. The author reviewed these 158 papers. Upon full-text review, 120 articles were excluded against the eligibility criteria. The most common reason for exclusion was that no subtest data was available due to battery findings not being reported by subtest. The other most common reasons for exclusion were not measuring cognitive decline (e.g., IQ assessment in younger adults) or no cognitive assessment (e.g., functional assessment). A total of 36 studies were included in the review.

1.7.3.2. PRISMA Flowchart.

This is summarised using a PRISMA flowchart below (Figure 1).

Figure 1

PRISMA Flowchart



From: Page MJ, McKenzie JE, Bossuyt PM, Boutron I, Hoffmann TC, Mulrow CD, et al. The PRISMA 2020 statement: an updated guideline for reporting systematic reviews. BMJ 2021;372:n71. doi: 10.1136/bmj.n71. For more information, visit: http://www.prisma-statement.org/

1.7.3.3. Study Methods.

This review identified 14 longitudinal studies, one Randomised Controlled Trial (RCT) that used longitudinal data, 19 cross-sectional studies, one that provided longitudinal and cross-sectional data, and one retrospective record review. The included studies were published from 1992 to 2022. The sample size across the studies ranged from 20 (Nelson et al., 2005) to 343 (Benejam et al., 2020). Of the longitudinal studies, follow-up ranged from one year (Cooper et al., 2016; Nelson et al., 2001; 2007; Pyo et al., 2011) to 6 years (Devenny et al., 1996). See Appendix B for complete details.

1.7.3.4. Study Characteristics.

All samples included participants with Down syndrome, including 11 studies that also included participants with an intellectual disability from other causes. No studies included a selection of only people with an intellectual disability from other causes. Most studies included a range of severity of intellectual disability, though this was not reported in three studies. The method used to assess the level of intellectual disability varied across the studies, with some using IQ scores from general intelligence tests such as the WAIS-IV and ICD-10 criteria, medical record reviews, and some extrapolating from age-equivalent scores on reading tests, such as the BPVS.

Where a study included participants with dementia or if dementia was diagnosed during the follow-up period, the primary subtype of dementia identified was AD. However, this was not specified in ten studies. Five studies included or solely focused on early-stage dementia (MCI). Most studies used the Cambridge Mental Disorders of the Elderly Examination (CAMDEX) or the Cambridge Examination for Mental Disorders of Older People with Down's Syndrome and Others with Intellectual Disabilities (CAMDEX-DS), DSM or ICD criteria, or a combination of these to diagnose dementia. Dementia diagnostic criteria were not specified in six studies.

The mean duration of dementia was not reported in any study. This omission likely reflects the difficulty in detecting dementia early in people with an intellectual disability. Of those studies that reported it, the overall age of participants ranged from 18 to 91. The overall proportion of female study participants ranged from 41% (Hom et al., 2021) to 70% (Powell et al., 2014), though this varied for subgroup analyses. The ethnicity of the study population was reported in six studies. All were majority White. Verbal ability for inclusion in the study was not reported in most studies. Still, two studies reported excluding participants who were either non-verbal or had severe or profound intellectual disabilities to avoid floor effects (see Appendix B for complete details).

The studies used a wide range of instruments, including a mixture of directly administered and informant-report measures, questionnaires, cognitive tests, or tasks that assess single cognitive domains, or batteries, including multiple subtests assessing a range of cognitive domains. This review focuses only on the results for directly administered single-domain cognitive tests used to assess cognitive decline (including battery subtests where this data is provided). See Appendices C and D for a comprehensive list of instruments used in the studies and their abbreviations. Eighty-one directly administered single-domain tests and tasks were found across all studies. Full details can be found in Appendix C. The findings from the directly administered single-domain subtests and tasks are reported below according to the primary cognitive domain they were developed to assess.

1.7.3.5. Orientation and Arousal.

Six studies included a subtest of orientation, five of which were taken from existing batteries, including the MMSE (not designed for people with an intellectual disability) and the CAMCOG (which uses items from the MMSE), but none were included in more than one study. Orientation tests usually assess a person's orientation to person, place, time, and situation; the higher the score, the greater the person's awareness. Typical questions include "What is your name?" "What year is it?" or "What is the name of this building?".

Most studies noted a decrease in scores associated with cognitive decline but were also affected by the severity of intellectual disability. Oliver et al. (1998) found the CAMCOG orientation subtest to be sensitive to early cognitive decline, whereas Sano et al. (2005) found the MMSE version to have floor effects for those with moderate to severe intellectual disability. Pyo et al. (2009), using an orientation test designed for adults with moderate to severe intellectual disability, found the AD group to score lower than the control group at baseline but showed poor sensitivity and specificity. However, age and intellectual disability aetiology did not significantly affect scores, but there were floor effects for those with a severe intellectual disability. Overall, orientation tests appear to be sensitive measures of decline, but questions need to be suitable for or designed specifically for people with intellectual disabilities to avoid floor effects.

1.7.3.6. Attention and Processing Speed.

Four studies used a specific measure of attention and processing speed. Two studies used a cancellation task developed by Krinsky-McHale et al. (2008) to assess dementia in people with Down syndrome. Two further studies used the Coding subtest of the Wechsler Intelligence Scale for Children-Revised (WISC-R; Wechsler, 1974), designed for children without an intellectual disability. Both of these tests showed promise in detecting dementia in this population.

Cancellation tasks

Cancellation tasks require finding and marking target items (e.g., the letter X) presented on paper, where targets are mixed with several distracters (e.g., a range of other letters). Both studies (Cooper et al., 2016; Krinsky-McHale et al., 2008) provided evidence of the test's utility in identifying dementia. Krinsky-McHale et al. (2008) found that Down syndrome adults showed progressive impairment in selective attention approximately two years before a diagnosis of MCI. Performance varied with the stage of dementia and level of intellectual disability, and it showed reasonably good sensitivity (80%) and specificity (82%) for identifying MCI and was easy to administer.

Coding task

On the Coding subtest of the WISC-R, both studies (Devenny et al., 1996, 2000) reported significant differences between those with and without dementia, including early-stage dementia.

1.7.3.7. Executive Function.

A total of sixteen subtests were found under this domain. Given that executive functioning incorporates multiple functions, it is unsurprising that various tests and types of tests were found under this domain, including subtests from the CEFA and CANTAB batteries, designed for people with intellectual disabilities. The most commonly used tests were verbal fluency (*n* = 8), 'Tower of London' tests (*n* = 5), and Cats and Dogs (*n* = 4) from the CEFA battery.

Verbal fluency tests – verbal fluency, set-shifting, and working memory

Verbal fluency tests usually incorporate phonemic (letter) and semantic (category) tasks. The verbal fluency tests found in the review were the Controlled Oral Word Association Test (COWAT; Lezak et al., 2012), also known as 'FAS', the Category Fluency Test (Benton, 1968), the category fluency task from the CEFA, and the McCarthy Category Fluency Test (M-CFT; McCarthy, 1972).

The category fluency task (animals) from the CEFA (*n* = 3) was the most commonly used version. Sinai et al. (2016) found it helpful in assessing dementia in people with Down syndrome, whilst Ball et al. (2008) reported it to be less sensitive than some of the other subtests in the CEFA. Although Bevins and Hurse (2014) found it not to correlate with other measures known to indicate dementia, they still felt it was helpful because it was independent of verbal comprehension ability with no floor effects.

Brugge et al. (1994) and Palmer (2006) used the COWAT with a small sample of participants. Palmer (2006) found people with mixed aetiology of intellectual disability and dementia to show lower scores compared to the matched control group with no dementia.

However, Brugge et al. (1994) found no significant differences between those with memory impairment and those without in a group of people with Down syndrome.

Hom et al. (2021) and Krinsky-McHale et al. (2020) both used the M-CFT, an adapted version with a more generous scoring system and a shorter timeframe (20-seconds). Whilst Hom et al. (2021) found significantly lower scores in those with Down syndrome and MCI compared to those without MCI, Krinsky-McHale et al. (2020) found a significant decline only with dementia onset and considerable floor effects for those with severe intellectual disability or dementia. Furthermore, Hom et al. (2021) found it did not strongly load onto any factor and was dropped from further analyses.

However, Cooper et al. (2016) found the category fluency task from the CAMCOG-DS accessible and could detect change over time in a relatively small sample. These findings suggest that a verbal fluency task may be sensitive to detecting dementia but maybe less so for early-stage dementia and less useful for those with a severe intellectual disability or dementia. It is unclear from these findings whether a phonemic fluency task may be more helpful since most studies used only a category fluency task. However, phonemic fluency tasks are more challenging and will likely be impacted by verbal ability to a greater degree.

Tower of London – planning, problem-solving, and working memory

The Tower of London task (Shallice, 1982) consists of two boards with pegs and several beads with different colours, requiring different problems to be solved. Several test variants exist, including the London-Drexel University: 2nd Edition (TOL^{DX}; Culbertson & Zillmer, 2005), a version designed for use with intellectual disability.

On the TOL^{DX} version, Sinai et al. (2016) found no significant differences between the dementia and no-dementia groups and significant floor and ceiling effects, especially for those with dementia.

However, they used an older sample, so those without dementia may have also experienced changes in executive function indicative of a pre-clinical phase of dementia. The sample also included a wider range of levels of intellectual disability. Esteba-Castillo et al. (2022) found the 'Hit' factor to be the most discriminating variable between groups, including prodromal AD, which aligns with previous study findings in the typically developing population (Rainville et al., 2012). The 'Hit' factor is a binary variable that denotes the ability to finish an item or not, regardless of the number of movements it takes to do so. García-Alba et al. (2017) reported consistency across levels of intellectual disability (mild to moderate) and no floor effects. However, the results did not indicate that the test items got progressively more difficult.

On the CEFA version, Ball et al. (2008) found the ToL to be more sensitive than some of the other subtests but showed floor effects for those with AD and was affected by older age. Cooper et al. (2016) found it showed less change over time but with no floor effects. Therefore, these results are somewhat mixed but suggest that the ToL test may be a useful test to assess for early signs of dementia in individuals with mild to moderate intellectual disability but may be too difficult to detect deterioration in those with a more severe intellectual disability or advancing dementia.

Cats and Dogs – rule maintenance (working memory) and response inhibition

The Cats and Dogs task is based on the day-night Stroop-like task (Gerstadt et al., 1994) and was developed as part of the CEFA battery (Ball et al., 2008) for use with adults with Down syndrome and AD, discussed later. A sequence of 16 pictures (eight cats and eight dogs) are arranged in a particular order, and the participant is first instructed to name the animal as they see them and then in the inhibition condition, they are instructed to say 'dog' when they see a picture of a cat and vice versa. The score is the time taken to complete the naming condition (congruent condition) subtracted from the time taken stating the opposite animal (incongruent condition). The number of errors on the two conditions is also recorded.

Ball et al. (2008) found only a small effect, suggesting that this test is insensitive to the effects of AD, and Cooper et al. (2016) found it had a poor participant completion rate. In a sample of twenty-eight people with Down syndrome, Bevins and Hurse (2014) found ceiling effects for some participants and a narrow range of scores. However, this may suggest that this task would be suitable for those with a more severe intellectual disability. Bevins and Hurse (2014) found that the Cats and Dogs task negatively correlated with the cognitive DLD score, suggesting that lower Cats and Dogs task scores are linked to cognitive decline, as rated by carers. This finding confirms the ecological validity of the tool and provides further evidence for its usefulness in assessing dementia symptoms. They found no correlation between the Cats and Dogs and verbal comprehension tests, confirming its usefulness as an executive measure in people with limited language ability. However, on factor analysis, Hom et al. (2021) found that the Cats and Dogs task did not load onto any of the factors (including executive function) with a uniqueness of 0.96 (needing eigenvalue of ≥1 to be included in a single factor). The error score loaded on the memory factor, and 28% of the sample had less than 50% accuracy. Therefore, the authors felt this test did not accurately measure executive function and was removed from their analysis.

Overall, the results of this test are mixed. Findings suggest this test may be useful for those with less verbal ability and potentially a more severe intellectual disability. The fact that the error score loaded onto the memory factor in Hom et al. (2021) may reflect the executive function skills of working memory and rule maintenance, which naturally have a memory component.

Other executive function tests

The scrambled boxes task from the CEFA is a modified task initially developed to measure EF with young children. It has been used in animal lesion studies to validate its use as a frontal lobe measure. It is used to measure working memory and response inhibition. Whilst only reported in the original paper by Ball et al. (2008), these preliminary findings suggest this task may be a useful test for identifying dementia (d>0.5) and less affected by the severity of intellectual disability.

They also found that the number of informant-reported personality/ behaviour changes significantly predicted scores, indicating executive dysfunction.

Using the Colour Trails Test (CTT), Palmer (2006) found lower scores in those with dementia compared to those without but with floor effects for those with dementia. The CTT is a non-verbal adaptation of the Trail Making Test (TMT; Reitan, 1958) used primarily to measure attention but with an additional executive component (Part B) used to measure set-shifting and working memory. The remaining executive function tests showed little to no effects, often with considerable floor effects.

1.7.3.8. Visuospatial.

Three studies reported on visuospatial subtests. The children's Wechsler style block design test was used in all three studies, with one study reporting on two further non-verbal subtests from the WISC-R.

Block design

This test involves arranging blocks with various colours on each side to match specified patterns provided on cards or as a model made by the examiner. It is often included in IQ test non-verbal indices and scoring is usually based on accuracy and speed. Hom et al. (2021) found the task to load onto the visuomotor domain in factor analysis and to produce the most significant difference in scores between those with DS who were cognitively stable and those with a diagnosis of MCI. Krinsky-McHale et al. (2020) found lower scores for those with Down syndrome and MCI, or dementia compared to those who were cognitively stable, using an adaptation with less complex items from the DSMSE Block T-test (Haxby, 1989). However, despite the adaptations, they found significant floor effects in those with a more severe intellectual disability. Devenny et al. (2000) found a significant difference between those with Down syndrome and dementia and those without, and that it was sensitive to early decline.

Overall, this test seems promising in distinguishing between dementia and no-dementia groups and possibly detecting early-stage dementia in those with mild to moderate intellectual disability. However, it may be too difficult for those with a more severe intellectual disability.

Other visuospatial tests

Also reported by Devenny et al. (2000) were the Object Assembly and Picture Completion subtests from the WISC-R. For both subtests, they reported significant differences between the healthy Down syndrome group and all the other Down syndrome and dementia status groups. However, they could not differentiate between early- and middle-stage dementia groups or the healthy and 'questionable' groups, indicating they are insensitive to decline.

1.7.3.9. Language.

The most used language measures assessing cognitive decline were the PPVT/ PPVT-R (*n* = 3) and the BNT (*n* = 3). A further eight subtests and tasks were included under this domain, but none were reported in more than one study.

PPVT

The PPVT (Dunn, 1981) measures receptive vocabulary and comprehension in children without verbal expression. The person listens to a word spoken by the assessor and then selects one of four pictures that best describes the word's meaning. Brugge et al. (1994) and Pyo et al. (2007) found no significant differences between those with or without dementia. However, Nelson et al. (2001) found that levels of receptive vocabulary were significantly lower in participants with Down syndrome who had abnormal physical findings, based on MRI and neurological examination, compared to those with normal findings. Language comprehension appeared to *increase* over time for those with normal physical findings, but scores reduced for those with abnormal findings. It is unclear from these findings if this test could be useful in detecting dementia.

BNT

The BNT (Kaplan et al., 1976) is a brief (15-minute) test of confrontation word retrieval and was developed for adults with aphasia or other language disorders caused by dementia or stroke. The BNT includes 60 line drawings that increase in difficulty, from everyday items (e.g., tree) to rarer items (e.g., abacus). In a sample of mixed aetiologies of intellectual disability, Palmer (2006) found the Down syndrome-dementia group to show difficulties in naming equivalent to more than one standard deviation below the control group's mean. Hom et al. (2021) found it to load onto the language/ executive function domain on factor analysis. They found significant differences between a group of cognitively stable people with Down syndrome and those with MCI. Meanwhile, Brugge et al. (1994) only found a trend towards a diagnostic group effect, though this was with a small sample. Overall, these results suggest utility in this population.

Other language subtests

Four other verbal subtests from the WISC-R were reported by Devenny et al. (2000), specifically Information, Similarities, Verbal Comprehension, and Vocabulary. All of these subtests showed some differences between those with Down syndrome and dementia and those without but could not differentiate between the stages of dementia, indicating they are insensitive to decline. One study (Carr, 2003) reported longitudinal findings for two verbal subtests from the NAID: Picture Identification and Picture Naming. Carr (2003) found that scores decreased in Picture Naming from ages 30 to 35 years but slightly increased in Picture Identification. One study (Sano et al., 2005) reported on the Expressive One-Word Picture Vocabulary Test (EOWPVT) and found the Vocabulary score was sensitive to dementia status and age but not level of intellectual disability, suggesting it may be useful across the range of intellectual disability but would need further assessment.

1.7.3.10. Learning and Memory.

Not surprisingly, given that most of the studies focused on Down syndrome and AD, the highest number of tests identified in this review were for learning and memory, with a total of thirty-four subtests and tasks reported.

These include tests of verbal and visual, immediate, and delayed recall, and recognition and cued subtests. The Busckke Selective Reminding Test (BSRT) was the most commonly used test used in seven studies. Second to this was The Cued Recall Test (CRT; *n* = 6) and Fuld Object Memory Evaluation (FOME; *n* = 4). Four studies reported using a version of object memory from the NAID or the Stanford-Binet Intelligence Scale (SBIS; Thorndike, 1986). Three reported on the 'memory for sentences' subtest from the same batteries.

The Buschke Selective Reminding Test (BSRT)

The BSRT is a brief (15-minute) measure of verbal learning and memory using a list-learning procedure over multiple trials (Buschke, 1973). Hill et al. (1988) modified it for use with people with an intellectual disability to avoid floor effects (used by Devenny et al., 1992, 1996, 2000; Hom et al., 2021; Krinsky-McHale et al., 2002, 2008, 2020). The modified version includes familiar animals or familiar foods. After the list presentation, participants are asked to recall as many items on the list as possible, in any order. After this first trial, only the items not recalled are shown again.

Most studies found a significant difference between those with no dementia and those with possible dementia, as well as those in early-stage decline, with a decline in scores increasing with the progression of dementia. Furthermore, Krinsky-McHale et al. (2002) found it could distinguish age-related decline in explicit memory from the more significant decline associated with a diagnosis of AD. Hom et al. (2021) found it to show the most significant group difference within the memory tests used in that study. However, Devenny et al. (1992) found no difference using longitudinal assessment and showed improvements, likely due to practice effects. In addition, scores were found to be affected by the level of severity of intellectual disability and floor effects were noted (Krinsky-McHale et al., 2002, 2020). Overall, these findings suggest utility for people with a less severe intellectual disability.

The Cued Recall Test (CRT)

The CRT (Grober & Buschke, 1987; Tulving & Pearlstone, 1966) assesses memory retrieval with the help of cues. The Grober and Buschke version was later modified for adults with Down syndrome (Devenny et al., 2002; Zimmerli & Devenny, 1995). The modified version has a simpler and fewer items and categories. During the learning phase, twelve items representing distinct semantic categories are presented on three four-item cards, each accompanied by a unique category cue. This is repeated up to three times if necessary. The testing phase includes three free and cued recall trials, generating two scores (free immediate recall and total). A 20-minute delayed recall trial has since been included, generating two additional scores (free delayed recall and a total delayed score).

All studies found the CRT to be a sensitive indicator of dementia, particularly early-stage dementia. Devenny et al. (2000, 2002) and Sacco et al. (2022) proposed cut-off scores to identify early-stage dementia due to AD in Down syndrome and Benejam et al. (2015), and Devenny et al. (2002) found those with AD to make more semantic errors, even when provided with a cue. However, Sacco et al. (2022) found total free recall scores to be significantly impacted by the level of intellectual disability, and so risks incorrectly diagnosing dementia in people with moderate to severe intellectual disability. Similarly, other studies found some floor effects in those with a more severe intellectual disability and those with a more advanced stage of dementia. These results suggest that the modified CRT is a sensitive tool that may help diagnose AD in subjects with Down syndrome and mild to moderate intellectual disability.

Fuld Object Memory Evaluation/ Modified Fuld Object Memory Evaluation

The Fuld Object Memory Evaluation (FOME; Fuld, 1977) used multisensory encoding of objects and was designed for use with older adults. This was later modified for use with people with an intellectual disability (Seltzer, 1997) with verbal prompting for objects during the recall trials and a shorter interval delay.

One study used the original (Palmer, 2006), two used the mFOME (Pyo et al., 2007; Sano et al., 2005), and one study (Pyo et al., 2010) simplified the original test by reducing the number of items and removing the interference tasks, following considerable floor effects being found in the 2007 study for those moderate to severe intellectual disability. However, using this simplified version, they found no significant difference between the AD and control groups. Both Palmer (2006) and Sano et al. (2005) found significantly lower scores in those with dementia, and Sano et al. (2005) noted its effectiveness across all levels of intellectual disability in a large sample of people with Down syndrome and a wide range of severity. These findings suggest it may be a suitable test but may require further assessment.

Other learning and memory tests

One study (Carr, 2003) reported on six subtests of The Rivermead Behavioural Memory Test for Children (RBMT-C; Aldrich & Wilson, 1991) with no significant differences found on longitudinal assessment and considerable floor and ceiling effects found across the subtests. However, the age group of these participants is likely not to include those with cognitive decline. Of the remaining subtests, the Delayed Recall and Delayed Recognition subtests of the CAMCOG-DS were found to have medium to large effect sizes ($d \geq 0.05$) for distinguishing AD from no-AD by Ball et al. (2008). Also indicative of positive utility, Devenny et al. (1996) found the digit span subtest to show significant differences between those with possible AD and those without in a group of participants with Down syndrome, as well as showing decline using longitudinal assessment in the group with possible AD. This finding was replicated in a follow-up study (Devenny et al., 2000). However, they also found that it was insensitive to the early stages of decline.

1.7.3.11. Sensorimotor.

Nine subtests and tasks to measure sensorimotor abilities were reported in the studies. The Brief Praxis Test (BPT; *n* = 5) and the Beery Buktenica Developmental Test of Visual-Motor Integration (BBDT-VMI; *n* = 3) were the most commonly reported tools. No other measure was reported in more than one study.

Brief Praxis Test (BPT)

The BPT (Dalton, 2009a, 2009b) is a brief, 20-item measure of praxis that requires minimal verbal ability and is designed for use with adults with Down syndrome. Praxis tests typically involve completing a series of highly practised brief voluntary movements (e.g., “Show me how you would wave goodbye”).

All studies showed lower BPT scores associated with dementia with minimal floor and ceiling effects. This included distinguishing between no- and probable but not early-stage dementia (Wallace et al., 2021). Furthermore, Powell et al. (2014) found poorer performance on the BPT correlated with neuropathological findings of white matter reduction, mainly within frontoparietal regions, suggesting that late myelinating frontal pathways may be vulnerable in Down syndrome. Sano et al. (2005) found that the BPT showed sensitivity to change over time in people with Down syndrome on longitudinal assessment. Head et al. (2011) found it was affected by the severity of intellectual disability. These results suggest that the BPT test may be a valuable measure of functional decline in Down syndrome due to dementia.

Beery Buktenica Developmental Test of Visual-Motor Integration (BBDT-VMI)

The BBDT-VMI is a test of visual-motor integration, first developed by Beery and Buktenica in 1997 (Beery et al., 1997), with several subsequent revisions. It involves copying increasingly complex drawings of geometric shapes.

Burt et al. (2005) found lower scores to be associated with dementia in adults with Down syndrome. Similarly, Krinsky-McHale et al. (2020) found that lower scores were associated with dementia onset in Down syndrome but not MCI. Hom et al. (2021) found it to be associated with dementia as part of a visuomotor composite score in adults with Down syndrome. These results indicate that the BBDT-VMI has some utility in assessing for dementia in those with Down syndrome but may not be sensitive enough to detect it at a prodromal phase.

Other sensorimotor tasks

Minimal to no effects were found for any of the other sensorimotor subtests, except for the Purdue Pegboard Test (PPT; Tiffin & Asher, 1948). Hom et al. (2021) found the 'both hands' score on the PPT to be significantly lower in those with early-stage dementia compared to those who were cognitively stable.

1.7.4. Summary of Findings and Discussion

A total of 81 directly administered single-domain subtests and tasks for assessing cognitive decline in people with intellectual disabilities were found across the 36 studies. Due to the evidence for the association between Down syndrome and AD, most studies had focused on this group, as opposed to other aetiologies of intellectual disability or dementia disease processes such as LBD or VaD. Therefore, it is difficult to ascertain how applicable the findings are to people with an intellectual disability from causes other than Down syndrome.

No novel tasks have been reported since the review by Paiva et al. (2020). Similar to the findings from the earlier reviews, many of the tests were not developed for people with an intellectual disability nor to detect dementia. Most commonly, measures have been designed to detect dementia in adults with typical development or to assess children's cognition, with limited use of tests of verbal and non-verbal reasoning. Though these domains may have been covered by other tests or batteries not reported in this review, such as the WAIS. However, there were several measures specifically designed for people with an intellectual disability, with attempts to establish normative values for comparison. Despite this, many still had floor effects at baseline. Therefore, they were insensitive to change within the intellectual disability population and were only suitable for the mild to moderate intellectual disability level. None of the identified studies specifically reported data about the tolerability of the measures or chose measures specifically for those with a more severe intellectual disability, sometimes excluding participants at baseline. However, many of the tests showed utility for distinguishing between people with and without dementia, including early-stage dementia, which could potentially be further adapted to suit this population.

No studies reported assessing olfactory abilities, despite odour identification, recall and recognition tests having the potential to be an accessible and non-invasive method for early identification of AD in people with Down syndrome, as highlighted earlier. Although many executive function tests were found, many were from the CANTAB or CEFA batteries, and many were verbal fluency tasks. Despite many being developed for use with people with an intellectual disability, they continued to show considerable floor effects, evidencing the difficulty assessing executive function in this population. No studies reported using any subtests from the BADS-ID to assess dementia. Given the suggestion that executive function may be an early marker of dementia in people with an intellectual disability, it may be prudent to continue to develop new tests for this domain.

Limitations of this review include the need for a systematic collection of studies and a thorough quality assessment. Therefore, the results should be interpreted with that in mind. Grey literature was not included to ensure sufficient quality of the studies but may have missed some potentially helpful information and novel instruments. Psychometric data and information on acceptability and tolerability were limited, unsurprisingly. Many measures were reported in secondary or post-hoc analyses and may be underpowered to find an effect. Lastly, the data was extracted by a single researcher. Therefore, it cannot be excluded that errors in extraction may have occurred.

Overall, gaps remain in the literature for studies including participants with an intellectual disability from other causes, assessment of olfactory function, and the development of appropriate tests of executive function designed specifically for people with an intellectual disability in the assessment of neurodegeneration, with tailored norms.

1.8. The Current Study

1.8.1. Rationale

Given the mixed findings for the currently recommended batteries and the lack of a 'gold standard' valid and reliable measure explicitly developed for this population, a novel test set is proposed using the literature review outlined above. This measure would facilitate further research into treatments for dementia in people with an intellectual disability and the ability to provide proactive clinical services and differential diagnosis. Prasher (1997) has argued that a single tool is unlikely to be used to detect dementia in people with an intellectual disability. Still, combining the most predictive subscales or subtests of existing instruments within a test set may lead towards developing a more dependable and helpful dementia screening tool for people with an intellectual disability.

This particular study was part of a larger project of work. The test materials were developed jointly with the project supervisor (a clinical neuropsychologist) and another researcher. Whilst this paper focused on collecting data through the NHS with people with an intellectual disability from causes other than Down syndrome, the other researcher has collected data from people with Down syndrome recruited through the NHS and third sector and voluntary organisations. The aim is that between these two studies, an initial draft will be piloted and reviewed with a small group of participants to derive preliminary data on the acceptability and feasibility of the test set to inform a second draft, which will be co-normed as a set. It is intended to be a normative battery with low floors, allowing discrimination of change. This differs from the current criterion tests recommended by the BPS (2015b), such as the CAMCOG or NAID, meaning they have a pass-or-fail scoring system.

This rationale aligns with Fenn et al. (2020), who suggest a 15-stage test development and validation process. Phases one and two represent the literature review described earlier and early discussions with experts, a research proposal, and an ethical review (outlined in the next chapter). The development of a draft novel test battery (phases three, four and five) is described in the next section. This thesis

will focus on phases six and seven: preliminary data collection and item analysis. This data will include test and item acceptability using qualitative analysis of participant behaviour and verbal feedback. These will inform phase eight, the creation of a second draft. Implications for the remaining stages will be considered in the discussion.

1.8.2. Research Questions

The following research questions were developed to assess for the *acceptability* and *feasibility* of the novel test set:

1) Does this measure include items appropriate for and suited to this population?
2) Therefore, is it sufficiently engaging?
3) Are the language and stimuli used at the right developmental level?
4) Do the tests yield an acceptable range of scores with no ceiling or floor effects?

CHAPTER 2: METHODS

2.1. Epistemology and Researcher's Position

The epistemological and ontological perspectives assumed in this research will influence the chosen methodology, data analysis, and reporting (Barker & Pistrang, 2005). This research takes a critical realist perspective. Critical realism assumes there is a real world with regularities but that we cannot know it with certainty, essentially meaning that phenomena can be measured in a standardised way, but that knowledge must be held tentatively (Barker et al., 2016; Burr, 2003). This position emphasises that results should be replicable by other researchers through a detailed description of the methods used and the results found. Furthermore, the topic should be approached using different methods, with complementary strengths and weaknesses, to produce a range of results that can be 'triangulated' (e.g., quantitative, and qualitative data).

Critical realism's assumptions oppose the assumptions of social constructionism and narrative approaches, for example, which reject objective reality and are only concerned with a person's subjective interpretations (Cruickshank, 2012). The cognitive domains referred to in this research are assumed to exist but are recognised as constructs of a particular social and political context. They cannot be directly measured; they can only be inferred through subjective observation and operationalisation. Although all attempts are made to reduce subjectivity and bias in research (and, therefore, in cognitive testing), these factors are inextricable due to their development. Given my position, I will use a quantitative methodology complemented by qualitative feedback.

2.2. Study Design

This acceptability and feasibility study adopts a cross-sectional design. It was, therefore, exploratory, designed to develop an understanding of the usefulness and appropriateness of a novel measure of dementia for people with an intellectual disability by piloting it with a small sample of people with an intellectual disability.

Quantitative data was gathered through the scoring of the measure, and qualitative data was collected through verbal feedback on the participants' experience of completing the measure, and through observation of their engagement with the tasks. The results will inform modification of the measure for future piloting with further samples of people with an intellectual disability.

2.3. Ethics

Ethics are a fundamental consideration for people with an intellectual disability due to a perceived vulnerability and associated concerns around capacity and consent (Nicholson et al., 2013).

2.3.1. Ethical approval

This study was approved by:

1) The Health Research Authority (HRA) and The NHS Research Ethics Committee (NHS-REC; Appendix E).
2) The collaborating NHS Trust's own research and development team.
3) The Ethics and Integrity Sub-Committee (EISC) at The University of East London (UEL; Appendix F).

2.3.2. Informed Consent and Capacity

People with an intellectual disability are susceptible to social desirability and suggestibility and may change their answers to questions when provided with negative feedback (Clare & Gudjonsson, 1993; Everington & Fulero, 1999). This susceptibility was essential to factor into the consent process to ensure voluntary participation (Dobson, 2008). Therefore, consent was requested from the participant and their relative/ carer (where applicable) using the participant and carer consent forms (Appendices G and H). Attempts were made during the recruitment process to ensure voluntary engagement by involving relatives, carers, and support workers to aid communication and understanding. Plenty of time was allowed to for questions, and a video , which explained the study in simple and clear language was provided.

If it were felt the participant could not consent, according to criteria set out in the Mental Capacity Act (MCA; Department of Health [DoH], 2005), a relative would have been consulted. This is also recommended by The BPS (Dobson, 2008). If participants chose to participate in the study, they were asked to bring their trusted relative/ carer/ friend (where applicable) to the testing session so they could advocate for them if they wished.

2.3.3. Participant Safety

Participants were monitored for any signs of discomfort or dissent to participation throughout the testing session. Regular breaks were offered to avoid fatigue, and refreshments were offered throughout the session. The researcher explained that the test instrument would not give any useful information regarding performance or have any diagnostic significance as it is a pilot study. This information was given during recruitment and consenting to ease the person's mind and reduce any anxiety about needing to perform 'correctly' on the tests (Bennett-Levy et al., 1994). However, people may still have been affected if they believed their performance to be suboptimal. Words of encouragement were provided in the examiner's manual of the novel test battery to reduce this. Furthermore, the researcher emphasised throughout the session that this pilot battery will be adapted based on their perspectives and feedback. This information was also provided in the debrief letters.

This research occurred during the period following the COVID-19 pandemic, so safety procedures were adhered to in line with the current government advice. The testing took place in a large room where two-metre social distancing rules could be followed, and the researcher wore personal protective equipment (PPE) when compulsory. It could be worn by the participant and their trusted persons if they chose to. Using a lateral flow test, the researcher tested for COVID-19 before each testing session. Participants and their trusted persons were recommended not to come to the assessment if they were experiencing symptoms of COVID-19 or had tested positive on a COVID-19 lateral flow test within the last week.

2.3.4. Data Protection and Confidentiality

Each participant was assigned a number (e.g., P001, P002) to protect the anonymity of the data. Test scores and materials were stored using this anonymised number and kept separately from any identifiable data. This method ensured that the results were kept confidential. Any patient-identifiable data were kept in a separate spreadsheet and stored securely. Patient-identifiable data were kept for three weeks following data collection to allow participants time to request to withdraw their data from the study if they wished to. After these three weeks, it was destroyed. Testing sessions were video-recorded and stored as .mp4 files to review scoring and test accessibility later. Videos were uploaded to the UEL OneDrive for Business (a secure and encrypted online service) after collection through an encrypted UEL Microsoft Teams account. Once scored (within three weeks), these video files were destroyed. Upon uploading to OneDrive, any paper information was destroyed confidentially. Only the researchers and the supervisor had access to the data. No identifiable data is given in this thesis or will be included in any report or publication. Data is reported by group and kept anonymous per person. The anonymised data will be stored for ten years in UEL's data repository, accessible only to the research team and a limited number of library staff. After this time, it will be destroyed following UKRI (United Kingdom Research and Innovation) recommendations.

2.4. Recruitment

Participants were recruited through three London NHS adult learning disability services. Each of these services was provided with a 'recruitment pack' consisting of the Study Recruitment Poster (see Appendix I) and invitation letters; one for the participant (see Appendix J), one for the person's relative/ carer (see Appendix K) and an easy-read version (see Appendix L). A link to a YouTube video explaining the research was also available if requested. The psychologists at each of these services identified eligible participants from their caseloads, and the poster was displayed in the waiting area of each of the services so that people could self-refer by speaking to their clinician. Once identified, the recruitment pack was sent to each potential participant, along with an easy-read cover letter developed by one of the services (see Appendix M).

After a week, the service contacted each potential participant and their guardians to ask if they would be interested in participating in the study. If they agreed, their details were passed to the researchers to make contact to discuss their participation further and book in for an assessment if they agreed to participate.

2.5. Procedures

2.5.1. Consent

Potential participants or their relatives/ carers who had given consent for their details to be shared were contacted by the researcher to explain the research and what was required of them. The potential participants and their relatives/ carers were encouraged to ask questions and raise any concerns. If they decided to participate, they were offered a range of dates and times to attend a testing session at their usual NHS adult learning disability service.

At the testing session, participants (and their relatives or carers) were asked to read the invitation letters with the support of the researcher to allow any further questions to be answered. Participants' capacity to consent was assessed using guidelines from the MCA (DoH, 2005; i.e., the ability to understand, retain, weigh up, and communicate a decision). If it were felt the participant could not do any of these things despite efforts to increase capacity to consent (e.g., using adaptations for communication), the testing session did not commence. This process ensured that participants could meaningfully engage with the materials or were omitted ethically. If the participant could consent, the participant (and their relative or carer) were asked to complete the consent forms. All participants and their relatives or carers were thanked for their time with a £10 shopping voucher, regardless of whether or not the testing session was completed.

2.5.2. Pilot Session Procedure

The test experience and feedback sessions occurred in a private room at the participant’s usual NHS adult learning disability service or affiliated day centre. The participant and researcher sat at a table facing each other during the meeting. Any relative or carer was asked to sit behind the participant, out of their sight, to avoid distraction.

The laptop to video-record the session was set up on the researcher's right side, with a complete view of the participant, researcher, and materials on the table. Demographic information was collected, including date of birth, age, sex/ gender, handedness, nationality/ ethnicity, primary language, other language(s), and years of education. This data was used to describe the sample and consider the generalizability of the primary results. Breaks were offered throughout to minimise fatigue or boredom. Test administration is described below. Following testing, participants and their relatives/ carers were debriefed verbally and given a copy of the Participant Debrief Letter and Easy-Read version (see Appendices N and O, respectively) before requesting verbal feedback using the pre-prepared Semi-Structured Interview Schedule (see Appendix P). If it were unclear or ambiguous which task a participant referred to in the semi-structured interview, the researcher would show the participant the test materials and ask them to point to which one they were referencing.

2.6. Test Materials

2.6.1. The Novel Cognitive Test Battery

The novel cognitive test battery included an Examiner's Manual, Record Form, and Stimulus Book (not reproduced here to protect the validity of the test). Some tests were adapted from existing batteries, whilst some were created for this study. Existing tests used were thought to assess the relevant cognitive domains appropriately but needed adjustment to make them suitable for the needs and abilities of people with an intellectual disability. The novel test battery components and their cognitive domains are outlined below in Table 2, along with the included tests and their adaptations.

Table 2

Test Battery Cognitive Domains, Component Functions, Associated Tests, Main Sources and Adaptations

Domain and Function	Test Component	Adapted From	Adaptation(s)
Sensory, olfactory	Smell Detection	UPSIT (Doty, 1995)	Everyday household substances on cotton pads placed in jars
Motor, upper limb	Motor Function Part A Motor Function Part B	EMAS (Bak et al., 2015)	Eight of the simplest items with accessible instructions
Attention - receptive	Orientation & Information Sentence Repetition	MMSE (Folstein et al., 1975) 'Orientation' task Spreen and Strauss (1998)	Culturally-unbound questions, suited to contexts and simplified Adapted using common single-syllable words in simple sentences
Attention - expressive	Eight Detection Circle Search	KBNA Auditory Signal Detection Test (Leach, 2000) KBNA Symbol Cancellation Test (Leach, 2000)	Simplified shorter format using a friendly female voice and restricted range of stimuli (numbers) Larger outline of basic shapes with a familiar target (circles) and fewer distractors
Executive - receptive	Verbal Reasoning Visual Reasoning	Wechsler style 'Similarities' task (Wechsler, 1955) Raven's style 'Matrix Reasoning' task (Raven, 1995)	Easier items using simplified language Colour palette appropriate for people with colour blindness, simpler items
Executive - expressive	Word Generation Cat-Dog Inhibition Shopping List Motor Programming	Typical format 'category fluency' tasks (Lezak et al., 2012) CEFA (Ball et al., 2008) 'Cats and Dogs' task 'Zoo Map' task from BADS (Wilson et al., 1996) and BADS-ID (Webb et al., 2020) 'Shopping List' task Golden & Freshwater (2001)	Instructions simplified and prompts given to aid performance Realistic pictures and uniform colours using shorter format Novel format task to increase ecological validity, using realistic stimuli Simpler instructions and modelling of the tasks in practice trials
Verbal - comprehension	Verbal Comprehension A Verbal Comprehension B	BDAE (Goodglass & Kaplan, 1972) BDAE (Goodglass & Kaplan, 1972)	Instructions simplified, fewer items, and prompts given to aid performance Instructions simplified, fewer items, and prompts given to aid performance
Verbal - expression	Verbal Expression Picture Naming	BDAE (Goodglass & Kaplan, 1972) BDAE (Goodglass & Kaplan, 1972)	Quality of speech output assessed by observation of previous test responses Novel set of familiar items in colour photographs
Visual - perception	Angle Judgment	JLO (Benton et al., 1978)	Fewer target lines and simpler 5-point reference key
Visual - action	Matchsticks Copy Praxis	Novel task Heilman and Rothi (1993)	Novel task, using matchsticks to copy a model instead of drawing Limited to pantomime of tool and task sequences with supportive instructions
Learning and Memory - verbal	Word List Learning Word List Imm. Recall Word List Delayed Recall Word List Recognition	RAVLT and its modified and simpler formats (Lezak et al., 2012)	Fewer words per trial and fewer trials, using common, concrete single-syllable words
Learning and Memory - visual	Matchsticks Imm. and Delayed Recall	See above	See above
	Picture Recognition	Wilson and Antablin (1980)	See above; paired two option forced-choice responses, to items previously seen, with motor responses permitted
Learning and Memory - olfactory	Smell Detection Recognition	See above	See above

Note: BADS = Behavioural Assessment of Dysexecutive Syndrome; BADS-ID = Behavioural Assessment of Dysexecutive Syndrome – Intellectual Disabilities; BDAE = Boston Diagnostic Aphasia Examination; CEFA = Cambridge Executive Functioning Assessment; EMAS = Edinburgh Motor Assessment Scales; JLO = Judgment of Line Orientation; KBNA = Kaplan Baycrest Neurocognitive Assessment; MMSE = The Mini Mental State Examination; RAVLT = Rey Auditory Verbal Learning Test; UPSIT = The University of Pennsylvania Smell Identification Test

2.6.2. The Semi-Structured Interview Schedule

A Semi-Structured Interview Schedule was developed for this study to gather individual feedback from participants regarding their experiences of completing the test battery.

2.6.3. Test Administration

For each task in the battery, the researcher read the instructions verbatim, per the Examiner's Manual, aloud and recorded the responses in the draft Record Form. Stimuli were presented using the Stimulus Book as and when required. The tests given as part of the novel test battery were completed in the order shown below. The first section generally addressed fundamental communication and sensory and motor functions. The second section addressed verbal learning, memory, and visuospatial and executive functions. In section three, visual learning and memory were assessed alongside verbal functions.

Motor and Language Functions

- Orientation & Information
- Smell Detection
- Verbal Expression
- Motor Function Part A & Verbal Comprehension Part A
- Motor Function Part B
- Motor Programming
- Praxis
- Verbal Comprehension Part B
- Smell Recognition

Verbal Learning and Visual Functions

- Word List Learning
- Circle Search
- Angle Judgement
- Visual Reasoning
- Shopping List
- Cat-Dog Inhibition

- Word List Delayed Recall
- Word List Recognition

Visual Learning and Verbal Functions

- Matchsticks Copy & Immediate Recall
- Eight Detection
- Picture Naming
- Sentence Repetition
- Verbal Reasoning
- Word Generation
- Matchsticks Delayed Recall
- Picture Recognition

Although no discontinuation rules were included (due to no normative data being available), if (for any reason) a participant could not complete a test, the researcher discontinued that test and moved on to the next. For example, if the test proved too difficult or required a function in which the participant was demonstrably impaired, such as upper limb function. Similarly, tests were usually discontinued after three consecutive scores of zero. This process was to avoid any additional stress on the participant.

2.7. Data Analysis

Quantitative data were coded into a spreadsheet for analysis using IBS SPSS Statistics for Mac (version 27).

2.7.1. Acceptability

Qualitative data was collected from verbal responses to the tasks and feedback and was recorded verbatim and transcribed following the testing session. Any additional comments from watching the video recordings, along with non-verbal expressions of interest, engagement, or difficulty, were also recorded.

Mental State Examination guidelines (Voss & Das, 2023) and guidelines for communicating with people with an intellectual disability (Boardman et al., 2014) were followed to assess relevant verbal and non-verbal communication throughout the testing session, for example, looking for any agitation, avoidance, tearfulness, anxiety, signs of distress or happiness, refusal to talk, and whether or not the behaviour is appropriate and congruent with verbal communication (e.g. saying they are happy to continue but looking objectively tearful). See Appendix Q for example coding, though this list was not exhaustive. Participant feedback using the semi-structured interview was examined using manifest content analysis (Hsieh & Shannon, 2005). See Appendix R for examples of coding for yes/no questions and to identify which tests participants were referencing. The frequency of these occurrences was then calculated. Individual answers were reported verbatim for responses to “Why” questions and “What could we change about these tests to make them better?” This information was recorded as part of the assessment of the accessibility and feasibility of the measure and was not analysed further.

2.7.2. Feasibility

Assessment of feasibility includes test performance and item analysis.

2.7.2.1. Test Performance.

A preliminary analysis of central tendency, dispersion, skewness, and kurtosis was conducted for each test in the battery to assess the distribution of scores, looking for ceiling and floor effects. The Shapiro-Wilk test of normality is reported, given that the sample size is <50. A distribution is called approximately normal if the skewness or kurtosis of the data is between − 1 and + 1. SPSS provides ‘excess’ kurtosis results obtained by subtracting three from the kurtosis (proper). Mishra et al. (2019) state that for a small sample size (n < 300), a more reliable method is to calculate a z-score by dividing the skewness values or excess kurtosis value by their standard errors. For a sample of n <50, a z-score of ± 1.96 suggests a normal distribution of the data (Ghasemi & Zahediasl, 2012). All of the above are reported.

2.7.2.2. Item Analysis.

Exploratory qualitative and quantitative methods for item analysis were undertaken. In terms of qualitative, Fenn et al. (2020) suggest reviewing the order, type, and wording of the test items and instructions to ensure they encourage participants to provide accurate and adequate information and avoid discontinuation or refusal to answer specific questions. Fenn et al. (2020) also state that test item wording must be sensitive to the target population. This factor will be reviewed using participant feedback and observations made during testing.

For quantitative analysis, items will be assessed for their difficulty level by calculating the proportion (percentage) passing or P value (Urbina, 2014). This value assesses whether items are appropriately scaled, with items increasing with difficulty on each subtest, allowing the development of discontinuation rules. Since participants do not have dementia, it is impossible to calculate discriminative power in this current study, though this may be undertaken in future studies.

2.8. Participants

2.8.1. Inclusion and exclusion criteria

The inclusion and exclusion criteria were used to enable sufficient engagement with the test instructions and stimuli and avoid confounding (Lezak et al., 2012). The age range of 30-55 years was chosen to be lower than the average age of dementia onset in people with an intellectual disability (Lott & Head, 2019). Participants were actively under the care of an affiliated NHS adult learning disability service to ensure that any support needed following the testing session could be facilitated.

Inclusion:

- Any sex.
- Aged between 30 and 55 years.
- Diagnosis of an intellectual disability (non-Down syndrome) but not dementia.
- Under the care of an affiliated NHS adult learning disability service.
- English language speaker with good understanding and fluency.

Exclusion:

- Any currently active neurological illness or acquired brain injury.
- Cerebral Palsy.
- Down syndrome.
- Known or suspected dementia.
- Blindness or deafness.
- Diagnosis of severe or enduring mental illness.
- History of or current illicit substance misuse in the last six months.

2.8.2. Sample

As this is a pilot study assessing the acceptability and feasibility of a first draft, a small sample is preferred and does not require an apriori sample size calculation (Bowen et al., 2009). Therefore, to reduce unnecessary (potentially unethical) testing, this study aimed to recruit a small sample of 5-8 people with an intellectual disability from causes other than Down syndrome. Participants were identified through three adult learning disability services within a London NHS Trust. Across the three services, 17 people were contacted to request their involvement in the study. Out of these 17, nine consented to have their contact details shared with the research team. From these 9, seven people took part in the study between April and June 2023. An additional person was invited to participate but was excluded at the assessment time due to concerns around capacity and ineligibility. A further two people were eligible and willing to take part. However, they required an interpreter to contact their relatives, which was impossible to arrange within the timeframe available for this thesis. The final sample (N = 7) consisted of five male and two female participants, aged between 37-55, with an average age of 47 (5.92). All had mild to moderate intellectual disability and lived either independently or semi-independently. The majority identified as White British. See Table 3 for a summary of demographic variables.

Table 3

Sample Characteristics

Participant	Sex	Age (Years)	Ethnicity	Handedness	Years of Education	Sight Difficulties	Hearing Difficulties
P1	F	48	Black British	Right	11	Yes	No
P2	M	51	White British	Left	11	Yes	No
P3	M	50	White British	Right	13	Yes	No
P4	M	37	British Asian	Right	13	No	No
P5	M	43	White British	Right	13	Yes	No
P6	F	45	Black British	Right	11	Yes	Yes
P7	M	55	White British	Right	13	Yes	No

CHAPTER 3: RESULTS

1.1. Acceptability

3.1.1. Participant Feedback

Q1. Did you find any of the tests interesting?

All participants indicated that they had found some of the tests interesting. Subtests that were mentioned included 'Picture Naming' (*n* = 3), 'Eight detection' (*n* = 2), 'Smell Detection' (*n* = 1), 'Sentence Repetition' (*n* = 1), 'Visual Reasoning' (*n* = 1), 'Angle Judgement' (*n* = 1), 'Word List' (*n* = 1), with two people saying "all of them".

Q2. Did you find any of the tests boring?

Only one participant indicated "yes" to whether they found any tests boring, citing the 'Visual Reasoning' subtest as being "too confusing."

Q3. Did you find any of the tests too easy?

Five participants indicated that they found some tests too easy, citing 'Picture Naming' (*n* = 2) and 'Visual Reasoning' (*n* = 1). However, two participants could not specify which test. One participant gave the reason for this as they "knew them all," relating to 'Picture Naming' and one suggested we use "harder pictures," also relating to 'Picture Naming'. One participant suggested we use cards, similar to a game of 'snap' for 'Visual Reasoning'.

Q4. Did you find any of the tests too hard?

Three participants indicated that they found some tests too hard, including 'Visual Reasoning' (*n* = 2), 'Matchsticks' (*n* = 2), 'Cats and Dogs' (*n* = 2), 'Angle Judgement' (*n* = 1), 'Shopping Task' (*n* = 1), and 'Word List' (*n* = 1). The reasons for this included them being "too hard," "too confusing," or "fiddly" concerning 'matchsticks'. Only one person made a suggestion how we could improve these, which was to use "less words" ('Word Lists'), "less shapes" ('Visual Reasoning'), and "less matchsticks" ('Matchsticks').

Q5. Do you have anything else you would like to say about the tests you did?

Three participants said they "enjoyed it." One person said that some subtests may be challenging for those with visual impairments, and we needed to make the words and pictures bigger, particularly relating to the 'Shopping Task.' One person said that some of the tests had made her "not feel good" but that it had been ok overall.

3.1.2. Observations

Most of the participants were able to attempt all of the subtests and appeared to enjoy the majority of the tasks due to verbal feedback provided throughout the assessment (e.g., "This is fun!") and non-verbal signs of enjoyment, such as laughter. The test battery took one to two hours to administer, depending on how quickly the participant could complete the tasks. One participant was noted to appear fatigued during the assessment, but others tolerated the length of the test seemingly well. This participant likely had a more severe intellectual disability than many of the other participants due to his difficulty with the majority of the tasks. This finding suggests the test may only be suited to people with a mild intellectual disability, though this would need to be assessed in future studies. Breaks were repeatedly offered but most often not taken. No participants asked for the assessment to be discontinued or their data to be excluded from the study. However, 'Angle Judgment,' 'Visual Reasoning,' and the 'Shopping Task' subtests were often discontinued, either due to the participant scoring three consecutive zeros, showing demonstrable difficulty with the task, or being unable to comprehend the instructions in order to engage with the task successfully. Timing for the 'Cats and Dogs' subtest was unintentionally not recorded for one participant, and the 'Matchsticks Immediate' task was unintentionally missed for one participant.

One participant could not undertake the tests that relied on verbal expression ('Word List,' 'Verbal Reasoning,' and 'Sentence Repetition') due to dysarthria or tests of complex motor function ('Motor Programming') due to motor control difficulties. However, he could participate in tests that did not require complex verbal or motor performances, e.g., 'Cats and Dogs,' 'Picture Naming' and 'Picture Recognition.' As the case for this participant shows, additional or separate tasks and tests will be required to meet the needs of non-verbal people and people with marked motor difficulties.

‘Eight detection’ was also abandoned for one participant because they tapped for every number, despite being instructed again to only tap for the number ‘8’. It is unclear whether this was a miscomprehension of the instructions, difficulties in working memory, or response inhibition. This finding reminds us that subsequent revisions should add test discontinuation rules to all tests, including single-trial multi-item tests. The ‘Matchsticks’ subtests proved difficult for many participants; it is unclear whether this was due to visual impairment or visual-perceptual difficulties. The matchsticks were noted to be difficult to manipulate for many, likely due to their small size and potential the dexterity difficulties of the participants.

1.2. Feasibility

3.2.1. Test Performance

Descriptive data for performance on the subtests is given in Tables 4-6 (broken down by subsection of the battery), along with numbers of missing data (due to participants being unable to complete the test or omission, as discussed above). Exploratory analysis of central tendency, dispersion, skewness, and kurtosis were derived for each subtest to assess the normality of the data (see Figures 2-4 and Appendix S). Results show that several subtests were not normally distributed; some showed floor or ceiling effects in this sample. However, the small sample size means these results should be considered provisional and need replication before conclusions can be drawn. The ‘Cats and Dogs’ subtest was missing the instructions to perform the test twice, once congruently and again incongruently, and therefore, no comparison could be made between the time taken to complete each condition.

‘Orientation Subtotal A’, ‘Orientation Total’, ‘Verbal Expression’, ‘Motor Subtotal A’, ‘Motor Subtotal B’, ‘Motor Function Total’, ‘Circle Search’, ‘Word List Recognition’, ‘Picture Naming’, ‘Picture Recognition’, and ‘Eight Detection’ all showed a negative skew, indicating that participants were generally scoring highly on these subtests. A high proportion of correct answers on ‘Orientation Subtotal A,’ ‘Verbal Expression,’ and ‘Word List Recognition’ subtests is to be expected in a non-dementia sample of people with good verbal ability.

This is the same for people who have shown typical development on tests like these. Of particular note, five participants scored the maximum on ‘Circle Search’ and four participants scored the maximum on ‘Eight Detection’ indicating a ceiling effect for these particular tests. ‘Motor Subtotal A’ and ‘Motor Subtotal B’ showed ceiling effects for some participants. This finding was reflected in the total score for ‘Motor Function.’

‘Visual Reasoning’ and ‘Verbal Reasoning’ both showed a positive skew, with most people scoring similarly. Two participants scored zero, and three participants scored one out of a maximum of ten, indicating a floor effect on ‘Visual Reasoning’ for many participants. Meanwhile, for ‘Verbal Reasoning,’ most participants scored eight, with two people scoring nine and ten, indicating a low range of scores. Although ‘Angle Judgement’ did not show a floor effect, it was difficult for almost all participants to score full marks on each item.

Table 4

Descriptive Data for Performance by Subtest - Motor & Language Functions

Motor & Language Functions							
Subtest	***n***	**Maximum Score**	**Range (Min-max)**	***n* Minimum Score**	***n* Maximum Score**	**Mean (*SD*)**	**Median (*IQR*)**
Orientation Subtotal A	7	12	3-12	0	2	9.3 (3.5)	11.0 (6)
Orientation Subtotal B	7	4	2-4	0	1	2.9 (0.7)	3.0 (1)
Orientation Total (A+B)	7	16	5-16	0	1	12.1 (3.9)	13.0 (6)
Smell Detection	7	5	0-2	1	0	1.1 (0.7)	1.0 (1)
Smell Recognition	7	10	5-8	0	0	6.1 (1.4)	6.0 (3)
Verbal Expression	7	20	9-19	0	0	16.3 (3.6)	17.0 (4)
Verbal Comprehension A	7	5	2-5	0	2	3.6 (1.3)	4.0 (3)
Verbal Comprehension B	7	18	13-17	0	0	15.3 (1.6)	16.0 (3)
Verbal Comprehension Total (A+B)	7	23	15-22	0	0	18.9 (2.7)	19.0 (5)
Motor Function Subtotal A	7	5	3-5	0	4	4.4 (0.8)	5.0 (1)
Motor Function Subtotal B	7	12	4-12	0	2	9.7 (2.7)	10.0 (2)
Motor Function Total (A+B)	7	17	7-17	0	1	14.1 (3.3)	15.0 (2)
Motor Programming	6	12	0-11	1	0	6.2 (4.2)	8.0 (7)
Praxis	7	30	26-29	0	0	27.7 (1.0)	28.0 (1)

Table 5

Descriptive Data for Performance by Subtest - Verbal learning & Visual Functions

Verbal Learning & Visual Functions							
Subtest	***n***	**Maximum Score**	**Range (Min-max)**	***n* Minimum Score**	***n* Maximum Score**	**Mean (*SD*)**	**Median (*IQR*)**
Word List Immediate	6	36	13-29	0	0	21.3 (5.5)	22.0 (9)
Word List Learning	6	9	0-3	1	0	1.8 (1.2)	2.0 (2)
Word List Delayed Recall	6	9	2-6	0	0	4.2 (1.6)	4.0 (3)
Word List Recognition	6	18	9-18	0	2	15.2 (3.3)	15.5 (5)
Circle Search	7	26	24-26	0	5	25.6 (0.8)	26.0 (1)
Angle Judgement	6	20	3-20	0	1	10.5 (7.9)	9.0 (16)
Visual Reasoning	6	10	0-4	2	0	1.17 (1.5)	1.00 (2)
Shopping List Map 1	7	20	1-18	0	0	9.1 (5.8)	7.0 (8)
Shopping List Map 2	5	22	4-20	0	0	13.0 (6.9)	14.0 (14)
Shopping List Total	5	42	5-38	0	0	23.2 (13.4)	28.0 (25)
Cat-Dog Inhibition	7	32	14-32	0	4	26.4 (7.4)	32.0 (11)
Cat-Dog Inhibition Time (seconds)	6	NA	30-85	0	NA	50.0 (21.7)	45.0 (38.5)

Table 6

Descriptive Data for Performance by Subtest - Visual Learning & Verbal Functions

Visual Learning & Verbal Functions							
Subtest	***n***	**Maximum Score**	**Range (Min-max)**	***n* Minimum Score**	***n* Maximum Score**	**Mean (*SD*)**	**Median (*IQR*)**
Matchsticks Copy	7	24	7-24	0	1	15.6 (7.4)	18.0 (15)
Matchsticks Immediate	6	24	6-19	0	0	12.0 (5.7)	11.5 (11)
Matchsticks Delayed Recall	7	24	7-19	0	0	11.7 (4.7)	9.0 (9)
Picture Naming	7	16	8-16	0	2	13.3 (3.1)	14.0 (6)
Picture Recognition	7	16	7-14	0	0	11.4 (0.9)	12.0 (3)
Eight Detection	6	14	13-14	0	4	13.7 (0.5)	14.0 (1)
Sentence Repetition	6	12	2-10	0	0	6.3 (2.9)	7.0 (5)
Verbal Reasoning	6	12	8-10	0	0	8.5 (0.8)	8.0 (1)
Word Generation	7	NA	20-33	0	NA	24.4 (4.4)	23.0 (6)

Figure 2

Boxplots for Motor & Language Functions: Central Tendency, Dispersion, Skewness

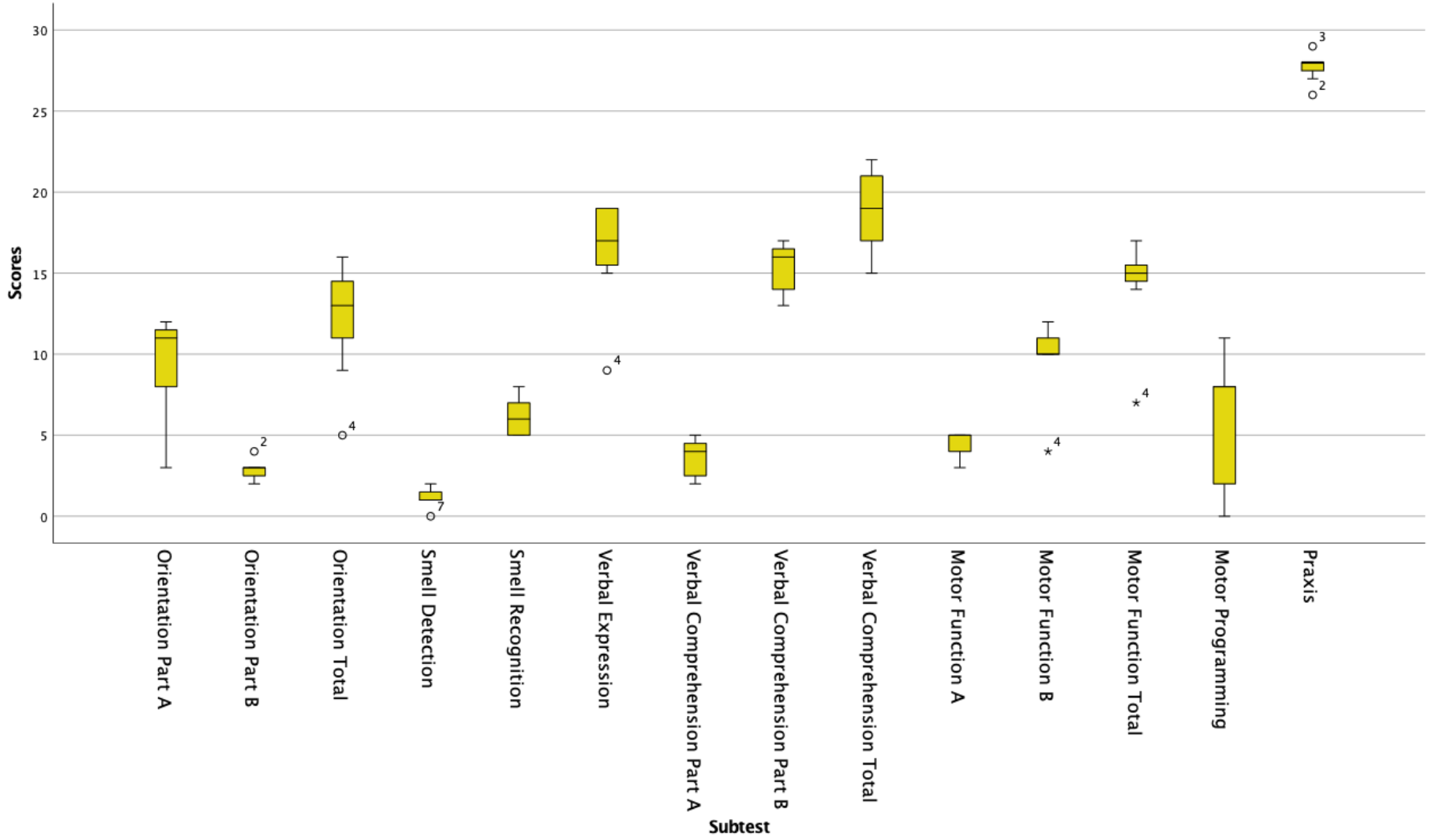


Figure 3

Boxplots for Verbal Learning & Visual Functions: Central Tendency, Dispersion, Skewness

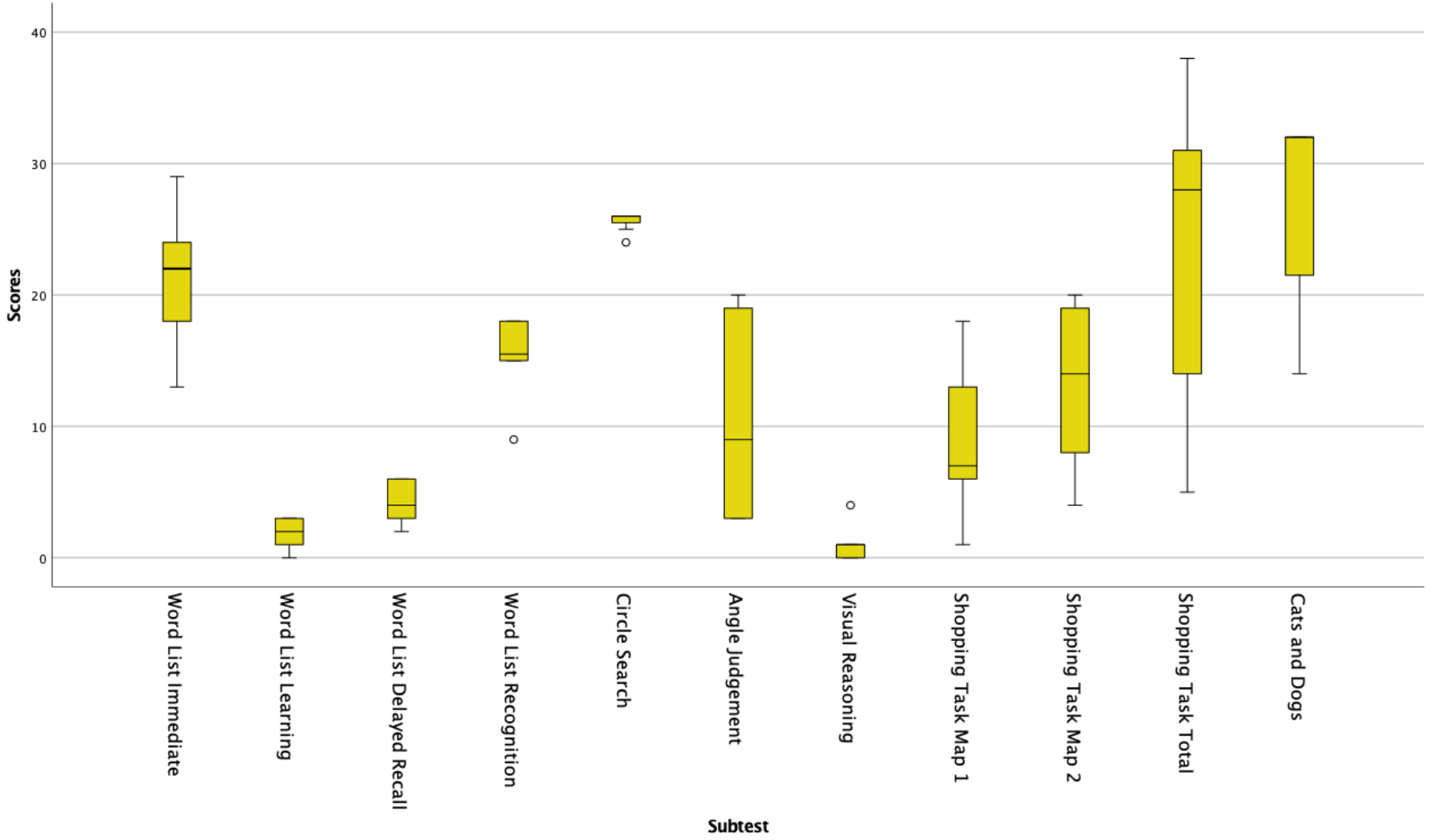
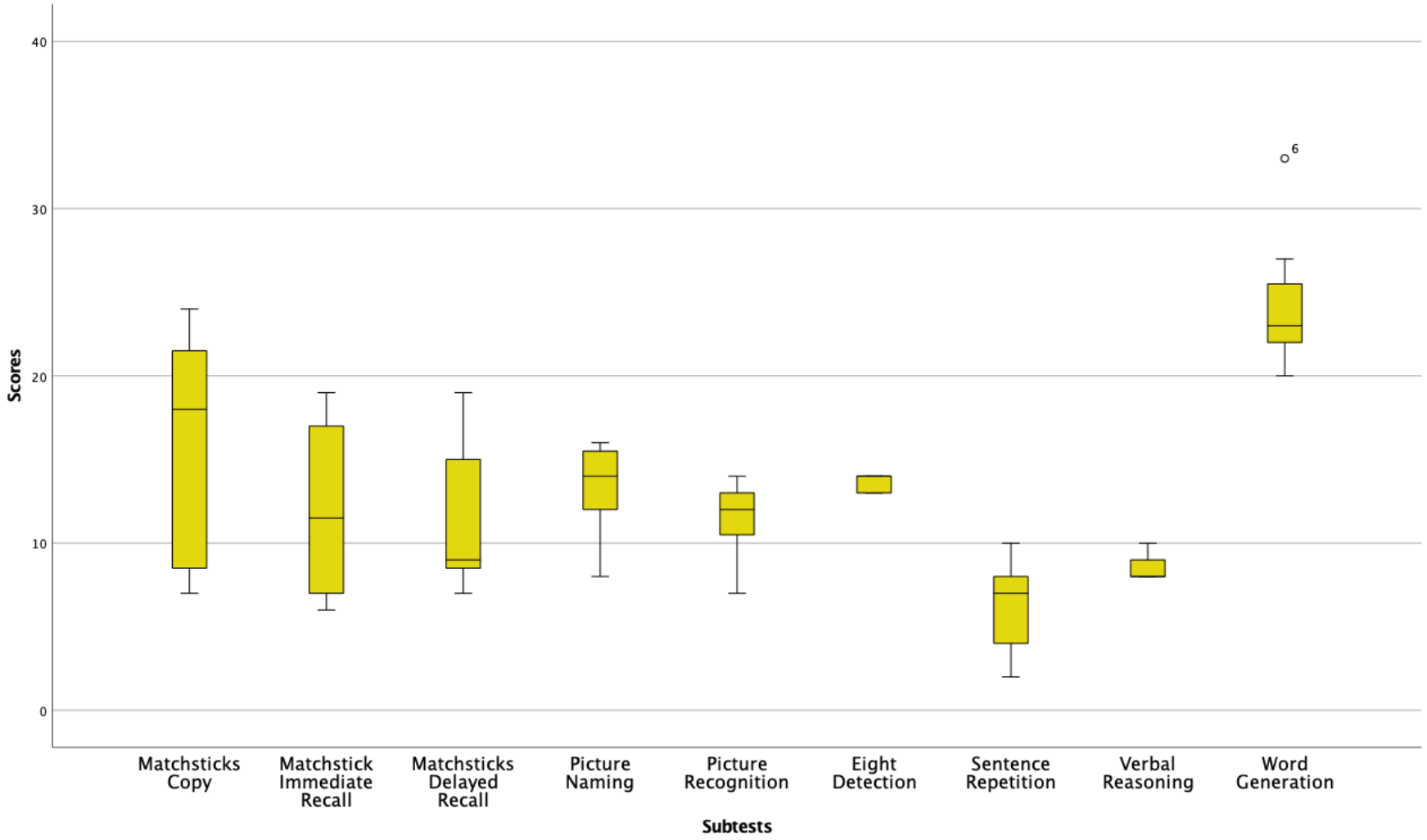


Figure 4

Boxplots for Verbal Learning and Visual Functions: Central Tendency, Dispersion, Skewness



3.2.2. Item Analysis

3.2.2.1. Qualitative.

Before the first assessment, the test materials were piloted with a person who has shown typical development to check for typos or other errors and ensure the instructions would encourage accurate engagement with the test materials. Some initial minor changes were made.

Acquiescence bias was observed for three participants on the ‘Smell Recognition’ subtest, equating to a score of 5/10 (50% correct by chance), and one participant said yes to all except one item. It is unclear whether this was due to poor differentiation between the smell items, difficulties with the olfactory function of the participants, the item instructions, or just that they are binary yes/no questions, which are known to elicit this response bias. However, this was not the same for the ‘Word List Recognition’ subtest, which indicates it may not have been the test format. Scores on the ‘Smell Detection’ task were generally low. Some of the fragrances used may be perceived as artificial (correctly) and, therefore, harder to name; that includes some powerful scents, such as mint. One of the most often given answers for the vanilla fragrance was “body cream” (or a similar name), which may indicate differences in knowledge and experience.

Some test items did not appear to encourage participants to provide adequate information to gain full marks, potentially discriminating unfairly. Fenn et al. (2020) state that the test item wording must be sensitive to the target population. For example, item one on the ‘Orientation Subtotal A’ asked, “What is your name?” The scoring instructions were one point for first and surname and zero points for anything else. Most people answered that question with just their first name, which would seem appropriate for people with an intellectual disability. Similarly, for ‘Orientation Subtotal B,’ a person would need to “give more than one, well-oriented, complete, and correct responses for each question” to receive the maximum score of two. Item two asked, “How did you get here today?” Most people answered this question accurately but with a single word or sentence, such as “bus” or “walk,” which again appeared to be difficult for participants with lower expressive verbal abilities (as opposed to their level of orientation).

Some items had complex instructions, such as ‘Motor Programming’ and ‘The Shopping Tasks.’ In ‘Motor Programming,’ items three and four are the knock-tap opposition and knock-tap inhibition tasks, respectively. These proved challenging for most participants to complete accurately, and the complexity of the instructions led to some minor errors in delivering those instructions, which had to be corrected. The ‘Shopping tasks’ instructions proved difficult for almost all participants to comprehend what exactly was asked of them. Many participants did not complete the map in a single move and would lift the pen and start at a new point for each item or draw through the aisles to get to other items. This finding may have been due to unfamiliarity with these sorts of ‘maze’ puzzles.

3.2.2.2. Quantitative.

The test items’ difficulty index levels are provided in Tables 7-9 to assess the appropriate scaling of each test item. The ‘Total Score’ indicates the total score of all participants combined. If tests were scaled appropriately, one would expect these numbers to decrease as the difficulty of the items increases (lower to higher number items). The P value indicates the ‘proportion (or percentage) passing.’ This value is only provided for the items with a binary correct or incorrect answer. An ideal range for an item difficulty index between 0.4 and 0.6 is chosen for norm-referenced tests (Urbina, 2014) and is highlighted in orange. Those items with a difficulty index below this level are highlighted in green, and items with a difficulty index above this range are highlighted in red. If tests were scaled appropriately, one would expect the lower items to be highlighted in green, indicating more accessible items, the central items to be orange, and the higher items to be highlighted in red, indicating more complex items. These values reflect the qualitative observations above, for example, ‘Orientation Subtotal A’ item one. Note that some tests, such as the ‘Orientation’ subtests and ‘Angle Judgement,’ were not designed to be scaled and therefore were not expected to show differentiation of difficulty between items and (as noted previously), ‘Angle Judgement’ was often discontinued early.

Item eight of ‘Praxis’ asked, “Show me how you would use scissors to cut through paper.” Most people pantomimed the action of scissors with their fingers and continued to do so, even after a request to imitate the researcher’s demonstration of a ‘squeezing’ motion.

Item 11 on ‘Verbal Comprehension Subtotal B’ proved difficult for many participants, with them often completing just one part of the two-part instruction or doing it in the wrong order. ‘Visual Reasoning’ proved too tricky for nearly all participants, including the sample items. Many participants scored low on ‘Smell Detection,’ and no participant answered items three (vanilla) and four (shoe polish) correctly. Item five of ‘Picture Naming’ was the picture of a hand and arm, with an arrow pointing to the wrist. People usually answered “hand” to this question. Performance improved by adapting the instructions to include “Yes, but what specific part is the arrow pointing to?” The final item, a butterfly, was easy to answer for all participants. ‘Sentence Repetition’ was challenging for most participants after the first two items, possibly due to verbal expressive difficulties. ‘Verbal Reasoning’ asked people to complete the sentence with an appropriate word to make the sentence true. Item five proved difficult for many participants: "A robin is a bird; a rabbit is…” Item 11 was quickly answered by most participants, which was “Pen is to writing as scissors is to…”. However, all participants answered this using a different tense, saying “cut” instead of “cutting.” This will require changes to the instructions to include this answer.

Table 7

Item Difficulty Level for Scalable Items – Motor & Language Functions

Motor & Language Functions																		
Orientation Subtotal A																		
Item #	1	2	3	4	5	6	7	8	9	10	11	12						
Total Score	3	6	5	5	4	5	6	6	7	7	6	5						
P value	0.43	0.86	0.71	0.71	0.57	0.71	0.86	0.86	1.00	1.00	0.86	0.71						
Orientation Subtotal B																		
Item #	1	2																
Total Score	10	10																
Smell Detection Total																		
Item #	1	2	3	4	5													
Total Score	2	2	0	0	4													
P value	0.29	0.29	0.00	0.00	0.57													
Verbal Comprehension Part A																		
Item #	1	2	3	4	5													
Total Score	5	6	6	4	4													
P value	0.71	0.86	0.86	0.57	0.57													
Verbal Comprehension Part B																		
Item #	1	2	3	4	5	6	7	8	9	10	11	12	13	14	15	16	17	18
Total Score	7	7	7	7	6	4	4	7	6	7	2	5	5	6	7	7	7	6
P value	1.00	1.00	1.00	1.00	0.86	0.57	0.57	1.00	0.86	1.00	0.29	0.71	0.71	0.86	1.00	1.00	1.00	0.86
Motor Function Part A																		
Item #	1	2	3	4	5													
Total Score	7	6	7	5	6													
P value	1.00	0.86	1.00	0.71	0.86													
Motor Function Part B																		
Item #	1	2	3	4														
Total Score	17	18	13	20														
Motor Programming																		
Item #	1	2	3	4														
Total Score	13	10	10	4														
Praxis																		
Item #	1	2	3	4	5	6	7	8	9	10	11	12	13	14	15			
Total Score	14	14	14	14	14	13	12	3	14	14	14	14	14	13	13			

Note. Total Score = The total score of all participants combined. P value = The proportion (or percentage) passing (this is only provided for the items with a binary correct or incorrect answer). Orange = Item difficulty index between 0.4 and 0.6; Green = <0.4; Red = >0.6

Table 8

Item Difficulty Level for Scalable Items – Verbal Learning & Visual Functions

Verbal Learning & Visual Functions										
Angle Judgement										
Item #	1	2	3	4	5	6	7	8	9	10
Total Score	10	8	8	4	7	6	6	6	6	4
Visual Reasoning										
Item #	1	2	3	4	5	6	7	8	9	10
Total Score	1	3	1	0	1	1	0	0	0	0
P value	0.14	0.43	0.14	0.00	0.14	0.14	0.00	0.00	0.00	0.00

Note. Total Score = The total score of all participants combined. P value = The proportion (or percentage) passing (this is only provided for the items with a binary correct or incorrect answer). Orange = Item difficulty index between 0.4 and 0.6; Green = <0.4; Red = >0.6

Table 9

Item Difficulty level for Scalable Items – Visual Learning & Verbal Functions

Visual Learning & Verbal Functions												
Sentence Repetition												
Item #	1	2	3	4	5	6	7	8	9	10	11	12
Total Score	6	6	3	2	5	3	1	4	2	2	4	0
P value	0.86	0.86	0.43	0.29	0.71	0.43	0.14	0.57	0.29	0.29	0.57	0.00
Verbal Reasoning												
Item #	1	2	3	4	5	6	7	8	9	10	11	12
Total Score	7	7	7	6	0	6	6	6	2	1	6	0
P value	1.00	1.00	1.00	0.86	0.00	0.86	0.86	0.86	0.29	0.14	0.86	0.00

Note. Total Score = The total score of all participants combined. P value = The proportion (or percentage) passing (this is only provided for the items with a binary correct or incorrect answer). Orange = Item difficulty index between 0.4 and 0.6; Green = <0.4; Red = >0.6

CHAPTER 4: DISCUSSION

4.1. Summary of Results and Test Development

This study used an empirical exploratory method for establishing the acceptability and feasibility of a novel cognitive test set to address the following questions:

1) Does this measure include items appropriate for and suited to this population?
2) Therefore, is it sufficiently engaging?
3) Are the language and stimuli used at the right developmental level?
4) Do the tests yield an acceptable range of scores with no ceiling or floor effects?

Overall, feedback was positive, and participants engaged well with the tasks. The test results showed that most of the subtests were appropriate for most of the participants with an intellectual disability. However, some of the tests proved challenging and showed floor effects. Most notable were 'Visual Reasoning', 'Motor Programming', and the 'Shopping Task'. All of these are tests of executive function, suggesting that executive function difficulties are a prominent feature of people with an intellectual disability, even before any decline in functions due to neurodegeneration. This finding may reflect that differences in frontal lobe functioning are noted in people with an intellectual disability, regardless of aetiology (Ball et al., 2008; Cornish et al., 2009; Mervis & John, 2010). Therefore, the view that executive functions decline first in dementia may reflect baseline functioning.

Another option would be to include an adaptation of the ToL or scrambled boxes subtests from the CEFA described in the literature review. The scrambled boxes task may be preferable due to the need for a specific apparatus for the Tower of London test, which Ball et al. (2008) found was sensitive to detecting the cognitive changes associated with personality and behaviour changes reported by relatives or carers in the preclinical stages of AD. Willner et al. (2010) note that these working memory tests were presented visually (in comparison to the other tests presented verbally with smaller effect sizes) in the Ball et al. (2008) study.

Therefore, they may involve the temporal cortex, which is thought to be vulnerable to the impairment of visuospatial processing that occurs early in the course of AD (e.g., Schmidtke & Olbrich, 2007; Swainson et al., 2001), and how far these tests precisely assess executive function is still to be debated. The BADS-ID (Webb et al., 2020) could also be reviewed for other suitable tests. Whichever executive function tests are chosen, it is essential to remember that executive functions are not a unitary component. Combined tests should cover the breadth of executive functions to account for possible strengths and weaknesses.

Some tests were too easy and showed ceiling effects: 'Circle Search', 'Picture Naming', 'Picture Recognition', and 'Eight Detection'. This finding was reflected in the participant's feedback on the semi-structured interview. Tests of this type tend to be too easy for people who have shown typical development but might be potentially valuable tests in zones for sensitivity to decline (e.g., attention, language, memory, and processing speed).

The test instructions required a certain level of verbal comprehension and sometimes felt too long, requiring some adaptation and repetition for those with less verbal ability. Based on these results and the literature review findings, recommendations for adaptations to the test battery and subtests are provided (phases 8 and 9 of the 15-stage test development process). Also, according to the criteria suggested by Urbina (2014), some minor changes to item order are suggested. These changes would allow the creation of test discontinuation rules.

A general recommendation for scoring would be to include instructions to the examiners that for non-verbal tests, there should be a choice of response strategy (in some cases, for example, pointing out the answer) but that for spoken answers, the quality of the verbal response ought not to be considered in grading the answer (for example, allowing "cut" instead of "cutting" for 'Verbal-Reasoning' item 11).

4.1.1. Motor & Language Functions

Orientation Subtotal A + B

No verbal feedback was given relating to this subtest. People generally scored high on Orientation Part A. The scoring for 'Orientation Part B' would benefit from adaptation, requiring only one complete sentence for each question to gain the maximum points, with a partial or limited (but correct) response gaining one point. As mentioned, people without neurodegeneration would be expected to score highly on orientation tests. It may be that the 'orientation to place' items on 'Orientation Part A' and 'Orientation Part B' would be more challenging for people with dementia or a more severe intellectual disability, given that they require intact recent memory. Given that this sample mainly included participants with mild intellectual disability, further piloting may show that these tests are affected by the severity of intellectual disability, as found in the literature review. These subtests were not designed to be scaled, though it might be possible to include more complex questions towards the end to increase their difficulty if desired.

Smell Detection + Recognition

People generally responded favourably to this subtest, and one person named it a test they found interesting in the semi-structured interview. However, despite the existing literature suggesting olfactory assessment may have utility in the early detection of dementia (albeit in Down syndrome), scores were generally low. It is difficult to ascertain if the low scores were merely due to issues with the test stimuli or pre-existing olfactory deficits. For future piloting, the fragrances would benefit from being reviewed to ensure a true likeness to the target fragrance, especially items three (vanilla) and four (shoe polish), since no one answered these items correctly. The participants were likely unfamiliar with these smells, especially since shoe polish was quite prominent. This change would allow the five most suitable of the ten fragrances to be retained or with additions. From this selection, the most suitable smells to retain could be chocolate, mint, coffee (due to some participants answering correctly), and possibly eucalyptus ('vapour rub') due to having a prominent smell. Eucalyptus would need to be piloted to see if participants can name this smell, as it was only included in the recognition task.

The participants' plausible responses can be included in the range of response options. Smell recognition may need to be removed since most participants answered ‘yes’ to all items.

Verbal Expression

No feedback was available as this was an observational test whereby the researcher judged the participants’ verbal expressive skills. The purpose is to monitor change over time and inform the interpretation of scores on the other tests. Most participants scored reasonably well on this subtest, indicating good verbal ability. This test may show some variation in people with Down syndrome, given that difficulties with verbal expression are common in this cognitive phenotype (Das et al.,1995; Iacono et al., 2010) and in people with a more moderate to severe intellectual disability.

Verbal Comprehension A + B

No specific verbal feedback was given for these subtests; generally, participants scored highly, with an acceptable range of scores. This finding is likely due to the sample comprising mainly people with mild intellectual disability and good verbal comprehension skills (per the inclusion criteria). Again, this is likely due to the sample showing no signs of cognitive deterioration. ‘Verbal Comprehension Part B’ is split into three sections, firstly ‘Pointing’ (items one to seven), secondly ‘Instructions’ (items eight to 12), and thirdly ‘Meanings’ (items 13 to 18). Regarding difficulty, ‘Pointing’ seemed appropriately scaled, with items six and seven being more difficult than items one to five. For ‘Instructions,’ item 11 would benefit from being moved to item 12. For the ‘Meanings’ section, most participants could answer these easily, so they would benefit from more challenging questions added towards the end if this subtest is intended to be scaled.

Motor Function A + B

No specific verbal feedback was given for these subtests, and participants generally scored highly. However, some participants struggled with ‘Motor Function B’ due to motor impairments.

A recommendation would be to remove ‘Motor Function Part B’ since the ‘Motor Programming’ subtest appears suitable for assessing both motor and executive functions.

Motor Programming

There was no specific verbal feedback relating to this subtest, though it proved quite challenging for participants, presumably due to the additional executive function requirement. The participant with significant motor difficulties could not score on this test. For the ‘knock-tap’ style items (three and four), a recommended revision would be to change the opposition task (item three) to a simple copy item to allow the participants to familiarise themselves with the task and ensure a person’s ability to complete the task. Then, follow this with the inhibition task (item four), as the executive function component, and a comparison between the two. Therefore, a person with motor difficulties would expect to score poorly on both. However, low scores on only the second item would indicate difficulties with executive function, similar to the score on the Purdue Pegboard for using both hands, reported by Hom et al. (2021). Given the acceptable range of scores, this, and the Luria-style sequence of fist-edge-palm (item two), so-called ‘bedside’ tests of executive function, may be suitable tests of executive function in people with an intellectual disability without motor difficulties.

Praxis

No verbal feedback was given relating to this subtest. This test is broken down into three parts: ‘Gestures’ (intransitive; items one to five), ‘Object Use’ (transitive; items six to 10), and ‘Buccofacial’ (oro-motor; items 11 to 15). Most participants completed this task efficiently except for item eight (scissors). A recommendation would be to move this item to the end of the ‘Object Use’ section and include some more challenging items towards the end of each subsection if aiming to scale this test. However, a test of this type is also easy for people who have shown typical development but are thought to be sensitive to dementia.

For example, the BPT was found to be sensitive to changes in neurodegeneration in Down syndrome (Sano et al., 2005; Wallace et al., 2021) but not to early-stage decline (Wallace et al., 2021), so it may be appropriate to leave this test as it is.

4.1.2. Verbal Learning & Visual Functions

Word List Immediate, Learning, Delayed Recall + Recognition

One participant listed this as a test they found interesting. However, one person listed it as a test they found too complicated and suggested we use fewer words. This test was unsuitable for people with verbal expression difficulties. 'Word List Immediate' gave an acceptable range of scores, and participants tended to score well on 'Delayed Recall' and 'Recognition,' which is to be expected in a sample of people with no suggestion of neurodegeneration. This test will likely discriminate well between people with and without suspected dementia, given the positive findings for the BSRT in the literature review.

Circle Search

No verbal feedback was directed at this test. Participants were generally observed to enjoy this test and complete it easily, evidenced by a considerable ceiling effect. Suggested adaptations would be to increase the number of items (not decrease their size due to possible visual impairments) on the page or lower the allocated time. Though this may discriminate against those with a motor impairment, it may also be a good measurement of the extent of impairment. Again, tests of this type are quickly completed by people who have shown typical development but were found to discriminate between those with and without dementia and early-stage dementia in the literature review.

Angle Judgement

This test was notably tricky for participants, who often got only one of the target lines correct. One participant listed this test as one they found too challenging.

Although this item was not intended to be scaled, given the difficulty many people had, it was often discontinued to avoid unnecessary testing. A recommendation would be to start with fewer line options to choose from and increase the number as the item difficulty increases; this would allow the item to be scaled and encourage a broader range of scores.

Visual Reasoning

The 'Visual Reasoning' test had considerable floor effects and was discontinued early for most participants after three consecutive zeros. One person said they found this test interesting, and another found it easy. However, one participant said it was confusing, and two said it was too hard. One participant recommended using playing cards, similar to a game of 'snap', and one person said to use fewer shapes. Difficulties on this task compared to the typically developing population may be due to differences in education between mainstream and special education provisions or reflect difficulties with abstract reasoning that are known to affect people with an intellectual disability (APA, 2013; Hassiotis et al., 2012). In terms of recommendations, this test could be removed or altered to offer fewer response options and increase the number as the item difficulty increases or to simplify each item. However, this subtest may still be too unfamiliar and challenging, and based on participant feedback, it may be better to choose an alternative. Another option would be to use a simplified version of the Brixton and Hayling Test (Burgess & Shallice, 1997), notably the Spatial Anticipation Test, which does not require a verbal response and is considered suitable for a wide range of functioning.

Shopping List Map 1 + 2

The 'Shopping Task' was listed as a test that was too hard by one participant, and one person felt (correctly) that the pictures and text needed to be enlarged for people with visual impairments. Nearly all participants did not complete this test correctly, and often, Map 2 was not administered due to Map 1 proving too difficult. This finding may be because of the use of abstract concepts, complex instructions, unfamiliarity with this kind of 'maze puzzle,' and weakness in task sequencing.

The scoring does not appear to reflect participants' difficulty on this test since most points could be gained despite not being completed according to the instructions.

The design of this test can be improved by removing or simplifying the rules in Map 1, requiring participants to collect only the items on the shopping list in order of their choosing, representing an assessment of planning. Then, Map 2 introduces rules requiring the participants to do certain things to acquire full marks (e.g., only use the white paths once and visit the assistant), representing the executive function components of rule compliance, inhibition, and shifting. The difference in scores between the two conditions would indicate the additional load from the executive function components. It would be beneficial to keep the written instructions for participants visible throughout the task to avoid additional strain on working memory since this is seen as a related but distinct function from executive functions (The BPS, 2015c).

Cat-Dog Inhibition

This test was cited as being too hard by two participants and given that it is a test of executive function, this is not surprising. However, similar to what was found in the literature review (Bevins & Hurse., 2014), most participants could easily complete the ‘Cats and Dogs’ subtest, producing a ceiling effect for almost half the participants. This finding suggests it may be a suitable test of executive function for people with a more severe level of intellectual disability and neurodegeneration. Including a primary ‘congruent’ naming trial would be useful to measure processing speed and compare the time taken on the two tasks. The difference between the two times is the ‘Stroop Effect’ (cost of inhibiting the irrelevant task; Stroop, 1935), along with incongruent error rates (Balota et al., 2010; Fine et al., 2008; Hutchison et al., 2010). However, given the evidence provided by the factor analysis completed by Hom et al. (2021), whereby they did not find this task to load onto the factor of executive function, factor analysis of future revisions would need to be completed to assess whether this task truly measures executive function.

4.1.3. Visual Learning & Verbal Functions

Matchsticks Copy, Immediate + Delayed Recall

The 'Matchsticks' subtests were listed as being too hard by two participants due to being 'fiddly' and difficult for people with visual impairment, suggesting using fewer matchsticks. In terms of scores, this was variable. This finding may reflect the variation in visuospatial abilities within differing aetiologies of intellectual disabilities (Lott & Dierssen, 2010). Some participants could easily complete the copy task, whereas others produced wholly inaccurate copies. It is unclear whether this is due to visual impairment or visual-perceptual difficulties. However, it produced a good range of scores. A recommendation would be to use larger matchsticks or small pencils to allow participants to manipulate them more easily.

Picture Naming + Recognition

Verbal feedback told us that participants often found these subtests too easy (n = 2) but also interesting (n = 3) and suggested the use of "harder pictures". This finding was reflected in the scores, with most people scoring highly. Therefore, this test requires adding or substituting more challenging items, for example, lesser-known animals, such as a walrus or more obscure items, such as a thimble. This alteration would make it similar to the BNT (Kaplan et al., 1976), which showed good sensitivity for distinguishing people with an intellectual disability and MCI or dementia from those without in the literature review. 'Picture Naming' would not be suitable to scale since the recognition component requires all items to have been presented in the naming task.

Eight Detection

Two participants said they found this test interesting. However, this test showed a ceiling effect for most participants but had to be discontinued for one participant who tapped for every number. A suggestion to increase the task's difficulty could be to increase the test length or the inter-stimulus interval (ISI) to make it more boring and, therefore, vulnerable to a lapse in attention. No auditory attention tasks were found in the review.

However, a selective attention test is likely to discriminate between people with and without neurodegeneration, similar to people from the typically developing population.

Sentence Repetition

No specific feedback was given for this subtest, and although it did not show any floor effects, likely due to its inherent scaling (each item increased in word length), it proved challenging for all participants. This finding was likely due to difficulties with verbal expression (vocabulary and articulation), as well as in the short-term stores (STS), not specific to only people with Down syndrome (Das & Mishra, 1995), but for people with an intellectual disability more widely. This test may need replacing for it to have discriminative power. It will not be suitable for people with low verbal ability. If this test is retained and simplified, a recommendation would be to reduce the number of items for brevity.

Verbal Reasoning

No specific feedback was given for this test, and most participants scored well (8/12). However, a non-normal distribution was observed, indicating it did not provide a good range of scores. Participants were noted to answer some questions easily, and no one could answer others (e.g., item five). Recommendations would be to place item five at the end and introduce some questions of moderate difficulty. Item 11 will require “cut” instead of “cutting” being added to the list of acceptable answers.

Word Generation

This verbal fluency task was easily completed by all participants, as expected in a sample of participants with no neurodegeneration, similar to Cooper et al. (2016), who found it simple to complete and detect change over time. However, one recommendation would be to remove one of the items since animal/ food naming is a semantic verbal fluency task, providing unnecessary repetition. An option would be to include a phonemic fluency item, similar to the COWAT (Benton et al., 1983), since phonemic verbal fluency tasks represent a more significant executive function load than semantic tasks.

However, as mentioned earlier, phonemic fluency items are likely more difficult for people with an intellectual disability. The rules usually applied to these tests (e.g., no names of people or places) should be removed to reduce further executive function load. Including uncorrected repetitions in the scoring would also assess working memory and inhibition.

4.2. Clinical Implications

The findings of this study have implications for developing assessments of dementia in people with an intellectual disability. The range of scores (including floor and ceiling effects) within a young sample of people with an intellectual disability (but not suspected of neurodegenerative decline) indicates the wide range of functioning that must be provided for within this population. This finding provides support for critics of IQ, such as Bertelli et al. (2017), who feel cognitive capacity may be better seen as a profile (rather than unidimensional 'IQ') since people with the same IQ level will have different cognitive strengths and weaknesses, due to varying factors involved in the aetiology of each condition. Limitations of functioning and many biopsychosocial factors associated with intellectual disability are highly correlated with difficulties in specific cognitive functions (rather than with overall IQ).

This range of ability makes any assessment of cognitive decline within this population problematic unless compared to a person's own baseline. This finding further supports the need to assess people with an intellectual disability at several time points to establish a baseline point from which to monitor decline, which is preferable to comparison against a normative sample (as proposed by Rowe et al., 2006). Given that executive functions were challenging to assess in a sample of people with no signs of neurodegeneration (and non-Down syndrome), these findings support the notion that pre-existing difficulties in executive function exist for people with an intellectual disability, which may account for reports of early deficits in executive function in neurodegenerative decline.

4.3. Critical Review

One of the strengths of this study was that qualitative feedback from participants was sought to inform the development of the test set (alongside quantitative data). Combining different methods (with complementary strengths and weaknesses) produced a range of results that could be 'triangulated,' providing strong evidence. However, going forward, it would be preferable to form a focus group of people with intellectual disabilities and clinicians specialising in the assessment of intellectual disabilities (discussed below).

Another strength is the range of demographics of the participants in the sample. Piloting with participants for whom English is an additional language would be necessary for future research. Another strength of the study is the efforts made to ensure meaningful and ethical involvement in the study by involving relatives, carers, or trusted people (where possible) in both the recruitment and consenting processes.

The included sample is biased towards people who could consent to participate in the research, such as having a less severe intellectual disability and the required verbal abilities. This finding points to the need to develop a test battery specifically for people with a more severe intellectual disability and for those who are non-verbal or motor impaired. However, this method was the most practical in terms of the aims of this study.

The small sample size limits the ability to conclude the distribution of data and item difficulty levels. This was further compounded by the sample mainly including people with a milder level of intellectual disability, which is the level that has the highest rates of unexplained aetiology (Patel et al., 2020) and where heterogeneity in cognitive profiles is likely to be higher. However, these were exploratory analyses and given that the study aimed to assess preliminary acceptability and feasibility, these results have already provided helpful direction.

As noted previously, people with an intellectual disability can be more susceptible to suggestibility and may change his or her answers to questions when provided with negative feedback (Clare & Gudjonsson, 1993; Everington & Fulero, 1999), which Beail (2002) has suggested is linked to poorer memory. It cannot be ruled out that the researcher unwittingly provided verbal or non-verbal cues to performance that may have affected participants' answers on test items. Similarly, despite best efforts to maximise the participants' engagement and attention during the pilot sessions, the testing length was likely tiring. Though all participants refused a break, it cannot be ruled out that test performance was affected by these factors. Another limitation is that health problems or mental health difficulties were not assertively screened for, relying on clinicians' and informant views at the time of recruitment. Therefore, it cannot be ruled out that participants may have had an undiagnosed but treatable condition that affected their scores. Lastly, there was significant reliance on verbal ability to understand the instructions. Therefore, it cannot be ruled out that poor comprehension of the instructions may have affected the participants' performance.

4.4. Future Research

A suggested adaptation would be to review the materials with the help of a focus group or, more specifically, to use the Delphi Panel method (Jones & Hunter, 1995). The Delphi method is a systematic technique used to establish consensus on issues. This method would allow test items and instructions to be reviewed before piloting and afford meaningful participation in the test's development at every stage (Arnstein, 1969).

There was some difficulty in recruiting a good number of participants within the timeframe available. The feedback received from services was that the eligibility criteria excluded many of the people on their caseloads (e.g., due to severity of intellectual disability, presence of severe and enduring mental health difficulties, age, or requirement of sufficient verbal ability to ensure capacity to consent).

Therefore, a second recommendation would be to seek a non-clinical community-dwelling sample of participants for future research studies (instead of a clinical sample of participants currently using services). One way this could be achieved would be to approach local day centres, community centres, and supported living services, as well as local and national charities and voluntary organisations, for their assistance in recruitment and to allow advertisement directly to potential participants.

Other reasons for not wanting to participate included how people did not feel incentivised enough or could not spare the time due to the carers' prior commitments or the potential participants' routine daily activities. A higher token of appreciation may have provided further incentive, though this would need to be carefully balanced so as not to become coercion (Largent & Lynch, 2017). However, considering the minimum hourly wage, it is also essential to appropriately compensate people for their time and inconvenience.

Testing with a broader sample of participants, including those with a more severe intellectual disability, less verbal ability, and differing aetiologies of intellectual disability, would be another recommendation (following adaptation). Piloting future adaptations with people with Down syndrome specifically would be an essential step, considering they are thought to be at risk of early neuropathological changes and executive dysfunction as an early marker of dementia. Similarly, collecting longitudinal data from people with Down syndrome and non-Down syndrome would be necessary to understand differences and trajectories further. A larger sample size will be needed for further data distribution analysis and detailed item analysis before final decisions are made, given the central limit theorem (whereby as the sample size increases, the distribution continually approaches a normal distribution).

In order to assess discriminative power and predictive validity, the test set needs to be piloted with people with an intellectual disability and dementia and compared to participants with an intellectual disability without dementia.

This needs to consider the cognitive profiles of different dementias, that is, known-group studies of predictive validity. Longitudinal data would be required as evidence for sensitivity to change and to establish clinically relevant cut-offs. Note that it is preferable not to repeat tests of executive function. However, there is literature to support this method (e.g., Griffith et al., 1999), which may be the only way to evidence decline in this function.

Following the 15-stage process of test development suggested by Fenn et al. (2020), a second revision of the test set should be devised (phase eight) and piloted (phase nine), and validity and reliability determined (phase ten), followed by exploratory factor analysis (phase 11) and creation of a third draft (phase 12). This process should then be followed by a confirmatory factor analysis (phase 13) and the creation of the final test set (phase 14) and manual (phase 15). The manual should include assistance with the interpretation of non-typical patterns of performance. Exploring test concurrent validity could involve comparisons with previous assessment data routinely collected by services or compared to an existing test set with normative data administered simultaneously. Once a test set is agreed upon, this could be validated in different cultures and languages.

4.5. Conclusions

From the literature review, most instruments have been developed for and tested with people with Down syndrome, with variable success. It remains to be seen how suitable some of these instruments are for assessing people with an intellectual disability from other causes and what the trajectory of neurodegeneration is in this heterogeneous group.

The range of scores across the subtests in the empirical study highlights the variance in abilities in people with intellectual disabilities within the mild to moderate level.

Furthermore, the floor effects on specific subtests, especially those designed to measure executive function, confirm the difficulty in designing suitable tests for people with pre-existing impairments sensitive enough to detect change due to neurodegeneration in this population. Overall, this novel test set appeared to be both acceptable and feasible for use with people with an intellectual disability. However, several revisions are needed to make them adaptable for people with a range of severity of intellectual disability and for people with less verbal ability. It is essential to continue to work towards the development of a valid and reliable measure that can be used to assess for cognitive decline in people with an intellectual disability in order to support early detection and diagnosis and provide appropriate services, treatments and support for families and carers. This aim aligns with England's Dementia Strategy (DoH, 2009), which seeks to promote better awareness, earlier diagnosis and intervention, and a higher quality of care for individuals, their families, and carers.

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APPENDICES

Appendix A

List of Abbreviations

AAIDD = The American Association on Intellectual and Developmental Disabilities

ACE = Addenbrookes Cognitive Examination

ACTB = Arizona Cognitive Test Battery

AD = Alzheimer's Disease

ADLs = Activities of daily living

ADVM = Auditory delayed verbal memory

AMT = Autobiographical Memory Test

APA = American Psychological Association

ApoE = Apolipoprotein E

APP = Amyloid precursor protein

ASL or landmark = Allocentric spatial learning or landmark

ASM = Auditory sequential memory

Aβ = Amyloid-beta

BADS = Behavioural Assessment of the Dysexecutive Syndrome

BADS-ID = Dysexecutive Syndrome for Intellectual Disabilities

BBDT-VMI = Beery Buktenica Developmental Test of Visual-Motor Integration

BD = Block Design

BNT = The Boston Naming Test

BP = Block Patterns - Hiskey-Nebraska Test of Learning Aptitude subtest

BPS = The British Psychological Society

BPT = The Brief Praxis Test

BPVS = British Picture Vocabulary Scale

BSRT = The Busckke Selective Reminding Test

BT-ID = Barcelona Test - Intellectual Disability

BTS = Block tapping span

bvFTD = behavioural variant of frontotemporal lobar dementia

CaD = Cats and Dogs task

CAMCOG = Cambridge Cognition Examination

CAMCOG-DS = Cambridge Cognitive Examination adapted for individuals with Down Syndrome

CANTAB = Cambridge Neuropsychological Test Automated Battery

CEFA = Cambridge Executive Functioning Assessment

CFT = Category Fluency Test

Co = Coding subtest of the WISC-R

CoD = Copy of drawings

CODB/ NAID = Crayton and Oliver Dementia Battery/ Neuropsychological Assessment of Dementia in Intellectual Disabilities

COVID-19 = Coronavirus disease

COWAT = The Controlled Oral Word Association Test (also known as FAS)

CRT = The Cued Recall Test

CS = Cognitively stable

CT = Cancellation task

CTT = The Colour Trails Test

CVD = Cerebrovascular disease

CVLT-C = California Verbal Learning Test - Children's Version

DoH = Department of Health

DLD = Dementia Questionnaire for People with Learning Disabilities

DMTS = Delayed match-to-sample

DNMP = Spatial delayed non-match-to-position

DNMS = Object delayed non-match-to-sample

DRecog = Delayed Recognition

DRecall = Delayed Recall

DS = Down syndrome

DSpan = Digit Span

DSDS = The Down Syndrome Dementia Scale

DSM = Diagnostic and Statistical Manual of Mental Disorders

DSMSE = Down Syndrome Mental State Examination

DSQID = The Dementia Screening Questionnaire for Individuals with Intellectual Disabilities

DVM = Delayed visual memory

EF = Executive function

EISC = Ethics and Integrity Sub-Committee

EMS = Evaluation of Mental Status

EOWPVT/ EOWPVT-R = Expressive One-Word Picture Vocabulary Test/ Expressive One-Word Picture Vocabulary Test-Revised

FOME/ mFOME = The Fuld Object-Memory Evaluation/ Modified - Fuld Object Memory Evaluation

FS = Finger Sequencing

FT = Finger Tapping subtest from the Halstead-Reitan Battery
FSIQ = Full-Scale IQ
FXS = Fragile X syndrome
GA = Gait Assessment (Timed Get Up and Go Test)
HC = Healthy controls
HOM = Hidden Object Memory Test
HRA = Health Research Authority
ICC = Intraclass Correlation Coefficient
ICD = International Classification of Diseases
ID = Intellectual disability/ disabilities
IM = Immediate Memory
IQ = Intelligent Quotient
IQCODE = Informant Questionnaire on Cognitive Decline in the Elderly
IQR = Interquartile range
JLO = Judgment of Line Orientation
K-BIT = Kaufman Brief Intelligence Test
KBNA = Kaplan Baycrest Neurocognitive Assessment
LBD = Lewy body dementia
LM = Logical Memory subtest of the WMS-R
LTM = Long-term memory
LTR = Long-term recall
LTS = Long-term stores/ storage
MCA = Mental Capacity Act
M-CFT = McCarthy Scales of Children's Abilities - Category Fluency Test
MCI = Mild Cognitive Impairment
MfO = Memory for objects from the NAID
mMMSE-DS = Modified Mini Mental Status Evaluation—Down Syndrome
MMSE = Mini Mental Status Evaluation
MSCA = McCarthy Scales of Children's Abilities
NA = Not applicable
NART = National Adult Reading Test
NDT = New Dot Test
NEPSY = A Developmental NEuroPSYchological Assessment
NHS = National Health Service
NHS-REC = NHS Research Ethics Committee
NICE = National Institute for Health and Care Excellence

NINCDS-ADRDA = National Institute of Neurological and Communicative Diseases and Stroke/ Alzheimer's Disease and Related Disorders Association

NR = Not reported

O = Orientation

ODL = Object discrimination learning

OI = Object identification

oID = Intellectual disability from other causes than Down syndrome

O-MMSE = Orientation subtest from the MMSE

Opp = Opposites subtest of the McCarthy Scales of Children's Abilities

PAL = Paired-associate learning task

PHE = Public Health England

PN = Picture Naming

PNFA = progressive non-fluent aphasia

PPT = Purdue Pegboard Test

PPVT/ PPVT-R = Peabody Picture Vocabulary Test/ Peabody Picture Vocabulary Test-Revised

PPVT-R/ PPVT-III = Peabody Picture Vocabulary Test-Revised/ 3rd Edition

PRISMA = Preferred Reporting Items for Systematic Reviews and Meta-analyses

PRMT/ r-PRMT = Picture Recognition Memory Test/ revised Picture Recognition Memory Test

people with an intellectual disability = people with an intellectual disability

QoL = Quality of Life

RaB = ‘Remembering a belonging' subtest of the RBMT-C

RADD/ RADD-2 = The Rapid Assessment of Developmental Disabilities/Second Edition

RAVLT = Rey Auditory Verbal Learning Test

RBMT-C = Rivermead Behavioural Memory Test for Children

RCT = Randomised Controlled Trial

RL = Reversal learning

SB = Scrambled Boxes

SBIS = Stanford-Binet Intelligence Scale

SD = standard deviation

SIB = Severe Impairment Battery

SPSS = Statistical Package for the Social Sciences

SR = Spatial Reversal

Srep = Sentence repetition

STM = Short-term memory

STS = Short-term stores/ storage

StoryRT = Story Recall Test (adapted from the RBMT-C)

SVDL = Simple visual discrimination learning

TD = Typical development or typically developing

TO = Temporal Orientation

ToL = Tower of London

TOL^{DX} = Tower of London-Drexel University: 2nd Edition

TSI = Test for Severe Impairment

UEL = University of East London

UK = United Kingdom

UKRI = United Kingdom Research and Innovation

UPSIT = The University of Pennsylvania Smell Identification Test

USA = United States of America

VABS = Vineland Adaptive Behavior Scales

VaD = vascular dementia

VAT = Visual Association Test

VC = Verbal comprehension

VF = Verbal Fluency

VisMT = Visual Memory Test

VMI = Visual Motor Integration

VR = Visual Representation subtest of the WMS-R

VT = Vocabulary Test

WAIS/ WAIS-R/ WAIS-III = Wechsler Adult Intelligence Scale/ Revised/ 3rd Edition/ 4^{th} Edition

WASI = Wechsler Abbreviated Scale of Intelligence

WCFST = Weigl Colour-Form Sort Test

WG-MTB = Working Group Memory Test Battery

WG-O = Working Group's Orientation Test

WISC/ WISC-R = Wechsler Intelligence Scale for Children/Revised

WM = working memory

WS = Williams syndrome

Appendix B

Study and Sample Characteristics

Table B 1

Study Characteristics

Author, Year	Country	Type of study	Follow-up	Sample size (N)	Tests included	General findings
Ball et al., 2008	UK	Cross-sectional	NA	103	CAMDEX-DS; BPVS II; CAMCOG-DS (IM, DRecog, DRecall); RaB (from RBMT-C); MfO, MfS subtests from the CODB/ NAID; CEFA battery	DS-no-AD scored ↑ on all measures vs. DS-AD. Informant reported personality/ behaviour changes predicted performance on EF and 'executive memory' tests of the CEFA for DS-no-AD, but not on episodic memory tests. Informant-reported memory changes associated with ↓ on delayed recall task only. Evidence for a specific impairment in frontal lobe functioning in preclinical stages of AD in DS.
Benejam et al., 2015	Spain	Cross-sectional	NA	DS-no-AD = 75 DS-AD = 15	Spanish version of the m-CRT; K-BIT	DS-AD scored ↓ on free recall and total score and committed ↑ intrusion errors vs. DS-no-AD. Age main factor associated with ↓ m-CRT scores.
Benejam et al., 2020	Spain	Cross-sectional	NA	CAMCOG-DS sample = 343 m-CRT sample = 271	Spanish versions of the CAMDEX-DS; CAMCOG-DS; mCRT	Progressive ↓ on CAMCOG-DS and m-CRT > age 40, especially for moderate ID.
Bevins & Hurse, 2014	UK	Cross-sectional	NA	28	CaD, WCFST, and VF from the CEFA battery; BVPS-II; MfO from NAID; DLD	NA
Brugge et al., 1992	USA	Cross-sectional	NA	DS = 17 Non-DS = 7	WAIS-R; LM and VR subtests of the WMS-R; CVLT-C; BNT; PPVT-R; MfS and MfO from SBIS-IV; COWAT; BBDT-VMI; Opp subtest from MSCA; FT from Halstead-Reitan Battery	Memory-impaired-DS group showed ↓ in performance on various cognitive tests with advancing age vs. non-memory-impaired DS, and oID controls, who showed no evidence of decline with age.

Author, Year	Country	Type of study	Follow-up	Sample size (N)	Tests included	General findings
Carr, 2003	UK	Longitudinal	Age 30 and 35 years	Overall = 75 age 30 = 38 age 35 = 37	LIPS; BPVS; HBS; RBMT-C; CODB/ NAID	Little change found on all tests over the f/u period. ↓ on all subtests of RBMT-C and CODB/ NAID, but only orientation (and picture memory on the CODB/ NAID) was significant.
Cooper et al., 2016	UK	RCT (longitudinal comparison)	1 year	21 (13 completed full year)	MfO from NAID; CT; PR memory from CANTAB; CaD; ToL; CRT; CFT; StoryRT (adapted from RBMT-C); ABS	NA
Devenny et al., 1992	USA	Longitudinal	3-5 years mean=41 years	DS = 28 oID = 18	IBR-EMS; BSRT; VisMT	No significant changes in test scores between baseline and f/u up to 5 years later for any of the groups. All groups showed ↑ in performance tasks from 1st to 2nd testing on memory. No functional deterioration or age-related memory decline in adults with DS.
Devenny et al., 1996	USA	Longitudinal	Yearly f/u for 6 years	DS = 91 oID = 64	IBR-EMS; BSRT; VisMT; BD, DSpan, Co subtests of WISC-R	Repeat testing of verbal LTM = younger-DS small ↑ in scores vs. older-DS small ↓ in scores. Overall performance on verbal LTM and a speeded psychomotor task was poorer in groups aged 5O+ years (DS and oID). 4/91 met criteria for AD but also for reversible causes of decline (e.g., hypothyroidism). Suggests differences in scores due to normal but precocious ageing in DS.
Devenny et al., 2000	USA	Longitudinal	4+ years apart	oID= 40 DS Healthy = 44 ?AD = 10 Early-Stage AD = 5 Middle-Stage AD = 7	WISC-R; CRT; SRT	Differences across the groups (healthy-DS, ?AD, early-stage-AD, and middle-stage-AD) indicating a pattern of cognitive decline starting with memory loss and significant ↓ on BD and Coding, and Object Assembly, Picture Completion, Arithmetic and Comprehension subtests, then ↓ on Information, Vocabulary and DSpan subtests of WISC-R.
Devenny et al., 2002	USA	Longitudinal	2 years+	oID= 66 DS-no AD = 75 DS-AD = 19	Adaptation of CRT	AD had ↓ total scores vs. no-AD. IQ level and age also negatively affected scores. Poor performance on CRT (adapted) was associated with early-stage-AD.

Author, Year	Country	Type of study	Follow-up	Sample size (N)	Tests included	General findings
Esteba-Castillo et al., 2022	Spain	Longitudinal	12-month intervals for 3 years	63 (final sample)	TOL^{DX}; WCFST; BT-ID; CAMCOG-DS; CAMDEX-DS; BRIEF	MCI group showed cognitive ↓ in several domains incl. stories delayed verbal memory) and pictures delayed visual memory) but especially EF (TOL^{DX} (Hit), abstraction, semantic VF and BRIEF informant-report measure). Model composed of Behavioural Regulation Index (BRI) on BRIEF, abstraction and delayed verbal memory best at predicting MCI vs. controls.
García-Alba et al., 2017	Spain	Cross-sectional	NA	63	K-BIT-2; ABS-RC:2; CAMDEX-DS; BT-ID; WCFST; BRIEF-P; TOL^{DX}	NA
García-Alba et al., 2019	Spain	Longitudinal	3 times over 3-year period	DS = 41 DS-AD = 13 DS-MNI = 14 DS-Control = 14	CAMDEX-DS; CAMCOG-DS; ADVM; WM; DVM; TO; BRIEF-P	DS-AD ↓ scores on all tests, especially in delayed visual memory and WM and ↓ scores vs. DS-MCI group in WM and ADVM tests. DS-MCI ↓ scores vs. DS-controls on CAMCOG-DS and DVM. ↓ CAMCOG-DS and DVM scores and impairment in domains TO, WM, and ADVM = MCI in DS. Findings suggest transition from MCI to AD in DS = worsening in global cognition, ↑ in TO and, especially, by a marked amnesic deficit.
Head et al., 2011	USA	Cross-sectional	NA	Study 1: DS-no-AD = 17 DS-AD = 17 HC = 11 AD controls = 12 Study 2: DS-AD = 52 DS-no-AD = 78	BPT; SIB; DMR	No association between scores on SIB and DMR and plasma Aβ levels.
Hoekman & Maaskant, 2002	The Nether-lands	Cross-sectional	NA	329	DMR; DMTS; CLD	NA
Hom et al., 2021	USA	Cross-sectional	NA	Overall = 144 CS = 103 DS-MCI = 41	BBDT-VMI; BD from WISC-IV; BNT; M-CFT; CaD; CRT; DSMSE; mMMSE-DS; RADD-2; PPT; RBMT; SRT; TBGAT	Analyses of 17 variables from 10 tests of cognition indicated performance reflected 3 underlying factors (language/ EF, memory, and visuomotor). All 3 domain composite scores significantly predicted DS-MCI status. Path modelling = language/EF composite score was most affected by MCI. Structural equation modelling = memory most affected, followed by visuomotor, and then language/ EF.
Krinsky-McHale, Devenny & Silverman, 2002	USA	Longitudinal	3 or more over 3 years	Overall = 85 DS-AD = 14 DS-no-AD = 71	DSDS; BSRT	Early-stage-AD showed significantly greater ↓ 3-years before diagnosis, particularly in LTM and retrieval, prior to other symptoms of dementia.

Author, Year	Country	Type of study	Follow-up	Sample size (N)	Tests included	General findings
Krinsky-McHale et al., 2008	USA	Longitudinal	3 or more over 3 years	Overall = 30 DS-AD = 5 DS-no-AD = 25	DSDS; CT; BSRT	DS progressive ↓ in selective attention approx. 2 years prior to MCI diagnosis.
Krinsky-McHale et al., 2020	USA	Longitudinal	≈14- to 22-month intervals	561	BSRT; mMMSE-DS; TSI; M-CFT; BD from the WISC-R; DLD; ABSI; RSMB; BBDT-VMI; NI; CUSPAD	Several measures showed ability to distinguish MCI and dementia.
McPaul et al., 2017	UK	Cross-sectional	NA	40	WAIS-IV; VAT; CAMCOG-DS	Participants scored well on VAT, irrespective of age, gender, or IQ.
Nelson et al., 2001	USA	Longitudinal	1 year	26	NBAP; DSDS; PPVT-III	Frontal lobe dysfunction = likely early AD in DS (levels of depression, 'indifference' and 'inappropriateness'). Correlated with neuropathological findings. Abnormal physical findings showed ↓ scores for memory on DSDS and receptive language on PPVT. ↓ in pragmatic language functioning came later, after depression and indifference.
Nelson et al., 2005	USA	Cross-sectional	NA	20	WAIS-III; DMR; ODL; RL; DNMP; DNMS	Age related to memory and learning. FSIQ best predictor of object memory (DNMS). Scores on DMR strongest predictor of reversal learning errors.
Nelson et al., 2007	USA	Longitudinal	1 year	34 at baseline (19 retested at one year f/u)	WAIS-III; NBAP; DMR; SVDL; RL; DNMS; ASL or landmark	NBAP found to be the strongest predictor of dementia status.
Oliver et al., 1998	UK	Longitudinal	0, 6, 13, 20, 25 and 50 months	57	BVPS; VABS; VMT adapted from MST; O-CAMCOG; MfO; MfS; MfP from the CODB/ NAID; ON; praxis	Severe cognitive deterioration = 28.3% of participants > 30 years, which ↑ with age and level of ID. ↓ in orientation, learning and memory came prior to aphasia, agnosia, and apraxia.
Palmer, 2006	USA	Cross-sectional	NA	Dementia = 10 (DS = 6; oID= 4) No-dementia = 12 (DS = 4; oID= 8)	CTT; BNT; COWAT; FOME; ESDCL	↓ scores in dementia group in areas consistent with AD in TD population (i.e., memory and learning). Dementia group ↓ scores on memory and learning (Fuld Total, Fuld Retention, and Ineffective Reminders scores), agnosia (BNT), semantic verbal fluency (Animal Naming), and attention/ EFs (CTT 1 and 2). Overall scores on ESDCL were ↓ for dementia group vs. control group.
Powell et al., 2014	USA	Cross-sectional	NA	oID= 10 DS-no-dementia = 10 DS-dementia = 10	BPT; SIB	↓ scores on BPT correlated with neuropathological findings (frontoparietal regions). DS-related reductions in white matter integrity in associated with ↓ cognition.

Author, Year	Country	Type of study	Follow-up	Sample size (N)	Tests included	General findings
Pyo et al., 2007	USA	Cross-sectional	NA	AD = 13 HC = 31	WG-MTB (mFOME; TSI; AMT; O); PRMT; NEPSY Comprehension Test; PPVT-III	AD group scored ↓ vs. control group on AMT and O. No difference on TSI total score or immediate and delayed memory subtest scores – may not be sensitive to detect early AD.
Pyo et al., 2009	USA	Cross-sectional	NA	AD = 16 HC = 35	Working Group's O Test	AD group scored ↓ vs. control group at baseline. Changes in scores over 1-year f/u not significantly different between the groups = poor sensitivity and specificity. Age and ID aetiology did not significantly affect scores.
Pyo et al., 2010	USA	Cross-sectional	NA	AD = 26 No-AD = 33 DS = 9 oID= 24 DS-AD = 15 oID-AD = 11	r-PRMT; mFOME; TSI; NEPSY	NA
Pyo et al., 2011	USA	Longitudinal	12 months	ID-AD = 21 ID-no-AD = 42	Working Group's AMT	AD group scored ↓ than control group at baseline. Controls with DS considerable ↓ at f/u, but not others.
Sacco et al., 2022	France	Retrospective record review	NA	DS = 194 AD = 12 Co-occurring conditions = 94 HC = 88	French versions of m-CRT and DSQID	Total recall scores significantly ↓ ($P < 0.0001$) in AD vs. controls. Scores ↓ with age and severity of ID and made ↑ intrusion errors.
Sano et al., 2005	USA	Cross-sectional and longitudinal ADa	Various	316	BPT; mFOME; NDT; O-MMSE; VT adapted from EOWPVT-R	Verbal learning, memory, and DR highly associated with dementia.
Sinai et al., 2016	UK	Cross-sectional	NA	49	ACTB battery; MfO and MfS from the NAID; ToL; VF; F-NT; GA; DLD; BRIEF; K-BIT-2	Some significant differences between dementia vs. no-dementia and younger- and older-DS.
Wallace et al., 2021	USA	Cross-sectional	NA	Overall = 100 No-dementia = 68 Possible dementia = 16	SIB; BPT; DLD	DLD total and subscales all ↓ for dementia group vs. possible and probable groups. Scores did not vary according to age, gender, or level of IQ.

Author, Year	Country	Type of study	Follow-up	Sample size (N)	Tests included	General findings
				Probable dementia = 16		
Walsh et al., 2015	USA	Cross-sectional	NA	114 (62% dementia)	RADD; DMR; BADLS; SIB; BPT	Dementia ↓ scores on all measures.

Note. ↑ = higher/ increase; ↓ = lower/ decline; < = less than; > = more than
?AD = Possible AD; ABS = Adaptive Behavior Scale; ABS-RC:2 = Adaptative Behavior Scale-Residential and Comunity-2nd edition; ABSI = American Association on Mental Deficiency - Adaptive Behavior Scale; AD = Dementia of the Alzheimer's Type Dementia; ADVM = Auditory delayed verbal memory; AMT = Autobiographical Memory Test; AoR = Acting on request; ApoE = Apolipoprotein E; ASL or landmark = Allocentric spatial learning or landmark; Aβ = Amyloid-beta; BADLS = Bristol Activities of Daily Living Scale; BBDT-VMI = Beery Buktenica Developmental Test of Visual-Motor Integration; BD = Block Design; BMT = Buschke Memory test; BNT = The Boston Naming Test; BP = Block Patterns; BPT = The Brief Praxis Test; BPVS = British Picture Vocabulary Scale; BRIEF = Behaviour Rating Inventory of Executive Function; BSRT = Busckke selective reminding test and modified versions; BT-ID = Barcelona Test-ID; CaD = Cats and Dogs; CAMCOG = Cambridge Cognition Examination; CAMCOG-DS = Cambridge Cognitive Examination adapted for individuals with Down Syndrome; CAMDEX = Cambridge Mental Disorders of the Elderly Examination; CAMDEX-DS = Cambridge Examination for Mental Disorders of Older People with Down Syndrome and Others with Intellectual Disabilities; CANTAB = Cambridge Neuropsychological Test Automated Battery; CEFA = Cambridge Executive Functioning Assessment; CFT = Category Fluency Test; CLD = Checklist with Symptoms of Dementia; COWAT = The Controlled Oral Word Association Test; CRT = The Cued Recall Test; CT = Cancellation task; CTT = The Colour Trails Test; CUSPAD = The Columbia University Scale for Psychopathology in Alzheimer's disease; CVLT-C = California Verbal Learning Test - Children's Version; DLD/ DMR = Dementia Questionnaire for People with Learning Disabilities; DM = Delayed memory; DMTS = Delayed match-to-sample; DNMP = Spatial delayed non-match-to-position; DNMS = Object delayed non-match-to-sample; Drecall = Delayed recall; Drecog = Delayed recognition; DS = Down syndrome; DSDS = Dementia scale for Down Syndrome; DSMSE = Down Syndrome Mental State Examination; DSpan = Digit span; DSQID = Dementia Screening Questionnaire for Individuals with Intellectual Disabilities; DVM = Delayed visual memory; EF = executive function; EMS = Evaluation of Mental Status; EOWPVT/ EOWPVT-R = Expressive One-Word Picture Vocabulary Test/ Expressive One-Word Picture Vocabulary Test-Revised; ESDCL = Early Signs of Dementia Checklist; f/u = follow-up; FOME = The Fuld Object-Memory Evaluation; FS = Finger Sequencing; FT = Finger Tapping subtest from the Halstead-Reitan Battery; FSIQ = Full-Scale IQ; GA = Gait Assessment; HBS = Handicaps, Behaviour and Skills Schedule; HC = Healthy controls; IBR-MSE = Mental State Examination from the New York Institute for Basic Research; IM = Immediate memory; IQ = Intelligent quotient; K-BIT = Kaufman Brief Intelligence Test; LT = Long-term; LM = Logical Memory subtest of the WMS-R; LTM = Long-term memory; m = months; M-CFT = McCarthy Category Fluency Test; MCI = Mild cognitive impairment; mCRT = Modified Cued Recall Test; MfO = Memory for objects; MfPT = Memory for pictures; MfS = Memory for sentences; mMMSE-DS = Modified Mini Mental Status Evaluation— Down Syndrome; MMSE = Mini Mental Status Evaluation/Modified Mini Mental Status Evaluation; MSCA = McCarthy Scales of Children's Abilities; NA = Not applicable; NBAP = Neuropsychology Behavior and Affect Profile; NEPSY = A Developmental NEuroPSYchological Assessment; NI = Neuropsychiatric Inventory; O = orientation; oID = Intellectual disability from other causes than DS; Opp = Opposites subtest from the MSCA; PPVT/ PPVT-R = Peabody Picture Vocabulary Test/ Peabody Picture Vocabulary Test-Revised; PRMT/r-PRMT = Picture Recognition Memory Test/ revised Picture Recognition Memory Test; RaB = remembering a belonging subtest from RBMT-C; RAVLT = Rey Auditory Verbal Learning Test; RBMT-C = Rivermead Behavioural Memory Test for Children; RL = Reversal learning; RSMB = Reiss Screen for Maladaptive Behaviour; SB = Scrambled Boxes; SBIS = Stanford-Binet Intelligence Scales; SIB = Severe Impairment Battery; SRT/mSRT = The Selective Reminding Test/ Modified - The Selective Reminding Test; STM = Short-term memory; SVDL = Simple visual discrimination learning; TBGAT = Tinetti Balance and Gait Assessment Tool; TD = Typically Developing; ToL = Tower of London; TOL^{DX} = Tower of London-Drexel University: 2nd Edition; TSI = Test for Severe Impairment; VABS = Vineland Adaptive Behaviour Scales; VAT = Visual Association Test; VF = Verbal Fluency; VisMT = Visual Memory Test; VMT = Verbal Memory Test; VR = Visual Representation subtest of the WMS-R; WAIS/ WAIS-R = Wechsler Adult Intelligence Scale/Revised; WCFST = Weigl Colour-Form Sort Test; WISC/ WISC-R = Wechsler Intelligence Scale for Children/Revised; WM = Working Memory; WMS-R = Wechsler Memory Scale-Revised

Table B 1

Sample Characteristics

Author, Year	Subtype of ID	Level of ID	Criteria/ method used for assessing level of ID	Comparator / control group (if included)	Dementia subtypes (if included)	Diagnostic criteria for dementia	Mean age (years, *SD*) and/ or range (years)	% female	% white ethnicity	Verbal ability (as part of inclusion not testing)
Ball et al., 2008	DS	Mild = 35.2% Moderate = 49.2%	ICD-10 and BPVS-II	Between groups (dementia vs. no-dementia)	AD (25 diagnosed at point of ax)	CAMDEX-DS	49 36-72	42.0%	NR	NR
Benejam et al., 2015	DS	DS = upper25 (33%) / middle50 (67%) DS-AD = upper1 (7%) / middle14 (93%)	K-BIT and informant report for DS-AD group	Between groups (AD vs. no-AD)	AD	NR	DS = 36.1 (9.8) DS-AD = 51.1 (5.1)	DS = 44% DS-AD = 60%	NR	NR
Benejam et al., 2020	DS	CAMCOG-DS subgroup = 91/205/47 mCRT subgroup = 85/161/25	DSM-V and K-BIT	Between groups (MCI vs. AD vs. no-AD)	AD	CAMDEX-DS	CAMCOG-DS subgroup = 41 (18.5) mCRT subgroup = 39 (18.0)	CAMCOG-DS subgroup = 49.1% mCRT subgroup = 47.2%	NR	NR
Bevins & Hurse, 2014	DS + oID	FSIQ range 13–120	BVPS-II	Between instruments	AD (*n* = 2)	NR	49.5 (9.3) 21–66	57.1%	NR	Range (13–120 on BVPS-II)
Brugge et al., 1992	DS + oID	DS = FSIQ 60.9 (2.8) oID = FSIQ 55.3 (1.6)	WAIS-R	Between groups (memory-impaired vs. non-memory-impaired)	NA	Short delayed savings score from the CVLT-C used to indicate memory-impaired	DS = 31.1 (2.9) years 22 to 51 oID = 28.9 (2.8) 22 to 46	NR	NR	NR
Carr, 2003	DS	NR	Leiter IQ	Longitudinal comparison	NA	NA	30 and 35 years of age	NR	NR	
Cooper et al., 2016	DS	Mild = 36.0% Moderate = 33.0% Severe = 43.0% Profound = 5.0%	NR	Longitudinal comparison	NA	NA	54.15 (3.10)	48.0%	NR	NR
Devenny et al., 1992	DS + oID	Mild to moderate	NR	Longitudinal comparison and between groups (DS vs. no-DS)	NA	NA	DS <35 = 31.7 DS >35 = 41.8 27-55	NR	NR	NR
Devenny et al., 1996	DS + oID	Mild to moderate	Clinical records	Longitudinal comparison	AD	DSM-III-R	DS = 31-63 oID = 31-76	NR	NR	NR
Devenny et al., 2000	DS	Mild to moderate	NR	Longitudinal comparison and between groups (dementia status)	NR	ICD-10	ID-no-DS = 53.68 ± 11.03 DS = no overall given	NR	NR	NR

Author, Year	Subtype of ID	Level of ID	Criteria/ method used for assessing level of ID	Comparator / control group (if included)	Dementia subtypes (if included)	Diagnostic criteria for dementia	Mean age (years, *SD*) and/ or range (years)	% female	% white ethnicity	Verbal ability (as part of inclusion not testing)
Devenny et al., 2002	DS	Mild to moderate	NR	Between groups (DS-AD vs. DS-no-AD and oID-no-AD)	AD	ICD-11	ID-no-DS = 56.8 (11.4) DS-no-AD = 47.3 (7.1) DS-AD = 54.8 (6.3)	NR	NR	NR
Esteba-Castillo et al., 2022	DS	Mild to moderate	DSM-V	Longitudinal comparison and between groups (AD vs. no-AD)	MCI	NA	45.8 ± 4.64 (final sample)	54.0%	100%	NR
García-Alba et al., 2017	DS	Mild = 62.0% Mod = 38.1%	DSM-V	Psychometric properties of instruments and between groups (level of ID)	NA	Excluded with CAMDEX-DS (which includes diagnostic criteria from the DSM-IV and ICD-10)	NR All aged ≥ 39 years	47.6%	NR	NR
García-Alba et al., 2019	DS	Mild to moderate	DSM-V, K-BIT and Vineland II	Between groups (dementia status and DS vs. no-DS)	MCI and AD	Clinical judgment and assessment of cognition using CAMDEX-DS and adaptive skills	Control-DS = 44.64 (3.30) DS-MCI = 51.64 (3.95) DS-AD = 53.54 (6.58) HC = 45.21 (4.39)	DS = 61.0% HC = 71.4% Control-DS = 71.4% DS-MCI = 42.9% DS-AD = 69.2%	NR	NR
Head et al., 2011	DS	Mild, Moderate and Profound (IQ<45 excluded)	Case file reviews of historical full-scale IQ assessments	Between groups (dementia status and level of ID)	AD (in comparator group)	Neurological evaluation and DSM-IV	Study 1: DS-no-AD = 44.1 (1.4) 37-54 HC = 46.5 (2.0) 39-56 DS-AD = 75.3 (1.8) 61-91 AD-controls = 74.2 (1.3) 66-83 Study 2: DS-AD = 53.3 (0.7) 41-63 DS-no-AD = 45.1 (1.9	Study 1: DS-no-AD = 47.1% HC = 54.5% DS-AD = 11.8% AD-controls = 41.7% Study 2: DS-AD = 50% DS-no-AD = 34.6%	NR	NR

Author, Year	Subtype of ID	Level of ID	Criteria/ method used for assessing level of ID	Comparator / control group (if included)	Dementia subtypes (if included)	Diagnostic criteria for dementia	Mean age (years, *SD*) and/ or range (years)	% female	% white ethnicity	Verbal ability (as part of inclusion not testing)
							26-60			
Hoekman & Maaskant, 2002	DS (47%) + oID	Mild = 14.0% Moderate = 60.0% Severe = 23.0% Profound = 3.0%	NR	Between instruments (one directly administered vs. two informant-rated)	NR	Expert opinion + ICD-10	58.6 40–91	NR	NR	NR
Hom et al., 2021	DS	Mild = 53.5% Moderate = 46.5%	NR	Between groups (MCI vs. cognitively stable)	MCI	Consensus Review Conference	CS = 48.65 (6.27) MCI-DS = 52.88 (6.72) 40–82	41.0%	86.10%	NR
Krinsky-McHale, Devenny & Silverman, 2002	DS	Mild to moderate	NR	Longitudinal comparison and between groups (AD vs. no-AD)	AD	ICD-10	Baseline: DS-AD F = 52.23 (7.49) DS-AD M = 45.32 (5.55) DS-no-AD F = 42.06 (7.01) DS-no-AD M = 44.36 (6.64)	DS-AD = 71.4% DS-no-AD = 50.7%	NR	NR
Krinsky-McHale et al., 2008	DS	Mild to moderate	Records for WAIS-R or Stanford-Binet IQ scores or LIPS	Longitudinal comparison and psychometric properties of instruments	MCI + AD	Clinical assessment	Baseline: DS-AD = 51.44 (5.20) 45-58 DS-no-AD = 49.40 (4.57) 44-62	NR	NR	NR
Krinsky-McHale et al., 2020	DS	Mean FSIQ at baseline = 33.3 (*SD* = 7.3)		Longitudinal comparison and between instruments and groups	MCI + AD	Clinical assessment	Baseline: 51.6 (9.1)	NR	92%	Sufficient to assent
McPaul et al., 2017	Various	Mild to moderate (mean FSIQ = 59.10 (*SD* = 7.57), range 46-73)	WAIS-IV and record review	Psychometric properties of instruments	NA (excl. at baseline)	NA	31.08 (8.08) 18-44	47.5%	NR	Fluent in English inclusion criteria
Nelson et al., 2001	DS	NR	NR	Between groups (normal vs. abnormal findings)	NR	NR	40.03 (11.8)	61.5%	NR	NR
Nelson et al., 2005	DS	Mean FSIQ at baseline = 50.7 (*SD* = 6.5), range 45-66	WAIS-III	Between instruments and age	NR	NR	37.2 (9.5) 22–58	60.0%	NR	NR

Author, Year	Subtype of ID	Level of ID	Criteria/ method used for assessing level of ID	Comparator / control group (if included)	Dementia subtypes (if included)	Diagnostic criteria for dementia	Mean age (years, *SD*) and/ or range (years)	% female	% white ethnicity	Verbal ability (as part of inclusion not testing)
Nelson et al., 2007	DS	Mean FSIQ at baseline = 51.31 (*SD* = 6.58)	WAIS-III	Psychometric properties of instruments	NR	NR	40.45 (8.67) 24-55	52.9%	NR	NR
Oliver et al., 1998	DS	NR	NR	Between groups (cognitive deterioration status)	NR	NR	42.34 (7.26)	59.6%	NR	Participants were excl. if they had speech limited to only a few words or were unable to understand simple instructions (e.g., 'sit down')
Palmer, 2006	DS + oID	Mild to Moderate	NR	Between groups (DS-dementia vs. DS-no-dementia vs. no-DS)	AD	DSM-IV-TR	AD = 50.50 (6.77) 36–62 HC = 44.50 (9.07) 33–66	AD = 60.0% HC = 66.7%	AD = 90.0% HC = 91.7%	NR
Powell et al., 2014	DS	Classified as low, medium, or high functioning	Medical records	Between groups (DS-dementia vs. DS-no-dementia vs. no-DS)	AD	NINCDS-ADRDA and consensus review	All DS = 51.38 (6.48) DS-no-AD = 50.61 (5.53) DS-AD = 52.16 (7.54) HC = 51.07 (2.14)	70.0%	NR	NR
Pyo et al., 2007	DS + oID	Moderate to severe	Medical records	Between groups (AD vs. no-AD)	AD	DSM-IV-TR	AD = 53.13 (10.56) 43-74 HC = 49.95 (5.13) 40-59	AD = 15.4% HC = 24.4%	NR	NR
Pyo et al., 2009	DS + oID	Moderate to severe	Medical records	Between groups (AD vs. no-AD)	AD	DSM-IV-TR	AD = 53.99 (10.20) HC = 50.76 (5.76)	AD = 31.3% HC = 2.9%	NR	NR
Pyo et al., 2010	DS + oID	Moderate to severe	Medical records	Between groups (AD vs. no-AD)	AD	DSM-IV-TR	DS-controls = 49.21 (4.41) oID-controls =52.87 (5.25) DS-AD = 47.89 (4.18) oID-AD = 57.13 (10.52)	DS-controls = 0.0% oID-controls = 8.3% DS-AD = 13.3% oID-AD = 36.4%	NR	NR
Pyo et al., 2011	DS + oID	Moderate to severe	Medical records	Between groups (AD vs. no-AD)	AD	DSM-IV-TR	DS-controls = 47.71 (5.21) oID-controls =	DS-controls = 0.0%	NR	NR

Author, Year	Subtype of ID	Level of ID	Criteria/ method used for assessing level of ID	Comparator / control group (if included)	Dementia subtypes (if included)	Diagnostic criteria for dementia	Mean age (years, *SD*) and/ or range (years)	% female	% white ethnicity	Verbal ability (as part of inclusion not testing)
							51.93 (7.05) DS-AD = 48.26 (2.43) oID-AD = 57.99 (11.14)	oID-controls = 6.9% DS-AD = 20.0% oID-AD = 45.5%		
Sacco et al., 2022	DS	Mild = 44.3% Moderate = 32.5% Moderate-Severe = 23.2%	WAIS-IV	Between groups (AD vs. no-AD)	AD + co-occurring	French version of Dementia Screening Questionnaire for Individuals with Intellectual Disabilities and consensus review	46.9 (6.8) 32-65	49.5%	NR	NR
Sano et al., 2005	DS	Mild = 15.0% Moderate = 52.0% Severe = 29.0% Profound = 4.0%	Records review	Between instruments	NR	DSM-IV	48.7 (6.2) 33-77	51.3%	Partially available for one cohort = 96.3%	NR
Sinai et al., 2016	DS	Mild = 37.1% Moderate/Severe = 62.9%	Informant report/ case notes	Between groups (dementia vs. no-dementia)	NR	Informant report and clinical consensus	52.7 (6.06) 45-64 at 1st assessment	53.1%	85.4%	NR
Wallace et al., 2021	DS	No dementia = Borderline/ Mild = 39%; Moderate/ Severe = 29% Possible dementia = Borderline /Mild = 8%; Moderate/ Severe = 8% Probable dementia = Borderline/ Mild = 5%; Moderate/ Severe = 10%; Not documented = 1%	Prior diagnosis and medical records review	Between instruments and method of testing	NR	NINCDS-ADRDA and consensus review	No dementia = 37.98 (9.33); Possible dementia = 46.66 (9.74); Probable dementia = 51.50 (8.79) 25-64	No dementia = 54.4% Possible dementia = 68.8% Probable dementia = 62.5%	NR	English-speaking
Walsh et al., 2015	DS	Mild = 35% Moderate = 39% Severe = 23% Profound = 3%	Previous diagnosis	Psychometric properties of instruments	NR	ICD-10 and DSM-IV	49.8 (8.9)	45.0%	NR	NR

Note. AD = Dementia of the Alzheimer's Type; BPVS/ BPVS-II = British Picture Vocabulary Scale/ British Picture Vocabulary Scale-2nd Edition; CAMCOG = Cambridge Cognition Examination; CAMCOG-DS = Cambridge Cognitive Examination adapted for individuals with Down Syndrome; CAMDEX = Cambridge Mental Disorders of the Elderly Examination; CAMDEX-DS = Cambridge Examination for Mental

Disorders of Older People with Down Syndrome and Others with Intellectual Disabilities; CS = Cognitively stable; CVLT-C = California Verbal Learning Test - Children's Version; DS = Down syndrome; DSM-III-R/ DSM-IV/ DSM-IV-TR/ DSM-5 = Diagnostic and Statistical Manual of Mental Disorders-3rd Revised/ 4th Edition/ 4th Edition Text-Revised/ 5th Edition; FSIQ = Full-Scale IQ; HC = Healthy controls; ICD-10/ ICD-11 = International Classification of Diseases-10th Edition/ 11th Edition; ID = Intellectual disability; IQ = Intelligent Quotient; MCI = Mild Cognitive Impairment; mCRT = Modified Cued Recall Test; NA = Not applicable; NINCDS-ADRDA = National Institute of Neurological and Communicative Diseases and Stroke/Alzheimer's Disease and Related Disorders Association; NR = Not reported; oID = Intellectual disability from other causes than DS; WAIS/ WAIS-R/ WAIS-III/ WAIS-IV = Wechsler Adult Intelligence Scale/ Revised/ 3rd Edition/ 4th Edition

Appendix C

Cognitive Tests Included in the Review

List of Single Domain Tests/ Tasks by Domain: Population(s) Developed for, Included Studies and Findings

Abbreviations	List of Measures	Function/ Cognitive domain(s)	A	ID	DS	NV	CD	Author(s)	Test Findings
Single Domain Subtests/ Tasks by Domain									
Orientation and Arousal									
O-CAMCOG	Orientation subtest from the CAMCOG	Orientation	YES	NO	NO	NO	YES	Oliver et al., 1998	↓ scores at 1st assessment indicative of cognitive deterioration at later assessments. ↓ appears before or alongside ↓ in aphasia, agnosia, and apraxia. Significant ↓ scores in moderate and severe cognitive impairment groups vs. no impairment.
O-MMSE	Orientation subtest from the MMSE	Orientation	NO	NO	NO	NO	YES	Sano et al., 2005	Too difficult for moderate to severe ID, with floor effects.
O-OCDB	Orientation subtest from the OCDB/ NAID	Orientation	YES	YES	NO	NO	YES	Carr, 2003	↓ from age 30 to 35 years (Z=2.47, p<.05). Scores correlated with scores on memory tests from OCDB/ NAID at age 30 (rho=0.53, p<.01) and 35 years (rho=0.55, p<.01) and verbal IQ estimated on the BPVS at age 30 years (rho=0.62, p<.001) and at 35 years (rho=0.63, p<.001).
O-RBMT-C	Orientation subtest from the RBMT-C	Orientation	NO	NO	NO	NO	NO	Carr, 2003	Highly correlated at ages 30 and 35 (r=0.86, p≤.001). Significant decline from age 30 to 35 years (Z=2.80, p<.01).
TO	Temporal Orientation subtest of the BT-ID	Orientation	YES	YES	NO	NO	NO	García-Alba et al., 2019	Impairments in TO found to indicate MCI in DS.
WG-O	Working Group's Orientation Test	Orientation	YES	YES	NO	NO	YES	Pyo et al., 2009	AD group scored ↓ vs. control group at baseline. Changes in scores over 1-year f/u not significantly different between the groups = poor sensitivity and specificity. Age and ID aetiology did not significantly affect scores. Floor effects for those with severe ID.
Attention/ Processing Speed									
Co	Coding subtest of the WISC-R and performance IQ	Processing speed but also attention, visuomotor and visuo perception, STM	NO	NO	NO	NO	NO	Devenny et al., 1996	Significant differences between DS-possible-AD and DS-no-possible-AD groups (U[4,73]=273.0, p=.03).
								Devenny et al., 2000	Significant difference between healthy-DS vs. questionable-dementia, early-stage dementia, and middle-stage dementia groups ($F_{3,59}$=4.75, p=.001).
CT	Cancellation task	Selective attention and visuospatial function	YES	YES	YES	YES	YES	Cooper et al., 2016	The CT showed utility.
								Krinsky-McHale et al., 2008	Performance on CT task varied with stage of dementia. Reasonably good sensitivity (80%) and specificity (82%) for identifying MCI and easy to administer.
Executive Function									

Abbreviations	List of Measures	Function/ Cognitive domain(s)	A	ID	DS	NV	CD	Author(s)	Test Findings
A	Arithmetic subtest from the WISC-R	Mental arithmetic and working memory	NO	NO	NO	NO	NO	Devenny et al., 2000	Significant difference between healthy-DS vs. questionable-dementia, early-stage dementia, and middle-stage dementia groups ($F_{3,62}$ =6.56, p=.005) but early- and middle-stage dementia groups did not differ from each other, or the healthy and 'questionable' groups.
CaD	Cats and Dogs subtest from the CEFA	Executive function - response inhibition	YES	YES	YES	DK	YES	Ball et al., 2008	Small effect size (d<0.5) for identifying AD from no-AD groups = less sensitive to the effects of AD.
								Bevins & Hurse, 2014	CaD correlated with OM. CaD negatively correlated with DLD. CaD did not correlate with VC measures. However, ceiling effect for some participants.
								Cooper et al., 2016	Poor completion rates.
								Hom et al., 2021	CaD Switch score significantly ↓ in MCI-DS group vs. CS group (U=4.43, p=.0001). Did not load onto any factors and dropped from analysis.
CaF	Cats and frogs (modified dots task)	Executive function - inhibitory control and working memory	YES	YES	YES	YES	NO	Sinai et al., 2016	81.3% attempted the task. High % of participants at floor. Significant moderate correlation with VF and ToL stages completed (from 0.48 to 0.63). No significant difference between dementia and no-dementia groups.
CFT	Category Fluency Test	Executive function - verbal fluency	YES	NO	NO	NO	NO	Cooper et al., 2016	Ease of completion and sensitivity to change over time.
COWAT	The Controlled Oral Word Association Test (also known as 'FAS')	Executive function - verbal fluency; language in children	YES	NO	NO	NO	NO	Brugge et al., 1994	No significant differences between DS-memory-impaired and DS-non-memory-impaired groups.
								Palmer, 2006	Dementia group ↓ scores.
CTT	The Colour Trails Test	Executive function - cognitive flexibility and processing speed	YES	YES	NO	YES	NO	Palmer, 2006	Dementia group ↓ scores. Floor effects for those with dementia.
IED	Intra-Extra Dimensional shift subtest from the CANTAB	Executive function - set-shifting	YES	YES	NO	DK	YES	Sinai et al., 2016	'Stages completed' showed high % at floor. Some CANTAB IED scores showed a significant mild to moderate correlation with VF and ToL 'stages completed' scores (from 0.40 to +/-0.45). Only BRIEF WM scores showed a mild to moderate correlation with some of the IED outcome measures (from -0.44 to 0.35). No significant differences between DS-memory-impaired and DS-non-memory-impaired groups.
M-CFT	McCarthy Scales of Children's Abilities - Category Fluency Test	Executive function - verbal fluency; language in children	NO	NO	NO	NO	NO	Hom et al., 2021	Score significantly ↓ in MCI-DS group vs. CS group (U=2.21, p=.027). CFT loaded onto multiple factors, but the loadings were weaker in relation to the other tests within the same factor. Dropped from further analyses.
								Krinsky-McHale et al., 2020	Considerable floor effects, especially for those with severe/ profound ID and/ or dementia. Test re-test reliability estimate (Cronbach's α=.865). % scoring 2 SDs above the

Abbreviations	List of Measures	Function/ Cognitive domain(s)	A	ID	DS	NV	CD	Author(s)	Test Findings
									floor by ID severity = mild (91.4%), moderate (67.9%), severe (35.5%), profound (4.7%). On longitudinal assessment, significant declines found only with dementia onset, indicating insensitive to early decline.
PA	Picture Arrangement from the WISC-R	Social competence, reflection, planning and performance IQ	NO	NO	NO	YES	NO	Devenny et al., 2000	Floor effects at baseline across all groups
RL	Reversal learning	Executive function - response inhibition and set-shifting						Nelson et al., 2005	Marginally significant relationship observed between age (p=.08) and FSIQ (p=.08) and RL error scores.
								Nelson et al., 2007	Moderate test-retest reliability (r=.63, p<.01). IQ was inversely associated (r=.55, p<.03).
SB	Scrambled Boxes from the CEFA	Executive Function	YES	YES	YES	DK	YES	Ball et al., 2008	Large effect size (d>0.5) for identifying dementia groups. Small effect (d<0.5) indicating less sensitive to effects of ID severity. Number of informant-reported personality/ behaviour changes contributed significantly to the model for SB (p<.05)
Sreversal	Spatial Reversal from the CEFA	Executive Function	YES	YES	YES	DK	YES	Ball et al., 2008	Considerable floor effects. Small effect size (d<0.5) for identifying dementia groups but not related to severity of ID = ?measuring EF.
ToL	ToL	Executive function - planning	YES	NO	NO	DK	NO	Ball et al., 2008	Revised ToL showed no floor effects but less sensitive to change.
								Cooper et al., 2016	Minimal floor effects.
TOL$_{DX}$	ToL-Drexel University: 2nd Edition	Executive function - planning	YES	NO	NO	DK	NO	Esteba-Castillo et al., 2022	MCI group showed cognitive ↓ on Hit score.
								García-Alba et al., 2017	No floor effects. It could distinguish between mild and moderate ID and highly associated with other measures of EF. Results showed test items not appropriately scaled.
								Sinai et al., 2016	No significant differences between DS-memory-impaired and DS-non-memory-impaired groups. Floor and ceiling effects, particularly for dementia group.
VF	Verbal Fluency from the CEFA (semantic)	Executive function - verbal fluency						Ball et al., 2008	Small effect (d<0.5) suggesting less sensitive to effects of AD.
								Bevins & Hurse, 2014	No correlations between semantic fluency and the other measures known to indicate dementia. Independent of verbal comprehension ability. Lack of floor effects.
								Sinai et al., 2016	Significant differences between DS-memory-impaired and DS-non-memory-impaired groups for raw score (p=.006) and adjusted (p=.002).

Abbreviations	List of Measures	Function/ Cognitive domain(s)	A	ID	DS	NV	CD	Author(s)	Test Findings
WCFST	Weigl Colour-Form Sort Test	Executive function - sorting and set-shifting	YES	NO	NO	YES	YES	Ball et al., 2008	Considerable floor effects. Significant interaction between AD diagnosis and ID severity. Moderate ID performing at floor regardless of whether they have AD. Dropped from further analyses.
								Esteba-Castillo et al., 2022	No differences found between control-DS and prodromal-dementia-DS.
								García-Alba et al., 2017	No relevant data.
Visuospatial									
BD	Block Design	Visual spatial ability, constructional praxis, motor skill, and problem-solving skill ability and performance IQ	YES/ NO	NO	NO	YES	NO	Devenny et al., 2000	BD from WISC-R, significant difference between healthy-DS vs. questionable-dementia, early-stage dementia, and middle-stage dementia groups ($F_{3,62}$ =8.14, p<.001).
								Hom et al., 2021	BD significantly ↓ in MCI-DS group vs. CS group (U=3.11, p=.002).
								Krinsky-McHale et al., 2020	BD supplemented with less complex items from Block T subtest from the DSMSE. Significant floor effects in those with severe or profound ID. ↓ in scores in MCI-DS and dementia groups vs. CS.
OA	Object Assembly from WISC-R	Visual spatial ability and performance IQ	NO	NO	NO	YES	NO	Devenny et al., 2000	Significant difference between healthy-DS vs. questionable-dementia, early-stage dementia, and middle-stage dementia groups ($F_{3,61}$ =6.15, p=.001) but early- and middle-stage dementia groups did not differ from each other, or the healthy and 'questionable' groups.
PC	Picture Completion from the WISC-R	Visual spatial ability and performance IQ	NO	NO	NO	YES	NO	Devenny et al., 2000	Significant difference between healthy-DS vs. questionable-dementia, early-stage dementia, and middle-stage dementia groups ($F_{3,62}$ =6.56, p=.001) but early- and middle-stage dementia groups did not differ from each other, or the healthy and 'questionable' groups.
Language									
BNT	The Boston Naming Test	Language - sensitive to detecting compromised lexical retrieval abilities and aphasia through visual confrontation naming	YES	NO	NO	NO	YES	Brugge et al., 1994	DS ↓ scores vs. oID on BNT (p<.025). Showed trends for diagnostic group effects (p<.075, F=2.94).
								Hom et al., 2021	BNT significantly ↓ in MCI-DS group vs. CS group (U=2.04, p=.041).
								Palmer, 2006	Dementia group ↓ scores.
EOWPVT/ EOWPVT-R	Expressive One-Word Picture Vocabulary Test/ Expressive One-Word Picture Vocabulary Test-Revised	Expressive language; Knowledge	YES	NO	NO	NO	NO	Sano et al., 2005	Vocabulary score was sensitive to dementia status and age but not level of ID.

Abbreviations	List of Measures	Function/ Cognitive domain(s)	A	ID	DS	NV	CD	Author(s)	Test Findings
Info	Information subtest from the WISC-R	Language and verbal IQ	NO	NO	NO	NO	NO	Devenny et al., 2000	Significant difference between healthy-DS vs. middle-stage dementia groups ($F_{3,62}$ =6.58, p=.001) but no significant differences between the healthy, 'questionable' and early-stage dementia groups. Not sensitive to the early stages of decline but did indicate the involvement of additional cognitive functions as decline progressed.
Opp	Opposites subtest from the McCarthy Scales of Children's Abilities	Verbal Reasoning	NO	NO	NO	NO	NO	Brugge et al., 1994	No relevant data.
PI	Picture Identification from the OCDB/ NAID	Language	YES	YES	NO	DK	YES	Carr, 2003	Mean scores slightly ↑ from age 30 to 35 years.
PN	Picture Naming from the OCDB/ NAID	Language	YES	YES	NO	DK	YES	Carr, 2003	Mean scores ↓ from age 30 to 35 years.
PPVT/ PPVT-R	Peabody Picture Vocabulary Test/ Peabody Picture Vocabulary Test-Revised	Language - receptive	YES	NO	NO	NO	NO	Brugge et al., 1994	DS significantly ↓ scores vs. controls. Showed trends for diagnostic group effects (p<.075, F=2.94). Scores did not reduce with age.
								Nelson et al., 2001	Significantly ↓ scores in DS subjects with abnormal physical findings vs. normal results (p=.019 and p=.003 at times 1 and 2, respectively). Scores appeared to improve over time in the group with normal physical results.
								Pyo et al., 2007	No significant difference between AD and no-AD groups.
Si	Similarities subtest from the WISC-R	Language and verbal IQ	NO	NO	NO	NO	NO	Devenny et al., 2000	Floor effects at baseline across all groups.
VC	Verbal Comprehension subtest from the WISC-R	Language and verbal IQ	NO	NO	NO	NO	NO	Devenny et al., 2000	Significant difference between healthy-DS vs. questionable-dementia, early-stage dementia, and middle-stage dementia groups ($F_{3,59}$ =7.98, p<.001) but early- and middle-stage dementia groups did not differ from each other, or the healthy and 'questionable' groups.
VoC	Vocabulary subtest from the WISC-R	Language and verbal IQ	NO	NO	NO	NO	NO	Devenny et al., 2000	Significant difference between healthy-DS vs. middle-stage dementia groups ($F_{3,61}$ =6.46, p=.001) but no significant differences between the healthy, 'questionable' and early-stage dementia groups. Not sensitive to the early stages of decline but did indicate the involvement of additional cognitive functions as decline progressed.
Learning and Memory									
ADVM	Auditory delayed verbal memory	Memory - delayed verbal						García-Alba et al., 2019	DS-AD ↓ scores and ↓ scores vs. DS-MCI group.
AMT	Autobiographical Memory Test	Memory - autobiographical	YES	NO	NO	NO	NO	Pyo et al., 2011	AD group scored ↓ than control group at baseline. Controls with DS considerable ↓ at f/u, but not others. May be useful as dementia screening tool in moderate to severe ID and DS but needs further validation. Limited score variability – requires modification.
ASL or landmark	Allocentric spatial learning or landmark	Spatial memory						Nelson et al., 2007	The lowest level showed good test re-test reliability (r=.78, p<.002) but all other levels were not significant. Sensitivity of 75% was demonstrated for the fourth level (most complex).

Abbreviations	List of Measures	Function/ Cognitive domain(s)	A	ID	DS	NV	CD	Author(s)	Test Findings
BSRT	The Busckke Selective Reminding Test	Memory and learning - short-term and long-term verbal	YES	NO	NO	NO	NO	Devenny et al., 1992	No significant difference on longitudinal assessment for number of correct items on 1st presentation. All groups showed comparable improvements on longitudinal assessment for total number of items recalled after first 9 trials. Likely due to yearly assessments providing repeated presentation (i.e., 3-4 presentations in a 3-5 year period).
								Devenny et al., 1996	Significant effects of ID aetiology (t=-2.395, p=.017) and age (t=-3.462, p=.001) and no interaction. Older showed ↓ performance. DS ↓ scores than oID. Difference between younger and older groups larger for DS than oID. Significant difference between possible-AD-DS and no-AD-DS (U[4,74]=283.0, p=.002).
								Devenny et al., 2000	No data reported.
								Hom et al., 2021	Scores affected by age. Largest group difference on memory tests (also DSMSE memory).
								Krinsky-McHale, Devenny & Silverman, 2002	Using total recall score, early-AD showed sig ↓ over 3-year period prior to diagnosis vs. no-AD. Older group ↓ number of items recalled vs. younger, but greater ↓ in AD group. Higher baseline IQ associated with ↑ recall. 20%+ below an individual's highest previous total recall score during 2 consecutive test sessions = criterion level to indicate substantial decline. Sensitivity = 78.6% at time of diagnosis and 92.9% 1 year after diagnosis. LTS and LTR scores particularly affected by early-AD and prior to diagnosis.
								Krinsky-McHale et al., 2008	Participants with later diagnosis of MCI scored sig ↓ on total number of words recalled over longitudinal assessment at T2 and again at T3. Most errors in the MCI and early-AD groups were perseverations.
								Krinsky-McHale et al., 2020	↓ scores affected by severity of ID. Not suitable to track dementia in severe or profound group due to floor effects at baseline.
CRT	The Cued Recall Test	Memory - cued recall	YES	NO	NO	NO	NO	Benejam et al., 2015	DS-AD scored ↓ on free recall and total score and committed ↑ intrusion errors vs. DS-no-AD. Age main factor associated with ↓ m-CRT scores. m-CRT not found to be useful in severe ID or advanced DS-AD due to difficulties in comprehension. DS-no-AD scored ↑ after semantic (category) cues provided.
								Cooper et al., 2016	Ease of completion and sensitivity to change over time.

Abbreviations	List of Measures	Function/ Cognitive domain(s)	A	ID	DS	NV	CD	Author(s)	Test Findings
								Devenny et al., 2000	Preliminary findings suggested criterion of ≤23 on CRT (and decline of 20%+ on BSRT) predictive of dementia.
								Devenny et al., 2002	AD had ↓ total scores vs. no-AD. IQ level and age also negatively affected scores. Poor performance on CRT (adapted) was associated with early-stage-AD. Cut-off of ≤23 on total score gave sensitivity of 94.7% and specificity of 93.9% with a PPV of 81.9% when comparing DS-AD with oID-no-AD. DS-AD make semantic errors when given cue.
								Hom et al., 2021	CRT significantly ↓ in MCI-DS group vs. CS group (U=4.43, p=.0001).
								Sacco et al., 2022	Total recall scores significantly ↓ (p<.0001) in AD vs. controls. Scores ↓ with age and severity of ID and made ↑ intrusion errors. m-CRT showed good sensitivity to detect cognitive decline in DS. Though, total free recall impacted by level of ID. Profound ID were excl. due to being unable to complete the m-CRT.
CVLT-C	California Verbal Learning Test - Children's Version	Memory, short-term and long-term	NO	NO	NO	NO	NO	Brugge et al., 1994	DS participants sig ↓ scores vs. oID on short-delay savings (p<.001), long-delay savings (p<.01) and false positives (p<.025). Short and long free recall, no significant differences between DS and control groups due to floor effects. Short delayed savings inversely correlated with age in DS group but not controls. Memory-impaired group (on short delay savings) sig older than non-memory impaired.
Dmess	Delivering a message subtest from the RBMT-C	Memory	NO	NO	NO	NO	NO	Carr, 2003	Ceiling effects.
DMTS	Delayed match-to-sample	Visual matching ability and short-term visual recognition memory						Hoekman & Maaskant, 2002	Little to no agreement between the results of the DMTS, CLD and DMR. DMTS identified far less participants as having dementia than CLD and DMR and expert opinion. Only the specificity of the DMTS was good (89%).
DNMP	Spatial delayed non-match-to-position	Spatial memory						Nelson et al., 2005	Most had difficulty completing the spatial DNMP task.
DNMS	Object delayed non-match-to-sample	Object recognition memory						Nelson et al., 2005	Significantly < errors on the object DNMS (M=6.4) vs. spatial DNMP (M=12.3; t(10)=4.79, p<.001). 55% able to reach criterion on object DNMS. Age was significantly correlated with number of errors (r=0.68, p=.02). FSIQ significant predictor of errors on object DNMS (F(1,9)=8.18, p=.02). Only one participant could score on DNMS at both time points.
								Nelson et al., 2007	14 reached criterion on DNMS 10s delay (Time 1). Of those 14, 11 reached criterion on the more complex DNMS 20s delay (Time 2). Only 1 subject reached criterion and could perform DNMS at both time intervals, across both points in time. Test re-test reliability = 10s delay (r=.214, p<.38) and 20s delay (r=.42, p< .72).

Abbreviations	List of Measures	Function/ Cognitive domain(s)	A	ID	DS	NV	CD	Author(s)	Test Findings
DRecog	Delayed Recognition from the CAMCOG-DS	Memory - delayed recognition	YES	YES	YES	YES	YES	Ball et al., 2008	Large effect size (d≥0.05) for distinguishing AD from no-AD. Less sensitive to effects of ID severity (d<0.5). Increasing age = decrease in performance.
DRecall	Delayed Recall from the CAMCOG-DS	Memory - delayed recall	YES	YES	YES	YES	YES	Ball et al., 2008	Large effect size (d≥0.05) for distinguishing AD from no-AD.
DSpan	Digit Span	Memory - short-term and working memory (executive function) for backwards digit span	NO/ YES	NO	NO	NO	NO	Devenny et al., 1996	Moderate test re-test reliability (r=0.640). Sig differences between possible-AD-DS and no-AD-DS groups (U[4,55]=187.0, p=.02). Possible-AD-DS declined on longitudinal assessment vs. no-AD-DS.
								Devenny et al., 2000	Significant difference between healthy-DS vs. middle-stage dementia groups ($F_{3,58}$ =4.71, p=.005) but no significant differences between the healthy, 'questionable' and early-stage dementia groups. Not sensitive to the early stages of decline but did indicate the involvement of additional cognitive functions as decline progressed.
DVM	Delayed visual memory	Memory - delayed visual						García-Alba et al., 2019	DS-AD ↓ scores. DS-MCI ↓ scores vs. DS-controls.
FOME/ mFOME	The Fuld Object-Memory Evaluation/ Modified - Fuld Object Memory Evaluation	Uses multisensory (tactile, visual, and verbal) encoding of objects for assessing memory	YES	NO	NO	DK	YES	Palmer, 2006	Dementia group ↓ scores on memory and learning (Fuld Total, Fuld Retention).
								Pyo et al., 2007	mFOME too difficult for moderate to severe ID.
								Pyo et al., 2010	No significant difference between AD group and control group.
								Sano et al., 2005	mFOME useful across all levels of ID.
Frecog	Face recognition subtest from the RBMT-C	Memory	NO	NO	NO	NO	NO	Carr, 2003	Score unchanged from 30 to 35 years. Floor and ceiling effects.
IM	Immediate Memory – items from the CAMCOG and SIB	Memory - immediate	YES	NO	NO	NO	YES	Ball et al., 2008	Sensitive to level of ID (d≥0.5).
LM	Logical Memory from WMS-R	Narrative episodic memory	YES	NO	NO	NO	NO	Brugge et al., 1994	Scores affected by age for LM-IB (r=0.640, p<.05) in non-memory-impaired-DS group, similar to controls = improvement with advancing age. Memory-impaired-DS group showed decline.
MfO	Memory for objects (from the NAID or SBIS)	Memory - short-term, visual	YES	YES	NO	DK	YES	Ball et al., 2008	DAT showed ↓ performance vs. no-DAT. Found to relate to EF (p<.05). Informant reported personality/ behaviour changes remained a significant predictor of test score (beta=-0.33, t=-3.06, p<.05) after non-executive memory tasks removed from analysis.
								Brugge et al., 1994	No data reported.
								Carr, 2003	Mean scores ↑ over time, though not sig.

Abbreviations	List of Measures	Function/ Cognitive domain(s)	A	ID	DS	NV	CD	Author(s)	Test Findings
								Cooper et al., 2016	MfO test from the NAID most appropriate cognitive instrument due to ease of completion and sensitivity to change over time.
MfP	Memory for pictures (from the NAID)	Memory	YES	YES	NO	DK	YES	Carr, 2003	Sig ↓ in scores from ages 30 to 35 years (Z=-3.06, p<.05). Only memory test to correlate with another (memory for sentences) at age 35 (rho=0.44, p<.5).
								Oliver et al., 1998	Scores sig ↓ for moderate and severe deterioration vs. no deterioration groups ($\chi 2(2)$=16.88, p<.001). Decline greatest for moderate deterioration group.
MfS	Memory for sentences (from the NAID or SBIS)	Memory – verbal short-term	YES	YES	NO	DK	YES	Ball et al., 2008	Affected by ID severity. DS sig more impaired than control group.
								Brugge et al., 1994	Memory-impaired-DS group scored sig lower (p<.01) than non-memory-impaired-DS group. Non-memory-impaired-DS group did not score differently than controls.
								Carr, 2003	Mean scores ↑ over time, though not sig.
NDT	New Dot Test	Memory - visuospatial memory						Sano et al., 2005	Too difficult for moderate to severe ID, with floor effects.
ODL	Object discrimination learning	Learning - conditioned learning						Nelson et al., 2005	Relationship between errors and DMR approached significance (p=.053). Errors not associated with age.
PAL	Paired Associate Learning task from the CANTAB	Spatial associative memory	YES	YES	NO	DK	YES	Sinai et al., 2016	Stages completed score significant difference between dementia and no-dementia group (p=.011), but floor effects noted in dementia group.
Precog	Picture recognition subtest from the RBMT-C	Memory	NO	NO	NO	NO	NO	Carr, 2003	Mean scores declined over time but not sig. Floor and ceiling effects noted.
PRMT/ r-PRMT	Picture Recognition Memory Test/ revised Picture Recognition Memory Test	Memory - immediate and delayed recognition	NO	NO	NO	DK	YES	Pyo et al., 2007	Majority with AD could be differentiated from controls using PRMT.
								Pyo et al., 2010	DS-controls showed ↑ scores, vs. AD on r-PRMT, with no overlap. oID-controls scored much ↓ (significant overlap) vs. AD group. r-PRMT discriminated between AD and no-AD in DS and moderate to severe ID, but not oID, when assessed at a single time point.
RaB	Remembering a belonging subtest of the RBMT-C	Memory – prospective	NO	NO	NO	NO	NO	Ball et al., 2008	Floor and ceiling effects. Not suitable for tracking decline over time due to floor effects. Less sensitive to effects of severity of ID (d<0.5). Scores ↓ with advancing age. Found to be associated with EF (p<.05). Informant reported personality/ behaviour changes remained a significant predictor of test score (beta=-0.28, t=-2.78, p<.05) after non-executive memory tasks removed from analysis.
								Carr, 2003	Slight ↑ in scores between ages 30 and 35 years. Some floor and ceiling effects, but not sig.

Abbreviations	List of Measures	Function/ Cognitive domain(s)	A	ID	DS	NV	CD	Author(s)	Test Findings
Rapp	Remembering an appointment subtest from the RBMT-C	Memory	NO	NO	NO	NO	NO	Carr, 2003	Slight ↑ in scores between ages 30 and 35 years, but not sig. Some floor and ceiling effects.
Rnames	Remembering first and second names subtest from the RBMT-C	Memory	NO	NO	NO	NO	NO	Carr, 2003	Floor and ceiling effects. Slight ↑ in scores between ages 30 and 35 years, but not sig.
Rroute	Remembering a route from the RBMT-C	Memory	NO	NO	NO	NO	NO	Carr, 2003	Some floor effects.
StoryRT	Story Recall Test (adapted from the RBMT-C)		NO	NO	NO	NO	NO	Carr, 2003	Floor effects. Lowest number of ceiling effects, indicating more difficult than the other memory subtests in the RBMT-C. Mean scores declined over time but not sig.
								Cooper et al., 2016	Poor completion rates.
SVDL	Simple visual discrimination learning	Learning and memory - visual discrimination and conditioned learning						Nelson et al., 2007	Poor test re-test reliability (r =.24, p<.33).
VAT	Visual Association Test	Short-term memory	YES	NO	NO	NO	YES	McPaul et al., 2017	Participants scored well on VAT, irrespective of age, gender, or IQ. No significant correlations between VAT and CAMCOG-DS. VAT = easy and quick test, though showed poor internal consistency. No floor effects on VAT.
VisMT	Visual Memory Test – delayed-match-to-sample	Memory - visual matching						Devenny et al., 1992	No significant changes in test scores between baseline and f/u up to 5 years later for any of the groups. All groups showed ↑ in performance tasks from 1st to 2nd testing on memory. No functional deterioration or age-related memory decline in adults with DS.
								Devenny et al., 1996	Affected by level of IQ. Ceiling effects on the simultaneous condition. Longer delays between target presentations and response choices produced ↓ performance. No differences between diagnostic or age groups with respect to overall proficiency of performance, participants with DS had a significantly steeper slope on the delay condition (r=-1.982, p=.471), indicating that ↑the amount of delay produced greater ↓ in performance.
VR	Visual Reproduction subtest from WMS-R		YES	NO	NO	NO	NO	Brugge et al., 1994	Memory-impaired-DS group scored sig lower (p<.05) than non-memory-impaired-DS group on immediate recall.
Sensorimotor									
AoR	Action on request subtest from the CODB/ NAID	Praxis - learned motor activity	YES	YES	NO	DK	YES	Carr, 2003	Mean scores declined over time but not sig.
BPT	The Brief Praxis Test	Praxis - learned motor activity	YES	YES	YES	DK	YES	Head et al., 2011	A diagnosis of dementia (F(1,55)=9.08, p=.004) and more profound cognitive impairment (F(2,55)=21.32, p<.0005) both contribute to lower mean BPT test scores. ↑ plasma Aβ1-40 and Aβ1-42 was associated with ↓ scores.
								Powell et al., 2014	

Abbreviations	List of Measures	Function/ Cognitive domain(s)	A	ID	DS	NV	CD	Author(s)	Test Findings
									↓ scores on BPT correlated with neuropathological findings (frontoparietal regions). BPT was sensitive to functional ↓ in DS due to dementia.
								Sano et al., 2005	
									BPT showed sensitivity to change over time with minimal floor and ceiling effects.
								Wallace et al., 2021	
									BPT showed fair discriminative ability between no/ possible vs. probable dementia. Could not discriminate between no- and possible dementia. No floor effects.
								Walsh et al., 2015	
									Dementia ↓ scores.
BBDT-VMI	Beery Buktenica Developmental Test of Visual-Motor Integration	Visual-motor Integration	YES	YES	NO	YES	YES	Brugge et al., 1994	No data reported.
								Hom et al., 2021	No sig difference between MCI-DS group vs. CS group (*U*=1.75, *p*=.081).
								Krinsky-McHale et al., 2020	Significant declines emerged only with dementia onset, indicating insensitive to early decline.
FS	Finger Sequencing	Upper limb co-ordination						Sinai et al., 2016	No significant differences between dementia and no-dementia groups. Minimal floor and ceiling effects.
FT	Finger Tapping from the Halstead-Reitan Battery	Sensorimotor	YES	NO	NO	NO	NO	Brugge et al., 1994	The memory-impaired-DS group was significantly worse than control subjects on DifferTap (*p*<.01), but no sig difference found between memory-impaired-DS and non-memory-impaired-DS. Affected by advancing age.
GA	Gait Assessment (Timed Get Up and Go Test)	Gait assessment						Sinai et al., 2016	No significant differences between the dementia and no dementia groups.
PPT	Purdue Pegboard Test	Dexterity and coordination	YES	NO	NO	YES	NO	Hom et al., 2021	PPT both hands score significantly ↓ in MCI-DS group vs. CS group (*U*=3.14, *p*=.002]).
SRTime	Simple Reaction Time from the CANTAB	Cerebellar function and attention	YES	YES	NO	DK	YES	Sinai et al., 2016	Statistically significant differences between the dementia and no dementia groups on CANTAB Simple Reaction Time median latency (*p*=.049).
VP	Visuomotor precision from the NEPSY	Visuo-motor tracking and hand-eye coordination	NO	NO	NO	YES	NO	Sinai et al., 2016	Statistically significant differences between the dementia and no dementia groups for Car and Motorbike (*p*=.013) but not Train and Car (*p*=.303).

Note. Top row - A = developed for the adult population; CD = developed to detect cognitive decline/ dementia; DS = developed specifically for people with Down syndrome; ID = developed specifically for people with an intellectual disability; NV = suitable for people who are non-verbal

↑ = higher/ increase; ↓ = lower/ decline; < = less than; > = more than; ?AD = Possible AD; AD = Dementia of the Alzheimer's Type Dementia; Aβ = Amyloid-beta; CS = cognitively stable; DS = Down syndrome; EF = executive function; f/u = follow-up; HC = Healthy controls; IM = Immediate memory; IQ = Intelligent quotient; LT = Long-term; LTM = Long-term memory; m = months; MCI = Mild cognitive impairment; oID = Intellectual disability from other causes than DS; PPV = Positive Predictive Value; SBIS = Stanford-Binet Intelligence Scales; SD = standard deviation; TD = Typically Developing; WM = Working Memory; WMS-R = The Wechsler Memory Scale-Revised

Appendix D

Full List of Instruments (Including Informant-Rated and Adaptive)

Abbreviation	Name of Instrument or Task	Author(s)
A	Arithmetic subtest from the WISC-R	Devenny et al., 2000
ABS	Adaptive Behavior Scale	Cooper et al., 2016
ABS-RC:2	Adaptative Behavior Scale-Residential and Comunity-2nd edition	García-Alba et al., 2017
ADVM	Auditory delayed verbal memory	García-Alba et al., 2019
AMT	Autobiographical Memory Test	Pyo et al., 2007; Pyo et al., 2011
AoR	Acting on request	Carr, 2003
ASL or landmark	Allocentric spatial learning or landmark	Nelson et al., 2007
BADLS	Bristol Activities of Daily Living Scale	Walsh et al., 2015
BBDT-VMI	Beery Buktenica Developmental Test of Visual-Motor Integration	Hom et al., 2021; Krinsky-McHale et al., 2020
BD	Block Design	Hom et al., 2021; Krinsky-McHale et al., 2020
BMT	Buschke Memory test	Devenny et al., 1992
BNT	The Boston Naming Test	Hom et al., 2021; Palmer, 2006
BPT	The Brief Praxis Test	Head et al., 2011; Powell et al., 2014; Sano et al., 2005; Wallace et al., 2021; Walsh et al., 2015
BPVS	British Picture Vocabulary Scale	Ball et al., 2008
BRIEF	Behaviour Rating Inventory of Executive Function	Esteba-Castillo et al., 2022; García-Alba et al., 2017; Sinai et al., 2016
BSRT	Busckke selective reminding test and modified versions	Devenny et al., 1992; Devenny et al., 1996; Devenny et al., 2000; Hom et al., 2021; Krinsky-McHale, Devenny & Silverman, 2002; Krinsky-McHale et al., 2008; Krinsky-McHale et al., 2020
BT-ID	Barcelona Test - ID	Esteba-Castillo et al., 2022; García-Alba et al., 2017
CaD	Cats and Dogs	Ball et al., 2008; Bevins & Hurse, 2014; Cooper et al., 2016; Hom et al., 2021
CaF	Cats and frogs (modified dots task)	Sinai et al., 2016
CAMCOG	Cambridge Cognition Examination	Ball et al., 2008; Esteba-Castillo et al., 2022; Oliver et al., 1998
CAMCOG-DS	Cambridge Cognitive Examination adapted for individuals with Down Syndrome	Benejam et al., 2020; Esteba-Castillo et al., 2022; García-Alba et al., 2019; McPaul et al., 2017
CAMDEX	Cambridge Mental Disorders of the Elderly Examination	Ball et al., 2008
CAMDEX-DS	Cambridge Examination for Mental Disorders of Older People with Down Syndrome and Others with Intellectual Disabilities	Benejam et al., 2020; Esteba-Castillo et al., 2022; García-Alba et al., 2017
CANTAB	Cambridge Neuropsychological Test Automated Battery	Cooper et al., 2016; Oliver et al., 1998; Sinai et al., 2016
CFT	Category Fluency Test	Cooper et al., 2016; Hom et al., 2021
CLD	Checklist with Symptoms of Dementia	Hoekman & Maaskant, 2002
COWAT	The Controlled Oral Word Association Test	Palmer, 2006

Abbreviation	Name of Instrument or Task	Author(s)
CRT	The Cued Recall Test and modified versions	Benejam et al., 2015; Cooper et al., 2016; Devenny et al., 2000; Devenny et al., 2002; Hom et al., 2021; Sacco et al., 2022
CT	Cancellation task	Cooper et al., 2016; Krinsky-McHale et al., 2008
CTT	The Colour Trails Test	Palmer, 2006
CUSPAD	Columbia University Scale to Assess Psychopathology in Alzheimer's Disease	Krinsky-McHale et al., 2020
DLD/ DMR	Dementia Questionnaire for People with Learning Disabilities	Bevins & Hurse, 2014; Head et al., 2011; Hoekman & Maaskant, 2002; Krinsky-McHale et al., 2020; Nelson et al., 2005; Nelson et al., 2007; Sinai et al., 2016; Wallace et al., 2021; Walsh et al., 2015
DMTS	Delayed match-to-sample	Hoekman & Maaskant, 2002
DNMP	Spatial delayed non-match-to-position	Nelson et al., 2005
DNMS	Object delayed non-match-to-sample	Nelson et al., 2005; Nelson et al., 2007
Drecall	Delayed recall	Ball et al., 2008
Drecog	Delayed recognition	Ball et al., 2008
DSpan	Digit Span	Manning et al., 1998
DSDS	Dementia scale for Down Syndrome	Krinsky-McHale, Devenny & Silverman, 2002; Krinsky-McHale et al., 2008; Nelson et al., 2001
DSMSE	Down Syndrome Mental State Examination	Hom et al., 2021; Krinsky-McHale et al., 2020; Manning et al., 1998
DSQID	Dementia Screening Questionnaire for Individuals with Intellectual Disabilities	Sacco et al., 2022
DVM	Delayed visual memory	García-Alba et al., 2019
EMS	Evaluation of Mental Status	Devenny et al., 1992
EOWPVT/ EOWPVT-R	Expressive One-Word Picture Vocabulary Test/ Expressive One-Word Picture Vocabulary Test-Revised	Sano et al., 2005
ESDCL	Early Signs of Dementia Checklist	Palmer, 2006
F-NT	Finger-Nose Test	Sinai et al., 2016
FOME	The Fuld Object-Memory Evaluation	Palmer, 2006; Pyo et al., 2007; Sano et al., 2005
Frecog	Face recognition subtest from the RBMT-C	Carr, 2003
FS	Finger Sequencing	Sinai et al., 2016
FSBS	Frontal Systems Behavior Scale	Fonseca et al., 2019b
GA	Gait Assessment	Sinai et al., 2016
HBS	Handicaps, behaviour, and skills schedule	Carr, 2003
IED	Intra-Extra Dimensional shift subtest from the CANTAB	Sinai et al., 2016
IM	Immediate Memory	Ball et al., 2008
Info	Information subtest from the WISC-R	Devenny et al., 2000
K-BIT	Kaufman Brief Intelligence Test	Benejam et al., 2015; García-Alba et al., 2017
M-CFT	McCarthy Category Fluency Test	Krinsky-McHale et al., 2020

Abbreviation	Name of Instrument or Task	Author(s)
MfO	Memory for objects	Ball et al., 2008; Carr, 2003; Cooper et al., 2016; Oliver et al., 1998
MfP	Memory for pictures	Carr, 2003; Oliver et al., 1998
MfS	Memory for sentences	Ball et al., 2008; Carr, 2003; Sinai et al., 2016
mMMSE-DS	Modified Mini Mental Status Evaluation— Down Syndrome	Hom et al., 2021; Krinsky-McHale et al., 2020
mOMT	Modified Objective Memory Test	Pyo et al., 2010
NBAP	Neuropsychology Behavior and Affect Profile	Nelson et al., 2001
OA	Object Assembly from WISC-R	Devenny et al., 2000
O-CAMCOG	Orientation subtest from the CAMCOG	Oliver et al., 1998
O-MMSE	Orientation subtest from the MMSE	Sano et al., 2005
O-OCDB	Orientation subtest from the OCDB/ NAID	Carr, 2003
PA	Picture Arrangement from the WISC-R	Devenny et al., 2000
PAL	Paired Associate Learning task from the CANTAB	Sinai et al., 2016
PC	Picture Completion from the WISC-R	Devenny et al., 2000
PN	Picture naming	Carr, 2003
PPVT/ PPVT-R	Peabody Picture Vocabulary Test/ Peabody Picture Vocabulary Test-Revised	Manning et al., 1998; Nelson et al., 2001; Pyo et al., 2007
PR	Pattern recognition	Cooper et al., 2016
Precog	Picture recognition subtest from the RBMT-C	Carr, 2003
PRMT/r-PRMT	Picture Recognition Memory Test/ revised Picture Recognition Memory Test	Pyo et al., 2007; Pyo et al., 2010
RADD/RADD-2	The Rapid Assessment of Developmental Disabilities/Second Edition	Hom et al., 2021; Walsh et al., 2015
RaB	Remembering a belonging subtest of the RBMT-C	Ball et al., 2008; Ball et al., 2010; Carr, 2003
Rapp	Remembering an appointment	Carr, 2003
RBMT-C	Rivermead Behavioural Memory Test for Children	Ball et al., 2008; Carr, 2003; Cooper et al., 2016; Hom et al., 2021
RL	Reversal learning	Nelson et al., 2005; Nelson et al., 2007
Rnames	Remembering Names subtest from the RBMT-C	Carr, 2003
Rroute	Remembering a route from the RBMT-C	Carr, 2003
RSMB	Reiss Screen for Maladaptive Behaviour	Krinsky-McHale et al., 2020
SB	Scrambled Boxes	Ball et al., 2008
Si	Similarities subtest from the WISC-R	Devenny et al., 2000
SIB	Severe Impairment Battery	Head et al., 2011; Powell et al., 2014; Wallace et al., 2021; Walsh et al., 2015
Sreversal	Spatial Reversal	Ball et al., 2008

Abbreviation	Name of Instrument or Task	Author(s)
SRT/mSRT	The Selective Reminding Test/ Modified - The Selective Reminding Test	Cooper et al., 2016; Devenny et al., 2000; Hom et al., 2021; Krinsky-McHale, Devenny & Silverman, 2002; Krinsky-McHale et al., 2008; Krinsky-McHale et al., 2020; Sinai et al., 2016
SRTime	Simple Reaction Time from the CANTAB	Sinai et al., 2016
SVDL	Simple visual discrimination learning	Nelson et al., 2007
TBGAT	Tinetti Balance and Gait Assessment Tool	Hom et al., 2021
TO	Temporal Orientation	García-Alba et al., 2019
ToL	Tower of London	Ball et al., 2008; Cooper et al., 2016; Sinai et al., 2016
TOL^{DX}	Tower of London-Drexel University: 2nd Edition	Esteba-Castillo et al., 2022; García-Alba et al., 2017
TSI	Test for Severe Impairment	Krinsky-McHale et al., 2020; Pyo et al., 2007; Pyo et al., 2010
VABS	Vineland Adaptive Behaviour Scales	Oliver et al., 1998
VAT	Visual Association Test	McPaul et al., 2017
VC	Verbal comprehension subtest from the WISC-R	Devenny et al., 2000
VF	Verbal Fluency	Ball et al., 2008; Sinai et al., 2016
VisMT	Visual Memory Test	Devenny et al., 1992
VMT	Verbal Memory Test	Oliver et al., 1998
VoC	Vocabulary subtest from the WISC-R	Devenny et al., 2000
VP	Visuomotor precision from the NEPSY	Sinai et al., 2016
VT	Vocabulary Test	Sano et al., 2005
WAIS/WAIS-R	Wechsler Adult Intelligence Scale/Revised	McPaul et al., 2017; Nelson et al., 2005; Nelson et al., 2007
WCFST	Weigl Colour-Form Sort Test	Ball et al., 2008; Bevins & Hurse, 2014; Esteba-Castillo et al., 2022; García-Alba et al., 2017
WGTA	Wisconsin General Testing Apparatus	Nelson et al., 2005
WISC/WISC-R	Wechsler Intelligence Scale for Children/Revised	Devenny et al., 2000; Hom et al., 2021; Krinsky-McHale et al., 2020
WM	Working Memory	García-Alba et al., 2019
WMS-R	The Wechsler Memory Scale-Revised	Brugge et al., 1994
WR	Word Recall	Ball et al., 2008

Appendix E
HRA Ethics Approval Letter

Ymchwil Iechyd a Gofal Cymru
Health and Care Research Wales

NHS
Health Research Authority

Dr Matthew Jones-Chesters
Senior Lecturer
The University of East London
UEL School of Psychology
Water Lane
London
E15 4LZ

Email: approvals@hra.nhs.uk
HCRW.approvals@wales.nhs.uk

10 August 2022

Dear Dr Jones-Chesters

HRA and Health and Care Research Wales (HCRW) Approval Letter

Study title:	**Assessment of cognition in people with intellectual disabilities using a novel set of neuropsychological tests**
IRAS project ID:	**295654**
REC reference:	**22/WA/0238**
Sponsor	**University of East London**

I am pleased to confirm that **HRA and Health and Care Research Wales (HCRW) Approval** has been given for the above referenced study, on the basis described in the application form, protocol, supporting documentation and any clarifications received. You should not expect to receive anything further relating to this application.

Please now work with participating NHS organisations to confirm capacity and capability, in line with the instructions provided in the "Information to support study set up" section towards the end of this letter.

How should I work with participating NHS/HSC organisations in Northern Ireland and Scotland?
HRA and HCRW Approval does not apply to NHS/HSC organisations within Northern Ireland and Scotland.

If you indicated in your IRAS form that you do have participating organisations in either of these devolved administrations, the final document set and the study wide governance report (including this letter) have been sent to the coordinating centre of each participating nation. The relevant national coordinating function/s will contact you as appropriate.

IRAS project ID	295654

Information to support study set up

The below provides all parties with information to support the arranging and confirming of capacity and capability with participating NHS organisations in England and Wales. This is intended to be an accurate reflection of the study at the time of issue of this letter.

Types of participating NHS organisation	Expectations related to confirmation of capacity and capability	Agreement to be used	Funding arrangements	Oversight expectations	HR Good Practice Resource Pack expectations
There is only one participating NHS organisation therefore there is only one site type.	Research activities should not commence at participating NHS organisations in England or Wales prior to their formal confirmation of capacity and capability to deliver the study in accordance with the contracting expectations detailed.	An Organisation Information Document has been submitted and the sponsor is not requesting and does not expect any other agreement to be used with participating NHS organisations of this type.	The sponsor has detailed its proposals with respect to whether any study funding will be provided to participating NHS organisations of this type in the relevant Organisational Information Document. This should be read in conjunction with the relevant Schedule of Events/SoECAT which details the cost implications of the study for participating NHS	In line with HRA/HCRW expectations a Local Collaborator should be appointed at participating NHS organisations of this type.	No Honorary Research Contracts, Letters of Access or pre-engagement checks are expected for local staff employed by the participating NHS organisations. Where arrangements are not already in place, research staff not employed by the NHS host organisation undertaking any of the research activities listed in the research application would be expected to obtain a Letter of Access based on standard DBS checks and occupational health clearance.

			organisations.		

Other information to aid study set-up and delivery

This details any other information that may be helpful to sponsors and participating NHS organisations in England and Wales in study set-up.
The applicant has indicated that they do not intend to apply for inclusion on the NIHR CRN Portfolio

Appendix F
UEL EISC Ethics Approval Letter

University of East London
Pioneering Futures Since 1898

4th April 2023

Dear Danielle,

Project Title:	**Assessment of cognition in people with intellectual disabilities using a novel set of neuropsychological tests**
Researcher(s):	**Danielle Pearce**
Principal Investigator:	**Danielle Pearce**

I am writing to confirm that the application for the aforementioned NHS research study reference **22/WA/0238,** IRAS project ID: **295654** has received ethical approval from the Ethics and Integrity Sub-Committee (EISC) and is sponsored by the University of East London.

The lapse date for ethical approval for this study is **4th March 2027**. If you require EISC approval beyond this date you must submit satisfactory evidence from the NHS confirming that your study has current NHS R&D ethical approval and provide a reason why EISC approval should be extended.

Please note as a condition of your sponsorship by the University of East London your research must be conducted in accordance with NHS regulations and any requirements specified as part of your NHS R&D ethical approval.

Please ensure you retain this approval letter, as in the future you may be asked to provide proof of ethical approval.

With the Committee's best wishes for the success of this project.

Yours sincerely,

Catherine Hitchens, Ethics, Integrity and Compliance Manager
For and on behalf of
Professor Winston Morgan
Deputy Chair, Ethics and Integrity Sub-Committee (EISC)
Email: researchethics@uel.ac.uk

Appendix G
Participant Consent Form

CONSENT FORM v1.03 (07.08.2022) IRAS ID: 295654

Participant Identification Number:

University of East London
School of Psychology
Assessment of Cognition in People with Intellectual Disabilities – Participant Consent Form
V07.08.2022

		Please Initial
1.	I confirm that I have read and understood the information sheet for this study (Participant Invitation Letter V1.03), and I have been given a copy of this to keep.	
2.	I confirm that the nature and purposes of this study have been explained to me, and I have been able to ask questions that have been answered to my satisfaction.	
3.	I understand that my involvement in this study and data produced will remain strictly confidential. I understand that only the researcher conducting this study will have access to identifiable information. The researcher has explained what will happen to my data once the research study has been completed. I understand what will happen to my data once the research study has been completed.	
4.	I understand that my participation in this study is entirely voluntary and that I am free to withdraw from the study at any time without having to give a reason.	
5.	I understand that I am entitled to a break in the middle of testing, where snacks and drinks will be provided for me. I understand that I am also entitled to unlimited additional rest breaks upon request.	
6.	I understand that I will be video recorded during my participation, and this will be used for data analysis. I consent to being video recorded for participation in this research.	
7.	I understand that the recording device or tests can be stopped at any time without giving a reason. I understand that if I request to stop the recording devices or tests, tat I will be offered a debrief and my data will be safely destroyed. I understand that this will not affect my receipt of a £10 Amazon gift voucher.	

Appendix H
Carer Consent Form

CARER CONSENT FORM v1.03 (07.08.2022) IRAS ID: 295654

Participant Identification Number:

University of East London
School of Psychology
Assessment of Cognition in People with Intellectual Disabilities – Carer Consent Form
V: 07.08.2022

		Please Initial
1.	I confirm that I have read and understood the information sheet for this study (Carer Information Letter V1.03), and I have been given a copy of this to keep.	
2.	I confirm that the nature and purposes of this study have been explained to me, and I have been able to ask questions that have been answered to my satisfaction.	
3.	I understand that I have been asked to accompany my child/ relative/ friend during the study as their guardian and advocate, to ensure the study treats them fairly and with respect at all times.	
4.	I understand that my child/ relative/ friend's involvement in this study and data produced will remain strictly confidential. I understand that only the researcher conducting this study will have access to identifiable information. The researcher has explained what will happen to my child/ relative/ friend's data once the research study has been completed. I understand what will happen to my child/ relative/ friend's data once the research study has been completed.	
5.	I understand that my child/ relative/ friend's participation in this study is entirely voluntary and that they are free to withdraw from the study at any time without having to give a reason.	
6.	I understand that my child/ relative/ friend is entitled to a break in the middle of testing, where snacks and drinks will be provided. I understand that they are also entitled to unlimited additional rest breaks upon request.	
7.	I understand that my child/ relative/ friend will be video recorded during their participation, and this will be used for data analysis. I consent to my child/ relative/ friend being video recorded for participation in this research.	

Participant Identification Number:

8.	I understand that the recording device or tests can be stopped at any time without giving a reason. I understand that if my child/ relative/ friend requests to stop the recording devices or tests, that we will be offered a debrief and their data will be safely destroyed. I understand that this will not affect their receipt of a £10 Amazon voucher.	
9.	I understand that my child/ relative/ friend can choose to withdraw their data from this study at any point **up to 3 weeks after participating.** I understand that after 3 weeks from their participation date, the researcher reserves the right to use their anonymous data in the analysis for this study.	
10.	**Given the above points, I hereby freely consent to my accompaniment to my child/ relative/ friend's participation in this study.**	

Participant's Name (BLOCK CAPITALS)	Date	Signature
Carer's Name (BLOCK CAPITALS)	Date	Signature
Researcher's Name (BLOCK CAPITALS)	Date	Signature

Appendix I
Study Recruitment Poster

PARTICIPANT POSTER v1.03 (07.08.2022) IRAS ID:295654

Are you a person with a learning disability aged between 30 and 55 years old?

Are you currently receiving care from an [redacted] NHS Learning Disabilities service?

Hello! My name is Danielle. I am a student in the School of Psychology at the University of East London, and I am studying for a Doctorate in Clinical Psychology. As part of my studies, I am doing the research described in this poster. To do my research, I need people with a learning disability who are aged between 30 and 55 years old, who do not have a diagnosis of dementia, and who come to an [redacted] NHS Learning Disability Service, to take part in my study.

What would I have to do?

If you take part in my study, you would participate in some tasks that I have created named 'cognitive tests'. Don't worry, this is not like a school test! I will ask you to do some short tasks, like answering questions, drawing things or copying what I do back to me. I would then ask for your feedback on the test. If you participate, your data will be anonymous, which means nobody but me will know who you are when the data is published in my research paper.

Why are you doing this research?

There are currently no tests for dementia that have been created for people with a learning disability that accurately look at all the different things that the brain can do accurately. I would like to make a test for people with a learning disability that feels engaging and takes into account what people with a learning disability think about it, so I can make the tests better.

What will I get for taking part?

If you choose to take part, you will help to improve the tests I have created for other people with a learning disability in the future. You will also get a £10 Amazon gift voucher to say thank you for your time.

How can I take part?

If you would like to take part, please speak to your health care professional at [redacted] Learning Disability Service or ask you parent/carer/guardian to let them know you are interested. They will put you in touch with me.

Thank you for reading! ☺

Appendix J
Participant Invitation Letter

INVITATION LETTER v1.03 (07.08.2022) IRAS ID: 295654

Participant Identification Number:

University of East London
School of Psychology
Assessment of Cognition in People with Intellectual Disabilities – Participant Invitation Letter
V07.08.2022

You are being invited to participate in a research study. Before you agree to take part, it is important that you understand what your participation would involve. Please read the following information carefully before deciding.

Who am I?
My name is Danielle, I am a student in the School of Psychology at the University of East London and am studying for a Doctorate in Clinical Psychology. As part of my studies, I am conducting the research you are being invited to participate in.

What is the research?
I am conducting research into making an assessment tool which can see if somebody with a learning disability may also be experiencing dementia. Dementia is when someone experiences a loss of memory, language, problem-solving and other thinking abilities that may make daily life more difficult. There are many different types, the most common kind of dementia is called Alzheimer's. I would like to investigate whether dementia looks different in people who have a learning disability, so that we can identify it sooner and help people who experience it have better support and quality of life.

My research has been approved by an independent NHS Research Ethics Committee. This means that my research follows the standard of research ethics set by the British Psychological Society.

Participant Identification Number:

Why have you been asked to participate?

You have been invited to participate in my research as I am looking to involve people who have a learning disability and are aged between 30-55, to help me explore my research topic.

You will not be judged or personally analysed in any way and you will be treated with respect at all times.

You do not have to say 'yes' to taking part, and there will be no consequences if you decide not to take part. You are free to choose what feels most comfortable to you.

What will your participation involve?

You will be asked to attend a 'testing session' with me. I will ask you to complete a series of short tasks including questionnaires and other short exercises exploring various skills and abilities including language, thinking and puzzle-solving. Some of these will involve me asking you questions, and others are pen-and-paper tasks, and some may involve you following instructions. Tasks with verbal answers will be video recorded so that your answers can be accurately scored and analysed. This will be safely stored on a password-protected computer and destroyed once the research has finished.

This will take around 1 hour. We will take a break in the middle where you can have some snacks and drinks that I will provide for you, and you can also take short breaks in between the different tasks if you wish. If you need, we could have two shorter sessions on two different days. I will also ask you to tell me what you thought of the tests, including what you think worked well and how you think I could make any of them better. This will take around half an hour and can be done on the same day as the tests or a different day.

This will take place in a private room at xxxxx NHS Learning Disability Service at a time we decide in advance, that fits for us both.

What are the potential risks and disadvantages of taking part?

Though we do not anticipate any negative effects of participation, some may arise. Testing may make you feel tired, which could lead to headaches. Taking part in some of the tests could also feel stressful. We will remind you throughout to take breaks if you need and provide refreshments while you take part in the testing session. We will also provide you with services and organisations you can contact at the bottom of this sheet, and in a debrief letter.

Participant Identification Number:

What are the potential benefits of taking part?

As a thank you for your time, you will be given a £10 Amazon voucher. There may not be any specific benefits to yourself in participating, but by taking part you can help to create tests for dementia which are better suited to other people with a learning disability in the future.

How will we use information about you?

We will need to use information from you for this research project.
This information will include:

- Your name
- Your mobile number or email address (to get in touch with you)
- The responses you give to the tests we try out together.

People will use this information to do the research or to check your records to make sure that the research is being done properly. People who do not need to know who you are will not be able to see your name or contact details. Your data will have a code number instead. We will keep all information about you safe and secure.

Once we have finished the study, we will keep some of the data so we can check the results. We will write our reports in a way that no-one can work out that you took part in the study.

The video recordings that we take of you completing the verbal tests will not be fully transcribed and will only be used to write down the answers you give on the tests. We will use this video to record your answers within one week of you completing the tests. After one week, the video recording will be safely destroyed.

What are your choices about how your information is used?

You can stop being part of the study at any time, without giving a reason, but we will keep information about you that we already have. We need to manage your records in specific ways for the research to be reliable. This means that we won't be able to let you see or change the data we hold about you.

After the study has been completed, your data will continue to be stored in a secure location, only accessible by the research team, for 10 years, as recommended by the UK Research and Innovation (UKRI) guidelines. After this, all data will be destroyed.

If you wish, I can provide you with a copy of the results of this study once it is finished.

What if you want to withdraw?

You are free to withdraw from the research study at any time without explanation, disadvantage or consequence. If you tell me that you would like to stop the video recording, any of the tests, or the discussions we are having at any point, we will stop these immediately. You will be offered the chance to have a talk about how you are feeling with me (this is called a debrief) and I will give you some resources of other people to speak to also. Any data collected about you, on paper, computer, or video, will be immediately and safely destroyed.

Separately, you may also request to withdraw your data even after you have participated, provided that this request is made within **3 weeks** of the data being collected. After 3 weeks, your name and other identifiable information will be deleted and your data will only be referred to by a numerical code, meaning we will no longer be able to identify which is your data.

If during your participation in the study you lose the ability to consent, we will immediately stop testing and offer you and your carer/guardian a debrief. We will then immediately and safely destroy your data, and you will no longer be included in the study. You will still receive a £10 Amazon voucher for your time.

Where can you find out more about how your information is used?

You can find out more about how we use your information

- at www.hra.nhs.uk/information-about-patients/
- by asking one of the research team
- by sending an email to xxxxx, or
- by ringing us on xxxxx

If you have any questions or concerns about how the research has been conducted, please contact:

- The research supervisor:
  Dr. Matthew Jones-Chesters,
  School of Psychology, University of East London, Water Lane, London E15 4LZ
  Email: xxxxx
  Phone: xxxxx

or

- Chair of the School of Psychology Research Ethics Sub-committee:
  Dr Trishna Patel

Participant Identification Number:

School of Psychology, University of East London, Water Lane, London E15 4LZ.
Email: xxxxx

Appendix K
Carer Invitation Letter

CARER INVITATION LETTER v1.03 (07.08.2022) IRAS ID: 295654

Participant Identification Number:

University of East London
School of Psychology
Assessment of Cognition in People with Intellectual Disabilities – Carer Invitation Letter
V07.08.2022

Your child/ relative/ friend is being invited to participate in a research study. We have asked you to accompany them as their guardian and advocate. Before they agree to take part, it is important that you understand what their participation would involve. Please read the following information carefully before deciding.

Who am I?
My name is Danielle, I am a student in the School of Psychology at the University of East London and am studying for a Doctorate in Clinical Psychology. As part of my studies, I am conducting the research your child/ relative/ friend is being invited to participate in.

What is the research?
I am conducting research into making an assessment tool which can see if somebody with a learning disability may also be experiencing dementia. Dementia is when someone experiences a loss of memory, language, problem-solving and other thinking abilities that may make daily life more difficult. There are many different types, the most common kind of dementia is called Alzheimer's. I would like to investigate whether dementia looks different in people who have a learning disability, so that we can identify it sooner and help people who experience it have better support and quality of life.

My research has been approved by an independent NHS Research Ethics Committee. This means that my research follows the standard of research ethics set by the British Psychological Society.

Participant Identification Number:

Why has my child/ relative/ friend been asked to participate?
Your child/ relative/ friend has been invited to participate in my research as someone who has a learning disability and is aged between 30-55.

Your child/ relative/ friend will not be judged or personally analysed in any way and will be treated with respect at all times.

They do not have to say 'yes' to taking part, and there will be no consequences if they decide not to take part. They are free to choose what feels most comfortable to them.

What will their participation involve?
They will be asked to attend a 'testing session' with me. I will ask them to complete a series of short tasks including questionnaires and other short exercises exploring various skills and abilities including language, thinking and puzzle-solving. Some of these will involve me asking them questions, and others are pen-and-paper tasks, and some may involve them following instructions. Tasks with verbal answers will be video recorded so that their answers can be accurately scored and analysed. This will be safely stored on a password-protected computer and destroyed once the research has finished.

This will take around 1 hour. We will take a break in the middle where you and your child/ relative/ friend can have some snacks and drinks that I will provide for you, and your child/ relative/ friend can also take short breaks in between the different tasks if you wish. If your child/ relative/ friend needs, we could have two shorter sessions on two different days. I will also ask your child/ relative/ friend to tell me what they thought of the tests, including what they think worked well and how they think I could make any of them better. This will take around half an hour, and can be done on the same day as the tests or a different day.

This will take place in a private room at xxxxx NHS Learning Disability Service at a time we decide in advance, that fits for us all.

What are the potential risks and disadvantages of taking part?

Though we do not anticipate any negative effects of participation, some may arise. Testing may make your child/ relative/ friend feel tired, which could lead to headaches. Taking part in some of the tests could also feel stressful. We will remind your child/ relative/ friend throughout to take breaks if they need, and provide refreshments while they take part in the testing session. We will also provide you both with services and organisations you can contact at the bottom of this sheet, and in a debrief letter.

Participant Identification Number:

What are the potential benefits of taking part?
As a thank you for your child/ relative/ friend's time, they will be given a £10 Amazon voucher. There may not be any specific benefits to them in participating, but by taking part they can help to create tests for dementia which are better suited to other people with a learning disability in the future.

What will happen to the information that my child/ relative/ friend provides?
We will need to use information from your child/ relative/ friend for this research project.

This information will include:

- Their name
- Their mobile number or email address (to get in touch with them)
- The responses they give to the tests we try out together.

We will also ask for information from you, which will include:

- Your name
- Your mobile number or email address (in case it is preferable to get in touch with you)

People will use this information to do the research or to check your records to make sure that the research is being done properly. People who do not need to know who you are will not be able to see your name or contact details. Your child/ relative/ friend's data will have a code number instead.

We will keep all information about your child/ relative/ friend safe and secure.
Once we have finished the study, we will keep some of the data so we can check the results. We will write our reports in a way that no-one can work out that your child/ relative/ friend took part in the study.

The video recordings that we take of your child/ relative/ friend completing the verbal tests will not be fully transcribed, and will only be used to write down the answers they give on the tests. We will use this video to record their answers within one week of them completing the tests. After one week, the video recording will be safely destroyed.

What are your choices about how your information is used?
Your child/ relative/ friend can stop being part of the study at any time, without giving a reason, but we will keep information about your child/ relative/ friend that we already have. We need to manage your records in specific ways for the research to be reliable. This means that we won't be able to let you see or change the data we hold about you.

After the study has been completed, your data continue to be stored in a secure location, only accessible by the research team, for 10 years, as recommended by the UK Research and Innovation (UKRI) guidelines. After this, all data will be destroyed.

If you wish, I can provide you with a copy of the results of this study once it is finished.

What if my child/ relative/ friend wants to withdraw?

Your child/ relative/ friend is free to withdraw from the research study at any time without explanation, disadvantage or consequence. If they tell me that they would like to stop the video recording, any of the tests, or the discussions we are having at any point, we will stop these immediately. They will be offered the chance to have a talk about how they are feeling with me (this is called a debrief) and I will give you both some resources of other people to speak to also. Any data collected about either of you, on paper, computer, or video, will be immediately and safely destroyed, and they will no longer be a participant in the study. They will still receive a £10 Amazon voucher for their time.

Separately, your child/ relative/ friend may also request to withdraw their data even after they have participated, provided that this request is made within **3 weeks** of the data being collected. After 3 weeks, names and other identifiable information will be deleted and their data will only be referred to by a numerical code, meaning we will no longer be able to identify which is their data.

If during your participation in the study your child/ relative/ friend loses the ability to consent, we will immediately stop testing and offer you both a debrief. We will then immediately and safely destroy their data, and they will no longer be included in the study. They will still receive a £10 Amazon voucher for their time.

Where can you find out more about how your information is used?

You can find out more about how we use your information

- at www.hra.nhs.uk/information-about-patients/
- by asking one of the research team
- by sending an email to xxxxx, or
- by ringing us on xxxxx

If you have any questions or concerns about how the research has been conducted, please contact:

- The research supervisor:
  Dr. Matthew Jones-Chesters,
  School of Psychology, University of East London, Water Lane, London E15 4LZ

Participant Identification Number:

Email: xxxxxx
Phone: xxxxx

or

- Chair of the School of Psychology Research Ethics Sub-committee:
  Dr Trishna Patel
  School of Psychology, University of East London, Water Lane, London E15 4LZ.
  Email: xxxxxx

Appendix L
Easy-Read Participant Invitation Letter

Participant Identification Number:

University of East London
School of Psychology
Assessment of Cognition in People with Intellectual Disabilities – Participant Information Letter (Easy-Read)
V07.08.2022

You are being invited to participate in a research study. Before you say yes, it is important that you understand what you would be doing. Please read this carefully before you decide if you want to take part.

Who am I?

Hello! My name is Danielle, I am a student in the School of Psychology at the University of East London, and I am studying for a Doctorate in Clinical Psychology. As part of my studies, I am doing the research you are being invited to take part in.

This is a picture of me.

What is the research about?

I want to find out how to know if someone with a learning disability may also be experiencing something called dementia. Dementia is something that happens to some people when they get older. Dementia is when someone finds it hard to remember things, to think, and to do things they used to do like go out on their own, use the toilet on their own, or eat and drink. I would like to see if dementia looks different in people who have a learning disability than people who do not have a learning disability. This will help us to know what is going on sooner, so we can help people who do get dementia have better support. To do this, I have made some tests that I think might be better suited to people with a learning disability than the ones we have already. I must see if the tests we have made can be carried out with people who have a learning disability and do not have dementia, to make sure the tasks are not too easy or too hard. I also need to find out whether different people who have a learning disability experience the tests I have

made differently, so I am first asking for people with Down Syndrome to try them out and tell me what they think of them.

My research has been approved by the NHS Ethics Committee. This means that the NHS and the British Psychological Society feel my study is safe and fair for you to take part in.

Why did you ask me?

You have been invited to participate in my research because I am looking to involve people who have a learning disability, who are aged between 30-55 years old and do not have dementia, to help me understand my study.

You will not be judged in any way and you will be treated with respect at all times.

You do not have to say 'yes' to taking part if you don't want to, and nothing will happen to your care if you say no. You are free to choose what feels most comfortable to you.

What will I have to do?

I will ask you to give me your name, and your phone number or email address, so that I can get in touch with you. People who do not need to know who you are will not be able to see your contact details. You will then be asked to attend a 'testing session' with me. Don't worry, this is not like a school test! I will ask you to do some short tasks, like answering questions, drawing things or copying what I do back to me. We will video record you completing the test so that your answers can be accurately scored. We will also ask you afterwards how you found the tests, what you liked about it and what you think

we could do to make the tests better. We may video-record you doing some of the tests, so that we can score it properly. Your data will be safely stored on a password-protected computer and destroyed once the research has finished. Only me, another researcher (her name is xxxxx) and my supervisor (a supervisor is like my boss) will be able to see your data.

These tests will take around 1 hour. We will take a break in the middle where you can have some snacks and drinks that I will get for you. You can also take more short breaks in between the different tasks if you want to. If you need, we could have two shorter sessions on two different days. After the tests are done, I will ask you what you thought of the tests, including what you think was good about them and how you think I can make them better. This will take around half an hour and can be done on the same day as the tests, or on a different day.

This will take place in a private room at the xxxxx NHS Learning Disability Service at a time we agree upon and plan in advance.

If you decide to take part, you will be given a £10 Amazon voucher as a thank you for your time.

If you take part, everything you say and do will be safe and nobody else will know you took part.

Your privacy and safety will always be respected. Nobody apart from the research team will be able to tell who you are in the data or the write up of the research. You can decide to stop taking part at any time, and you do not need to tell me why you would like to stop. We don't think you will feel upset or stressed by taking part, but if you

are, we have put some names of people you can talk to at the bottom of the sheet.

What will happen to the information that I give you?

All information you provide will be kept strictly confidential. This means that nobody apart from the research team will be able to see it. Your data will be stored on a safe storage device at UEL. After I put it on that device, all paper information will be destroyed. Nobody will be able to tell who you are because I will give your data a number instead of using your name. For up to 3 weeks after you take part, I will keep a secure record of which number links to your name. This is in case you decide you have changed your mind about taking part, so I can find which data is yours and destroy it. After 3 weeks, your name will be deleted from our records.

Your anonymised data will be seen by myself, the other researcher on the team (xxxxx) and my supervisor (Dr. Matthew Jones-Chesters). Group data will be included in my research report, which will be read by examiners, and will be in a paper that other people can see. In this paper, you will only be known as a number, so nobody will know it is you.

After the study has finished, your data will be stored in a secure location, only accessible by the research team, for 10 years, as recommended by the UK Research and Innovation (UKRI) guidelines. After this, all data will be destroyed.

If you want, I can give you a copy of the results of this study once it is finished.

What if I change my mind?

You are free to withdraw from the research study at any time without telling me why. If you tell me that you would like to stop the video recording, any of the tests, or the discussions we are having at any point, we will stop these immediately. You will be offered the chance to have a talk about how you are feeling with me (this is called a debrief) and I will give you some resources of other people to speak to also. Any data collected about you, on paper, computer, or video, will be immediately and safely destroyed, and you will no longer be a participant in the study. You will still receive a £10 Amazon voucher for your time.

If you change your mind after you take part, you have 3 weeks to change your mind and ask me to delete your data. After 3 weeks, your name and other information linked to who you are will be deleted and your data will only be referred to by a number, meaning we will no longer be able to identify which is your data. This means that after 3 weeks, you cannot change your mind and take your data out of the study.

Contact Details

If you would like further information about my research or have any questions or concerns, please ask me using my phone number or my email:

- Telephone: xxxxx, email: xxxxx

If you have any questions or concerns about how the research has been conducted please contact:

- The research supervisor:
  Dr. Matthew Jones-Chesters,
  School of Psychology, University of East London, Water Lane,
  London E15 4LZ
  Email: xxxxx

or

- Chair of the School of Psychology Research Ethics Sub-committee:
  Dr Trishna Patel
  School of Psychology, University of East London, Water Lane,
  London E15 4LZ.
  Email: xxxxx

You can also visit this website to learn more about how your information is used: www.hra.nhs.uk/information-about-patients/

Appendix M
Easy-Read Cover Letter

Dear (Person's name),

You have had recent (within the last five years) contact with the [name of community team].

You have taken part in an assessment where we asked you lots of questions.

University of East London

Our team have been approached by the University of East London.

They are doing some research into the tests used to assess dementia in people who have a learning disability.

To carry out this research they are looking for participants:

- Aged 30 – 55 years old
- Who have a learning disability
- Who do <u>not</u> have dementia

They would like people to give feedback on some new assessments.

Please see the attached information leaflet.

If this is something you might be interested in doing please let me know and I can put you in touch with Danielle, the researcher.

I can be contacted on the office number [number] or e-mail [email]

[person's name] will follow-up with a phone call in the next couple of weeks to check if this is something you are interested in.

If you do not wish for us to contact you by phone, you can send a text to [number] with your initials and "no"

Or let me know by phone or e-mail.

Please be assured that none of your information has been or will be shared without your permission and you do not have to take part in this study if you don't want to.

Insert image of psychologist

With best wishes,

Psychologist

Appendix N
Participant Debrief Letter

University of East London
School of Psychology
Assessment of Cognition in People with Intellectual Disabilities – Participant Debrief Form
V08.06.2022

Thank you so much for participating in my research study on creating a test set to look for dementia in those who have a learning disability. This letter offers information that you might find important now that you have now taken part.

What will happen to the information that you provide?
All the information you provide will be kept strictly confidential. That means that only the researchers (Danielle Pearce and xxxx) and their supervisor (Dr. Matthew Jones-Chesters) will be able to see it. Your data will be stored on the UEL OneDrive, which is a secure and encrypted online service. After uploading your information to the UEL OneDrive all paper information will be destroyed. Your data will be anonymised by using a numerical code instead of your name. For up to 3 weeks after you participate in the study, a separate document will be kept to link your name to your numerical code (this is in case you decide you want to withdraw your data from the study during this period), and after 3 weeks your name will be deleted from our records.

Your anonymised data will be seen by me, the other researcher on the team (xxxx) and my supervisor (Dr. Matthew Jones-Chesters). Data will be analysed in groups, and will be incorporated into my thesis paper. This will be read by examiners and will be made available to the public, the NHS Learning Disability service involved in your care, and to you. If the study is published it will appear in an academic journal. No individual or identifiable information will be included in any report or publication.

After the study has been completed, your data continue to be stored in a secure location, only accessible by the research team, for 10 years, as recommended by the UK Research and Innovation (UKRI) guidelines. After this, all data will be destroyed.

If you wish, I can provide you a copy of the results of this study when it is finished.

What if you want to withdraw?
You are free to withdraw from the research study at any time during participation without telling me why, and there will be no consequences for doing so.

Additionally, you may also request to withdraw your data even after you have participated, provided that this request is made **within 3 weeks** of the data being collected. After 3 weeks, your name and other identifiable information will be deleted and your data will only be referred to by a numerical code, meaning we will no longer be able to identify which is your data.

What if you have been adversely affected by taking part?
We do not anticipate that you will be negatively affected by taking part in the research, and all reasonable steps have been taken to minimise potential harm. Nevertheless, it is still possible that your participation – or its after-effects – may have been challenging, distressing or uncomfortable in some way. If you have been affected in any of those ways you may find the following resources/services helpful for information and support:

Down's Syndrome Association
The Down's Syndrome Association is dedicated to helping everybody with Down's Syndrome to feel included and empowered. They are a community of people which will provide support, advice, friendship and advocacy.
Tel: +44 (0)333 1212 300– Monday to Friday 10:00am – 4:00pm
Website: https://www.downs-syndrome.org.uk/

British Institute of Learning Difficulties (BILD)
BILD (British Institute of learning difficulties) informs you of the types of advocacy available for people with learning difficulties. They work in partnership with people with learning difficulties and families enabling them to get the right support to make informed choices about their own lives.
Tel: 0121 415 6960– Telephone line open Monday-Friday 9am-5pm
Website: http://www.bild.org.uk/about-bild
e-Mail: enquiries@bild.org.uk

Mencap
Mencap offers a range of personal and unique services for people with a learning disability, families and carers. Mencap's Empower Me service gives personalised advocacy support for people with a learning disability, helping to develop skills, confidence and knowledge needed to voice concerns and secure rights.
Tel: 0808 808 1111 Phoneline is open 9am to 3pm, Monday to Friday
Website: https://www.mencap.org.uk/our-services/personal-support-services/advocacy

You are also very welcome to contact me or my supervisor if you have questions or concerns.

Contact Details

If you would like further information about my research or have any questions or concerns, please ask me:

- Danielle Pearce
  E-Mail: xxxx

If you have any questions or concerns about how the research has been conducted please contact:

- The research supervisor:
  Dr. Matthew Jones-Chesters,
  School of Psychology, University of East London, Water Lane, London E15 4LZ
  Email: xxxx
  Phone: xxxx

or

- Chair of the School of Psychology Research Ethics Sub-committee:
  Dr Trishna Patel
  School of Psychology, University of East London, Water Lane, London E15 4LZ.
  Email: xxxx

Or to find out more about how we use your information:

www.hra.nhs.uk/information-about-patients/

Appendix O

Easy-Read Participant Debrief Letter

EASY-READ DEBRIEF LETTER v1.03 (23.05.2023) IRAS ID: 295654

Participant Identification Number:

University of East London
School of Psychology
Assessment of Cognition in People with Intellectual Disabilities – Participant Debrief Letter (Easy-Read)
V23.05.2023

Thank you for taking part in our research study!

What were the tests for?

We think that the tests you did might be better suited to people with a learning disability than the tests we have already.

What will you do with my information?

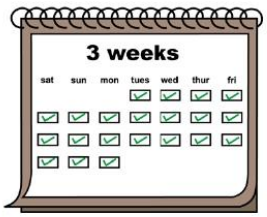
All the information you provided will be kept strictly confidential. This means that nobody apart from the research team will be able to see it. Your data will be stored on a safe

storage device at UEL. Nobody will be able to tell who you are because we will give your data a number instead of using your name.

For up to 3 weeks from now, we will keep a secure record of which number links to your name. This is in case you decide you have changed your mind about taking part, so we can find which data is yours and destroy it. After 3 weeks, your name will be deleted from our records.

Your anonymised data will be seen by the researchers (Elicia McGregor and Danielle Pearce) and our supervisor (Dr. Matthew Jones-Chesters). Group data will be included in our research report, which will be read by examiners, and will be in a paper that other people can see. In this paper, you will only be known as a number, so nobody will know it is you.

After the study has finished, your data will be stored in a secure location, only accessible by the research team, for 10 years, as recommended by the UK Research and Innovation (UKRI) guidelines. After this, all data will be destroyed.



What if I change my mind?

If you decide you would no longer like to be a part of the study, you have 3 weeks from today to let us know. You do not need to tell us why. Any data collected about you, on paper, computer, or video, will be immediately and safely destroyed, and you will no longer be a participant in the study. You will still keep your £10 Amazon voucher for your time.

If you change your mind after 3 weeks, your name and other information linked to who you are will be deleted and your data will only be referred to by a number, meaning we will no longer be able to identify which is your data.

This means that after 3 weeks, you cannot change your mind and take your data out of the study.

Do you have any questions?

We hope that you have not felt stressed doing these tests, but if you have and would like someone to talk to about it, you or your carer/ guardian can contact the person that you usually see at your ELFT Learning Disability Service for support. Or you may find the following resources/services helpful for information and support:

Down's Syndrome Association

They can provide support, advice, friendship and advocacy.

Tel: +44 (0)333 1212 300– Monday to Friday 10:00am – 4:00pm

Website: https://www.downs-syndrome.org.uk/

British Institute of Learning Difficulties (BILD)

They can help people with learning difficulties and families to get the right support to make informed choices about their own lives.

Tel: 0121 415 6960– Telephone line open Monday-Friday 9am-5pm

Website: http://www.bild.org.uk/about-bild

e-Mail: enquiries@bild.org.uk

Mencap

Mencap offers a range of services for people with a learning disability, families and carers. Mencap's Empower Me service gives support for people with a learning disability, helping to develop skills, confidence and knowledge needed to voice concerns.

Tel: 0808 808 1111 Phoneline is open 9am to 3pm, Monday to Friday

Website: https://www.mencap.org.uk/our-services/personal-support-services/advocacy

If you would like further information about our research or have any questions or concerns, please ask us using our phone number or email:

- Telephone: 020 3576 1343, email: u1945505@uel.ac.uk

If you have any questions or concerns about how the research has been conducted please contact:

- The research supervisor:

  Dr. Matthew Jones-Chesters,
  School of Psychology, University of East London, Water Lane, London E15 4LZ
  Email: m.h.jones-chesters@uel.ac.uk

or

- Chair of the School of Psychology Research Ethics Sub-committee:

  Dr Trishna Patel
  School of Psychology, University of East London, Water Lane, London E15 4LZ.

Email: t.patel@uel.ac.uk

You can also visit this website to learn more about how your information is used:

www.hra.nhs.uk/information-about-patients/

Appendix P

Semi-Structured Interview Schedule

University of East London
School of Psychology
Assessment of Cognition in People with Intellectual Disabilities – Semi-Structured Interview Schedule
V07.08.2022

1. Did you find any of the tests interesting?
    a. Which ones in particular?
    b. Why?
2. Did you find any of the tests boring?
    a. Which ones in particular?
    b. Why?
3. Did you find any of the tests too easy?
    a. Which ones in particular?
    b. Why?
    c. What could we change about these tests to make them better?
4. Did you find any of the tests too hard?
    a. Which ones in particular?
    b. Why?
    c. What could we change about these tests to make them better?
5. Do you have anything else you would like to say about the tests you did today?

Appendix Q

Example Coding for Verbal and Non-Verbal Communication

Verbal	**Positive Examples**	**Negative Examples**
• Speech • Volume • Tone • Rate • Clarity • Fluency	• Verbal indications of enjoyment e.g. "This is fun!" • Jovial tone, laughter • Good speech output	• Verbal indications of distress e.g. "I don't like this" or "I don't want to do more." • Sighing, 'huffing' • Hesitancy • Limited speech output (though this is impacted by verbal ability) or refusal to talk
Non-Verbal	**Positive Examples**	**Negative Examples**
• Body language • Facial expression • Eye contact • Posture & gait • Gesture • Signing • Distance • Vocalisations / noises • Behaviour	• Facing towards examiner, open stance • Objectively happy, smiling. • Good eye contact • Engaged posture. • Vocalisations to indicate happiness, • Behaviour to indicate happiness, such as jumping up and down in excitement or clapping hands.	• Facing away from examiner, folded arms, retreating • Objectively unhappy, tearful • Poor eye contact, avoidant (though this is common in ASD) • Slumped shoulders • Vocalisations to indicate unhappiness, such as screaming. • Behaviour to indicate unhappiness, such as banging the table, pushing the test materials away, or hitting/ kicking, wringing hands, fidgeting, attempting to leave

Appendix R

Example Coding System for Semi-Structured Interview Feedback

	Example Verbal Responses	Example Non-Verbal Responses
Yes	"Yes," "yeah", "maybe", "some", "lots" (or something similar)	Nodding their head
No	"No," "none", "na" (or something similar)	Shaking of their head
Orientation	Responses indicating questions on name, place, time, how they got here	Pointing to test materials
Smell Detection & Recognition	"Smells," "jars" (or something similar)	Pointing to test materials, indicating smelling something, sniffing
Motor Function Part A & Verbal Comprehension Part A	Verbal expressions of the instructions (e.g., "touching my nose")	Pointing to test materials, imitations of the instructions (e.g., touching their nose, closing their eyes, and opening them)
Motor Function Part B	Verbal expressions of the instructions (e.g., "holding out my arms")	Pointing to test materials, imitations of the instructions (e.g., finger-to-nose, holding out their arm)
Motor Programming	Verbal expressions of the instructions (e.g., "knocking")	Pointing to test materials, imitations of the instructions (e.g., knocking on the table), (with qualifying questions)
Praxis	Verbal expressions of the instructions (e.g., "combing my hair")	Pointing to test materials, imitations of the instructions (e.g., combing their hair, waving, coughing)
Verbal Comprehension Part B	Verbal expressions of the instructions (e.g., "put the pen on the watch"), items included in the instructions (e.g., "watch," "pen", "keys")	Pointing to test materials, imitations of the instructions (e.g., moving the pen on to the paper)
Word List	"Lots of words," "memory one" (with qualifying questions), items included in the instructions (e.g., "dish", "shoe", "frog")	Pointing to test materials

Circle Search	"Shapes" (with qualifying questions), "circles," "drawing" (with qualifying questions) or something similar	Pointing to test materials, imitations of the instructions (e.g., imitating crossing out items with a pen/pencil)
Angle Judgement	"Lines," "matching lines", "angles"	Pointing to test materials
Visual Reasoning	"Shapes" (with qualifying questions), "patterns," "pictures" (with qualifying questions), "puzzle" (with qualifying questions)	Pointing to test materials
Cat-Dog Inhibition	"Cats," "dogs", "pictures" (with qualifying questions)	Pointing to test materials
Shopping List	"Shopping," "food" (with qualifying questions), "puzzle" (with qualifying questions), "drawing" (with qualifying questions), "maze", items included in the shopping list (e.g., "eggs"; with qualifying questions)	Pointing to test materials
Matchsticks	"Matchsticks," "picture" (with qualifying questions)	Pointing to test materials, imitations of the instructions (e.g., using the matchsticks)
Eight Detection	"Numbers," "8"	Pointing to test materials, imitations of the instructions (e.g., tapping on the table; with qualifying questions)
Picture Naming & Recognition	"Pictures" (with qualifying questions), items included in the instructions (e.g., "butterfly," "waterfall"; with qualifying questions)	Pointing to test materials
Sentence Repetition	"Words" (with qualifying questions), repetition of the instructions (e.g., "take that home"	Pointing to test materials
Verbal Reasoning	Repetition of the instructions (e.g., "Robin is a bird…")	Pointing to test materials
Word Generation	"Animals," "foods" (with qualifying questions)	Pointing to test materials

Appendix S

Data Distribution Tables

Table S 1

Data Distributions - Motor & Language Functions

Motor & Language Functions							
Subtest	**Skewness**			**Kurtosis**			***Normality***
	Value	**SE**	***Z***	**Value**	**SE**	***Z***	**Shapiro-Wilk test *p***
Orientation Subtotal A	**-1.33**	0.79	-1.67	0.54	1.59	0.34	**.043**
Orientation Subtotal B	0.17	0.79	0.22	0.34	1.59	0.21	.099
Orientation Total (A + B)	**-1.26**	0.79	-1.58	0.99	1.59	0.62	.202
Smell Detection	-0.17	0.79	-0.22	0.34	1.59	0.21	.099
Smell Recognition Total	0.80	0.79	1.01	**-1.28**	1.59	-0.81	**.029**
Verbal Expression	**-1.65**	0.79	**-2.08**	**2.96**	1.59	1.87	**.040**
Verbal Comprehension A	-0.22	0.79	-0.28	**-1.72**	1.59	-1.08	.215
Verbal Comprehension B	-0.31	0.79	-0.38	**-1.83**	1.59	-1.15	.224
Verbal Comprehension Total (A + B)	-0.37	0.79	-0.47	**-1.48**	1.59	-0.93	.519
Motor Function Subtotal A	**-1.12**	0.79	-1.40	0.27	1.59	0.17	**.020**
Motor Function Subtotal B	**-1.95**	0.79	**-2.45**	**4.58**	1.59	**2.89**	**.006**
Motor Function Total (A + B)	**-2.19**	0.79	**-2.75**	**5.32**	1.59	**3.35**	**.006**
Motor Programming Total	-0.70	0.85	-0.83	**-1.09**	1.74	-0.63	.201
Praxis Total	-0.86	0.79	-1.09	**1.25**	1.59	0.78	.183

Note. Items in bold highlight where criteria have been met for non-normal distribution.

Table S 2

Data Distributions - Verbal Learning & Visual Functions

Verbal Learning & Visual Functions							
Subtest	**Skewness**			**Kurtosis**			***Normality***
	Value	**SE**	***Z***	**Value**	**SE**	***Z***	**Shapiro-Wilk test *p***
Word List Immediate	-0.26	0.85	-0.31	0.39	1.74	0.22	.985
Word List Learning	-0.67	0.85	-0.79	-0.45	1.74	-0.26	.421
Word List Delayed Recall	0.41	0.85	0.49	**-1.31**	1.74	-0.75	.425
Word List Recognition	**-1.56**	0.85	-1.84	**2.92**	1.74	1.68	.093
Circle Search	**-1.76**	0.79	**-2.22**	**2.36**	1.59	1.49	**.001**
Angle Judgement	0.29	0.85	0.34	**-2.53**	1.74	-1.45	.118
Visual Reasoning	**1.84**	0.85	**2.18**	**3.91**	1.74	**2.25**	**.020**
Shopping List Map 1	0.27	0.79	0.34	-0.66	1.59	-0.41	.792
Shopping List Map 2	-0.37	0.91	-0.40	**-2.10**	2.00	-1.05	.529
Shopping List Total	-0.52	0.91	-0.57	**-1.45**	2.00	-0.73	.688
Cat-Dog Inhibition	-0.85	0.79	-1.07	-0.87	1.59	-0.55	**.024**
Cat-Dog Inhibition Time (seconds)	0.84	0.85	0.99	-0.41	1.74	-0.24	.363

Note. Items in bold highlight where criteria have been met for non-normal distribution.

Table R 3

Data Distributions - Visual Learning & Verbal Functions

Visual Learning & Verbal Functions							
Subtest	**Skewness**			**Kurtosis**			***Normality***
	Value	**SE**	***Z***	**Value**	**SE**	***Z***	**Shapiro-Wilk test *p***
Matchstick Copy Total	-0.23	0.79	-0.29	**-2.33**	1.59	-1.47	.137
Matchstick Immediate Total	0.15	0.85	0.18	**-2.64**	1.74	-1.52	.218
Matchsticks Delayed Recall Total	0.76	0.79	0.96	**-1.25**	1.59	-0.79	.183
Picture Naming Total	**-1.08**	0.79	-1.36	-0.23	1.59	-0.15	.099
Picture Recognition Total	**-1.14**	0.79	-1.43	**1.18**	1.59	0.74	.429
Eight Detection Total	-0.97	0.85	-1.15	**-1.88**	1.74	-1.08	**.001**
Sentence Repetition Total	-0.50	0.85	-0.59	-0.40	1.74	-0.23	.741
Verbal Reasoning Total	**1.54**	0.85	1.82	**1.43**	1.74	0.82	**.006**
Word Generation Total	**1.42**	0.79	1.78	**2.09**	1.59	1.32	.203

Note. Items in bold highlight where criteria have been met for non-normal distribution.