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

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ABSTRACT

***Background**:* There is no 'gold standard' for identifying dementia in people with an intellectual disability, which hinders access to early identification and appropriate support to members of this community. Differences in executive functioning may mean widely used cognitive assessments and tests are not accessible, acceptable, or feasible for use with people with intellectual disabilities. A literature review highlighted 114 available stand-alone measures and 37 batteries used to measure cognitive decline in people with an intellectual disability. Many did not show robust assessment of executive function and showed floor effects for people with more severe cognitive impairments. Research showed tests of olfaction may be an accessible format for use with people with intellectual disabilities.

***Methods**:* Responding to the need highlighted through the literature review, a novel draft cognitive battery was created, which included robust assessment of executive functions, and an olfactory measure of learning and memory. An exploratory method was adopted to assess feasibility and acceptability by piloting the battery with four people with Down Syndrome. Quantitative data were gathered through test performance, and qualitative data were collected through participant feedback and researcher observation.

***Results**:* Results indicated acceptability and feasibility of the battery for use with people with Down Syndrome, but many items require modification. Feasible tests of executive function proved most challenging to create, aligning with the literature. Implications for olfactory assessment with people with intellectual disabilities were identified.

***Conclusions**:* Results identify recommendations for revisions to the battery and candidate tasks which may improve feasibility and acceptability. The importance of gathering the opinions of people with intellectual disabilities to shape instruments for their care is highlighted. Results inform future piloting with larger and more diverse samples from the intellectual disability community.

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1 INTRODUCTION

1.1 Overview

This research aims to create a novel test battery to assess cognitive function with the intellectual disability (ID) community, and pilot this with people with Down Syndrome (PWDS). With revisions, this battery may go on to be useful as a screening tool for dementias in this community. In this chapter, I outline terminology used, review the literature focusing on cognitive functions, assessment, and dementias; both generally and for people with intellectual disabilities (PWID). Through a literature review, I critically review cognitive tests currently available for PWID and highlight gaps in the literature, particularly in the assessment of executive function (EF) and olfactory ability (OA). I explore how these may be included in the novel battery to improve accessibility feasibility and acceptability. I conclude with the rationale for the present study.

1.2 Terminology

The term ‘learning disability’ is used in the participant recruitment information for this research, as it is the most understood and commonly used term throughout services in the UK (Abbott & Burns, 2007). Throughout this thesis, the term ‘intellectual disability’ (ID) is used, as the most common term used academically and internationally (Schalock et al., 2007), which avoids confusion with conditions associated with ‘learning disability’, such as dyslexia. Terms used have evolved over time to create distance from previous terms and associated stigma, prejudice, and dehumanisation (Parmenter, 2011). After careful consideration, pejorative terms are included in the search string of the ‘Literature Review’, to avoid omission of important early research. This is the only time pejorative terms are used. Abbreviations are used throughout this thesis; a list of abbreviations is presented in Appendix A.

1.3 Personal Context

All research is directly or indirectly influenced by the values of the researcher through narratives they create (Stevenson, 1988). I have worked with PWID for most of my adult life, and noticed many instruments used in their care were not created

for, or with, PWID. This felt discriminatory, and I wanted to produce something that could become a ripple of change in tailoring care to PWID. I wanted to make this accessible by making it low-cost and easy to obtain and use. My brother-in-law, who had an ID, was also a huge inspiration for this thesis. He was non-verbal, but always had a lot to say if you learned the right way to listen. He sadly passed away in 2021. His influence carries through into my work, which I hope can benefit others in his community.

1.4 Intellectual Disabilities

ID is a neurodevelopmental condition characterised by cognitive, communication, behavioural, motor, and social functioning impairments, alongside an intelligence quotient (IQ) of <70 (WHO, ICD-10, 1992). Guidance on assessing and diagnosing ID by the British Psychological Society (BPS; 2015a) states that significant impairments in intellectual functioning and in social and everyday functioning must be present in childhood. Diagnosis and severity of ID is indicated through standardised IQ test scores and levels of independent functioning. In the typically developing (TD) population, IQ has a mean of 100 and a standard deviation of 15. An IQ score of 50-69 indicates mild ID, 35-49 indicates moderate ID, 20-34 indicates severe ID and <20 indicates a profound ID. As severity increases, characteristics become more pronounced and varied, requiring more continuous support in activities of self-care and daily living (WHO, ICD-10, 1992; Henry, 2001). Acquired brain injury (ABI) such as traumatic brain injury, or neurodegenerative disease in childhood, can also result in ID (Einfeld & Emerson, 2008). In the United Kingdom (UK), ID prevalence is 4.7 per 1,000; globally, prevalence is between 1% and 3% (Roeleveld et al., 1997; Health and Social Care Information Centre, 2013).

1.4.1 Health and Social Inequalities

PWID have a shorter life expectancy than TD individuals (Heslop et al., 2014). This can partly be attributed to a higher number of preventable and amenable deaths linked to social inequalities, diagnostic overshadowing of additional healthcare needs, and overuse of psychotropic medications; all leading to poorer health

outcomes (Glover & Ayub, 2010; Branford et al., 2018). Though this disparity in life expectancy is decreasing due to improved healthcare, policy changes and medical advancements (Coppus, 2013; Englund et al., 2013), PWID still experience barriers to healthcare, possibly due to communication difficulties between PWID and clinicians (Doherty et al., 2020) and lack of community engagement initiatives (Hendrix et al., 2020). Further, the opinions of PWID are largely omitted from research discourse, preventing them from contributing to decisions and resources made for their care (Coons & Watson, 2013; Beighton et al., 2017). Improving access to appropriate healthcare and developing tailored resources is fundamental to reducing unjust health inequalities and improving quality of life for PWID.

1.4.2 Causes and Subtypes

ID is a clinical feature of many aetiologically distinct genetic conditions, such as Fragile X Syndrome (FXS), Williams-Beuren Syndrome (WS) and Down's Syndrome (DS). There are also many aetiologically undetermined forms of ID. This subsection focuses on DS, FXS and WS, as these aetiologies and associated phenotypes are the most prevalent in the current literature base (Glasson et al., 2020). Non-syndromic causes of ID are also outlined. Differences in cognitive and behavioural profiles (relative strengths and weaknesses) associated with each aetiology are described.

1.4.2.1 Down Syndrome

Down Syndrome (DS) (also known as trisomy 21) is the most common genetic condition with ID as a clinical feature (WHO, ICD-10, 1992). ID severity ranges from mild to severe depending on phenotypical variation (Epstein, 1989; Roizen & Patterson, 2003; Bull, 2020). The extra chromosome on chromosome pair 21 creates distinct physical features, including almond-shaped eyes and poor muscle tone. This can also create physical health issues, such as congenital heart disease, which is present for around 54% of PWDS (WHO, ICD-10, 1992; Bergström et al., 2016). DS prevalence is 6.8/10,000 in males, and 5.9/10,000 in females in the United Kingdom,

with an estimated 417,000 PWDS living throughout Europe (Alexander et al., 2016; DeGraaf et al., 2021). Global prevalence of DS is difficult to establish, but is estimated at 1 in 750 (Kozma, 2008; Antonarakis et al., 2020).

PWDS have a unique cognitive profile. Strengths are found in non-verbal abilities, including associative learning, implicit long-term memory, and visuo-spatial short-term memory (Lott & Dierssen, 2010). Though visuo-spatial abilities are thought to be a relative strength, this may not be relative to mental age, and may show inter-individual variability (Yang et al., 2014). Difficulties are generally seen in verbal abilities, including expressive and receptive language, verbal working memory, production, and comprehension (Lott & Dierssen, 2010; Næss et al., 2012; Grieco et al., 2015; Fernández-Alcaraz & Carvajal, 2020). Das & Mishra (1995) report difficulties in phonological processing and verbal short-term memory for PWDS, which they posit as related to impairment of the 'phonological loop' (Baddeley & Hitch, 1974). However, some evidence indicates verbal fluency as a relative strength for PWDS (Conners et al., 2011). Cognitive variability may be linked to evidence suggesting that cognitive weaknesses are less severe in females and PWDS with mild ID (Määttä et al., 2006).

Executive function (EF) deficits are also shown, which may be linked to pre-existing frontal lobe abnormalities which are exacerbated by accelerated ageing and subsequent neurodegeneration (Crome & Stern, 1972; Holland et al., 2000). Deficits in olfactory ability (OA) may also be pre-existing, as those with DS have smaller olfactory bulbs than TD counterparts (Bianchi et al., 2014; Bontempi et al., 2020). When age and sex-matched to TD counterparts, PWDS show significant impairments in odour detection, identification, and recognition memory (Murphy & Jinich, 1996). PWDS show decreased olfactory function with age, and a more severe impairment than age and IQ-matched PWID of different aetiologies (Nijjar & Murphy, 2002). However, research concerning olfactory impairment in DS is scarce, and much is outdated (Windsperger & Hoehl, 2021).

1.4.2.2 Other Genetic Causes of Intellectual Disability

Fragile X Syndrome (FXS)

Fragile X Syndrome (FXS) is related to the silencing or expansion of the FMR1 gene found on the X chromosome, which is responsible for producing a protein fundamental to brain development (WHO, ICD-10, 1992). Population prevalence is estimated at around one in 2,500 (Crawford et al., 2001; Hagerman, 2008). ID is a clinical feature in approximately 80% of FXS males and 70% of females. Physical features include: hyperflexible joints, large ears, and flat feet (WHO, ICD-10, 1992; Sherman et al., 1996; Scharfenaker et al., 1996; Hagerman & Hagerman, 2002).

The cognitive profile of FXS includes strengths in expressive language, verbal reasoning, verbal immediate memory (Edgin et al., 2010), verbal comprehension and visual-motor coordination (Freund & Reiss, 1991; Kogan et al., 2009), whilst difficulties are found in spatial object discrimination and spatial learning abilities (Kogan et al., 2009), short-term memory and EF (Reiss & Hall, 2007; Van der Molen et al., 2010).

Williams-Beuren Syndrome (WS)

WS is caused by the deletion of part of chromosome 7q11.23., and a gene responsible for elastin production (WHO, ICD-10, 1992; Lowery et al., 1995). ID can be a clinical feature of WS, alongside characteristics related to elastin deficiency including connective tissue abnormalities and lax skin (Morris et al., 1990; Vaux et al., 2003). Individuals with WS are at greater risk of cardiovascular disease than the TD population (Honjo et al., 2022). Population-based data on WS incidence is limited, but prevalence is estimated at one in 7,500 (Strømme et al., 2002).

The cognitive phenotype of WS includes delayed speech, but strengths in immediate recall and verbal conceptual abilities (Mervis et al., 2000; Mervis & Pitts, 2015), and sustained attention (Atkinson & Braddick, 2011). People with WS show significant difficulties in selective attention (Fung et al., 2012) visuospatial expression, and EF (Bellugi & Wang, 1998). Both WS and DS youth show strengths in sustained

attention and weakness in selective attention, with PWDS showing particular strength in auditory sustained attention (Breckenridge et al., 2013).

1.4.2.3 Other Aetiologies and Subtypes

Syndromic causes do not account for up to 80% of ID cases (Rauch et al., 2006). Perinatal risk factors for non-syndromic ID include maternal alcohol and/or drug use, malnutrition, and birth complications (such as preeclampsia) (Huang et al., 2016). Postnatal risk factors include neonatal meningitis, ABI, neglect, and abuse (Buchanan & Oliver, 1977; Shree & Shukla, 2016; Oh et al., 2019). In acquired ID, cognitive profile varies depending on age of onset, brain injury severity and neural areas affected (Slomine & Locascio, 2009). Up to two thirds of IDs are of an unknown cause and may be due to a complex interplay of socioeconomic inequalities affecting prenatal, perinatal, and neonatal outcomes (Abdelaziz & Abdelmageed, 2021). Therefore, cognitive profiles are highly variable in such cases.

1.4.2.4 Differences in Executive Functions Between Syndromic IDs

PWID show deficits in EF in comparison to TD counterparts (Ball et al., 2008; Alloway, 2010; Peltopuro et al., 2014). Differences in EF between ID aetiologies are also indicated. Research by Costanzo et al. (2013) with WS and DS adults indicated that although both groups presented with EF deficits, PWDS showed deficits in task shifting, verbal fluency and verbal inhibition, whilst participants with WS exhibited specific weakness in task planning. Further, individuals with FXS and DS experience deficits in selective and sustained attention, and task shifting, in comparison to TD counterparts and people with other subtypes of ID (Munir et al., 2000). However, when matched for mental and chronological age, PWDS perform better in tests of task setting and shifting compared to FXS counterparts (Van Der Molen et al., 2012). PWDS also show deficits in simultaneous and successive processing, alongside sequenced motor responses, in comparison to counterparts with non-DS ID (Snart et al., 1982; Lincoln et al., 1985).

A review study comparing the cognitive profiles of PWDS, FXS and WS by Conners et al. (2011) concluded that individuals with FXS show weakness particularly in both visual and verbal working memory. For people with WS, relatively good performance in visual and verbal working memory was found, with difficulties in reading comprehension. In PWDS, severe deficits were seen in verbal working memory, alongside strengths in semantic fluency and immediate visual recall. There is some discrepancy as to whether EF deficits in PWDS are age-related, as research indicates these processes are preserved in childhood (Pennington et al., 2003; Lanfranchi et al., 2010).

1.4.3 Summary

Evidence suggests PWID have unique cognitive profiles. Differences between ID aetiologies are also found. PWDS show weaknesses in verbal functions, people with WS show relative weaknesses in visuospatial abilities and selective attention, and individuals with FXS show impairments in visual-spatial functions and memory. All show weakness in EF, but these are found in different functions between syndromes. These differences in cognitive and behavioural phenotypes may have implications for one of the most common age-related diseases: dementias.

1.5 Dementias

The ICD-10 defines dementias as progressive neurodegenerative diseases, characterised by difficulties in cognitive function, including learning, memory, reasoning, calculation, EF, comprehension, attention, visuo-spatial and verbal-conceptual abilities. These deficits are often indicated by changes in social behaviour, activities of daily living (ADLs), emotional control or regulation, and motivation (WHO, ICD-10, 1992). Diagnosis is ascertained through neuropsychological testing, where scores must have declined to two standard deviations below the person's expected baseline. This decline must impede functioning in instrumental ADLs and must not be better attributed to emotional or motivational influences, such as depression, nor to physical illness.

Neurodegenerative cognitive impairment below the clinical threshold for dementia may be classified as prodromal dementia or mild cognitive impairment (MCI), which may lead to routine follow-up to monitor advancement towards dementia (NICE, 2018). As dementia progresses, impairments become global and clinical features overlap, creating difficulties in accurate diagnosis (Karantzoulis et al., 2011). Currently, there are 900,000 people experiencing dementia in the UK, which is projected to increase to 1.6 million by 2040 (Alzheimer's Society, 2022).

1.5.1 Causes and Subtypes

There are several disparate forms of dementia, each with different aetiologies and trajectories influencing their neuropsychological profiles. The main forms include Alzheimer's Disease (AD), vascular dementia (VaD), and dementia with Lewy Bodies (DwLB) (including Parkinson's Disease Dementia; PDD) (WHO, ICD-10, 1992). Other cortical conditions are also described below.

1.5.1.1 Alzheimer's Disease

AD is associated with ageing, and is the most common dementia globally, affecting ~20 million individuals currently and projected to affect ~150 million by 2050 (Wisniewski & Goñi, 2015; Alzheimer's Society, 2022). AD is characterised initially by increased amyloid-β protein plaques, neurofibrillary tangles and cortical atrophy in the hippocampus and temporo-parietal regions of the brain (Ferri et al., 2005; Fjell et al., 2014). This affects neurotransmitter function between brain structures, and can lead to a deficit of acetylcholine, a crucial neurotransmitter related to learning and memory (Piggott, 2013). Though dementias are generally associated with memory difficulties, these are most prominent and have the earliest onset in the AD phenotype (Bowler et al.,1997). In early AD, neuropsychological decline in episodic memory, learning, recall and recognition is seen. As neurodegeneration progresses through the medial temporal lobes, executive dysfunction in task setting and switching emerge, alongside poor semantic fluency. Later, the person experiences global neuropsychological decline, including further EF impairments, apraxia and

agnosia, reflecting widespread cortical degeneration (WHO, ICD-10, 1992; Markowitsch & Staniloiu, 2012).

1.5.1.2 Vascular Dementia and Other Subtypes

VaD accounts for around 20% of dementia diagnoses (Alzheimer's Society, 2022). Onset is related to restrictions in cortical blood supply due to one or multiple large cerebral strokes, several small strokes, or cerebrovascular disease. VaD is therefore not necessarily related to age, and cognitive impairment is sudden or gradual (Verdelho et al., 2021). This creates a "patchy" neuropsychological presentation dependant on the location of injury, where memory is relatively preserved, but decline is seen in EF, attention, and cognitive flexibility (O'Brien & Thomas, 2015).

DwLB is categorised by Lewy bodies throughout the brain, and accounts for 10-15% of dementia diagnoses (Alzheimer's Society, 2022). DwLB acts as an umbrella term for the related diagnosis of PDD. Clinical features overlap, but DwLB is diagnosed if cognitive impairment precedes parkinsonism (Gomperts, 2016). DwLB can create fluctuating attention difficulties in task-switching, impulse control and working memory. Individuals with DwLB may experience resting tremors, rigid limbs, and slow movement (Jellinger, 2018; Alzheimer's Society, 2022). This differs from AD, as executive dysfunction, visuospatial, attentional, and working memory deficits are early indicators of DwLB pathology, yet language and memory are relatively preserved until later stages (Galvin, Pollack & Morris, 2006). This may be due to Lewy body accumulation in the limbic system and neocortex, with preservation of the medial temporal and hippocampal lobes in the early stages (Salmon & Bondi, 2009; Elder et al., 2017).

1.5.1.3 Other Cortical Conditions

Other cortical conditions include posterior cortical atrophy (PCA) and frontotemporal dementias (FTDs). PCA can be resultant of AD or DwLB neurodegeneration, with atrophy beginning in the dorsal and ventral streams of the primary visual cortex. This

results in visuospatial, reading, and writing impairments, yet relative preservation of language and memory (McMonagle et al., 2006; Crutch et al., 2012). PCA is rare, accounting for ~5% of dementia cases, with an earlier onset than AD (Crutch et al., 2012).

FTDs are a collective term for three variants of rare dementias affecting the frontal lobes: behavioural-variant FTD (bvFTD), non-fluent primary progressive aphasia and semantic-variant primary progressive aphasia (Bang et al., 2015; Alzheimer's Society, 2022). FTD is categorised by white matter degradation and has a point prevalence range of 0.01-4.61 in 1000 persons, with bvFTD being four times more common than the primary progressive variant (Bang et al., 2015; Hogan et al., 2016).

In bvFTD, characteristic signs include behavioural changes (e.g., disinhibition and apathy) with notable EF deficits. Visuospatial ability and language are often preserved, though speech output is reduced. Around 12.5% of people may also develop motor-neuron disease, and experience dysphagia, dysarthria and/or pseudobulbar affect (Bang et al., 2015; Burrell et al., 2011). Decline in other cognitive areas can be slow, and individuals may show no atrophy through magnetic resonance imaging (MRI) or positron emission tomography (PET) until later stages (Davies et al., 2006; Kipps et al., 2010). The progressive aphasias are characterised by early prominent language dysfunction, with preserved memory, motor, and EF. The non-fluent variant is associated with left-hemisphere degeneration, and the semantic variant is linked to temporal lobe atrophy (Snowden et al., 2002; Bang et al., 2015).

1.6 Cognitive Assessment

Cognitive assessment can inform dementia diagnosis, outline a cognitive profile of strengths and weaknesses after ABI, inform ID diagnoses through IQ testing and contribute to education health care plans. This is achieved by using a combination of domain-specific tests, or multiple tests in a pre-defined "battery", which examine each cognitive domain and their functions. Cognitive *domains* are thought as

separate entities associated with certain brain regions, though cognitive *functions* may involve multiple brain regions. Cognitive functions are thought to be receptive (input) or expressive (output). Domains (and their receptive and expressive functions) include:

- Sensorimotor (sensory input; motor expression)
- Attention (orientation, short-term stores; selective, sustained)
- Executive Function (abstraction and goal direction; task setting and task switching)
- Verbal-conceptual (comprehension; expression)
- Visuospatial (perception; construction)
- Learning and Memory (registration, encoding; recognition, retrieval)

Tests chosen must be acceptable, feasible, reliable, and valid in the domain or process they aim to measure (BPS, 2009;2015a; 2015b). *Feasibility* to an intended population is established by examining whether the test avoids floor effects (indicating it is too difficult) and ceiling effects (indicating it is too simple). If most scores fall between 'floor' and 'ceiling', it can be assumed as an accurate measure of ability (Liu & Wang, 2021). *Acceptability* is explored through ease of administration, comprehension of test instructions and good completion rates with the target population (Yardley et al., 2015). *Reliability* is the consistency and stability of a test over time (test-retest reliability), between administrators (inter-rater reliability) and across test items (internal consistency). *Validity* is the extent to which test criterion measure the phenomena of interest which it purports to measure (Price at al., 2015). However, many cognitive assessments are developed by, and normed within, Western, English-speaking, educated, industrialised, rich, and democratic (WEIRD) populations, which can restrict cross-cultural validity (Heine & Norenzayan, 2010).

Critically, comprehensive cognitive assessment must include a physical health assessment and thorough clinical interview exploring the sociocultural and historical context of the person, alongside self-reported mood, pre-morbid function, and informant report and/or observed declines in ADLs (APA, 2021).

1.6.1 Assessment of Cognitive Functions

Evidence suggests that many cognitive functions overlap and create a positive manifold, making it difficult to measure specific domains (Kovacs & Conway, 2016; Burgoyne et al., 2022). Therefore, cognitive assessments must access a wide range of domains. Descriptions of each cognitive domain and its functions, alongside typical tests used to assess these with TD individuals, are discussed below.

1.6.1.1 Attention

Attention is required for task focus, thus can be difficult to locate in a specific neural area, or isolate through testing (Hommel et al., 2019; Lindsay, 2020). Literature is conflicting regarding definitions of attention. The current study adopts a working definition of attention as a largely automatic system related to the parietal lobes, which operates separately, controlling limited mental processes flexibly across domains. This system is divided into two separate but related functions: orientation and short-term stores and selective and sustained attention (Posner, 1995; Huang et al., 2023). The receptive functions relate to consciousness (whether a person is awake, aware, and oriented to time, place, person, and situation) and short-term stores (Posner, 1995).

Selective attention is the voluntary process of identifying and recognising a target amongst distractors (Posner, 1995; Posner et al., 1998). Tasks assessing this function include visual target searching and matching such as the WAIS Coding task (Wechsler, 2012) and Kaplan Baycrest Neurocognitive Assessment (KBNA) Symbol Cancellation (Leach, 2000); or span ability such as the WAIS Digit Span Forward and Backward (Wechsler, 2012). Sustained attention can be understood as maintaining accuracy, persistence, and speed of information processing toward a target (Posner, 1995; Posner et al., 1998). Tests of this function include the Lottery and Elevator Counting tasks found in the Test of Everyday Attention (TEA) battery (Robertson et al., 1996), and the KBNA Auditory Signal Detection task (Leach, 2000), which requires the examinee to identify a target letter (e.g., X) throughout an audiotape amongst distractors.

1.6.1.2 Sensorimotor

This domain refers to the higher-order process of perceiving sensory input, for example through smell, sight, proprioception (where the body is in space) or hearing, and the verbal or motor responses to such input (Freund, 2001; Hurley & Noë, 2003). It is related to the frontal and parietal lobes, and typically assessed within tests focused on other domains which require basic perception and upper limb movement. Such tests are often given early in testing, as one of the primary processes to assess before progressing is whether, or how well, a person can understand, communicate, speak, and move their body to make marks and signal (Li & Lindenberger, 2002). Typical tests include screening measures for abnormal motor signs such as the Edinburgh Motor Assessment Scale (EMAS; Bak et al., 2015), or tests focusing on gross motor skills and manipulative dexterity, such as the Purdue Pegboard Test (Tiffin & Asher, 1948). Praxis tests involving executing highly practiced sequences of movements (e.g., “show me how you would give me the thumbs up”) can fall under this domain (overlapping with assessment of visuospatial ability). Tests of olfactory function are also available, including the University of Pennsylvania Smell Identification Test (UPSIT; Doty, 1984).

1.6.1.3 Executive Functions

EF is a set of higher-order cognitive processes thought to be related to adaptive behaviour and associated with the anterior frontal lobes (Lezak, 2012; Witt et al., 2021). These are involved in planning, self-monitoring and purposeful action (Lezak, 2012). There is no agreed model of EF, though much evidence supports a tripartite structure of working memory and monitoring (updating), self-generative behaviour and task-shifting (shifting) and inhibition (Miyake et al., 2000; Collette et al., 2005; Gross & Grossman, 2010). Working memory (WM) is the process of holding information and manipulating/working with it in the absence of stimuli, with manipulation creating a distinction from short-term memory. WM can be divided into nonverbal (visual-spatial) and verbal (Smith & Jonides, 1999).

EF assessment is fundamental to accurate dementia diagnosis and implementing effective and appropriate treatment interventions (Lezak et al., 2012; Salmon &

Bondi, 2009). Robust assessments include tests of receptive and expressive functions (Lezak, 2012), and may work best if resembling ‘everyday’ tasks (Burgess et al., 2006). EF includes employment and coordination of multiple brain systems, thus batteries containing several tests are needed to examine all functions, such as the Behavioural Assessment of the Dysexecutive Syndrome (BADS; Wilson, 1996) or the Delis-Kaplan Executive Function System (D-KEFS; Delis et al., 2012).

Single tests have been developed which claim to examine one or more executive functions. Abstraction (abstract thinking) refers to concept formation, and the ability to identify superordinate relationships between stimuli (e.g., a dog and cat are both animals). Induction is the process of noticing patterns and rules which underpin recurring events and shifting behavioural responses in accordance to rule changes (Lezak, 1982; 2012). Tests exploring these include the Temporal Judgement from the BADS (Wilson, 1996), Raven’s Progressive Matrices (Raven, 1936), the Tower of London Test (TOL; Shallice, 1982), and the ‘Frog Hop’ from the Hayling and Brixton Tests (Burgess & Shallice, 1997). Tasks used in IQ or educational testing may also be employed, such as the Miller Analogies Test (MAT; Miller, 1960) or adaptations with lower floor scores as seen in the Wide Range Intelligence Test (WRIT; Glutting et al., 2000). Tests of inhibition include the Stroop test (Stroop, 1935), which has been adapted for inclusion in cognitive batteries such as the Colour Word Interference Test (CWIT) within the D-KEFS (Delis et al., 2001).

Task setting involves other processes (e.g., attention) to create feasible plans with available resources to complete a task or problem-solve (Lezak, 1982; 2012). Word generation tasks such as category fluency are generally used, which may assess executive components of verbal expression (Benton, 1968; Lezak, 2012). Task switching is the process of recognising and assigning the priority of mental sets and revising this in response to changes in task priority. This involves inhibition to uphold priority in sequencing. These functions are related to purposeful action and effective performance (Lezak, 2012). Tests examining these functions include the ‘Zoo Map’ from the BADS (Wilson et al., 1996) and the rule shift trials of ‘Stroop-like’ tests.

EF assessment can include tests of frontal lobe integrity, such as Luria-style tasks (Golden & Freshwater, 2001) known as ‘bedside’ tasks due to their ease of administration. These can require repetition of a modelled motor sequence (e.g.: bimanual hand alternation), learning a motor response to a rule (e.g.: “when I tap once, you tap once”), and engaging inhibition and WM by adapting to a rule shift (go/no-go) (e.g.: “now when I tap once, you clap twice”). These formats have been adapted and included in batteries such as the Developmental Neuropsychological Assessment (NEPSY; Korkman et al., 1998).

1.6.1.4 Learning and Memory

Learning and memory relate to the processes of registration, encoding, storing, and retrieving new or learned information. It includes immediate and long-term memory, recognition, and delayed recall. Encoding is the receptive storage of information into long-term memory through rehearsal; though salient information may be encoded directly (Lezak, 2012). Recognition is implicit (learned procedures needed to complete tasks), while recall is explicit. Recall is episodic (personally experienced events), or semantic/declarative (memory for concepts that have been learned). Episodic recall can be retrograde or anterograde (Lezak, 2012). Though other lobes are implicated depending on the sensory qualities of information, these processes are generally linked to the temporal lobe and limbic system (Squire at al., 2020).

Good tests of memory employ both visual and verbal tasks (BPS, 2015a). Verbal tasks include the Rey Auditory Verbal Learning Test (AVLT; Lezak, 2012), the Buschke Selective Reminding Test (BSRT; Buschke, 1973), and the Cued Recall Test (CRT; Tulving & Pearlstone, 1966). Visual tests include the Pattern Recognition Test of the Cambridge Neuropsychological Test Automated Battery (CANTAB-PRM; Sandberg, 2011), and verbal tests include the Buschke Selective Reminding Test (BSRT; Buschke, 1973), or picture recognition format tasks (e.g., Wilson & Atantablin, 1980). Tests can also include materials learned previously within a battery, as in the Prudhoe Cognitive Function Test (PCFT; Kay et al., 2003).

1.6.1.5 Visuo-Spatial Functions

Visuospatial ability is related to the ventral ("what") and dorsal ("where") systems in the primary visual cortex. The ventral stream continues to the temporal lobe for object perception, recognition and naming, and the dorsal stream extends to the parietal lobe for spatial location and locomotion. Processes involve discrimination and recognition of object form, colour, distinction, and location in space, alongside construction and praxis (Goodale & Milner, 1992; Hebart & Hesselmann, 2012). The latter are tests involving voluntary motor movements. These processes involve organisation and mental manipulation, thus may involve EF (Harvey, 2019).

Typical tasks of visuospatial perception include the Judgment of Line Orientation (JLO; Benton et al., 1978), requiring examinees to determine target lines from a reference graphic of 11 drawn lines. Tasks of construction include Block Design (BD; Wechsler, 1989) which require recreating a stimulus design outlined on paper with a set number of patterned blocks; and draw-copy tasks such as the Rey-Osterrieth Complex Figure Test (Rey, 1941). Many widely used tests may be culturally and experientially bound, therefore tasks such as Matchstick Copy may be used with people of cultural global majority, or those with limited writing ability (Jones Chesters, 2021). Praxis may also fall under this domain, with many tasks derived from those which explore apraxia (impairment in execution of sequenced movements and/or gestures) (Heilman et al., 1993).

1.6.1.6 Verbal Functions

Verbal functions are generally related to the left temporal lobe, and include understanding patterns of sound, comprehension of graphic and symbolic images, and the production of speech or writing (Mesulam et al., 2013). Comprehension refers to perception of phonological input patterns; or creating meaning out of visual arrays. These are converted into meaningful words and phrases, with input from context and grammar (Mesulam et al., 2013).

Assessments focused on specific impairment of these skills include the Boston Diagnostic Aphasia Examination (BDAE; Goodglass & Kaplan, 1972), and can involve ensuring appropriate response to requests (e.g., “touch your head”). Typical stand-alone tests include the British Picture Vocabulary Test (BPVS; Dunn et al., 1982) which measures verbal comprehension, and simple naming tests such as the Boston Naming Test (BNT; Kaplan et al., 1976). Verbal ability is often measured through IQ assessments, such as within the ‘Verbal Comprehension Index’ subsection of the WAIS-IV (Wechsler, 2012), which includes ‘Vocabulary’, ‘Similarities’ and ‘Information’ (answering questions of general knowledge). However, these may not be valid cross-culturally, as performance may depend on economic, linguistic, and cultural background (Lonigan et al., 2013; Cockcroft et al., 2015).

1.6.1.7 Estimating Optimal Ability

To understand cognitive decline, assessment outcomes must be compared to a measure of a person’s optimal ability. This can be done through comparison of a previous score on the same cognitive assessment. If unavailable, tests of premorbid ability such as The Test of Premorbid Functioning (TOPF; Holdnack et al., 2013) can be employed, which assesses cognitive functions that are largely preserved in dementia (e.g., highly practiced vocabulary and phonemes).

1.6.2 Dementia Assessment

As mentioned, cognitive assessment can inform dementia diagnoses. Cognitive tests are created for (and normed within) certain populations. In the TD population, test results are compared against such norms, to understand performance against ‘typical’ members of that population with similar characteristics, alongside comparison with an estimation of the individual’s optimal ability. Severity and rate of decline can differentiate between normative, or dementia-related, decline. Some tests are designed for brief assessment of key domains specifically to identify dementia-related decline, such as Addenbrooke’s Cognitive Examination III (ACE-III; Mathuranath et al., 2000).

1.7 Dementias in Adults with Intellectual Disabilities

In the UK, dementia prevalence is three-four times higher for PWID than the TD population (BPS, 2015b). This may increase due to the accelerated ageing seen in PWDS and increase in life expectancy for PWID discussed previously (Bittles & Glasson, 2004; Patterson & Cabelof, 2012; Zigman, 2013). As mentioned, establishing dementia-related cognitive decline requires comparison measures of the individual's baseline cognitive abilities, or premorbid estimation of ability, based on a normative sample of similar age, premorbid ability, and education. PWID have unique cognitive profiles compared to TD counterparts, and between ID aetiologies, which has implications for clinical presentations and trajectories in dementia in this community.

1.7.1 Alzheimer's Disease in Down Syndrome

Research indicates amyloid plaques and neurofibrillary tangles are present in the brains of PWDS by age 35 (Zigman et al., 2008). This may be explained by accelerated ageing and the function of chromosome 21 in contributing to the development of amyloid precursor protein (APP), which is involved in AD-related neurodegeneration (Hampel et al. 2021). Subsequently, AD risk is significantly elevated for PWDS compared to TD individuals and other PWID, with a majority showing AD pathology by age 40 (Lott & Head, 2019).

A domain which may be crucial to accurate and timely AD diagnoses for PWDS is EF. Compared to TD individuals, executive dysfunction may emerge sooner, alongside behavioural and mood changes (Lautarescu et al., 2017), and before memory impairment (Ball et al., 2008; Adams & Oliver., 2010), thus showing a differing AD trajectory. This may be due to pre-existing neurodevelopmental deficits in the frontal lobes, which are more susceptible to earlier and more rapid neurodegeneration (Cooper & Prasher, 1998; Adams & Oliver, 2010; Dekker et al., 2015). This may mimic more behaviourally disordered forms of dementia (e.g., FTD, DwLB) in DS, highlighting the potential for diagnostic overshadowing. However,

some research indicates that visuospatial organisation and memory skills may first decline for PWDS in prodromal stages (Devenny et al., 2002; Krinsky-McHale & Silverman, 2013). Though there are discrepancies in which functions are first affected, evidence suggests that disease trajectory differs from the TD population.

Though typical cognitive assessment does not include olfactory ability (OA), evidence suggests that decline in OA may be a precursor to MCI or AD in the TD population (Wilson et al., 2007). This decline may be apparent earlier and to a more significant degree in PWDS than ID of other causes (Nijjar & Murphy, 2002). A recent systematic review by Manan and Yahya (2021), examining available research on the assessment of olfactory threshold, discrimination, and identification in PWDS, provides strong support that olfactory impairments are present and measurable before the age of 30, and increase in severity with age. This may support research suggesting that PWDS have smaller olfactory bulbs than TD counterparts, leading to earlier and more severe olfactory impairment with dementia-related neurodegeneration (Bianchi et al., 2014; Bontempi et al., 2020). Though OA tests have the potential to be a direct, accessible, and non-invasive way to identify AD in PWDS, research concerning this is scarce or outdated.

1.7.2 Presentation of Dementia in PWID

Research on dementias in other ID phenotypes with moderate-profound ID is scarce, perhaps due to greater heterogeneity (BPS, 2015a). However, research suggests that the diagnostic criteria for dementia does not accurately reflect the clinical presentations of PWIDs (Stanton & Coetzee, 2004). Sheehan et al. (2015) explored the diagnostic reliability and validity of the ICD-10 (WHO, 1992) and DSM-IV-TR (APA, 2013) for ascertaining a dementia diagnosis for PWDS. Results showed that dementia diagnoses were correctly identified in 70.3% of cases using the ICD-10, and 56.3% using the DSM-IV-TR. Accuracy rose to 84.4% if clinicians experienced in understanding dementia presentations in PWDS incorporated their clinical judgement, rather than diagnostic criteria alone. However, many healthcare professionals feel under-skilled in recognising the symptoms of dementia in people with severe-profound ID (Dekker et al., 2021). Despite this, there are no adapted

diagnostic criteria available for PWID. This creates significant difficulties in accurate cognitive assessment.

1.7.3 Cognitive Assessment of Dementia in PWID

The BPS (2009;2015a; 2015b) recommends a cognitive instrument which is validated for assessing dementia in PWID, that assesses all domains and functions previously stated, with considerations to any additional tests needed (e.g., praxis). A supplementary informant report of symptoms, such as the Dementia Screening Questionnaire for Individuals with Intellectual Disabilities (DSQID; Deb et al., 2007), or the Dementia Questionnaire for People with Learning Disabilities (DLD; Evenhuis, 2018), is recommended (NICE, 2018). The most used cognitive tests in the UK are the Cambridge Cognitive Examination (CAMCOG; Roth et al., 1986), the CAMCOG adapted for PWDS (CAMCOG-DS; Ball et al., 2004), and the Neuropsychological Assessment of Dementia in Individuals with Intellectual Disabilities (NAID; Crayton et al., 1998) (BPS, 2015a). A list of available measures derived from the literature review is seen in Appendix B.

1.7.4 Difficulties in Cognitive Assessment of PWID

Dementia diagnosis is based on significant changes from baseline functioning, not a deviation from a level of functioning expected of the TD population. However, there is no ‘gold standard’ in diagnosing dementia in PWID (Krinsky-McHale et al., 2020), Typical assessment methods may not be appropriate due to comparison against TD norm data. Several other factors can impede cognitive assessment with PWID, which are discussed below.

1.7.4.1 Uniqueness of Cognitive and Behavioural Profiles

As discussed, differences across ID aetiology and severity are found in a wide range of cognitive domains. This can lead to trajectories and clinical presentations of dementia that differ from the TD population, rendering it inappropriate to apply single-domain assessments of function (e.g., memory, EF) when assessing for dementia (Krinsky-McHale & Silverman, 2013). This also impedes establishment of

norm data as a comparative baseline of premorbid cognitive and functional ability, which is necessary for understanding dementia-related cognitive decline (Moran et al., 2013). Presence of comorbid conditions can further increase disparities in cognitive profiles. Depression is thought to be experienced by 2.5-3.4% of PWID and manifest differently to TD individuals, with symptoms also presenting similarly to dementia (Oliver, 1999; Costello et al., 2006; Maiano et al., 2018). Further, overuse of psychotropic medications for behaviour that challenges can increase the presence of symptoms such as dyskinesia and tremors (BPS, 2015a; Branford et al., 2018), modifying dementia trajectories and presentations.

1.7.4.2 Over-Reliance on Informant Report

Informant report is crucial to best-practice dementia assessments, providing additional supportive information to cognitive testing (NICE, 2018). Most available cognitive assessment instruments for PWID are based at least partially on informant-led questionnaires (Zeilinger et al., 2013). This may be linked to research suggesting difficulty in validating self-reported cognitive and/or emotional states of PWID, potentially due to difficulties in establishing normative sample data (Finlay & Lyons, 2001; Moran et al., 2013). However, self-report measures of general mood, adaptability, interpersonal and intrapersonal domains have been validated for PWDS (Robles-Bello et al., 2020; Sánchez-Teruel et al., 2020). Therefore, over-reliance on informant report may be interpreted as discriminatory practice which privileges informant report over PWID. Further, informant reports can focus on ADLs, and reflect the extent that caring for a PWID affects the carer, rather than changes in cognitive functioning (Elliott-King et al., 2016). This may over or under-estimate the true level of impairment and functioning of PWID.

Informant report may also impede estimations of pre-morbid ability. To give a confident diagnosis of dementia, data on premorbid and current behavioural and cognitive functioning across at least 6 months must be available (Aylward et al., 1995). Current estimates such as the TOPF (Holdnack et al., 2013) assume a literacy level typical of the TD population, and proxy measures such as the British

Picture Vocabulary Scale (BPVT; Dunn et al., 1982) are based on TD levels of pre-morbid ability, thus both may be inappropriate for PWID. Informant report may instead be used, however many PWID in care homes may have a high turnover of carers, and variability in note-keeping, leading to incomplete or low-quality estimations of functioning (Holland et al. 2000).

1.7.4.3 Assessing Executive Function

Though EF may be an important preclinical indicator of dementia-related decline, there is no agreed test battery for assessment of EF in PWID (BPS, 2015a). Batteries such as the Cambridge Executive Functioning Assessment (CEFA; Sandberg, 2011), and an adaptation of the BADS (Wilson et al., 1996), the Behavioural Assessment of Dysexecutive Functioning for Intellectual Disabilities (BADS-ID; Webb et al., 2020), have been created for assessment of EF in PWID, and are comparable in reliability and validity (Webb et al. 2020). The CEFA was created in line with the tripartite concept of EF processes (Miyake et al. 2000; Collette et al., 2005; Gross & Grossman, 2010), which implies that executive functions in PWID also resemble this structure (Willner et al., 2010). However, neither are normed with PWID.

1.7.4.4 Floor Effects

As discussed previously, floor effects indicate tests are too difficult for the intended population, and likely not valid. PWID often show floor effects in normative tests used for dementia assessment (Lautarescu et al., 2017). This may be partly due to communication differences, as PWID (particularly PWDS) often show difficulties in verbal communication skills, with difficulties increasing with ID severity (Smith et al., 2020). PWID can also show difficulties in processing speed, verbal expression, comprehension, and abstract reasoning (Hassiotis et al., 2012). This may hamper their ability to engage with tests or score in timed tasks, contributing to floor effects. This suggests a range in abilities for PWID which are not accounted for in normative tests, impeding the ability of many available tests to detect dementia-related decline.

1.7.5 Summary

Several difficulties impede appropriate and accurate cognitive assessments with PWID. Considering the importance of examining cognitive functioning alongside informant report in dementia assessment, available instruments which are not informant-led must be identified. Appropriate tests of EF for PWID must also be identified, which consider the disparate cognitive and behavioural profiles within the ID population. OA may also be an accessible route of cognitive assessment of PWID, which should be explored. The following literature review explores available tests and cognitive batteries used in cognitive assessment with PWID. It will consider their appropriateness to this community and their sensitivity to normative and dementia-related cognitive decline. Findings inform the construction of a novel draft cognitive battery for PWID.

1.8 Literature Review

1.8.1 Aims

The aims of the review were to:

1) Identify global cognitive test sets, assessments, and instruments used with PWID to assess cognitive decline, which are administered directly (not via informants).
2) Examine included tests for acceptability, feasibility, validity and/or reliability when used with PWID.
3) Identify any novel/adapted tests produced since the review by Paiva and colleagues (2020).

The review does not replicate a systematic review, or to identify all available research, due to the breadth of the topic areas and the scope of this thesis. Therefore, quality assessment of papers was not undertaken, though the Preferred Reporting Items for Systematic Reviews and Meta-Analyses (PRISMA) (Moher et al., 2009) checklist and flow diagram were used as a guideline to report findings (see Appendix C).

1.8.2 Methods

A systematic review concerning this topic was undertaken by Zeilinger et al. (2013), covering research published between 1948-2010, and expanded upon by Paiva et al. (2020), without limiting the date or language of publications. Therefore, the current review spans from 1980- April 2021 to build upon previous reviews while excluding papers where constructs and tests have since been revised. The literature search was conducted through EBSCO, including CINAHL, APA PsycArticles and APA PsycINFO electronic databases. Further relevant papers were identified through citation-searching and consultation with researchers in the field. Full details of the search strategy method (and a summary of methods of included studies) are seen in Appendix D.

67 studies were identified. Studies were classified as 'cross-sectional' if the sample was assessed at a single time point and 'longitudinal' if assessed at multiple time points. Data concerning relevant outcomes (cognitive assessment results and psychometric properties), study methods and characteristics, and sample characteristics were extracted (Mann, 2003). A total of 114 direct cognitive tests and 37 batteries were identified across the studies identified for inclusion. IQ measures were not addressed, as they were largely not used for dementia assessment. Full details of included studies are shown in Appendix E. Results indicated that instruments fall into four main categories: single domain tests, brief instruments, screening tests, and comprehensive assessments. The most commonly appearing tests are described below.

1.8.3 Single Domain Tests

1.8.3.1 Attention

Many studies used a stand-alone test for arousal and orientation from an existing battery, such as the 'orientation' subtest of the Mini Mental State Examination (MMSE-O; Folstein et al., 1975), or Working Group's 'Orientation' test (WG-O; Burt and Aylward, 2000). These measure awareness through orientation to person, time, situation, and place, using questions such as "What is your name?", "How did you get here today?" or "What month is it?".

The Symbol Cancellation Task (SCT)

The SCT is a subtest within the Kaplan Baycrest Neurocognitive Assessment (KBNA) (Leach, 2000). This was created for TD individuals and requires examinees to search for a visual target amongst distractors. Krinsky-McHale et al. (2008) adapted the SCT, repeating administration over two years with PWDS. This paper-based task includes target English letters to 'strike out' amongst a field of distractors. Findings showed sensitivity to progressive impairment in selective attention up to two years prior to meeting criteria for diagnosis of AD, showing discriminative ability between participants with or without dementia through good specificity (correct detection of non-AD cases) and sensitivity (correct identification of AD cases). The task was easy to administer, suggesting acceptability. However, letter recognition may be culturally bound, and rely on level of support with phonetic strategies in schooling (Næss et al., 2012). Therefore, this test may not be appropriate to all PWID.

1.8.3.2 Sensorimotor

The most common test was the Brief Praxis Test (*n*= 4) followed by the Beery Buktenica Developmental Test of Visual-Motor Integration (*n*= 2). Though not used with PWID, relevant literature regarding the UPSIT was included through citation-searching as a potential novel testing avenue for the novel battery.

The Brief Praxis Test (BPT)

The BPT (Dalton & Fedor, 1997) is a 20-item test developed for PWDS. The BPT asks participants to follow instructions to lift body parts, alongside placing coins in a jar in specific ways. It does not require extensive verbal ability, but relies on finer motor abilities, which may be inappropriate for PWID with motor impairments. Diagnoses of dementia and severe ID are reported to be related to lower BPT scores in PWDS, and sensitive to changes in cognitive profile over time (Sano et al., 2005; Head et al., 2011). Further, Powell et al. (2014) found that PWDS with reduced white matter integrity in frontoparietal regions correlated with poorer BPT scores. This was more evident in individuals with a dementia diagnosis. This suggests that these affected pathways are a preclinical indicator of AD in PWDS, indicating the BPT may be sensitive to prodromal decline. Though study samples are largely comprised of those with mild ID, all studies found minimum floor or ceiling effects, suggesting acceptability and validity for some PWDS.

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

The BBDT-VMI (Beery et al., 1997) and its revisions were created for the TD population, requiring drawing copies of increasingly complex geometric shapes. Krinsky-McHale et al. (2020) found lower scores are associated with AD in PWDS, but not to MCI. Burt et al. (2005) reported that decline in scores on the BBDT-VMI may be associated with dementia in PWDS, though slope of decline, rather than score difference, may be a more reliable indicator. No floor effects were noted, suggesting the BBDT-VMI is a feasible test to PWDS, but may not be sensitive to prodromal dementia-related decline.

University of Pennsylvania Smell Identification Test (UPSIT)

The UPSIT (Doty et al.,1984) is a standardised test of olfactory function created for TD individuals, with strong reliability shown in clinical trials (Juniper et al., 2005). Examinees smell an odorant strip and identify the odour from four multiple choice answers. UPSIT performance strongly predicts advancement from MCI to AD in TD

adults, indicating sensitivity to dementia-related decline (Tabert et al., 2005). Schmitt et al. (2010) identified UPSIT scores are a significant moderate correlate of outcome scores on the immediate and delayed memory indexes in the RBANS, but not premorbid IQ. This indicate it is sensitive to dementia-related decline regardless of premorbid cognitive ability, which may be useful in communities with high cognitive variability such as PWID. Though no research was found with PWID samples, the UPSIT shows potential as a non-invasive approach suitable to a population with unique pre-existing cognitive impairments.

1.8.3.3 Language

The most common tests were the PPVT (*n*= 6) and BNT (*n*= 4).

Boston Naming Test (BNT)

The BNT (Kaplan et al., 1976) is a 15-minute test of word retrieval developed for TD adults with aphasia or acquired language disorders, often included in the Boston Diagnostic Aphasia Examination (BDAE; Goodglass & Kaplan, 1972). It includes 60 monochrome line drawings varying in difficulty of recognition and familiarity. Scores are number of items correctly named, regardless of semantic cue given.

Palmer (2006) found that PWDS and dementia scored over one standard deviation below a control group of PWID without dementia on the BNT. However, Jozsvai et al. (2002) found similar BNT scores between PWDS over the age of 40, regardless of dementia status. BNT scores were most affected by age, indicating sensitivity to normative rather than dementia-related decline. Pulsifer et al., (2020) found BNT scores to decrease significantly with increasing dementia severity in PWDS but did not significantly predict dementia status. However, findings may reflect unique cognitive profiles and patterns of dementia-related neurodegeneration in PWDS, and changes may be found if used longitudinally in a younger cohort, considering the earlier onset of dementia in PWDS (Zigman, 2013; Lott & Head, 2019).

Peabody Picture Vocabulary Test

The PPVT (Dunn, 1981) was developed to measure receptive language and comprehension in TD children. Examinees listen to a word, and then select a picture which best matches the word. Therefore, the PPVT measures language ability without requiring verbal expression. Das et al. (1995) found that PPVT scores effectively discriminated between older PWDS or non-DS ID, and their younger (40-49 years) counterparts. However, no data on dementia status was collected, so it is difficult to establish whether the PPVT is sensitive to *age*-related or *dementia*-related decline. Alexander et al. (1997) found no differences between older (41-61 years) and younger (22-38 years) DS adults on PPVT scores when controlling for ID severity. However, Nelson et al. (2001) found that PWDS who showed prefrontal lobe atrophy in MRI scans scored significantly lower on the receptive language test of the PPVT than those with 'typical' MRI findings. Mixed evidence makes it difficult to establish the clinical utility of the PPVT, though accessibility to PWID seems high.

1.8.3.4 Visuospatial

The most used measure (*n=5*) was the Wechsler Block Design test (BD; Wechsler, 1989), which appears in several IQ batteries. The examinee uses patterned blocks to recreate a model presented to them, with scores based on speed and accuracy. Alexander et al. (1997) found significantly lower BD performance in older PWDS than a younger DS comparator group. In a recent longitudinal study, Hartley et al. (2020) found neocortical APP concentration was significantly associated with BD scores of PWDS, but scores could not consistently distinguish between preclinical (asymptomatic) and prodromal AD cases, indicating limited reliability and sensitivity to early dementia-related decline.

1.8.3.5 Executive Function

The most common tests were the TOL (and its revisions; *n*= 8), CaD (*n*= 5) and the CFT (*n*= 3).

Tower of London and Tower of London-Drexel University: 2nd Edition (TOL^{DX})

The Tower of London Test (TOL; Shallice, 1982) was created originally for TD individuals; a revision of this (TOL^{DX}; Culbertson & Zillmer, 2005) appears in the CEFA (Ball et al., 2008), and in a computerised format in the Cambridge Neuropsychological Test Automated Battery (CANTAB; Sandberg, 2011). The TOL measures abstraction, cognitive flexibility, planning and problem-solving, requiring examinees to move coloured beads across pegs on a board to solve different problems.

Willner et al. (2010) found the TOL correlated highly with BPVS (Wechsler, 1981) scores, indicating performance may rely on verbal comprehension. Further, Cooper et al. (2016) found little change over time for TOL scores, indicating poor sensitivity to cognitive decline. An adapted TOL for PWID with reduced difficulty scaling was created by Masson et al. (2010), which correlated with carer scores on the standardised dysexecutive questionnaire from the BADS (Wilson, 1996), indicating sensitivity to EF impairment. García-Alba et al. (2017) examined the psychometric properties of the TOL^{DX} with PWDS with mild-moderate ID. Results showed high reliability and consistency across ID severity, good discrimination between ID severity groups and high association with other measures of EF. An absence of floor effects was noted, though difficulty did not seem to increase as the test progressed.

Sinai et al. (2016) found no significant difference in TOL scores between PWDS aged 45+ with and without dementia, indicating poor sensitivity to dementia-related decline. However, findings may be explained by evidence suggesting that EF decline is an early feature of AD in PWDS (Lautarescu et al., 2017), which may be present throughout the older sample of Sinai and colleagues' research (2016) regardless of dementia diagnosis. Findings suggest that, with adaptations, the TOL may be appropriate for use with people with mild-moderate DS-ID. However, this test may not represent 'everyday' EF functions, nor be appropriate as a non-verbal EF

measure. Further, task materials are not easily replicated, which may limit accessibility to services.

Cats and Dogs Task (CaD)

CaD is a Stroop-like (Stroop, 1935) subtest of the CEFA (Ball et al., 2008). CaD was created for PWDS and examines WM and inhibition through two trial conditions. The examinee moves through a sequence of 16 pictures of dogs and cats, naming them congruently, then incongruently (i.e.: 'dog' as 'cat' and vice-versa). Scores are the summation of time taken in condition one subtracted from condition two (indicating the cognitive 'cost' of inhibition).

Willner et al. (2010) evaluated the utility of the CaD as a predictor of dementia in PWID. Findings showed an absence of floor effects, and no correlation with BPVS scores, indicating it does not rely on verbal ability and is appropriate when considering the variety of verbal comprehension ability in PWID. Bevins and Hurse (2014) found the CaD was sensitive to cognitive decline, as scores correlated negatively with informant-rated decline scores on the DLD (Evenhuis, 2018). However, findings showed ceiling effects and narrow score ranges, indicating this task may be too easy for those with mild-moderate ID. Conversely, Cooper et al. (2016) noted PWID of varying severity had difficulties completing the CaD. Though findings are mixed, they imply the CaD has sufficient ecological validity, acceptability, and dementia-related sensitivity for use with PWID.

Category Fluency Test (CFT)

The CFT (Benton, 1968) is a task of semantic fluency created for TD adults, requiring examinees to give as many category-specific words as possible in one minute. It seems ecologically valid, using 'everyday', widely familiar tasks (e.g., creating a shopping list). It is usually accompanied by a task of phonemic fluency. An adapted version created for TD children, the McCarthy Category Fluency Test (M-CFT; McCarthy, 1972) uses a shorter timeframe (20 seconds) and more lenient

scoring criteria. These tasks are thought to measure EF through inhibition, task planning (strategy of word retrieval) and monitoring, as words produced must be unique (no repetition). Pulsifer et al. (2020) found declining M-CFT scores to be a strong predictor of AD in PWDS, and Cooper et al. (2016) note it is easy to complete. Findings suggest that CFT-like tasks are accessible, with good clinical utility.

1.8.3.6 Learning and Memory

The most common tests were the CRT (*n*= 9) and the BSRT (*n*= 3).

The Cued Recall Test (CRT)

The CRT (Tulving & Pearlstone, 1966; Grober & Buschke, 1987) was created for TD adults and assesses memory for previously learned words or phrases (episodic memory), using semantic cues. A modification with simpler semantic categories (CRT-M; Devenny et al., 2002; Zimmerli & Devenny, 1995) was created for PWDS and shown to be reliable and valid for this population. In the learning (encoding) phase, examinees are given 12 pictures linked to unique categories to learn; this can be repeated up to three times if necessary. Examinees then engage in free and cued recall trials. A delayed recall trial for both cued and free conditions is given after an interval, creating four separate scores: free immediate recall, total, free delayed recall, and total delayed recall.

Benejam et al. (2015) found that CRT-M free recall and intrusion error scores distinguished between healthy PWDS and PWDS with AD, but difficulties in task comprehension for PWDS with severe ID or late-stage AD were seen. Devenny et al. (2000) found introducing a cut-off score of <23 for the CRT-M total score produced sensitivity of 94.7% and specificity of 93.9%. A positive predictive value of 80.9% was found, with many participants who scored poorly at baseline on the CRT-M later receiving a dementia diagnosis. Hartley et al. (2020) identified that CRT-M scores were significantly associated with greater neocortical APP, and accurately indicated transition of participants from preclinical to prodromal AD. However, all included studies recruited samples of people with mild-moderate ID and/or with early-stage

AD. Findings indicate the CRT-M is an accessible task which is sensitive to early AD dementia-related decline for PWDS with mild-moderate ID.

The Buschke Selective Reminding Test (BSRT)

The BSRT (Buschke, 1973) is a list-learning test created for TD individuals, which assesses verbal learning and memory. A list of words is read, and the examinee is asked to immediately recall as many items as possible, in any order. Floor effects are noted in the BSRT for PWID, and this was modified to include more familiar list stimuli (i.e., a list of eight animals) (BSRT-M; Hill et al., 1988). Any missed items are re-presented after the initial recall trial; therefore, it can be considered both a measure of short and long-term memory.

High test-retest reliability is shown for the BSRT-M (Devenny et al., 1996). Krinsky-McHale et al. (2002) found BSRT-M scores could discriminate between healthy PWDS and those with early AD. It was found to be sensitive to AD-related and age-related decline in verbal explicit memory. In a further study, Krinsky-McHale et al. (2008) showed a relationship between AD severity/progression and BSRT-M scores, further supporting evidence of dementia-related sensitivity. Findings suggest the BSRT-M is an appropriate and clinically useful measure of learning and memory for PWID.

1.8.4 Brief Instruments of Cognitive Performance

The most commonly used brief global assessments were the CEFA (n=7), SIB (*n*= 7), DSMSE (n= 6), TSI (*n*= 6) and PCFT (*n*= 5). Though only used in one identified study, the BADS-ID was included to explore utility in the draft battery, considering the paucity of EF batteries designed for PWID. The TESTAD was also reported as it showed robust assessment of EF.

The Cambridge Executive Functioning Assessment (CEFA)

The CEFA (Ball et al., 2008) was created to assess EF in PWID. It comprises of six tests: verbal fluency, Weigl Sorting (WST; Weigl, 1927), CaD, TOL, Scrambled Boxes and Spatial Reversal. The CEFA is administrated alongside the CAMCOG-DS (Ball et al., 2004), which is discussed later. Administration takes around one hour.

Ball et al. (2010) and Fonseca et al. (2019a, 2019b) report changes in adaptive behaviour predict scores on CEFA tasks, indicating it may be a valid assessment of EF. Ball et al. (2008) found PWDS with AD showed consistently poorer performance across all tests than non-AD DS adults. However, the WST, Verbal Fluency, and CaD tests were less sensitive to discrimination between dementia group condition. Performance on TOL was significantly affected by ID severity and increasing age, with floor effects found for the ID-AD group. Spatial reversal was the only task not significantly affected by ID severity. Willner et al. (2010) reported WST produced significant floor effects and was difficult to administer with PWID. However, Verbal Fluency and CaD seem accessible to PWID, with simplistic instructions and minimal resources required for administration. Similarly, Bevins and Hurse (2014) found the WST was too complex for PWID, but relative usefulness of the CaD and Verbal Fluency tasks. The Verbal Fluency task correlated with BPVT scores, which may imply increased cognitive demand on lower-order processes known to be impaired for PWDS, such as verbal abilities (Lott & Dierssen, 2010). This may have been reduced with additional prompts or cues. However, many studies report floor effects, and some ceiling effects in tasks (e.g., CaD), particularly for people with severe ID.

The Severe Impairment Battery (SIB)

The SIB (Panisset et al., 1994) examines orientation to name, memory, social interaction, language, attention, orientation, visuospatial and constructional abilities, and praxis. It was created for TD individuals with severe dementia, using simple instructions and gesture cues. The SIB gives a total score and six major subscale scores in attention, construction, memory, visuospatial ability, orientation, and

language. Administration time is 30 minutes; the upper limits of concentration for those with late-stage dementia.

A shortened version (SIB-S) has been developed which retains the reliability and validity of the SIB and takes 10-15 minutes to administer, reducing cognitive load (Saxton et al., 2005). The SIB shows high criterion validity and test-retest reliability in healthy PWID, with minimal floor effects (Witts & Elders, 1998). Good concurrent criterion validity in comparison to the DLD is also shown in healthy PWDS (Hutchinson & Oakes, 2011). However, Head et al. (2011) found the SIB was not sensitive to dementia status or cognitive decline in a sample of PWDS, and McKenzie at al. (2002) found only the orientation domain showed discriminant validity of AD-related cognitive decline in this population. Therefore, the validity of the SID to detect dementia-related changes in PWID is unclear.

Down Syndrome Mental Status Examination (DSMSE)

The DSMSE Haxby (1989) was created for PWDS and examines age-related cognitive changes. Tests include measures of information, orientation, short-term recall and recognition, language (naming clothing and body parts), visuospatial construction and praxis (executing a sequence of tasks).

DSMSE performance has been shown to decline with age on all areas except praxis (Manning et al., 1998). Cosgrave et al. (1998) found DSMSE scores to distinguish between PWDS with and without dementia (in those with moderate ID), but showed score ranges, and floor effects for those with advanced dementia. Floor effects or the DSMSE are also seen for those with severe ID (Krinsky-McHale et al., 2020). McCarron et al. (2014) found evidence of predictive validity for the DSMSE, as score decline was present in a sample of PWDS up to one year prior to dementia diagnosis. However, DLD score decline was present up to five years prior, indicating informant-rated measures as more sensitive. Considering this, and the paucity of tasks examining other EF functions (such as abstraction and task-switching), the

DSMSE may show limited dementia-related sensitivity to PWDS without supplemental measures.

The Test for Severe Impairment (TSI)

The TSI (Albert & Cohen, 1992) was created for adults with severe cognitive impairment. It includes assessment of language comprehension (e.g., action on request) and production (e.g., body part naming), immediate and delayed object memory, visuospatial skills, and motor function (e.g., “Show me how you would wave hello”). It can be administered in 20 minutes, which may reduce testing fatigue, and is validated for use with PWID (Tyrrell et al., 2001).

Cosgrave et al. (1998) reported acceptable test-retest and inter-rater reliability for the TSI, alongside satisfactory convergent validity, with a good score range preferable in comparison to the DSMSE. Most individuals with moderate/severe ID can perform on the TSI but may fail to score if they have very advanced dementia (Cosgrave et al., 1998; Krinsky-McHale, 2020). The TSI seems accessible to PWID, as most tasks require non-verbal responses. However, there are no alternatives to scoring for non-verbal participants. McCarron et al. (2014) found the TSI could identify cognitive decline in PWDS up to one year prior to reaching clinical threshold for dementia. However, decline became more gradual post-diagnosis, with informant measures showing higher sensitivity (McCarron et al., 2017). Conflictingly, some research suggests no significant difference in test performance between PWID with and without dementia (Pyo et al., 2007; 2010). Results of sensitivity to dementia-related decline are mixed, and generally indicate the TSI as inappropriate for those with late-stage dementia.

The Prudhoe Cognitive Function Test (PCFT) and Prudhoe Cognitive Function Test-Short (s-PCFT)

The PCFT (Kay et al., 2003) is a 58-item measure created for PWID for administration by those without specialist knowledge. It assesses orientation, recall,

language expression and comprehension, praxis (e.g., “show me how you would wave hello”) and calculation. The s-PCFT consists of 21 items and can be considered a screening measure. The PCFT utilises motor responses for many items, which implies it is a suitable measure for non-verbal PWID.

Kay et al. (2003) found that all PCFT subtests were valid for use with PWID with mild-severe aetiologies, except for recall. Floor effects are seen in people with profound ID, and the PCFT’s ability to detect dementia-related cognitive decline is reduced with increasing ID severity (Tyrer et al., 2010). Margallo-Lana et al. (2003) found high test-retest reliability and inter-rater reliability in detecting cognitive deterioration when administered by non-specialists, though floor effects were found for BD (which was subsequently removed from the PCFT). Recently, the s-PCFT has been validated for PWID in Italian, with a wide range of scores, no floor effects for praxis and language subtests, and minimal ceiling effects (DeVreese et al., 2021). Studies indicate that low s-PCFT scores are associated with later dementia diagnosis, with scores being significantly related to ID severity but not age for PWDS, indicating sensitivity to dementia-related decline (Margallo-Lana et al., 2007; DeVreese et al., 2021). Findings imply the s-PCFT has potential to be cross-culturally valid, but may be inappropriate for people with severe ID.

The TESDAD Battery

The TESDAD (De Sola et al., 2015) was developed to ascertain the cognitive profile of PWDS as a baseline for interventional changes in clinical trials. Tests include much of the CANTAB battery, Digit Span Forward (Wechsler, 1981) to examine verbal attention, the CRT-M to assess verbal episodic memory, and a semantic fluency task. Visual and verbal WM was assessed with the Spatial Span backward recall (SSP, CANTAB) and the Digit Span backward, respectively. Planning was measured using the TOL^{DX} and mental flexibility with the WST. The CaD was used to assess response inhibition. Expressive and receptive language were assessed using the BNT and the Token Test, respectively. Notably, EF is well-assessed in this battery. De Sola et al. (2015) found floor effects for the WST, SSP and Digit Span

tests, alongside ceiling effects for the CaD and CRT-M is a sample of PWDS. Though not created for sensitivity to dementia-related decline over time, some tests could be adapted to increase feasibility and acceptability to PWID by examining the reasons behind floor and ceiling effects.

The Behavioural Assessment of the Dysexecutive Syndrome (BADS) and Intellectual Disabilities Adaptation (BADS-ID)

The BADS (Wilson et al., 1996) was created for assessment of EF in TD individuals following acquired brain injury. It includes an informant-report dysexecutive questionnaire (DEX) and six subtests administered directly: the 1) Rule Shift Cards, 2) Key Search, 3) Temporal Judgement, 4) Zoo Map, 5) Action Program and 6) Modified Six Elements. 1) requires subjects to say “yes” for red cards and “no” for black cards when presented, then say “yes” if two cards of the same colour are presented in sequence. 2) requires finding lost keys in a field. 3) involves estimating the time required for events to take place. 4) demonstrates route planning which adheres to a set of rules to visit set locations around a zoo. 5) requires retrieving a cork out of a tube using different objects. 6) involves completing three competing tasks. Tasks may reflect more day-to-day applications of EF, showing high ecological validity (Burgess et al., 2006).

A simplified version of the BADS (BADS-C, Emslie et al., 2003) produced floor effects with PWID (Willner et al., 2010). Therefore, Webb et al. (2020) explored the validity of an adaptation of the BADS for PWID (BADS-ID) in a sample of 101. Performance was compared with properties of the CEFA, alongside BADS-C data as found from Willner et al. (2010). Feedback of participant test experience was collected. Results showed that the BADS-ID ‘Supermarket Map’ with life-like images reduced floor effects seen in the ‘Zoo Map’ from 87.5% to 2.8%, and significantly increased the proportion of ceiling effects from 2.5% to 59.2% for the second trial. Qualitative feedback indicated participants better related these tasks to their everyday lives, to aid them in task completion. Evaluations of internal consistency, face validity and inter-rater reliability for the BADS-ID were comparable or superior to

the CEFA. Though promising, the study excluded PWDS, and no further research using the BADS-ID or subtests was identified. The BADS-ID also showed poor internal consistency, meaning the extent to which the BADS-ID measures EF is unknown. Further validation of the BADS-ID with PWDS, and with healthy and dementia comparator groups, is needed.

1.8.5 Screening Tests for Dementia

The Neurological Assessment of Dementia in Intellectual Disabilities (NAID)

The NAID (Crayton et al., 1998) was designed for assessment of dementia in PWDS, but lacks measures of concept formation, abstraction, and non-verbal EF. The NAID comprises of the informant-report Vineland Adaptive Behaviour Scales (VABS; Sparrow & Chicchetti, 1985), BPVS (Dunn et al. 1982) to measure receptive language, CANTAB (Sandberg, 2011) to measure visual learning and memory, Sentence Repetition test (SRT; Spreen & Strauss, 1998) to examine attention and immediate memory, 'orientation' subsection of the CAMCOG (Roth et al., 1986), and three novel tasks of motor praxis, visuospatial EF and picture naming. Many tests rely on verbal ability.

Crayton et al. (1998) reported 18.6% (*n*= 70) of PWDS experienced floor effects at baseline, implying item difficulty is inappropriately scaled. Adams and Oliver (2010) used the Reliable Change Index statistic (Jacobson et al., 1984; Christensen & Mendoza, 1986) to explore the reliability of changes in NAID scores to indicate cognitive deterioration in PWDS with mild-moderate ID. Though significant change was found on two NAID subtests, it is unknown whether this is due to normative or dementia-related decline. Carr and Collins (2018) found floor effects for the NAID for PWID with a dementia diagnosis by age 50, and those with severe-ID, and Cooper et al. (2016) found the object memory test to be easy to complete and sensitive to change over time; however, similarly to Adams and Oliver (2010), it is unknown whether findings are due to cognitive variability or dementia-related decline. Evidence suggests the NAID as an appropriate 'snapshot' measure of cognitive deterioration for PWDS and mild-moderate ID, but not of dementia-related decline over time.

The Learning Disabilities Dementia Battery (LDDB)

The LDDB (Broxholme & Jahoda, 2000) was created for PWID and consists of 22 direct subtests, measuring: orientation, visual, verbal and recognition memory (immediate and delayed), verbal fluency, new learning, planning, visuospatial ability, abstract thinking/concept formation, and language ability. Research by Poveda and Broxholme (2016) showed the LDDB is sensitive to cognitive change over time for PWDS and may distinguish between 'probable' and 'no dementia' groups. However, this research was conducted with a small sample size, with very few participants with non-DS ID. Notably, this battery does not robustly assess EF, as it does not include a test of inhibition nor attention setting and shifting. Considering the importance of EF in establishing dementia-related cognitive changes (Lezak et al., 2012; Salmon & Bondi, 2009), this may limit the overall clinical utility of the LDDB.

1.8.6 Comprehensive Dementia Assessments

One battery aiming to provide a diagnosis of dementia was identified.

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

The CAMCOG-DS is the cognitive component of the CAMDEX-DS (Ball et al., 2004), and expands on previous revision (CAMCOG; Roth et al., 1986). The CAMCOG-DS uses various tests to assess orientation, language comprehension and expression (e.g., picture naming), memory, praxis (e.g., clock copy drawing), abstraction (e.g., similarities) and visual perception (e.g., naming pictures from unusual angles). Clock Copy may rely on pre-existing knowledge and skills, such as time-telling and writing. These experiences may be linked to educational opportunities, and thus differ across socioeconomic backgrounds and cultures (Lonigan et al., 2013). Further, the CAMCOG-DS lacks tests of planning, task setting or switching. Notably, the CAMCOG-DS-II (Beresford-Webb & Zaman, 2021) has recently been released as the cognitive component of the CAMDEX-DS-II, which shows broader assessment of EF functions than its predecessor and aims to establish pathological cognitive

change from baseline for PWID. However, this latest revision lacks tests of task setting and switching.

Several studies indicate the CAMCOG-DS detects prodromal and clinical AD in PWDS (García-Alba et al., 2019; Benejam et al., 2020; Fortea at al., 2020), and one study indicates the CAMCOG-DS correlated highly with the MMSE, with fewer floor effects in a near-population sample of PWDS aged 30-65 (Hon et al., 1999). It has been shown to be valid and reliable in predicting dementia diagnoses for PWDS cross-culturally (Fonseca et al., 2019a; Fonseca et al., 2019b). Evidence suggests the CAMCOG-DS is sensitive to dementia-related decline and predicts AD onset, however many with severe-profound ID were unable to complete the tests, showing floor effects (Benejam et al., 2020).

The CAMCOG-DS is not validated for those with severe-profound ID, and many studies report floor effects with these individuals (Hon et al., 1999; Ball et al., 2004; García-Alba et al., 2019; Fonseca, 2019a; Benejam et al., 2020). This may be linked to tests largely relying on verbal production ability and general knowledge, which show poor feasibility and acceptability to those with severe-profound ID. Notably, this is the only battery that evidences norm data for PWID, though this is specifically for PWDS and mild-severe ID.

1.8.7 Summary

No new instruments, screening tests or batteries seem to have been created since the publication of Paiva et al. (2020). However, as this was not a systematic review, some literature may have been missed. Of the reviewed literature, many tests and batteries used to assess cognitive abilities were not adapted for PWID, or for the identification of dementia-related decline in PWID. More commonly, tests were established for neurotypical adults, or children, and applied to PWID. Many of the tests identified suffer from floor effects, which may render them insensitive to the cognitive profiles of PWID/DS.

Only one study collected participant feedback to inform acceptability and feasibility. Additionally, no studies used tests of olfactory function with a sample of PWID to examine normative or dementia-related decline. No test identified was normed across ID aetiologies and severities, and the majority were not normed with PWID. No research used any subtests of the BADS-ID to assess for dementia-related decline. The literature review highlights the need for a cognitive battery which is acceptable, accessible, and feasible to PWID, with robust assessments of EF and OA.

1.9 The Current Study

As evidenced, there is no current consensus on how to best assess for dementia, or measure dementia-related decline, with PWID. Many available instruments seem inadequately sensitive to the heterogeneous nature of IDs, showing floor effects when used with this community. Many lack appropriate, robust assessment of EF. This may indicate poor clinical utility as evidence suggests that executive dysfunction may be an early indicator of AD in PWID (particularly for PWDS). There seems to be no robust, norms-based cognitive test set designed for and acceptable to PWID which appropriately measures EF (Zeilinger et al., 2013). Though the BADS-ID is a promising assessment of EF, the sample excluded PWDS. Further, no research explored utility of the BADS-ID or its subtests in exploring dementia-related decline in PWID.

Further, no established battery utilised OA in tests, which has potential to be an accessible, non-invasive route to identifying early signs of dementia-related neurodegeneration. The literature review highlights the need for a novel cognitive battery which robustly assesses all domains, including executive function (EF), and which explores the utility of olfactory function assessment. This should include scalable test items to detect cognitive impairment, which are feasible, acceptable, and accessible for PWID. This battery may be revised and validated, contributing to research in providing early diagnosis, intervention and understanding of the different phenotypes of dementia in IDs. This is crucial to increasing quality of life and appropriateness of support for PWID, their families, and carers.

1.9.1 Aims

The need highlighted in the literature review informed the aims and objectives for the current study as follows:

- To develop a draft cognitive battery for use with PWID that is feasible and acceptable to this community.
    - For this battery to appropriately access all cognitive domains, including executive and olfactory functioning.
    - For the administration of this battery to be comprised of free and low-cost materials, for ease of distribution to low resource services.
- To use the understanding gained from the present study to inform future research and development of the draft battery.

1.9.2 Objectives

Related to the aims of exploring feasibility and acceptability of the battery, the following objectives were outlined:

- To investigate performance on the novel battery in adult participants with DS.
    - To use performance data to evaluate the feasibility of items within the battery by any floor and ceiling effects of the novel battery.
- To interview participants and gain feedback on:
    - The difficulty of the novel battery,
    - The appropriateness, acceptability, and suitability of the battery,
    - The perceived feelings of engagement with the battery,
- To further explore acceptability through researcher observation of engagement with the battery.

2 METHODS 1: TEST DEVELOPMENT

2.1 Epistemological and Ontological Stance

The philosophical stance of research is important to consider and comprises the epistemological and ontological perspectives. Epistemology is an area of philosophy which explores the nature and limits of how people acquire knowledge (Ferrier, 1854; Burr, 2003). Several epistemological stances attempt to understand how knowledge is ascertained, and to define the relationships between concepts related to this such as objectivity, subjectivity, and truth (Young, 2007; Willig, 2013). Ontology is a differing branch of philosophy concerned with the conceptualisation of reality, which explores the concepts of what is and what 'could be' (Smith, 2012). Researchers must have an explicit awareness of their epistemological and ontological stance, as this influences the methodological approach and subsequent data analysis (Barker & Pistrang, 2005). Though the areas of ontology and epistemology are rich with debate, a full exploration is beyond the scope of this thesis. However, an outline of the epistemological and ontological stance adopted by the researcher is discussed below.

Critical realism builds upon the ontological position of scientific realism, which assumes that the world is seen as real between independent observers, and therefore that phenomena can be measured in a standardised way (Burr, 2003). Critical realism assumes that knowledge itself is not objectively acquired, but that reality can be captured and understood through critical examination of observable phenomena (Bhaskar et al., 1998). For example, cognitive domains cannot be directly observed, but must be inferred. Critical realism allows for the effects of human error, bias, and subjectivity in attempting to quantify and understand constructs and their social contexts (Trochim & Donnelly, 2001). Much previous research focusing on disability tends to take either a social constructionist or a realist epistemological stance, with the former exploring how some facets of disability may be disabling by societal norms and narratives imposed by the neurotypical majority, and the latter seeking to understand the origins of disability through focusing on areas such as genetic and biomarker research (Bhaskar & Danermark, 2006; Burr, 2003).

Therefore, the current study takes a critical realist epistemological stance, assuming neuropsychological domains (e.g.: attention) may be defined through an interplay of behaviour and sociocultural contexts, which can be accessed, quantified, and measured. This approach acknowledges that although inferences about cognitive domains and performance can be obtained through behavioural and verbal responses, cognitive domains are constructs created by the consensus interpretations of instruments constructed by humans, and thus prone to error, bias, and subjectivity.

2.2 Overview

The current study aimed to develop a novel cognitive battery that is acceptable to PWID and shows feasibility by avoiding floor and ceiling effects.

The method involves two stages:

- The first describes the creation of novel task elements and the rationale behind task adaptations.
- The second describes the method used to capture feedback (and performance) on the battery tasks by participants.

The following section outlines the phases of battery development. As this is the first draft of the battery, created within the constraints of the professional doctoral thesis, it was decided that a small sample would be recruited. An exploratory method is used, testing the feasibility and acceptability of the battery with a narrow scope (Bowen et al., 2009). DS is the most common genetic condition with ID as a clinical feature, therefore, the decision was made to first pilot the battery with PWDS. Concurrently, research exploring the acceptability and feasibility of the novel battery with representatives from the wider ID communities is being undertaken within other doctoral thesis projects. It is anticipated that the development and pilot testing of this battery with different members of the ID community through these research projects will allow refinement and ‘beta’ testing with PWID of diverse aetiologies and severities and inform future objectives, such as validation of the battery.

In this chapter, I describe the development of this test set guided by stages three and four and five of 15 stages outlined by Fenn et al. (2020) namely:

- Test-format decision
- Item writing

Stages one and two (test-construction decision and investigation into concept) are fulfilled by discussions with experts in the field, submission of a research proposal, ethical approval, and the literature review of tests available for use in cognitive assessment with PWID, outlined in chapter one. Considering the scope of this thesis and the aim to create a pilot test set to explore feasibility and acceptability, the current study does not encapsulate all fifteen stages of development. Stage five (item review) is fulfilled using feedback from participants and researcher observation which is further detailed in chapter three.

Stages six and seven (preliminary data collection using draft test version and item analysis) are also initially explored in chapter three, and further explored in separate projects. Implications for the remaining stages are discussed at the end of this chapter. Battery development began with identifying cognitive domains of interest, including their receptive and expressive functions, and exploring which established tests are currently used for sensitivity to dementia-related decline in these areas. Following critical evaluation of the current literature and limitations of existing single-domain tests, brief instruments and comprehensive batteries used to screen for dementia in PWID, candidate tasks for the draft battery were established. This is outlined in table 2 at the end of this chapter.

2.3 Domains of Interest

A comprehensive (directly administered) neuropsychological test battery must include tests which sufficiently examine each cognitive domain and their functions. Receptive functions refer to how input is ‘taken in’ or ‘received’, and expressive functions relate to how this information is ‘acted upon’ or the ‘output’. A matrix of cognitive domains and their receptive and expressive processes is given in Table 1 below.

Table 1. *Cognitive Domains and Functions*

Domain		**Functions/ Test Focus**
Sensorimotor	Receptive	Basic sensation and perception
	Expressive	Basic movement (upper limbs)
Attention	Receptive	Orientation, short-term stores
	Expressive	Selective, sustained
Verbal-conceptual	Receptive	Comprehension of terms and syntax
	Expressive	Production of terms and syntax
Visuo-spatial	Receptive	Object perception, spatial perception
	Expressive	Construction, praxis
Executive Function	Receptive	Abstraction, induction
	Expressive	Task setting, task switching
Learning and Memory	Receptive	Learning, retention
	Expressive	Recall, recognition

Following the literature search (stage one: 'test-construction decision' and stage two: 'investigation into concept'), this subsection aligns with stages three and four of test development (Fenn et al., 2020), by drawing together existing materials, identifying adaptations if necessary, and creating novel stimuli and procedures for use in test sets which are suitable in complexity to the target population. Tests with normative structures (categories of performance, such as low, medium, high) are preferred for

inclusion over criterion tests with binary ‘pass/fail’ conditions, which may permit the collection of more discriminating data for the novel battery in future research (Urbina, 2004; Fenn et al., 2020).

As cognitive processes overlap (Kovacs & Conway, 2016; Burgoyne et al., 2022), this section presents tests as generally linked to the receptive and expressive functions of each domain. A brief outline of format and item description for each included test (Fenn et al., 2020) is provided. Tests that seem most acceptable, accessible and feasible to the ID community are identified, and their formats are used for adaptation into candidate tasks for the current study. Novel items are created in line with these test formats. This is to ensure that the novel battery can be widely available following further adaptations and revisions. Any adaptations made, or novel tasks created, are described. Due to many tests for PWID showing floor effects and/or inaccessibility to those with severe-ID or advanced dementia, several included tasks were adapted from widely-used tests used in the TD population. This chapter concludes with a summation of candidate item formats which were adapted and included in the battery for the current study, and implications for further stages of battery development.

2.3.1 Sensorimotor

Many cognitive batteries evidence the capacity to use limbs individually via other subtests. As such, many of the tests included in the draft battery (discussed in further subsections) provide evidence of limb use in participants. A simplified format of widely used assessment of basic motor function (e.g., EMAS) was included, utilising the eight most concrete items.

For motor sequencing, the ease of administration of Luria-style motor tasks may prove acceptable and accessible to PWID (providing motor skills are unimpaired). Therefore, an adapted Luria-style task (‘*Motor Programming*’) was included in the draft battery. This task includes four conditions: bimanual alternation, hand sequencing, knock-tap opposition (motor conflict) and knock-tap inhibition (go/no-

go). This task was simplified by including examiner modelling of the motor sequence at the beginning of each trial and simplifying verbal instructions to sequences given.

Tests of sensory perception (olfactory ability) have been created which may be sensitive to dementia-relate changes. However, research is scarce, concerns TD samples, and/or dated. To the best of my knowledge, there is no research which utilises OA tests to establish cognitive profile changes in PWID. Considering the potential of this approach as a measure of dementia-related decline (Tabert et al., 2005), and to be accessible to PWID (Manan & Yahya, 2021), a novel test of olfactory learning and memory (adapted from the UPSIT) was created by the researcher and their supervisor. This consists of low-cost and easily accessible materials. The olfactory learning task (*'Smell Recognition'*) consists of five target scents: mint, coffee, vanilla, shoe polish, and chocolate. Acceptable responses are provided for each scent (e.g.: for vanilla, sweets, ice-cream, chocolate, and almond are all accepted answers). Correctly naming the odour is not a key capacity, only discrimination between smells. These are created with solid materials in five separate jars with a lid and a single hole for participants to detect the scent, without seeing any substance.

2.3.2 Attention

2.3.2.1 Orientation and Short-Term Stores

No tests identified in the literature search were specifically created for PWID within this domain. The MMSE-O (Folstein et al., 1975) was identified as a brief and widely used test of receptive attention, though considering the floor effects identified for PWDS (Deb & Braganza, 1999), test items may not be feasible in their established iterations.

Therefore, the format of the MMSE-O was adapted to create the *Orientation* and *Information* sections. These simplify the wording of questions asked, and include 14 culturally unbound questions concerning person, time, place, and situation. This gathers further relevant background details and addresses the

examinee's general mental status. Aligned with the general approach to orientation in behavioural neurology, questions addressed awareness of personal information, time, place, and situation; along with information checks on sensory or motor impairments (e.g., 'Do you need glasses to read?') (Lezak et al., 2012). All questions of 'situation' could be modified according to client context (e.g., "How did you travel to get here today?" or "How are you feeling today?"). Questions tied to western culture (e.g.: who is the current prime minister?) were omitted. Questions to build rapport between the administrator and the participant were also included to reduce test-related anxiety (Thompson et al., 2018).

Though created for TD individuals, typical 'verbal repetition' format tasks may be a more ecologically valid and semantically structured method of accessing verbal short-term stores, in preference to other widely used measures (e.g., digit span forward; Weschler, 1986) which show floor effects for PWID (e.g., De Sola et al., 2015). Therefore, this format was adapted to create the '*Sentence Repetition'* task, with reduced syllable counts and simplified statements. This involves the examiner saying a declarative statement to the subject, which the subject must then repeat back precisely. The number of test items was reduced from 22 to 12, omitting the items which are the highest in scaled difficulty (i.e.: more statements and higher syllable counts) to reduce the potential of floor effects. Items increase in syllable and sentence length, to scale difficulty.

2.3.2.2 Selective and Sustained Attention

Cancellation format tasks (Leach, 2000), and adaptations (Krinsky-McHale et al., 2008) were identified as accessible and sensitive to functional decline. However, performance may be linked to knowledge of English reading and/or writing skills. Therefore, the format was adapted to become the '*Circle Search'* task. The target symbol was simplified and made culturally unbound (a circle) within a visual field that contains distinct distractor shapes (stars, triangles, and squares). The participant must 'strike out' all target shapes within 90 seconds. The start time, end time, 'hits' and false positives are recorded to create the task score. The shapes are larger, with

fewer distractors, to reduce the likelihood of floor effects. These adaptations were chosen to retain ease of administration whilst increasing acceptability to PWID.

Regarding sustained attention, instruments made for use with PWID seem sparse in the literature. Strengths in auditory sustained attention are seen for PWDS (Breckenridge et al., 2013), therefore including a test of this function may be beneficial for later validation of dementia-related decline. Therefore, an adaptation of signal detection format tasks (Leach, 2000), named '*Eight Detection'*, was created. This requires examinees to listen for a target number (eight) within a list of distractor numbers. Examinees must indicate the target number by tapping or other means (e.g.: speech, blinking). The examiner follows along and records 'hits' and false positives to produce a total score. Task duration was reduced to five minutes to reduce overall battery administration length, and reduce potential testing fatigue, while retaining validity.

2.3.3 Verbal-Conceptual

2.3.3.1 Comprehension

Evidence suggests that 'action on request' test formats of verbal comprehension traditionally used for aphasia and language impairment (BDAE, Goodglass & Kaplan, 1972) are most suitable for PWID, due to their brevity and lack of reliance on verbal expression. These were used in the CAMCOG-DS and TSI, though show floor effects with people with more severe ID and/or dementia. Therefore, an adaptation of this format felt appropriate to assess verbal-conceptual ability in the draft battery ('*Verbal Comprehension Part A & B'*). Adaptions included reducing items, simplifying instructions, and adding prompts to aid participant performance. Items which were the most complex (e.g.: items which required longer sequences, or those which had multimple conjunctions) were those chosen to be omitted, in an attempt to reduce any potential floor effects. Items include five actions on request ('*Part A'*) (e.g., "close your eyes and then open them") and 18 items ('*Part B*') requiring easily acquired apparatus (a coin, a set of keys, a watch, and a pen). These 18 items are split into three sections; items 1-7 are *'Pointing'* (e.g., "point to the watch"), 8-12 are '*Instructions*' (e.g., "touch the pen but not the watch") and 13-18 are *'Meanings'* (e.g.,

“which is like a clock?”). This aligns with Luria-style tasks of motor function used in neuropsychological assessment and allows the examiner to know whether to proceed with further items (Lezak et al., 2012).

2.3.3.2 Production of Terms and Syntax

Confrontation naming formats (e.g., BNT; Kaplan et al., 1976) seem acceptable and feasible to PWID, but stimuli may be culturally and experientially bound. Considering findings of Webb et al. (2020) concerning better performance and engagement when stimuli are ecologically valid for PWID, the ‘*Picture Naming’* test was made for inclusion in the current study. It includes novel real-life colour photographs of familiar culturally unbound stimuli (e.g., elbow, fire, the moon). These items were arranged in an order to reflect difficulty (i.e.: item one is ‘nose’, item 14 is ‘ostrich’). Participants are given a semantic cue if the item was obviously misperceived and prompts if they have named part of the picture correctly, but not the target identifier (e.g.: named ‘arm’ instead of ‘elbow).

A widely used assessment of verbal expression similar in format to that seen in the BDAE was added to analyse verbal output in responses to the ‘*Orientation A & B’* and ‘*Smell Detection’* subtests. This examined meaning, errors, words used, prosody, articulation, information and speech volume and rate. For example, a response of “Mum drove me here in the car” would generate an information unit point for ‘Mum’, ‘drove’, ‘me’, ‘here’ and ‘car’, respectively, as unique mentions of meaningful inference. Scoring involves the examiner rating responses to each domain from 0-2, where zero indicates extreme difficulty (e.g., <2 information units) and two indicates ‘typical’ performance (e.g., >6 information units).

2.3.4 Executive Function

EF is a component which draws on and coordinates many other cognitive domains (Lezak, 2012). Many EF tasks used with PWID in the literature showed floor effects, which may be unsurprising as deficits in EF are well-observed in PWID. As evidence

indicates a lack of robust EF assessment in cognitive batteries for PWID, several tests were considered for incorporation.

2.3.4.1 Abstraction and Induction

Suitable tests of verbal abstraction for PWID were sparse. Tasks such as Temporal Judgement showed limited validity and floor effects when used with PWID (Webb et al., 2020). Therefore, a reformatting of a widely used task of verbal abstraction for TD individuals, the Miller Analogies Test (MAT) as shown in the WRIT (Miller, 1960; Glutting et al., 2000) was constructed. The task ('*Verbal Reasoning*') includes simplified items and language, which give longer sentence prompts as guidance towards acceptable responses (Lezak, 1982) (e.g.: "A hat goes on the head, a shoe goes on the…" [acceptable response of 'foot' or 'feet']). These sentences scale in difficulty (e.g.: item nine reads "The moon is to the earth as the earth is to the…") Accepted responses score one point per item.

The TOL (Shallice, 1982) and TOL^{DX} (Culbertson & Zillmer, 2005) were identified as tests of non-verbal implicit EF, though the former was found to be related to verbal ability (Willner et al., 2010). Both revisions showed floor effects with increased ID severity and require manufactured materials. Therefore, a Matrices-like format task (Raven, 1995) was created ('*Visual Reasoning'*). This format is widely used to access receptive EF functions, with adaptations to such tasks being incorporated into dementia screening tools for the TD population. The 'rules' underpinning items in this new format were simplified; though item numbers were not reduced as a discontinuation rule was applied (after three consecutive scores of zero and/or if the person is demonstrably struggling). This may help to understand any floor or ceiling effects, while avoiding unneccessary continuation of the test if a participant is finding it too difficult. Discussions with researchers in the field highlighted the higher incidence of colour-blindness in PWID (Dwyer, 1991). Therefore, the colours of the stimuli were changed to a palette appropriate for those with colour blindness (colours on the opposite ends of the colour wheel spectrum), which may increase task validity.

2.3.4.2 Inhibition, Task Setting and Switching

Regarding inhibition, the CaD (Ball et al., 2008) showed no floor effects for PWDS, and no correlation with verbal ability (Willner et al., 2010). Evidence also suggests sensitivity to dementia-related decline (Bevins & Hurse, 2014), though may be too difficult for non-DS ID individuals (Cooper et al., 2016). Therefore, an adaptation of '*Cats and Dogs*' was included. To retain and improve upon ecological validity, realistic photos of dogs and cats on a white background were used. The images were all in the same colour (tan) and of the same size. The task involves a practice trial of eight photographs (not scored), which the participant names once congruently to check understanding, then once incongruently. The trial condition consists of 32 photos which are to be named incongruently. Scores are based on the number of correct responses.

Word generation format tasks, such as semantic fluency, are widely used to assess task setting in the TD population and may involve executive components of verbal output (Benton, 1968; Lezak, 2012). Adapted formats such as the CFT and M-CFT were commonly used with PWDS (e.g., De Sola et al., 2015; Ball et al., 2004) and seem easy to administer and sensitive to dementia-related changes across ID severities (e.g., Ball et al., 2004; Cooper et al., 2016; Fonseca et al., 2019a; Fonseca et al., 2019b). Therefore, a semantic fluency task was included in the novel battery ('*Word Generation'*). This task requires participants to generate as many animals (trial one) and foods (trial two) as possible, each within a one-minute time limit. The score is the number of items generated, excluding errors and repetitions.

Regarding planning and task switching, promising validity, acceptability and feasibility evidence was found for the 'Zoo Map' (shopping list) adaptation of the BADS-ID (Webb et al., 2020). However, no further research has utilised these tasks. Therefore, the '*Shopping Lists'* task was included in the final battery. The task format and stimuli were adapted to resemble a 'real-life' format to further increase ecological validity and acceptability (Burgess et al., 2006). The task consists of two different maps, each printed in colour on A4 paper. Map one depicts a 'supermarket'

with three aisles, an entrance, and a checkout. The aisles include real-life photo images of items that one may purchase at a supermarket, including: bread, broccoli, eggs, tomatoes, a toothbrush, toilet paper, oranges, bananas, strawberries, chocolate, apples, and carrots. The participant is asked to collect certain items from their 'shopping list' in any order they choose. The rules were that participants:

- Must begin at the 'entrance'
- Cannot use a path more than once
- Must finish at the 'checkout'.

Participants are scored two points per correct item acquired. One point is deducted each time a participant uses a path more than once or acquires an item not on their list. The second map is more complex, with six 'aisles'. Participants follow the same rules as before with the additional rule that they must also visit the 'shopping assistant' on the map once. Scoring is as seen for map one, with an additional two points given for visiting the shopping assistant once.

2.3.5 Visuo-Spatial Perception

Tests, such as Clock Copy (CAMCOG-DS; Ball et al., 2008) are shown to be accessible and sensitive to dementia-related decline in PWID but may rely on culturally bound experiential knowledge. Therefore, a simplified adaptation of a line judgement format task (e.g., Benton et al., 1978), '*Angle Judgement*', was created for inclusion, with fewer target lines and a reference key of five angle points. This provides a culturally unbound task which may increase accessibility to, and validity with, PWID.

2.3.5.1 Construction and Praxis

Regarding construction, block design (BD; Wechsler, 1981) tasks are widely used, though were not indicated as sensitive to dementia-related decline and showed floor effects with PWID (Alexander et al., 1997; Margallo-Lana et al., 2003). Therefore, a novel format was created for the current study: '*Matchstick Copy'*. This novel task has shown to be acceptable and feasible task of construction in a small sample of

TD older adults with limited writing ability (Jones Chesters, 2021). It involves arranging 12 matchsticks to recreate a design presented on A4 paper. The inclusion of low-cost, familiar materials may increase ecological validity. Delayed trials were also comprised and are discussed below. Scoring is as follows: One point is given per matchstick if placed in the correct orientation *or* place as in the presented design, and two points are given if the orientation *and* placement are both correct.

Concerning praxis, 'gesture to command' formats (e.g., Heliman et al., 1993; PCFT, Kay et al., 2003) are commonly used, and shown to have high reliability and validity with PWID. These tests also showed wide score ranges and minimal floor effects across cultures, suggesting good feasibility and acceptability (Margallo-Lana et al., 2003; DeVreese et al., 2021). However, adaptations may be necessary to replicate findings for people with severe ID. Therefore, the current study battery included an adapted task format (*'Praxis'*) examining gestures (intransitives); (e.g.: "show me how you would wave goodbye"), object use (transitives); (e.g.: "show me how you would use a comb to comb your hair") and buccofacial movements (oro-motor); (e.g.: "show me how you would lick your lips"). Adaptations included limiting the complexity and number of test items to pantomime of tool and task sequences and providing support/prompts if the person incorrectly mimicked the tool rather than use of the tool. Items which were included in the adaptation were those identified as well-rehearsed and familiar 'every day' motor sequences for PWID through discussions with key stakeholders as discussed previously.

2.3.6 Learning and Memory

2.3.6.1 Learning and Retention

Good tests of memory employ both visual and verbal tasks (BPS, 2015a). Evidence shows that list learning formats, such as the CRT-M (Devenny et al., 2002; Zimmerli & Devenny, 1995), show good sensitivity and accessibility, but only to those with mild-moderate ID. To decrease task demand and likelihood of floor effects, simplified word list learning tasks were created. These tasks ('*Word List Learning'* and '*Word List Immediate Recall'*) and their counterparts ('*Delayed Recall'* and

‘*Recognition*’, discussed below) were created with common, single-syllable tangible (physical in form) words. The item number was reduced to nine, to reduce the cognitive load in hopes of improving acceptability and accessibility to PWID.

2.3.6.2 Recall and Recognition

Recall and recognition tasks are typically paired within tasks given previously in cognitive assessments (Lezak et al., 2012). Regarding visual recall, the ‘*Matchsticks Copy’* task was accompanied by immediate and delayed recall conditions. This required the examinee to reproduce the graphic of a matchstick design presented to them earlier, using matchsticks, through free recall. Scoring was the same as for ‘*Matchstick Copy’* discussed earlier.

Items presented in ‘*Picture Naming’* were included as a confrontation visual recognition test (‘*Picture Recognition’*). Participants were presented with two colour photographs of the same stimuli (e.g.: two pictures of horses), with one picture having been one presented previously in ‘picture naming’. They were asked to choose the picture they saw earlier from two images. The pictures differ in small degrees, rather than with obvious distinctions (e.g.: both horses presented will have the same colour coat, but are oriented slightly differently). Motor responses are permitted (i.e.: the participant may point to the item or otherwise indicate their response).

Verbal recall and recognition are explored in the ‘*Word List Delayed Recall’* and ‘*Word List Recognition’* subtests. The former uses a free recall format of the nine words presented in ‘*Word List Learning’* after a 10–15-minute interval. The latter involves cued recall of this list, by reading through a word list and asking the participant whether each word was in the list presented earlier.

A novel format of olfactory recall was also created: ‘*Smell Recognition’*. In this task, participants are presented the five target smells presented in ‘*Smell Detection’*; and five new smells. After an interval, the participants are asked if they recognise each scent from those presented to them at the beginning of testing, and responses are

recorded (yes or no). Participants were not scored on their ability to 'correctly' label the scents, but rather on whether they accurately identified smelling this scent in the *'Smell Detection'* subtest (e.g.: if a participant incorrectly labelled a scent in *'Smell Detection'*, but then used the same incorrect label in *'Smell Recognition'*, this would still garner a point).

2.4 Draft Battery Composition

The current study is focused on test development and piloting, by identifying and operationalising psychological constructs of interest, to give rise to appropriate candidate tasks and items for PWID. These items are then explored for *feasibility* (whether the tests give a good range of scores, and whether they show floor or ceiling effects), and *acceptability* (whether participants find that the items and tasks are appropriate for their developmental and cultural needs) in a small sample of PWDS.

Outcomes will inform revision of the battery. All test items chosen for inclusion have potential for further normative structure and scaling to produce scoring criteria relative to categorisations of interest (e.g., ID severity), alongside employment of discontinuation rules. It is anticipated that tests which show poor performance will be omitted or replaced for further revisions. Item-level analysis of tasks included in the novel battery (chapter three) will inform removal of items of inappropriate difficulty. This shortened revision (and/or subsequent formats) may go on to be standardised, with normative data collection in a larger and more diverse sample of PWID to gather evidence which seeks to establish reliability and validity of the novel battery.

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, 1984)	Everyday household substances on cotton pads placed in jars
Motor, upper limb	Motor Function Part A & Part B	Edinburgh Motor Assessment Scales (EMAS; Bak et al., 2015)	Eight of the simplest items with accessible instructions
Attention - receptive	Orientation & Information	MMSE (Folstein et al., 1975) 'Orientation' task	Culturally-unbound questions, suited to contexts and simplified
	Sentence Repetition	Spreen & Strauss, 1998	Adapted using common single-syllable words in simple sentences
Attention - expressive	Eight Detection	KBNA Auditory Signal Detection Test (Leach, 2000)	Simplified shorter format using a friendly female voice and restricted range of stimuli (numbers)
	Circle Search	KBNA Symbol Cancellation Test (Leach, 2000)	Larger outline of basic shapes with a familiar target (circles) and fewer distractors
Executive - receptive	Verbal Reasoning	Traditional analogies-style task (Miller, 1960; WRIT, Pearson, 2020)	Concrete items using simplified language
	Visual Reasoning	Raven's style 'Matrix Reasoning' task (Raven, 1995)	Colour palette appropriate for people with colour blindness, simpler items
Executive - expressive	Word Generation	Typical format 'category fluency' tasks (Lezak et al., 2012)	Instructions simplified and prompts given to aid performance
	Cat-Dog Inhibition	CEFA (Ball et al., 2008) 'Cats and Dogs' task	Realistic pictures and uniform colours using shorter format
	Shopping List	'Zoo Map' task from BADS (Wilson et al., 1996) and BADS-ID (Webb et al., 2020) 'Shopping List' task	Novel format task to increase ecological validity, using realistic stimuli
	Motor Programming	Golden & Freshwater, 2001	Simpler instructions and modelling of the tasks in practice trials
Verbal comprehension	Verbal Comprehension A & B	BDAE (Goodglass & Kaplan, 1972)	Instructions simplified, fewer items, and prompts given to aid performance.
Verbal expression	Verbal Expression	BDAE (Goodglass & Kaplan, 1972)	Quality of speech output assessed by observation of previous test responses
	Picture Naming	BDAE (Goodglass & Kaplan, 1972)	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-spatial construction	Matchsticks Copy	Novel task	Novel task, using matchsticks to copy a model instead of drawing
	Praxis	Heilman & Rothi, 1993	Limited to pantomime of tool and task sequences with supportive instructions
Verbal Learning and Memory	Word List Learning Word List Immediate Recall Word List Delayed Recall Word List Recognition	Rey Auditory Verbal Learning Test and it's modified and simpler formats (Lezak et al., 2012)	Fewer words per trial and fewer trials, using common, concrete single-syllable words
Visual Learning and Memory	Matchsticks Immediate and Delayed Recall	Novel task	See above
	Picture Recognition	Wilson & Antablin (1980)	See above; paired two option forced-choice responses, to items previously seen, with motor responses permitted
Olfactory Learning and Memory	Smell Detection Recognition	See above	See above

3 METHODS 2: STUDY DESIGN AND PARTICIPANTS

3.1 Study Design

The current study adopts an exploratory research design to address acceptability, feasibility, and performance on the draft battery for PWID, by piloting this battery with PWDS. Quantitative data were collected through task scores and analysed through item-level analysis. Qualitative data were collected on the experiences of participants in completing the battery, including questions on difficulty and engagement with tests. Non-verbal indications of interest, difficulty or engagement that may inform acceptability were also recorded. For candidate items which do not receive specific feedback from participants, the guidelines for interpretation from the Mental State Examination (Voss & Das, 2023) and for communicating with PWID (Boardman et al., 2014) were followed to interpret participant response, and assess congruence between non-verbal behaviours and any verbal responses. This data was not formally analysed but is included to understand participant experience with the battery and used to understand floor and ceiling effects, and item order suitability. The results of the current study will be used to inform further development of the battery, for future research to consider piloting with larger and more diverse samples of PWID.

3.2 Ethical Considerations

3.2.1. Ethical Approval

This study was approved by:

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

Additionally, to increase public involvement in the process of the research, key stakeholders were consulted during stages one and two of test development (Fenn et al.,

2020) to understand their views and gather any potential revisions on the current study and proposed battery. This included the submission of a research proposal to the People's Committee at the University of East London, and discussions with the heads of affiliated NHS Adult ID services and charities before proceeding with recruitment. Virtual calls were also held with groups of potential participants before they decided whether they would like to be contacted to take part in the research, to hear an explanation of the study from the researcher and ask any questions they may have. A video with the researcher explaining the research aims, rationale and process in accesible language was also created and distributed to any potential participants.

3.2.2 Consent and Mental Capacity

Potential participants were individuals with ID from DS. Importantly, PWID may be socially naïve and vulnerable to coercion and/or suggestion and may change answers to provide one deemed socially desirable (Everington & Fulero, 1999; Khemka et al., 2009). Power relations between participant and researcher can also make it difficult for participants to communicate discomfort (Spears & Smith, 2001; Khemka et al., 2009). To lessen any impact of these factors, participants were approached for participation alongside their trusted parents, advocates, carers, or guardians. If all consented to receiving more information, prospective participants and their guardians were given an information sheet (Appendix H and I), an easy-read information sheet (Appendix J), and a .mp4 video file explaining the rationale and procedures of the study in jargon-free language, to facilitate informed consent.

Considering differences in processing speed for PWID, prospective participants were allowed at least seven days after receiving these materials to consider any potential benefits or risks to participation, and what taking part may involve, before being contacted. Participants were encouraged to bring any questions or concerns to the researcher before consenting. Adjustments were made for communication between the researcher and participants to increase independent responses if required. If prospective participants consented to involvement with the current study, they were asked to bring their trusted guardian with them to the meeting. No decisions regarding involvement with the study or

contact with the researcher occurred without this trusted person present to advocate for them if necessary.

3.2.3 Participant Wellbeing

Though the COVID-19 pandemic had reduced by the time testing occurred, measures were implemented to protect the physical wellbeing of participants, their guardians, and the researcher. Each testing room was a large, COVID-secure, well-ventilated room to maintain social distancing. The researcher wore full personal protective equipment, and each participant and carer wore personal protective equipment unless exempt. The researcher was fully vaccinated and regularly used lateral flow tests. Participants and their guardians were encouraged not to attend testing sessions if they tested positive within a week of the meeting date or were experiencing any symptoms of COVID-19. A risk assessment for this was undertaken and approved by the School of Psychology at UEL.

To counteract potential testing fatigue, participants were offered unlimited breaks, and reminded they could take these at any time throughout the meeting. Refreshments were also provided. Importantly, participants may have felt anxiety whilst completing the testing session for several reasons (Bennett-Levy et al., 1994). Participants and their guardians were informed before participation that the meeting outcome was not diagnostic, which may have lessened any anxiety. However, participants may have been negatively emotionally impacted if they perceived their performance to be poor. Words of encouragement were incorporated into the examiner's manual of the novel battery to counter this, and to focus on test experience rather than performance. The debrief and questions given at the end of testing surrounding participant opinions on task difficulty may also have helped to ease these uncomfortable feelings, by emphasising that this is a draft battery to be shaped in future by their feedback.

3.2.4 Data Protection and Confidentiality

Participants were allocated a numerical code (e.g.: 001) to keep data unidentifiable. This was kept in a spreadsheet separate to identifiable information which was collected from

participants, for the purposes of contacting participants to arrange testing sessions. Participant performance was video-recorded and stored as .mp4 files, to ensure accurate scoring and interpretations of test accessibility after the meeting. Data collection and management was undertaken in accordance with the Data Protection Act (1988); a data protection plan was completed and approved in line with UEL data management guidelines (see Appendix K).

3.3 Participants

3.3.1 Recruitment

A study poster (Appendix L) was created and sent to the psychologist contact within a London NHS Trust. This poster was circulated in ID services within the trust. The psychologist agreed to identify eligible prospective participants, and their guardians, to gauge interest. This was also circulated to the managers of a Hampshire ID charities, who spoke with potential participants and their carers to ascertain interest and eligibility. All participants involved were recruited either through identification by the psychologist or manager, or by responding to the study poster.

3.3.2 Sample

This research adopts an exploratory feasibility design and does not require an *a-priori* sample size calculation based on statistical power. Feasibility and acceptability studies work best with a narrow scope (Bowen et al., 2009), therefore this study aimed to recruit 5-8 PWDS (a sample of people with non-DS ID were also recruited as part of a separate related study). Participants were identified using convenience (non-probability) sampling (Ireland et al., 2005). This sampling method has limitations for valid applicability of data to the general population (Etikan et al., 2016), and may introduce response bias considering the type of individuals and/or carers who may be more likely to participate in research. However, this sampling method also has high practicality and ease of use. Considering this research was exploratory and conducted within a limited timeframe, this sampling method was deemed appropriate for the aims of the current study. 15 people were

contacted to request their involvement in the study, of which five consented to having their contact details shared with the research team. One person was excluded at the meeting due to concerns around capacity and ineligibility. Four took part in the study between June and September 2023. All had mild ID, lived either independently or semi-independently, and attended the meeting with a trusted carer or guardian. Difficulties in recruiting the anticipated sample are discussed later. The final sample (*n*=4) was majority female, with an average age of 38. See table three 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	40	White British	Right	13	Yes	No
P2	M	44	White British	Right	11	No	No
P3	F	32	White British	Left	14	Yes	No
P4	F	36	White British	Right	15	Yes	No

3.3.3 Inclusion and Exclusion Criteria

To meet the aims and objectives of the current study, some limitations were made to recruitment criteria. The age range was limited to 30-55 years, to be representative of the earlier age of dementia onset in PWDS, and therefore likely age of routine cognitive assessment (Lott & Head, 2017). Participants must also have sufficient verbal/motor ability to ensure capacity to consent and have been actively under the care of the affiliated NHS trust, or in regular attendance of the affiliated charities.

This research aims to pilot the draft battery on the 'typical' DS population. Therefore, participants who have a current severe/enduring mental illness, known/suspected dementia, substance misuse and/or neurological injury/trauma were excluded. This was done to avoid confounding impacts on initial test outcomes (Lezak et al., 2012). Good understanding and fluency in English was also required, as many established tests and cognitive batteries also require this (Lezak et al., 2012). Limited/no verbal production ability was not an exclusion criterion.

3.3.3.1 Inclusion Criteria

- Aged between 30-55 years
- DS
- Currently under the care of the NHS-ID affiliated services or in regular attendance of activities run by the affiliated charities.
- Good understanding of, and communication in, English
- Capacity to consent through speech or other means

3.3.3.2 Exclusion Criteria

- History of and/or current illicit substance misuse in the last 6 months
- Known diagnosis of, or suspected, dementia
- Experiencing a current severe/enduring mental illness
- Have a neurological injury/ experienced neurological trauma

3.4 Procedure

3.4.1 Consent

Individuals interested in participating were contacted via phone or email, where the researcher would introduce themselves and encourage prospective participants and

guardians to raise any questions they may have around the research. They were then given a choice of dates and times to attend the NHS-ID service or charity to participate in the meeting.

On the day of the meeting, participants were assessed for their ability to consent autonomously. As research alongside PWID requires sensitivity to informed consent, this was guided by the considerations outlined by the Mental Capacity Act (2005), namely: whether a person can understand, retain, and weigh-up information to communicate a decision. If the participant was felt to be unable to do any of these processes, the session was terminated, and the participant and their guardian were thanked for their time. This was to avoid unethical inclusion of the participant in the study and avoid compromising the participant's ability to engage meaningfully with the test set. If the participant did show capacity to consent, the researcher discussed both the information sheet and the easy-read information sheet with the participant in the presence of their guardian. Participants and guardians were given another opportunity to ask questions before deciding whether to take part. If the participant consented, they were asked to sign the consent form (Appendix M), and their guardian was asked to sign the guardian consent form (Appendix N). Participants were thanked with a £10 voucher, regardless of whether the testing session was completed and given a debrief and easy-read debrief letter after the meeting (Appendix O and P). One participant declined the voucher, stating they "do not need it". This request was respected, as it was declined in front of their parent, who agreed with their refusal.

3.4.2 Testing Set-Up

During testing, the researcher sat across from the participant on a table two meters apart in line with COVID-19 measures. The guardian sat behind the participant, out of their view, so the participant would not be distracted. For each test, the researcher either read the questions aloud and recorded the response or showed the participant a stimulus from the stimuli book.

3.4.3 Demographic Information

Information was collected on participant age in years, primary language, other spoken languages, handedness, ethnicity, years of education acquired, and identified gender. This was gathered to understand what (if any) affect this information may have on generalisability (Lezak et al., 2012).

3.4.4 Test Administration Order

The battery begins with simple tests of language comprehension and expression, upper limb movements, and olfaction. This is followed by verbal learning immediate and delayed trials, with visual tests in the delay interval. Finally, visual learning and memory tests were presented, with mainly visual tests in the interval. The decision was made not to present the manual, record form and stimuli, as they may be vulnerable to use outside of this thesis when not adequately validated. Test administration order is as follows:

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

If participants were unable to complete a test for any reason (e.g.: because the test was too difficult for them, or because the test required motor function that the participant could not perform), this test was discontinued and the researcher moved to the next test. This was done to reduce participant distress.

3.4.5 Qualitative Feedback Interview

Participants were asked questions from a semi-structured interview schedule regarding their experience of the test battery. This was done either straight after the meeting, or at a later date if preferred. The semi-structured interview schedule is seen in Appendix Q.

3.5 Analysis

3.5.1 Quantitative

Quantitative data was analysed using IBS SPSS Statistics for Windows (version 29). Descriptive statistics, central tendency and dispersion were gathered to inform item level analysis, to understand the quality of each test item. Items were assessed for their difficulty level by calculating the percentage of the sample who got the item correct

(percentage passing, or ‘Pass’ Value) (Urbina, 2004). This was to understand if items were scaling appropriately in difficulty for this population. Reliability was explored narrowly through measures of internal consistency. Discriminative power could not be explored, as no comparative sample of individuals with dementia was recruited. However, study data will contribute towards future research which may seek to establish preliminary norm and discriminative data.

3.5.2 Qualitative

Verbal responses to tests and feedback from the post-test interview were recorded verbatim through writing as they responded. These were then transcribed onto the encrypted excel spreadsheet alongside their anonymous numerical code identifiers. As mentioned previously, qualitative feedback data on the accessibility and difficulty of the tests was collected for tool refinement and was not formally analysed. Objective descriptions of how accessible and engaging the participants found the test set through researcher observation of participant testing sessions and video recordings were also gathered.

4 RESULTS

4.1 Acceptability

4.1.1 Participant Feedback

After completion of the pilot battery, participants engaged in a semi-structured qualitative interview regarding their experience. The feedback given from participants to these questions was limited, which may have implications for the questions used to gather such feedback, and when these questions are asked, in future research.

Q1. Did you find any of the tests interesting?

Three participants responded “yes” to this question. One participant cited ‘Matchsticks Memory’ as interesting, as they enjoyed trying to put together the design from memory. Another commented that ‘Picture Naming’ was most interesting, enjoying the variety of pictures given. ‘Smell Detection’ and ‘Smell Recognition’ were also mentioned as interesting (“I have never done something like that before”).

One person responded that they did not find any of the tests interesting but did not specify reasons why (“they just were”).

Q2. Did you find any of the tests boring?

Three participants answered “no” to this question, with one adding: “it was really fun!”.

One participant indicated “yes” to this question, citing the ‘Smell Detection’ and ‘Smell Recognition’. When asked, the participant did not elaborate on what could be done to make these more engaging.

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

One participant answered “no” to this question.

One participant found that ‘Matchsticks Memory’ and “remembering the words” (referring to ‘Word List’ Memory’) were tasks they found too easy. This participant suggested we use harder words in the list. Another participant named ‘Shopping List’ as a test they found too easy, saying the map was “easy to follow”. They suggested changing the instructions for the shopping list, as there were “too many words”. ‘Cats and Dogs’ and ‘Picture

Recognition' were 'too easy' for two participants, with one suggesting we make all pictures, particularly the 'butterfly' pair, harder.

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

Only one participant answered "yes" to this question and indicated that the 'Motor Programming' test was "too confusing". They suggested we include easier movements and learn the parts of the sequence one at a time.

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

Two participants indicated they enjoyed the tests, with both describing their experience as "fun". One participant commented "don't change it, I like it". Two participants commented that the test was "too long" and should be made shorter. One participant shared that they felt this research was important and were happy to be taking part in it. Feedback was also gathered on the way task information is presented, with one person suggesting that task information should be aligned to the left and highlighted in subtests that required reading (e.g.: 'Shopping Task'). This format was stated as more familiar, and easier to read, for PWID and is an important recommendation for future revisions.

4.1.2 Researcher Observations

The battery took one to two hours to administer in full, depending on the speed in which participants were able to proceed with tasks, or whether asks were discontinued based on the ability of participants to engage or complete them. All participants who completed the battery were able to attempt all subtests. Though test length seemed to be tolerated well by most participants, administration time is too long and requires reduction to avoid testing fatigue.

As mentioned, one participant did not complete the battery and did not wish to return to complete the remaining portion. This participant seemed to find the 'Motor Programming' subtest difficult after item one and could not execute the sequences to imitation. However, they successfully gesticulated all 'Praxis' items to command. It is therefore unknown

whether this indicated difficulties in motor sequencing, motor inhibition, or a misunderstanding of task instructions. After completing ‘Smell Recognition’, they appeared fatigued. Though breaks were offered throughout, these were not taken up. The participant was asked whether they would like to stop testing, which they confirmed. It may have been that the battery was not engaging enough to motivate them to continue. This may be supported by the fact that they consented to staying and providing feedback about the battery after the tests were abandoned. Up until ‘Smell Recognition’, the participant attempted all tests, scored well, and showed signs of non-verbal enjoyment (e.g.: smiling) and verbal humour (e.g., responding: “Woah! That’s strong!” to an item from ‘Smell Recognition’) throughout the meeting.

The other three participants were able to attempt all tasks in the battery and showed similar non-verbal (e.g., laughing) and verbal (e.g., “that was fun”) signs of engagement and enjoyment throughout. Laughter was noted most often from participants in response to the ‘Praxis’ task item of gesturing *‘show me how you would threaten me with your fist’*.

One participant seemed to find the ‘Matchsticks’ subtests difficult, being unable to create the design from the stimulus in the ‘copy’ trial and finding it hard to pick up and move the matchsticks; instead opting to push them into place with their finger. This may have been due to a sight difficulty, impairment in visual-perceptual ability, or that the matchsticks were too small. Another participant was observed moving their face close to the page in ‘Angle Judgement’ and commented that the images were ‘very small’.

‘Shopping Lists’ and ‘Visual Reasoning’ were discontinued for two participants, as they had demonstrable difficulty with these tasks (or in comprehension of task instructions) or scored three consecutive zeros. This contrasts with the feedback of one participant who indicated that ‘Shopping Lists’ was too easy. This may indicate either a misunderstanding of task instruction and/or social desirability in giving feedback. Only one participant was able to collect only the correct items, and all participants used a path more than once, suggesting this task may need revision to be appropriate for PWDS.

4.2 Feasibility

4.2.1 Test Performance

Tables 4-6 provide descriptive data for test performance per domain. Missing data due to termination of the session or subtest discontinuation (due to demonstrable participant difficulty with the task, as discussed above) are indicated, alongside any reduction in data sample size. Time for the 'Cats and Dogs' task was only recorded for the incongruent trial, and so between-trial comparisons could not be made.

Planned analysis included exploratory analysis of data through central tendency, dispersion, and skewness. However, these measures require at least five data sets (Nuzzo, 2016), therefore data was unable to be analysed as planned. Due to the small sample, results are provisional and require extensive replication. Cautious interpretations of this data are therefore made based on item-level analysis, participant feedback and researcher observations. These are used to consider modifications to the next draft of the battery, including removal or substitution of subtests.

Two participants scored the maximum on 'Orientation Subtotal A', 'Verbal Comprehension A', 'Motor Function A' and 'Picture Recognition' which may indicate ceiling effects, though these are 'easy' tests to complete, with ceiling effects also seen in the TD population. On 'Visual Reasoning', two participants scored zero, and another scored just two, which may indicate a floor effect requiring further analysis. Scores were generally high, with a narrow range, on 'Orientation Subtotal A', 'Orientation Total', 'Verbal Expression', 'Motor Function A' and 'Praxis'. 'Word List Recognition' and 'Circle Search' (n= 3), scores were also generally high. These findings are similar to those seen in TD people without dementia, with intact verbal and motor ability.

Table 4. *Descriptive Data for Performance by Subtest – Verbal, Visual, Motor and Olfactory Functions*

Verbal, Visual, Motor and Olfactory Functions							
Subtest	***n***	**Maximum Score**	**Range (min-max)**	***n* Minimum Score**	***n* Maximum Score**	**Mean**	***SD***
Orientation Subtotal A	4	12	9-12	0	2	10.75	1.50
Orientation Subtotal B	4	4	2-4	0	1	2.75	0.96
Orientation Total (A+B)	4	16	11-16	0	1	13.50	2.28
Smell Detection	4	5	2-3	0	0	2.25	0.50
Smell Recognition	4	10	5-8	0	0	6.25	1.50
Verbal Expression	4	20	14-20	0	1	16.75	2.50
Verbal Comprehension A	4	5	3-5	0	2	4.25	0.96
Verbal Comprehension B	4	18	12-18	0	1	15.25	2.36
Verbal Comprehension Total (A+B)	4	23	16-23	0	1	19.50	3.00
Motor Function Subtotal A	4	5	4-5	0	2	4.50	0.58
Motor Function Subtotal B	4	12	9-10	0	0	10.25	1.26
Motor Function Total (A+B)	4	17	13-15	0	0	14.75	1.71
Matchsticks Copy	3	24	1-23	0	0	13.33	11.24

Table 5. *Descriptive Data for Performance by Subtest – Verbal and Visual Attention and Executive Functions*

Verbal and Visual Attention and Executive Functions							
Subtest	***n***	**Maximum Score**	**Range (min-max)**	***n* Minimum Score**	***n* Maximum Score**	**Mean**	**(*SD*)**
Circle Search	3	26	23-26	0	1	24.67	1.53
Angle Judgement	3	20	10-19	0	0	13.00	5.20
Visual Reasoning	3	10	0-2	2	0	0.67	1.20
Verbal Reasoning	3	12	6-9	0	0	8.00	1.73
Shopping List Map 1	3	20	2-14	0	0	8.00	6.00
Shopping List Total	3	44	8-28	1	0	18.00	14.14
Cat-Dog Inhibition	3	32	17-31	0	0	26.00	7.81
Cat-Dog Inhibition Time (seconds)	3	N/A	40-125	N/A	N/A	86.00	43.02
Eight Detection	3	14	12	0	1	12.67	1.15
Sentence Repetition	3	12	2-8	0	0	5.00	3.00
Motor Programming	4	12	1-6	1	0	5.00	3.92
Praxis	4	30	27-30	0	1	28.5	1.29

Table 6. *Descriptive Data for Performance by Subtest – Verbal and Visual Learning and Memory*

Verbal and Visual Learning and Memory							
Subtest	***n***	**Maximum Score**	**Range (min-max)**	***n* Minimum Score**	***n* Maximum Score**	**Mean**	**(*SD*)**
Word List Immediate	3	36	13-30	0	0	22.67	8.74
Word List Learning	3	9	3	0	0	3.00	0.00
Word List Delayed Recall	3	9	3-8	0	0	5.33	2.52
Word List Recognition	3	18	14-18	0	1	16.67	2.31
Matchsticks Immediate	3	24	0-21	1	0	9.67	10.60
Matchsticks Delayed Recall	3	24	4-20	0	0	11.33	8.09
Picture Naming	3	16	13-14	0	0	13.67	0.58
Picture Recognition	3	16	23-26	0	2	12.67	2.89
Word Generation	3	NA	17-48	0	N/A	29.33	16.44

4.3 Item-Level Analysis

The utility of battery items were analysed narrowly through item-level analysis. Aligning with Urbina (2004), items are analysed for difficulty by reviewing scores, and indicating the proportion of participants who were able to answer the item correctly. This was done to inform scaling of tests in future battery revisions. For norm-referenced tests, the ideal item difficulty is between 0.4 and 0.6. (Urbina, 2004). An item which all participants passed would result in a difficulty index of 1. This may indicate feasibility for early subtest items, or ceiling effects for later scalable test items. A suitably scaled subtest may be expected to begin with a lower difficulty index score for its earlier items (0.6 or higher), scores of 0.4-0.6 for the mid-test items, and higher difficulty (0.39 or lower) for the later and final items. This is shown in the table below as the 'pass value' (PV), where items of lower difficulty are highlighted in green, those between 0.4 and 0.6 are highlighted in orange, and those of higher difficulty highlighted in red. This is shown in table seven below.

Importantly, this difficulty index is only applicable to subtest items which produce a binary (incorrect/correct; yes/no), answer. Scores are generally reflective of the acceptability feedback and researcher observations (presented previously). Some subtests (e.g., Smell Detection) were not designed to be scaled, and so differences in item difficulty index scores were not expected. As previously stated, one participant discontinued the test after 'Smell Recognition'. All subsequent item analysis is done with the other three participant data sets.

Tasks with more complex instructions proved more difficult for participants, with 'Shopping List Map Two' being discontinued for two participants due to demonstrable difficulties in understanding task instructions. This may have been due to familiarity with other 'map-like' tasks with fewer 'rules' or related to feedback regarding how the 'shopping list' was presented to participants in text format.

Scores were generally low for 'Smell Detection', with no participant scoring at maximum. One participant initially identified many scents as 'cream'. This may have been because smells were in jars that may resemble skincare products. In subsequent revisions, semantic relationships to containers in which smells are presented should be considered. No participants were able to name item four (shoe

polish) and did not seem to recognise the scent (answers given included: ‘perfume’, ‘sugar’ and ‘burnt’), suggesting that answers given were ‘best guesses’. This may suggest that scents such as shoe polish require specific prior cohort and/or cultural experiences or knowledge. This may be supported by findings that item one (mint) was successfully identified by all participants (i.e.: presumably, all participants have prior experience of teeth-brushing, but not of polishing shoes). A similar explanation may underlie the finding that no participants could name item 14 (ostrich) of the ‘Picture Naming’ subtest. The final item (butterfly) was named correctly by all participants, suggesting that shifting the test items may reflect a more suitable difficulty scaling.

In ‘Smell Recognition’, two participants correctly identified being presented the ‘shoe polish’ odour previously, which may mean lack of previous experience with an odour does not affect performance on this subtest. However, both participants also scored 5/10 in this subtest, suggesting that correct answers were possibly given by chance. This may be due to acquiescence bias, as both participants achieved maximum score on another binary format subtest (‘Word Recognition’). Therefore, this may be better attributed to either pre-existing olfactory sensitivity, misunderstanding of task instructions, poor differential quality of target odours, or lack of prior experience with certain odours.

All participants scored highly on ‘Praxis’, which may be due to the ‘everyday’ familiarity of motor sequences in items. Alternatively, these tasks may have been felt to be more ‘engaging’ by participants, corresponding to researcher observations of perceived fun and enjoyment. This may have increased researcher-participant rapport. The exception to this was item eight (“Show me how you would use scissors to cut through paper”), where most participants would pantomime the action of the scissors themselves rather than of holding and ‘squeezing’ of scissors, despite prompts and imitation of the sequence by the researcher. This may be due to the ‘Makaton’ sign for scissors being an imitation of the tool, rather than the action, which may be a rehearsed and familiar motor sequence to PWID that was difficult to inhibit.

Notably, scoring for ‘Orientation Subtotal B’ may be too restrictive for PWDS. Item two requires asking participants about their current situation (e.g.: “how did you travel to get here today?” or “how are you feeling today?”), which often garnered correct one-word responses (e.g.: “bus” or “good”). To score well or at maximum, the examiners manual states that participants must *“give more than one, well-oriented, complete, and correct response for each question”*. Though not indicated in the manual, prompts were given in response to encourage participants to give more information (e.g.: “where did you get the bus from?”), which often increased scores. Initial responses will reflect verbal expressive ability rather than orientation; the addition of prompts is an important recommendation for future revisions of this subtest.

Table 7. *Item Difficulty Levels for All Scalable Test Items*

Motor & Language Functions												
Orientation Subtotal A												
Item #	1	2	3	4	5	6	7	8	9	10	11	12
Total Score	3	4	4	4	3	4	4	3	4	3	4	3
PV	0.75	1.00	1.00	1.00	0.75	1.00	1.00	0.75	1.00	0.75	1.00	0.75
Orientation Subtotal B												
Item #	1	2										
Total Score	7	5										
Smell Detection												
Item #	1	2	3	4	5							
Total Score	3	2	1	0	3							
PV	0.75	0.5	0.25	0.00	0.75							
Verbal Comprehension Part A												
Item #	1	2	3	4	5							
Total Score	3	4	4	2	4							
PV	0.75	1.00	1.00	0.5	1.00							
Motor Function Part A												
Item #	1	2	3	4	5							
Total Score	4	4	4	2	4							

PV	1.00	1.00	1.00	0.5	1.00													
Motor Function Part B																		
Item #	1	2	3	4														
Total Score	11	10	10	10														
Motor Programming																		
Item #	1	2	3	4														
Total Score	6	3	7	3														
Praxis																		
Item #	1	2	3	4	5	6	7	8	9	10	11	12	13	14	15			
Total Score	8	8	8	8	7	8	8	4	8	8	8	8	8	8	8			
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	4	4	4	3	1	2	3	4	4	4	2	3	4	4	4	4	4	3
PV	1.00	1.00	1.00	0.75	0.25	0.5	0.75	1.00	1.00	1.00	0.5	0.75	1.00	1.00	1.00	1.00	1.00	0.75
Smell Recognition																		
Item #	1	2	3	4	5	6	7	8	9	10								
Total Score	4	1	2	3	1	4	0	4	4	2								
'PV	1.00	0.25	0.5	0.75	0.25	1.00	0.00	1.00	1.00	0.5								
Verbal Learning and Visual Functions																		
*Angle Judgement**																		

Item #	1	2	3	4	5	6	7	8	9	10
Total Score	4	6	5	5	3	2	4	4	4	5

Word List Recognition*																		
Item #	1	2	3	4	5	6	7	8	9	10	11	12	13	14	15	16	17	18
Total Score	2	3	3	2	3	2	3	3	3	3	2	3	3	3	3	3	3	3

Matchstick Copy*												
Item #	1	2	3	4	5	6	7	8	9	10	11	12
Total Score	4	3	3	4	4	4	4	2	2	2	4	3

Matchstick Learning*												
Item #	1	2	3	4	5	6	7	8	9	10	11	12
Total Score	3	3	3	3	3	3	4	1	1	1	2	2

Picture Naming*																
Item #	1	2	3	4	5	6	7	8	9	10	11	12	13	14	15	16
Total Score	3	2	3	3	2	3	3	3	2	2	3	3	3	0	3	3
PV	1.00	0.66	1.00	1.00	0.66	1.00	1.00	1.00	0.66	0.66	1.00	1.00	1.00	0.00	1.00	1.00

Visual Learning and Verbal Functions												
*Sentence Repetition**												
Item #	1	2	3	4	5	6	7	8	9	10	11	12
Total Score	3	3	2	1	1	2	0	0	1	0	0	0

PV	1.00	1.00	0.66	0.33	0.33	0.66	0.00	0.00	0.33	0.00	0.00	0.00				
*Verbal Reasoning**																
Item #	1	2	3	4	5	6	7	8	9	10	11	12				
Total Score	3	3	3	2	1	2	2	3	1	1	2	1				
PV	1.00	1.00	1.00	0.66	0.33	0.66	0.66	1.00	0.33	0.33	0.66	0.33				
Picture Recognition*																
Item #	1	2	3	4	5	6	7	8	9	10	11	12	13	14	15	16
Total Score	2	3	1	2	2	3	3	2	3	3	2	3	3	1	2	3
PV	0.66	1.00	0.33	0.66	0.66	1.00	1.00	0.66	1.00	1.00	0.66	1.00	1.00	0.33	0.66	1.00

** indicates subtest item data presented with discontinued participant removed (n=3). Total Score= total score of all participants combined. PV= percentage of participants passing (only provided for items with binary correct or incorrect answers). Item Analysis Key: Green= item difficulty index <0.4; Orange= item difficulty index 0.4-0.6; Red= >0.6.*

5 DISCUSSION

5.1 Overview and Summary of Results

A literature search highlighted the need for a novel cognitive battery which robustly assesses all domains, including executive function (EF), and which explores the utility of olfactory function assessment. This informed the aims of the current study:

- To develop a draft cognitive battery for use with PWID that is feasible and acceptable to this community.
    - For this battery to appropriately access all cognitive domains, including executive and olfactory functioning.
    - For the administration of this battery to be comprised of free and low-cost materials, for ease of distribution to low resource services.
- To use the understanding gained from the present study to inform future research and development of the draft battery.

Psychometric and acceptability findings of available tests were critically reviewed, and tests with few floor effects and suggested acceptability for PWID were considered for inclusion. Word generation, Luria-style motor tasks and Stroop-like formats were implemented with ease into the battery. Matrix reasoning, signal detection and line orientation formats required modification to simplify task items. Novel formats were created for the task of planning and task setting and switching, a visual task of learning and memory, and for odour detection and delayed recognition. Through this process, a draft battery was successfully created.

Using an exploratory method, the current study addressed the following objectives based on exploring the feasibility, acceptability, and accessibility of the draft battery:

- To investigate performance of PWDS on the novel battery.
    - To use preliminary performance data to evaluate the feasibility of items within the battery by any floor and ceiling effects of the novel battery.
- To interview participants and gain feedback on the following:
    - The difficulty of the novel battery,
    - The appropriateness, acceptability, and suitability of the battery,
    - The perceived feelings of engagement with the battery.

- To further explore acceptability through researcher observation of engagement with the battery.

This is the first draft of the battery, and therefore includes a wide range of candidate tasks and items which will be refined in future revisions. Generally, feedback indicates that all subtests were well-received, and all could be attempted, indicating test instructions could largely be comprehended and executed. However, administration of the battery took 1-2 hours, indicating most subtests must be shortened in further revisions to reduce likelihood of testing fatigue. The matrix reasoning format measure of visual abstraction did not prove feasible for participants. The Luria-style task of proxy executive functioning, and the adapted BADS-ID format of planning, task setting and switching, were also challenging for participants. As these are all tasks of EF, findings may support evidence of pre-existing EF impairment in PWDS which are more susceptible to earlier and more rapid neurodegeneration (Cooper & Prasher, 1998; Adams & Oliver, 2010; Dekker et al., 2015). Tentatively, this may also support theories that executive dysfunction occurs as an earlier symptom of AD in PWDS than seen in the TD population (Lautarescu et al., 2017).

Verbal feedback given for tests was limited, which may reflect inappropriate interview questions, difficulties in verbal expression seen in PWDS, and/or effects of social desirability bias (Everington & Fulero, 1999; Khemka et al., 2009; Lott & Dierssen, 2010; Grieco et al., 2015; Fernández-Alcaraz & Carvajal, 2020). Alternatively, this may be associated with timing of questions asked, as all participants opted to complete the interview immediately after completing the battery, when they may have been fatigued.

5.2 Feedback and Subtest Development

Findings for the acceptability and feasibility of each subtest from feedback and item-level analysis (Urbina et al., 2004) are discussed below. Implications for stages six and seven (preliminary data collection using draft test version and item analysis) and suggested revisions to subtests to inform a second draft (as per phase eight-fifteen of test development outlined by Fenn et al., 2020) are also given.

5.2.1 Motor and Language Functions

5.2.1.1 Orientation A & B

This included questions of orientation to time, place, person, and situation. No specific feedback was given for these subtests. Scores were generally high for 'Orientation Part A', as expected for PWDS with mild ID and without dementia. 'Orientation Part B' scores were similarly high, though additional prompts were needed to ensure responses were rich enough to reflect orientation alongside verbal expressive ability. This indicates tentative feasibility and acceptability of this format for PWDS. Adaptations should be made to scoring criteria and examiner instructions (e.g., *If a one word answer such as "bus" is given, ask a follow-up question related to the answer such as "where did you get the bus from?")* in future revisions, to increase validity.

5.2.1.2 Smell Detection & Recognition

These subtests involved smelling five odours and naming them. Participants all reported (and were observed) enjoying this subtest, and one named it as interesting in the semi-structured interview. All could complete these subtests, suggesting good acceptability. However, scores were generally low in the 'detection' subtest, which might reflect pre-existing olfactory impairments in PWDS, which exacerbate with age (Nijjar & Murphy, 2002; Bianchi et al., 2014; Bontempi et al., 2020; Manan & Yahya, 2021). However, scores may also be linked to familiarity with target scents. While nobody could answer item four (shoe polish) correctly, item five (chocolate) and one (mint) were the most reliably identified scents. Chocolate and mint may have been identified as forms of these scents are included in everyday items (e.g., toothpaste), and previous experience with these scents may have increased their salience and recognisability. However, it may be that correct naming is not necessary for this test, only that participants generate a consistent label for each item.

The recognition subtest involved indicating whether odours were presented earlier in '*Smell Detection'*. Though discrimination of smells rather than identification was the key capacity in this subtest, recognition scores were poor. All participants incorrectly identified item seven (cinnamon) as being presented previously, which may indicate it as too similar to a target smell and should be removed. Though chocolate and mint seem suitable smells to retain, most participants answered "yes" to most items, which may imply acquiescence bias. Though seemingly acceptable to PWDS, refinements are needed to

increase feasibility before piloting in a larger sample. To explore whether findings are due to olfactory impairment or issues with test stimuli, a wider array of plausible responses typically given from PWDS to identify these scents should also be collected and incorporated into scoring criteria. Alternatively, multiple choice picture answers could be presented.

5.2.1.3 Verbal Expression

This test was examiner rated thus no feedback was given. Most participants scored well, indicating good verbal ability, which contrasts with difficulties in verbal expression commonly reported for PWDS (Lott & Dierssen, 2010; Grieco et al., 2015; Fernández-Alcaraz & Carvajal, 2020). Findings may reflect the largely female sample (Määttä et al., 2006), self-selection bias, or additional prompts given in 'Orientation Subtotal B'.

5.2.1.4 Verbal Comprehension A & B

These tasks involved 'action on request' formats of language comprehension and motor response. No specific feedback was given for these subtests. Participants scored well, which again may be due to sample characteristics of healthy, mostly female PWDS. Item-level analysis indicates that item four of subtest 'A' ("Before you touch your ear, tap your shoulder) should be the final item (as the most difficult) in place of item five ("Now, look at the ceiling, then the wall, and then the floor"). In part 'B', '*Pointing*' item five ("Point to the buckle") was only answered correctly by one participant, so should be placed as item seven. Similarly, in '*Instructions'*, item 11 ("Before touching the coin, turn over the keys") may be better placed as the final item. This could be shortened by removing the simplest item per section from part 'B'.

5.2.1.5 Motor Function A & B

No specific feedback was given for this test. Participants scored well, indicating acceptability and feasibility. Items could be reduced and incorporated into '*Motor Programming*'.

5.2.1.6 Motor Programming

Feedback indicated this task was too confusing for some participants. Most participants struggled with 'hand sequencing' and 'inhibition' components, requiring several periods of task practice until the sequence was learned. Scores were generally low; as these are Luria-style tasks of frontal lobe integrity, this may be expected considering evidence of pre-existing frontal lobe abnormalities for PWDS (Holland et al., 2000; Peltopuro et al., 2014). Alternatively, findings may suggest differences in motor control and sequencing for PWDS. Considering time spent learning the sequence, a full copy trial of the 'knock-tap' sequence may be helpful before inhibition. This can help ensure performance on the 'inhibition' trial is related to EF (or proxy) functions, rather than difficulties in motor control and/or learning.

5.2.1.7 Praxis

This subtest involved mimicking object-use to command or imitation. No specific feedback was given, though participants showed the most verbal and non-verbal signs of enjoyment in this task. All participants scored well on this test, though all required a prompt for item eight (scissors) which may be best placed as the final item of '*Object Use'*. Similar formats are indicated as sensitive to cognitive degeneration over time for PWDS (Sano et al., 2005; Head et al., 2011; DeVreese et al., 2021), and current findings support an absence of floor effects for PWDS, so can be assumed as feasible and acceptable in its current iteration.

5.2.2 Verbal Learning & Visual Functions

5.2.2.1 Word List Immediate, Learning, Delayed Recall & Recognition

This subtest involved learning and recall of a list of eight familiar words. One participant identified these tasks as too easy and suggested including more difficult words. The task may have been understood as a word repetition task, rather than a learning task. Superficially, score ranges were generally good. Participants scored well in immediate and delayed recall trials, and in recognition, with 'slope' scores indicating good information acquisition as expected in this sample (Devenny et al., 1992). Similar tests indicate good test-retest reliability and sensitivity to AD-related decline in PWDS (Devenny et al., 1996; Krinsky-McHale et al., 2002; 2008), and so this test should be retained.

5.2.2.2 Circle Search

This task involved 'striking out' target shapes amongst a field of distractors. No specific feedback was given by participants for this test, though ease of administration and completion were high, suggesting inclusion of these stimuli are acceptable to PWDS. Acceptability findings support Krinsky-McHale and colleagues (2008), yet a narrow range of scores was seen, suggesting this task may be too easy for PWDS. As this format has shown good specificity and sensitivity (Krinsky-McHale et al., 2008), it should be retained with adaptations to include a higher number of distractors, or distracters of greater similarity to the target item (e.g., ovals), to increase task difficulty and widen score range.

5.2.2.3 Angle Judgement

This task involved identifying target numbered lines with reference to a five-point reference key. No specific feedback was given for this test, and performance was variable with one participant completing this task with ease, and fair scores seen for the other two participants. Researcher observations indicated that stimuli are too small for participants and may be difficult for those with visual impairments. Tentatively, this task showed a good range of scores for PWDS, though stimuli should be made larger to be accessible to people with visual impairments.

5.2.2.4 Visual Reasoning

This task involved completing a shape sequence with an item that fit the sequence pattern, from a multiple choice selection. Though no feedback was given, scores and performances indicated demonstrable difficulty on this task, with considerable floor effects and early employment of the discontinuation rule. Results indicate this task as not feasible in its current iteration, and scores may reflect difficulties in abstraction for PWID (Hassiotis et al., 2012). As the participant sample were aged 32-40, difficulties may have been exacerbated by age (Crome & Stern, 1972; Holland et al., 2000). However, all participants could complete the practice items, suggesting simplification of test items may improve feasibility. Alternatively, low face validity may underpin results, similar to participant feedback on EF tasks reported by Webb and colleagues (2020). Therefore, this test may be better substituted with a different, more 'life-like' task of abstraction. The BADS-ID could be reviewed for other suitable candidate tasks, though acceptability and feasibility to PWDS has not been explored. The 'frog hop' task from the Hayling and Brixton tests

(Burgess & Shallice, 1997) may be useful as a substitution, to assess pattern detection and response to rule shifts. Alternatively, series-format item of 'everyday' abstraction (e.g., "what comes next?") could be considered, such as steps in making a meal (Burgess et al., 2006).

5.2.2.5 Shopping List Map 1 & 2

This task involved collecting items on a map whilst adhering to a set of rules. One participant indicated this task was too easy, though item-level analysis indicated a high level of difficulty. Most participants completed map one incorrectly, therefore map two was often not given. This may reflect poor task acceptability and feasibility, which conflicts with previous research indicating a wider range of scores with acceptable ceiling scores for this format with PWID (Webb et al., 2020). However, as Webb and colleagues (2020) did not include PWDS, current findings may instead indicate pre-existing difficulties in executive functions such as task sequencing in this population (Snart et al., 1982; Lincoln et al., 1985; Costanzo et al., 2013). Despite this, scores were fair, and may reflect scoring criteria being too generous for parts of instructions that were completed (e.g., specific item collection) compared to penalties for deviation of task rules (e.g., using a path only once). This task may benefit from gradual introduction of additional EF load, as seen in Luria-style tasks (Korkman et al., 1998; Golden & Freshwater, 2001). This may be achieved in future revisions by first presenting a simplified trial/practice map to assess task planning and task understanding, then a second map with rules which engage inhibition and task switching. Additionally, feedback indicated that presented task rules were not in an appropriate format. This may have left participants holding instructions in mind, increasing strain on WM (Smith & Jonides, 1999). Aligned with feedback, task instructions should be highlighted, aligned left, and in larger font. However, considering floor effects and the long duration of battery administration, this test may be best omitted in future revisions.

5.2.2.6 Cat-Dog Inhibition

This task involved congruent and incongruent naming of photos of cats and dogs. One participant indicated this test was too easy. Though no participants scored at maximum (no errors), two scored very highly, and one scored at just above 50% correct, indicating variability in task performance. Findings tentatively support evidence of the Stroop-like task format as feasible and easy to administer for PWDS (Bevins & Hurse, 2014). Incongruent naming was not timed, so between-trial comparisons could not be made;

though this was not imperative in the current study, as an exploration of acceptability and feasibility to PWDS. However, to understand the 'cost' of inhibition in this task (Stroop, 1935), a timed congruent trial from which the incongruent time is accounted for (e.g., by subtraction or division) should be added to further revisions.

5.2.3 Visual Learning & Verbal Functions

5.2.3.1 Matchsticks Copy, Immediate & Delayed Recall

This task involved copying a matchstick design to reference and from memory. One participant found this task fun and interesting, though some participants were observed finding it difficult to manipulate matchsticks into position. A wide range of scores were shown for all trials, with one participant completing accurate matchstick designs across trials, and another who showed difficulty recreating the design across all trials. This may reflect variability in visuospatial skills for PWDS independent of mental age (Yang et al., 2014), or observations of difficulty in manipulating matchsticks due to their small size. Alternatively, as the design presented in the 'copy' trial was a printed line drawing of 'matchsticks', instead of using a 'real life' model (such as in block design tasks), it may employ elements of abstraction. Therefore, future revisions should use larger materials to reduce reliance on fine motor skills and consider replacing the target design with a 'real life' model (e.g., matchsticks glued to a piece of card to represent the target design), which may increase face validity.

5.2.3.2 Picture Naming & Recognition

One participant indicated this test was too easy and suggested that pictures were 'harder' to create more challenge. This may have been a shared experience, as all participants scored highly. No participants could identify 'ostrich'; possibly as it is a lesser-known animal which is more appropriately placed as a later test item. There appears a need for 'mid-difficulty' items in this subtest. Lesser-known animals could be included as substitutes to mid-late items to increase task difficulty, more closely replicating the difficulty scaling of the BNT (Kaplan et al., 1976). Considering difficulties in verbal expression seen in the cognitive profile of DS (Lott & Dierssen, 2010; Grieco et al., 2015; Fernández-Alcaraz & Carvajal, 2020), lesser-known items should not also be too phonetically challenging to ensure assessment of confrontation word retrieval rather than verbal ability. Animals such as 'leopard' or 'rhino' could be considered, but not 'axolotl' for

example. As evidence is mixed regarding whether confrontation word retrieval tasks are sensitive to dementia-related decline in PWID (Palmer, 2006; Pulsifer et al., 2020), further revisions should be piloted longitudinally to inform inclusion in the battery.

5.2.3.3 Eight Detection

This task involved signalling a target number in an audio recording. No specific feedback was given, and all participants scored well. This was interesting, as signal detection was presented towards the end of the battery, where lapses in attention may be expected. This may reflect strengths in auditory sustained attention for PWDS (Breckenridge et al., 2013). Any 'misses' were generally towards the end of the task. This may indicate that the test is not sufficiently derailing (boring) for PWDS, and test length may need to be extended, or have longer intervals between numbers.

5.2.3.4 Sentence Repetition

This task involved repeating a sentence back verbatim, with items increasing in syllable length. No specific feedback was given for this test, though item analysis suggests that task difficulty is inappropriately scaled for PWDS, with many failing to score after item three (four syllables), and all failing to score after item six (eight syllables). This may relate to difficulties in verbal expression and short-term memory seen for PWDS (Das & Mishra, 1995; Næss et al., 2012), implying this task is unlikely to be sensitive to dementia-related decline in PWDS. As it would be difficult to simplify this task further (item one has three syllables), it may need to be substituted in further revisions. Other widely-used tasks, such as digit-span (Wechsler, 1986), have shown floor effects for PWDS (De Sola et al., 2015). Therefore, a non-verbal substitute task format such as immediate memory for objects (e.g., TSI; Albert & Cohen, 1992) could be considered as a substitute candidate task. This may prove feasible considering strengths in visuospatial short-term memory seen in PWDS (Lott & Dierssen, 2010).

5.2.3.5 Verbal Reasoning

This task involved completing a sentence with a fitting word. No specific feedback was given, and participants generally scored well, with all items answered correctly by at least one participant. For appropriate difficulty scaling, item 11 ("Pen is to writing as scissors is

to…") should be swapped with item five ("A robin is a bird, a rabbit is a…"), and item eight ("An aeroplane goes in the sky, a boat goes on the…") should be an early test item.

5.2.3.6 Word Generation

One participant found this task interesting, and enjoyed seeing how many foods they could think of. All participants scored well, which is expected within the characteristics of the study sample, though there was high variability in scores (17-48). Ease of administration reflects the findings of Cooper and colleagues (2016), and high scores support research indicating semantic fluency as a strength of PWDS (Conners et al., 2011). Semantic fluency task formats indicate sensitivity to AD-related decline in PWDS (Pulsifer et al., 2020), therefore high scores may not indicate poor feasibility or potential validity of this format. However, this task may need revision or substitution to increase difficulty. Phonemic category tasks are unlikely to be appropriate, considering difficulties for PWDS in phonological encoding and the influence of differences in schooling experience (Næss et al., 2012) which may discriminate against examinees. A shorter time frame could be employed, similar to McCarthy (1972) to increase brevity and task difficulty. Alternatively, an 'action fluency' format could be considered.

5.3 Clinical Implications

Even in this small sample of healthy PWDS, variability in test performance is seen, supporting research indicating a wide range of functioning within a single aetiology of ID (Krinsky-McHale et al., 2008; Conners et al., 2011; Yang et al., 2014). Though data on ID severity was not gathered, it is likely that all participants had mild ID, indicating variation in cognitive abilities despite similar IQ. This highlights the need for cognitive tests which are feasible and acceptable to people of each ID aetiology, alongside IQ severity, with appropriate adaptations for individual differences. Considering the difficulty in establishing norm data for the ID population, findings of high individual variability also support the need to routinely assess PWDS across their lifespan, to establish baselines which support identification of dementia-related decline (Moran et al., 2013). Challenges in assessment of EF and OA may support previous research reporting pre-existing impairments in frontal lobes (Crome & Stern, 1972; Holland et al., 2000) and olfactory bulbs (Bianchi et al., 2014; Bontempi et al., 2020) for PWDS. This suggests routine cognitive assessments should

begin at an earlier age for PWID/DS, to establish a baseline of these functions to identify pathological cognitive decline.

This study highlights the importance of including the opinions of PWID in research concerning the development of cognitive instruments, to shape acceptability of tests as shown in the current study and by Webb and colleagues (2020). Many crucial implications for test item feasibility and acceptability would not have been understood through interpretation of performance data alone. Further, results demonstrate the feasibility of creating a low-cost battery with readily available resources which (with further refinement) may be used widely in low resource services.

5.4 Critical Review

This study gathered qualitative feedback from participants alongside quantitative data to ascertain feasibility and acceptability, for purposes of test refinement. This can be considered a strength, as combined data can create stronger evidence towards interpretations of quantitative task performance. This also includes the voices and experiences of PWDS in influencing the instruments created for their care (Coons & Watson, 2013), which is largely missing in existing literature. However, this could have been amplified by involving PWDS at far earlier stages, such as stage two of test development (Fenn et al., 2020; Hendrix et al., 2020). Further, questions specifically around comprehension of task instructions could have been included to improve acceptability, as feedback indicated that some tasks were confusing. Questions could also have been added to gather feedback on each test specifically, alongside test duration, to inform acceptability and feasibility. However, this would have increased administration time, and may have been better collected through a follow-up focus group. However, this may also affect participant recall of their experience.

Although efforts were made to reduce feelings of coercion, suggestion, or power imbalance, these still may have been present, as many participants did not take breaks despite these being offered regularly. It may have been difficult for participants to express any discomfort during testing meetings (Spears & Smith, 2001; Khemka et al., 2009). This may have also led participants to give feedback that they felt the researcher ‘wanted to hear’ regarding the tests and complete the battery in a single meeting, which may have

been perceived as a ‘correct’ or socially desirable behaviour (Everington & Fulero, 1999; Khemka et al., 2009). However, as trusted guardians were present, and one participant was able to give constructive criticism around the tests (and end the test early at their preference) effects may not have been too deleterious.

The small sample size is a significant weakness of the current study, and the anticipated sample size was not achieved within the timeframe available. Therefore, firm conclusions and inferences from the data cannot be drawn. Difficulties in recruiting PWDS have been reported in many similar studies (e.g., Sinai et al., 2016), and feedback from the affiliated clinical services indicated that the eligibility criteria excluded many people known to them, particularly the requirement of having sufficient verbal/motor ability to consent and no severe and/or enduring mental health difficulties. Upon reflection, this was to be expected for many PWDS whose needs are such that they are under the care of an NHS service. Though this barrier was not seen as prominently when recruiting from the non-clinical third-sector services, the charities were small and created for local community members, with far fewer potential participants available to contact. Further, few charities for DS adults were open to contact. A recommendation is to approach several third-sector and community organisations across the UK and spend time with PWDS and their carers in these settings to raise awareness and understanding of the research directly with potential participants. This will also improve meaningful opportunities for PWID to contribute to instruments created for their care.

Additionally, all participants who completed the battery were female, with mild ID, which are shown to have milder cognitive weaknesses in comparison to the wider DS community (Määttä et al., 2006). Further, the sample included only white individuals, which does not inform accessibility or feasibility of the battery cross-culturally. A recommendation for is to ensure a community-led approach is made to engaging with PWDS and PWID of other representations of varying cultural and ethnic identities, to facilitate a more representative participant group to understand the experiences of PWID in relation to the novel battery. This will also ensure that the underlying narrative of research which infers that WEIRD individuals are the ‘standard’ is challenged, as individuals who are WEIRD are shown to have vastly different experiences to the rest of the general population (Heine & Norenzayan, 2010).

Finally, as all participants in the current study were PWDS, it can be reasonably assumed that any potential floor or ceiling effects shown in this small sample may be specific to a

general cognitive profile of DS, and not indicative of performance which may be seen from representatives of the wider ID communities. This limits generalisability, though variation in task performance was still shown. Indeed, the current study is an exploratory analysis of feasibility and acceptability and did not seek to establish norm data. As a primary aim of this research was to use the understanding gained from the present pilot study to inform future research and development of the draft battery, it can be argued that findings are valuable directions for further revisions despite the small sample.

5.5 Future Research

Aligned with the aims of the study, avenues for future research are highlighted which have been briefly discussed previously. Considering the small sample in the current study, item-level analysis and feedback data will be incorporated into a further revision of the battery and piloted again in a larger sample of PWDS, and people with ID of other aetiologies. One such project is currently being undertaken as a separate doctoral thesis. This can better establish feasibility of tests through item analysis for PWDS/ID, with the potential for deriving preliminary norm data.

Future research should focus on the remaining phases of test development; revision of the battery, piloting, determination of validity and reliability (through comparison with other standardised tests) and exploratory factor analysis (Fenn et al., 2020). Results should inform a third revision of the battery, where confirmatory factor analysis can inform creation of a final battery and accompanying examiner manual. Sensitivity to dementia-related decline should be established, accounting for differences in cognitive profile and dementia trajectories seen in the ID community. This can be done through undertaking longitudinal research with samples of healthy PWID of multiple aetiologies and matched comparator groups with dementia of different subtypes. Data can be compared with existing appropriate normative data from assessments for this population such as the CAMCOG-DS (Ball et al., 2008) or CAMCOG-DS-II (Beresford-Webb & Zaman, 2021), to establish concurrent validity.

In these further phases of test development, future research should include PWID at earlier stages of development; to gather opinion and acceptability data which cannot be found through test performance alone. This should be more meaningfully done at the

coproduction and participatory action levels (Arnstein, 1969; Coons & Watson, 2013). This may be achieved through focus groups and/or creating links with charities and community initiatives created for the DS and ID communities. This may also improve likelihood of recruiting sample sizes with sufficient statistical power to make meaningful inferences about utility of candidate tasks. Findings can inform further adaptation of the battery, to eventually establish validity in different languages, and across cultures. Information resulting from this which may be useful to the ID community, their carers and ID services should be shared in several formats, including 'easy read'. Such avenues are crucial to address unmet healthcare needs for PWID, and reduce the unjust disparities seen in health outcomes and life expectancies for PWID (Glover & Ayub, 2010).

5.6 Conclusions

Though many instruments have been developed for PWID, several show floor effects and lack robust, acceptable assessment of executive function (EF). Evidence for impairments in olfactory ability in PWDS are well-documented, though this has not been exploited for use in detecting dementia-related neurodegeneration. The current study aimed to create a draft novel battery of comprehensive cognitive assessment for PWID, including measures of EF and olfactory function. Results highlighted significant challenges in the development of tests which appropriately examine EF and olfactory ability, alongside relative acceptability of the battery to PWDS. Feasibility was tentatively implied for some subtests, and several indications for revisions to the battery were identified, most notably shortening of overall administration time and revisions to EF tasks. Findings in the current study show limited generalisability to the wider ID community and to the DS community, as the sample was small, and largely comprised of white women. However, valuable directions for revisions to the novel battery were identified and reasonably congruent even within this small sample of PWDS. Revisions of this battery may go on to create a format which is valid and reliable to PWID. Such instruments are crucial to development of a 'gold standard' in diagnosing dementia in PWID, to reduce disparities in health outcomes by providing early diagnosis and intervention. Aligned with NHS and Department of Health priorities (Zeilinger at al., 2013; Ham & Murray, 2015), this can provide improved quality of life and access to meaningful support for the PWID, their families and carers.

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APPENDICES

Appendix A: List of Abbreviations

List of Abbreviations

ABCD = Arizona Battery for Communication Disorders of Dementia

ABI= Acquired Brain Injury

ACE = Addenbrookes Cognitive Examination

ACTB = Arizona Cognitive Test Battery

AD= Alzheimer's Disease Dementia

ADLs = Activities of daily living

ADVM = Auditory delayed verbal memory

AMT = Autobiographical Memory Test

APA = American Psychological Association

APP = Amyloid precursor protein

ASL or landmark = Allocentric spatial learning or landmark

ASM = Auditory sequential memory

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

BPS = The British Psychological Society

BPT = The Brief Praxis Test

BPVS = British Picture Vocabulary Scale

BSRT = The Buschke 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

CAS = Das–Naglieri Cognitive Assessment System

CEFA = Cambridge Executive Functioning Assessment

CFA = Confirmatory factor analysis

CFT = Category Fluency Test

CMS = The Children's Memory Scale

CoD = Copy of drawings

COVID-19 = Coronavirus disease

COWAT = The Controlled Oral Word Association Test

CRT = The Cued Recall Test

CS = Cognitively stable

CT = Cancellation task

CTT = The Colour Trails Test

D-KEFS = Delis-Kaplan Executive Function System

DLD = Dementia Questionnaire for People with Learning Disabilities

DM-ID = Diagnostic Manual — Intellectual Disability

DMVMT = Dalton-McMurray Visual Memory Test

DNMP = Spatial delayed non-match-to-position

DNMS = Object delayed non-match-to-sample

DRS = Dementia Rating Scale

DS = Down syndrome

DSpan = Digit Span

DSpan-B = Digit Span - backwards

DSDS = The Down Syndrome Dementia Scale

DSpan-F = Digit Span - forwards

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

DwLB = Dementia with Lewy bodies

EF = Executive function

EFA = Exploratory factor analysis

EISC = Ethics and Integrity Sub-Committee

EMAS= Edinburgh Motor Assessment Scale

EMS = Evaluation of Mental Status

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

FMR1 = Fragile X messenger ribonucleoprotein 1 gene

FMRP = Fragile X messenger ribonucleoprotein

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

FS = Finger Sequencing

FSIQ = Full-Scale IQ

FTLD = Frontotemporal lobar degeneration

FXS = Fragile X syndrome

GA = Gait Assessment

HOM = Hidden Object Memory Test

HRA = Health Research Authority

ICAT = Iowa Cognitive Abilities Test

ICC = Intraclass Correlation Coefficient

ICD = International Classification of Diseases

ID = Intellectual disability/ disabilities

IQ = Intelligent Quotient

IQCODE = Informant Questionnaire on Cognitive Decline in the Elderly

IQR = Interquartile range

ITPA = Illinois Test of Psycholinguistic Ability

JLO = Judgment of Line Orientation

K-BIT = Kaufman Brief Intelligence Test

KBNA = Kaplan Baycrest Neurocognitive Assessment

LIPS = Leiter International Performance Scale

LTM = Long-term memory

LTR = Long-term recall

LTS = Long-term stores/ storage

MAT = Matrix Analogies Test-Expanded Form

MCA = Mental Capacity Act

M-CFT = McCarthy Scales of Children's Abilities - Category Fluency Test

MCI = Mild Cognitive Impairment

MEAMS = Middlesex Elderly Assessment of Mental State

MfO = Memory for objects from the NAID

mMMSE-DS = Modified Mini Mental Status Evaluation—Down Syndrome

MMSE = Mini Mental Status Evaluation

MMSE-O = Orientation subtest from the MMSE

MoCA = The Montreal Cognitive Assessment

MSCA = McCarthy Scales of Children's Abilities

N/A = Not applicable

NAID = Neuropsychological Assessment of Dementia in Intellectual Disabilities

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

NR = Not reported

ODL = Object discrimination learning

OI = Object identification

oID = Intellectual disability from other causes than DS

OPS = Object-Pointing Span

PAL = Paired-associate learning task

PCFT/s-PCFT = Prudhoe Cognitive Function Test/Prudhoe Cognitive Function Test – Short Forms

PD = Parkinson's disease

PDD= Parkinson's disease dementia

PHE = Public Health England

PN = Picture Naming

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

PV= Percentage/proportion passing

PWID = People with an Intellectual Disability

PWDS= People with Down Syndrome

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

RBANS = The Repeatable Battery for the Assessment of Neuropsychological Status

RBD = REM (repetitive eye movement) sleep behavioural disorder

RBMT-C = Rivermead Behavioural Memory Test for Children

RCPM = Raven Coloured Progressive Matrices

RCT = Randomised Controlled Trial

RL = Reversal learning

SBIS = Stanford-Binet Intelligence Scales

SD = Standard deviation

SIB = Severe Impairment Battery

S-MMSE = Shultz Mini Mental State Exam

SPSS = Statistical Package for the Social Sciences

SR = Sentence repetition

STM = Short-term memory

STS = Short-term stores

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

SVDL = Simple visual discrimination learning

TACL-III = Test of Auditory Comprehension of Language-3

TD = Typical development or typically developing

TEA= The Test of Everyday Attention

TEA-Ch= The Test of Everyday Attention for children

TO = Temporal Orientation

TOL = Tower of London

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

TSI = Test for Severe Impairment

TT = Token Test

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 Behaviour Scales

VaD = vascular dementia

VAT = Visual Association Test

VC = Verbal comprehension

VF = Verbal Fluency

VisMT = Visual Memory Test

VMI = Visual Motor Integration

VT = Vocabulary Test

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

WASI = Wechsler Abbreviated Scale of Intelligence

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

WPPSI = Wechsler Preschool and Primary Scale of Intelligence

WS = Williams syndrome

WST = Weigl Colour-Form Sort Test

WTAR= Wechsler Test of Adult Reading

Appendix B: Cognitive Tests in Included Studies

Table B 1. *List of Batteries: Cognitive Domains and Associated Functions, Population(s) Developed for, and Studies Used Within*

Abbreviations	List of Measures	Function/ Cognitive domain(s)	Developed for Adult	Developed for ID	DS-specific	Developed for Dementia/ Cognitive deterioration	Author(s)
ABCD	Arizona Battery for Communication Disorders of Dementia	General cognitive ability: mental state; episodic memory; linguistic expression; linguistic comprehension; and visuospatial construction	YES	NO	NO	YES	Carvalho et al., 2018
ACTB	Arizona Cognitive Test Battery	Cognitive function of prefrontal, hippocampal and cerebellar areas (often associated with cognitive difficulties in PWDS)	YES	YES	YES	NO	Sinai et al., 2016
BADS	Behavioural Assessment of the Dysexecutive Syndrome	Six tasks of executive functioning	YES	NO	NO	YES	Wilson et al., 1996
BADS-C	Behavioural Assessment of the Dysexecutive Syndrome adapted for children	Six tasks of executive function adapted from the BADS	NO	NO	NO	NO	Emslie et al., 2003
BADS-ID	Behavioural Assessment of the Dysexecutive Syndrome adapted for people with Intellectual Disabilities	Six tasks of executive function adapted from the BADS	YES	YES	NO	NO	Webb et al., 2020
CAMCOG	Cambridge Cognition Examination	Dementia assessment battery: orientation; attention and perception; language; language and memory; praxis; abstract thinking; and calculation	YES	NO	NO	YES	Fonseca et al., 2014; Hon et al., 1999
CAMCOG-DS	Cambridge Cognitive Examination adapted for PWDS	Dementia diagnosis tool for PWDS. Orientation; attention and perception; language comprehension and expression;	YES	YES	YES	YES	Ball et al., 2004; Ball et al., 2008; Ball et al., 2010; Benejam et al., 2020; Fonseca et al., 2019a;

Abbreviations	List of Measures	Function/ Cognitive domain(s)	Developed for Adult	Developed for ID	DS-specific	Developed for Dementia/ Cognitive deterioration	Author(s)
		learning and memory (visual and verbal); praxis; abstract thinking					Fonseca et al., 2019b; Fortea et al., 2020; García-Alba et al., 2019
CANTAB	Cambridge Neuropsychological Test Automated Battery	Assesses cognitive changes: working memory; learning and executive function; visual, verbal and episodic memory; attention, information processing and reaction time; social and emotion recognition, decision making and response control	YES	YES	NO	YES	Cooper et al., 2016; Oliver et al., 2005
CAS	Das–Naglieri Cognitive Assessment System	Originally a measure of cognitive ability in TD children. Measures attention; planning; and 'simultaneous and successive cognitive processes' (based on PASS theory of intelligence)	NO	NO	NO	NO	Das et al., 1995; Das & Mishra, 1995
CEFA	Cambridge Executive Functioning Assessment	Developed to aid dementia detection for PWDS: eight EF (two executive memory) subtests, four memory subtests	YES	YES	YES	YES	Adams & Oliver, 2010; Ball et al., 2008; Ball et al., 2010; Bevins & Hurse, 2014; Fonseca et al., 2019b; Willner et al., 2010; Web et al., 2020
DSMSE	Down Syndrome Mental State Examination	Screening for cognitive deterioration: orientation (days of the week, seasons); personal information; short-term memory; language (confrontation naming of clothing and body parts); visuospatial construction and praxis	YES	YES	YES	YES	Alexander et al., 1997; Cosgrave et al., 1998; Krinsky-McHale et al., 2020; Manning et al., 1998; McCarron et al., 2014; McCarron et al., 2017
MEAMS	Middlesex Elderly Assessment of Mental State	Cognitive functioning: orientation and memory; new learning; naming; comprehension and arithmetic; visiospatial skills; and perception	YES	NO	NO	YES	Thompson, 1994
mMMSE-DS	Modified Mini Mental Status Evaluation—Down Syndrome	Screening for cognitive deterioration	YES	YES	YES	YES	Krinsky-McHale et al., 2020
MMSE	Mini Mental Status Evaluation	Screening for cognitive deterioration: orientation, registration (immediate memory), short-term memory (but not long-term memory) as well as language functioning	YES	NO	NO	YES	Gutman et al., 2016; Hon et al., 1999

Abbreviations	List of Measures	Function/ Cognitive domain(s)	Developed for Adult	Developed for ID	DS-specific	Developed for Dementia/ Cognitive deterioration	Author(s)
MoCA	The Montreal Cognitive Assessment	Screening for dementia and MCI: short term memory; visuospatial abilities; executive functions; attention, concentration and working memory; language; orientation to time and place	YES	NO	NO	YES	Carvalho et al., 2018
NAID	Neuropsychological Assessment of Dementia in Intellectual Disabilities	Assesses cognitive changes: early stages of dementia (working memory) and later stages (agnosia, aphasia, and apraxia); orientation and language	YES	YES	NO	YES	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; Sinai et al., 2016
LDDB	Learning Disabilities Dementia Battery	Assesses cognitive changes: orientation, visual, verbal and recognition memory (immediate and delayed), verbal fluency, new learning, perceptual/planning, visuospatial, abstract thinking/concept formation, and language ability.	YES	YES	NO	YES	Poveda & Broxholme, 2016
PCFT/s-PCFT	Prudhoe Cognitive Function Test/Prudhoe Cognitive Function Test – Short Forms	Assesses cognitive function in PWID. Repeated administration over time may indicate cognitive deterioration.	YES	YES	NO	YES	De Vreese et al., 2021; Kay et al., 2003; Margallo-Lana et al., 2003; Margallo-Lana et al., 2007
RADD/RADD-2	The Rapid Assessment of Developmental Disabilities/Second Edition	General cognitive ability and cognitive decline in ID	YES	YES	NO	YES	Walsh et al., 2015
SIB	Severe Impairment Battery	Assesses behavioural and cognitive deterioration in severe dementia: orientation; attention; language; learning and memory; visuospatial ability; construction	YES	NO	NO	YES	Ball et al., 2010; Head et al., 2011; Hutchinson & Oakes, 2011; Powell et al., 2014; Walsh et al., 2015; Witts & Elders, 1998
S-MMSE	Shultz Mini Mental State Exam	Screening for cognitive deterioration: orientation; personal knowledge; immediate and delayed memory; and language comprehension	YES	YES	NO	YES	Shultz et al., 2004
TESTAD	A Neurocognitive Battery for Clinical Trials in DS adults	Characterisation of cognitive function in young adults with DS. May be used to assess cognitive change in intervention studies.	YES	YES	YES	NO	De Sola et al., 2015

Abbreviations	List of Measures	Function/ Cognitive domain(s)	Developed for Adult	Developed for ID	DS-specific	Developed for Dementia/ Cognitive deterioration	Author(s)
TSI	Test for Severe Impairment	Cognitive function for people with severe cognitive impairment	YES	NO	NO	YES	Cosgrave et al., 1998; Krinsky-McHale et al., 2020; McCarron et al., 2014; McCarron et al., 2017; Pyo et al., 2010
WG - MTB	Working Group Memory Test Battery	Memory	YES	YES	NO	YES	Pyo et al., 2007
BT-ID	Barcelona Test – Intellectual Disability	Cognitive function across eight cognitive domains	YES	YES	NO	NO	García-Alba et al., 2017
BPVS	British Picture Vocabulary Scale	Language	NO	NO	NO	NO	Adams & Oliver, 2010; Ball et al., 2008; Ball et al., 2010; Crayton et al., 1998; Oliver et al., 2005
EMS	Evaluation of Mental Status	General cognitive status: orientation to person, place and time; object naming; visuomotor coordination; and concentration.	YES	YES	NO	NO	Devenny et al., 1996
K-BIT	Kaufman Brief Intelligence Test	Verbal and non-verbal IQ	YES	NO	NO	NO	Benejam et al., 2015; García-Alba et al., 2017; Sinai et al., 2016
LIPS	Leiter International Performance Scale	Non-verbal IQ - wide variety of functions from memory to nonverbal reasoning	YES	NO	NO	NO	Burt et al., 2005; Carr & Collins, 2018
MAT	Matrix Analogies Test-Expanded Form	Non-verbal IQ	NO	NO	NO	NO	Das et al., 1995
MSCA	McCarthy Scales of Children's Abilities	General cognitive ability: verbal, perceptual-performance; quantitative, general cognitive, memory, and motor	NO	NO	NO	NO	Burt et al., 1998
NEPSY	A Developmental NEuroPSYchological Assessment	Assesses language; motor; social, emotional, behavioural; play; adaptive skills; academic skills	NO	YES	NO	NO	Pyo et al., 2007; Pyo et al., 2010
PPVT-R/ PPVT-III	Peabody Picture Vocabulary Test-Revised/ 3rd Edition	Receptive language	YES	NO	NO	NO	Alexander et al., 1997; Das et al., 1995; Manning et al., 1998; Nelson et al., 2001
RCPM	Raven Coloured Progressive Matrices	Non-verbal IQ, abstract reasoning (sequences)	YES	NO	NO	NO	Thompson, 1994
SBIS	Stanford-Binet Intelligence Scales	Verbal and non-verbal IQ: fluid reasoning; general knowledge; quantitative reasoning; visuospatial processing;working memory	NO	NO	NO	NO	Alexander et al., 1997; Das et al., 1995
WAIS/ WAIS-R/ WAIS-III	Wechsler Adult Intelligence Scale/ Revised/ 3rd Edition	Global cognitive ability, IQ	YES	NO	NO	NO	Das et al., 1995; Nelson et al., 2005; Nelson et al., 2007

Abbreviations	List of Measures	Function/ Cognitive domain(s)	Developed for Adult	Developed for ID	DS-specific	Developed for Dementia/ Cognitive deterioration	Author(s)
WISC/ WISC-R	Wechsler Intelligence Scale for Children/Revised	Global cognitive ability, IQ in children	NO	NO	NO	NO	Devenny et al., 2000; Krinsky-McHale et al., 2020

Table B 2. *List of Single Domain Tasks Per Domain: Associated Functions, Population(s) Developed for, and Studies Used Within*

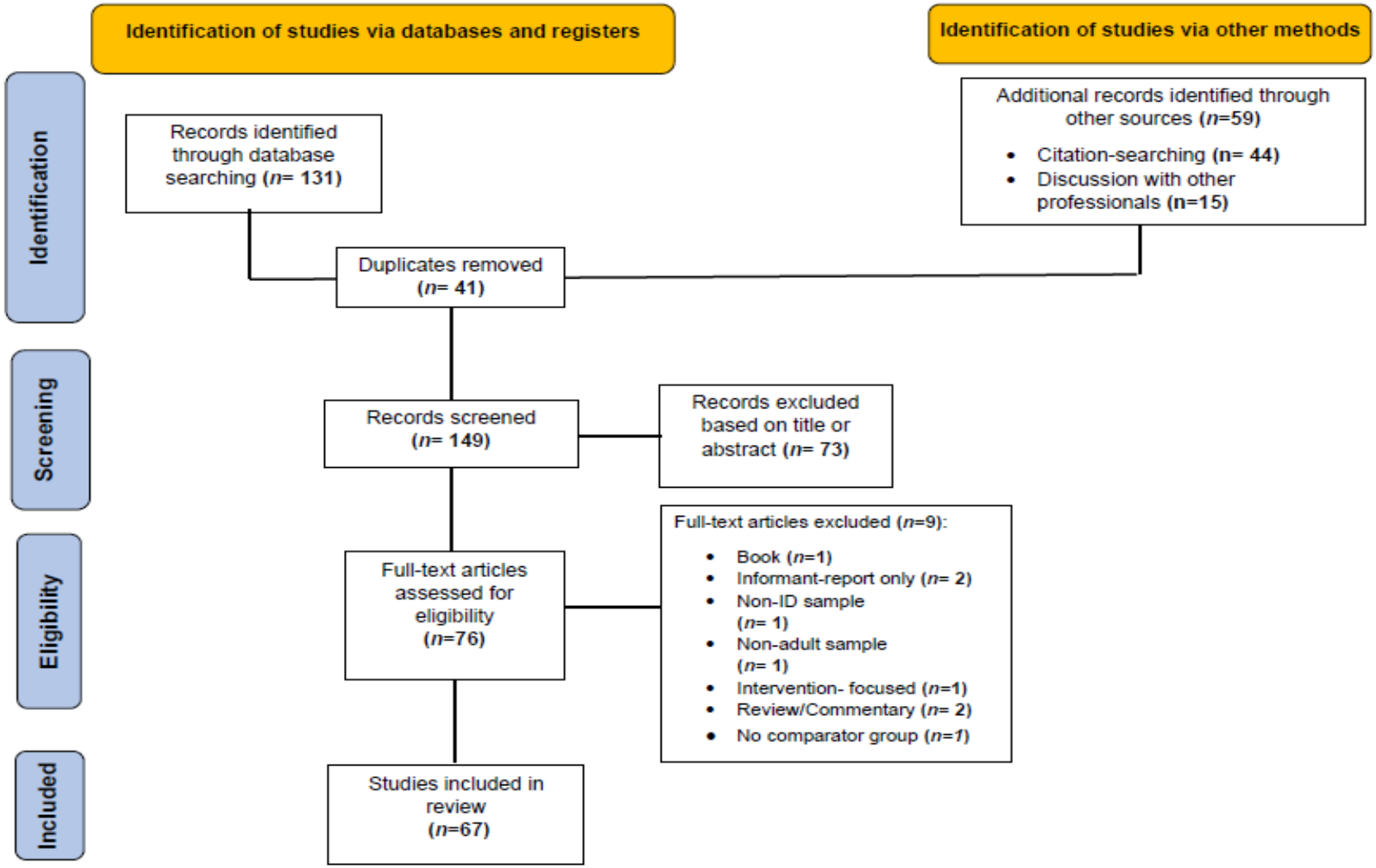
Abbreviations	List of Measures	Function/ Cognitive domain(s)	Developed for Adult	Developed for ID	DS-specific	Includes non-verbal	Developed for Dementia/ Cognitive deterioration	Author(s)
Single Domain Tests/ Tasks by Domain								
Orientation and Arousal								
MMSE-O	Orientation subtest from the MMSE	Orientation	NO	NO	NO	NO	YES	Jozsvai et al. 2002; Kinsky-McHale et al., 2002; Sano et al., 2005
TO	Temporal Orientation	Orientation						García-Alba et al., 2019
WG-O	Working Group's Orientation Test	Orientation	YES	YES	NO	NO	YES	Pyo et al., 2009
Attention								
CT	Cancellation task	Selective attention and visuospatial function	YES	YES	YES	YES	YES	Cooper et al., 2016; Krinsky-McHale et al., 2008
Executive Function								
CaD	Cats and Dogs task	Executive function - response inhibition						Bevins & Hurse, 2014; Cooper et al., 2016; De Sola et al., 2015; Hartley et al., 2020
CFT	Category Fluency Test	Executive function - verbal fluency	YES	NO	NO	NO	NO	Cooper et al., 2016
COWAT	The Controlled Oral Word Association Test	Executive function - verbal fluency	YES	NO	NO	NO	NO	Palmer, 2006
CTT	The Colour Trails Test	Executive function - cognitive flexibility and processing speed	YES	YES	NO	YES	NO	Palmer, 2006
M-CFT	McCarthy Scales of Children's Abilities - Category Fluency Test	Executive function - verbal fluency	NO	NO	NO	NO	NO	Krinsky-McHale et al., 2020; Pulsifer et al., 2020;
PAL	Paired-associate learning task	Executive function and short term visuospatial memory						Shultz et al., 2004
RL	Reversal learning	Executive function - response inhibition and set-shifting						Nelson et al., 2005; Nelson et al., 2007
TOL	Tower of London	Executive function - planning	YES	NO	NO	DK	NO	Cooper et al., 2016; Masson et al., 2010
TOL^{DX}	Tower of London-Drexel University: 2nd Edition	Executive function - planning	YES	NO	NO	DK	NO	De Sola et al., 2015; García-Alba et al., 2017; Sinai et al., 2016
VF	Verbal Fluency	Executive function - verbal fluency						Sinai et al., 2016
WST	Weigl Colour-Form Sort Test	Executive function - sorting and set-shifting	YES	NO	NO	YES	YES	Bevins & Hurse, 2014; García-Alba et al., 2017

Abbreviations	List of Measures	Function/ Cognitive domain(s)	Developed for Adult	Developed for ID	DS-specific	Includes non-verbal	Developed for Dementia/ Cognitive deterioration	Author(s)
Visuospatial								
BD	Block Design	Constructional, motor skill, problem-solving	YES	NO	NO	YES	NO	Alexander et al., 1997
BP	Block Patterns - Hiskey-Nebraska Test of Learning Aptitude subtest	Visuospatial ability	NO	NO	NO	YES	NO	Alexander et al., 1997
Language								
BNT	The Boston Naming Test	Language - confrontation naming retrieval	YES	NO	NO	NO	YES	Jozsvai et al. 2002; Palmer, 2006; Pulsifer et al. 2020
BPVS	British Picture Vocabulary Scale	Language- naming	NO	NO	NO	NO	NO	Crayton et al., 1998; Ball et al., 2008; Bevins & Hurse, 2014; Carr & Collins, 2018; Oliver et al, 2005; Willner et al., 2010
EOWPVT/EOWPVT-R	Expressive One-Word Picture Vocabulary Test/ Expressive One-Word Picture Vocabulary Test-Revised	Expressive language	YES	NO	NO	NO	NO	Sano et al., 2005
ITPA	Illinois Test of Psycholinguistic Ability	Measure of children's spoken and written language	NO	NO	NO	NO	NO	Alexander et al., 1997
OI	Object identification	Language						Alexander et al., 1997
PN	Picture Naming	Language						Oliver et al., 2005
PPVT/ PPVT-R	Peabody Picture Vocabulary Test/ Peabody Picture Vocabulary Test-Revised	Language - receptive	YES	NO	NO	NO	NO	Burt et al., 1998; Pyo et al., 2007
Srep	Sentence repetition	Language						Alexander et al., 1997
TT	Token Test	Verbal comprehension	YES	NO	NO	NO	NO	De Sola et al., 2015
VT	Vocabulary Test	Language						Sano et al., 2005
Learning and Memory								
ADVM	Auditory delayed verbal memory	Memory - delayed verbal						García-Alba et al., 2019
ASL or landmark	Allocentric spatial learning or landmark	Spatial memory						Nelson et al., 2007

Abbreviations	List of Measures	Function/ Cognitive domain(s)	Developed for Adult	Developed for ID	DS-specific	Includes non-verbal	Developed for Dementia/ Cognitive deterioration	Author(s)
BSRT	The Busckke Selective Reminding Test	Memory and learning - short-term and long-term verbal	YES	NO	NO	NO	NO	Devenny et al., 1992; Devenny et al., 2000; Krinsky-McHale, Devenny & Silverman, 2002; Krinsky-McHale et al., 2008; Krinsky-McHale et al., 2020
BTS	Block tapping span	Memory - short-term						Alexander et al., 1997
CRT	The Cued Recall Test	Memory - cued recall	YES	NO	NO	NO	NO	Benejam et al., 2015; Cooper et al., 2016; Devenny et al., 2002; Devenny et al., 2000; Oliver et al., 2005;
DNMP	Spatial delayed non-match-to-position	Spatial memory						Nelson et al., 2005
DNMS	Object delayed non-match-to-sample	Object recognition memory						Nelson et al., 2005; Nelson et al., 2007
DSpan	Digit Span	Memory - short-term						Manning et al., 1998
DSpan-B	Digit Span - backwards	Memory - short-term (WM component)						De Sola et al., 2015
DSpan-F	Digit Span - forwards	Memory - short-term						Alexander et al., 1997
DVM	Delayed visual memory	Memory - delayed visual						García-Alba et al., 2019
FOME/mFOME	The Fuld Object-Memory Evaluation/ Modified - Fuld Object Memory Evaluation	Verbal, visual and touch (tactile) stimuli to encode objects	YES	NO	NO	DK	YES	Palmer, 2006; Pyo et al., 2010; Sano et al., 2005
HOM	Hidden Object Memory Test	Memory - short-term, visual	YES	NO	NO	YES	YES	Alexander et al., 1997
MfO	Memory for objects from the NADIID	Memory - short-term, visual						Burt et al., 2005; Cooper et al., 2016; Oliver et al., 1998
NDT	New Dot Test	Memory - visuospatial memory						Sano et al., 2005
ODL	Object discrimination learning	Learning - conditioned learning						Nelson et al., 2005
OPS	Object-Pointing Span	Immediate memory						Manning et al., 1998
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; Pyo et al., 2010
RAVLT	Rey Auditory Verbal Learning Test	Auditory attention, memory, and learning	YES	NO	NO	NO	NO	Manning et al., 1998
RBMT-C	Rivermead Behavioural Memory Test for Children	Memory in children	NO	NO	NO	NO	NO	Carr & Collins, 2018
RaB	'Remembering a belonging' subtest of the RBMT-C	Memory	NO	NO	NO	NO	NO	Ball et al., 2008; Ball et al., 2010

Abbreviations	List of Measures	Function/ Cognitive domain(s)	Developed for Adult	Developed for ID	DS-specific	Includes non-verbal	Developed for Dementia/ Cognitive deterioration	Author(s)
StoryRT	Story Recall Test (adapted from the RBMT-C)							Cooper et al., 2016
SVDL	Simple visual discrimination learning	Learning and memory - visual discrimination and conditioned learning						Nelson et al., 2007
VisMT	Visual Memory Test	Memory - visual matching						Devenny et al., 1992
WG-AMT	Autobiographical Memory Test	Memory - autobiographical	YES	NO	NO	NO	NO	Pyo et al., 2011
Sensorimotor								
BPT	The Brief Praxis Test	Praxis – highly-practiced motor sequences	YES	YES	YES	DK	YES	Head et al., 2011; Powell et al., 2014; Sano et al., 2005; Walsh et al., 2015
BBDT-VMI	Beery Buktenica Developmental Test of Visual-Motor Integration	Visual-motor Integration	YES	YES	NO	YES	YES	Burt et al., 2005; Krinsky-McHale et al., 2020
FS	Finger Sequencing	Upper limb co-ordination						Sinai et al., 2016
GA	Gait Assessment (Timed Get Up and Go Test)	Assessment of walking style (gait)						Sinai et al., 2016
PPT	Purdue Pegboard Test	Motor coordination and dexterity	YES	NO	NO	YES	NO	Burt et al., 2005
UPSIT	University of Pennsylvania Smell Identification Test	Olfactory recognition ability	YES	NO	NO	YES	NO	Doty et al., 1984; 1995; Khan et al., 2006; Schmitt et al., 2010; Tabert et al., 2005

Appendix C: PRISMA Flow Diagram



Appendix D: Literature Review Strategy

Date Conducted: April 2021

Search Terms Used

Search terms were adapted from Zellinger et al. (2013) and Paiva et al. (2020).

The following search string was used to conduct the searches: (Adult* OR older adult*) AND (cognit* task or cognit* test OR neuropsych* test) AND (instrument OR questionnaire OR screening) AND (dementia OR alzheimer* OR cognit* impair*) AND (intellectual* disabilit* OR mental* retar* OR general learn* disabilit*). This search was performed a second time to include the search terms: (Down Syndrome OR Trisomy 21 or Down's Syndrome OR Down's or Trisom*). Limiters were applied to return studies written in English, published in peer-reviewed journals, those related to the adult population and to exclude papers published before 1980. Filters were also applied for the key search terms: NOT (child* OR adolesc* OR youth*).

Inclusion Criteria

Studies were included if they met the following criteria:

1) Studies written in English;
2) ID sample or TD sample if a measure of olfactory ability is used
3) Examined cognitive changes related to age and/or dementia
4) Reported comparisons between groups (e.g., dementia status, intellectual disability aetiology, intellectual disability severity), or longitudinally, or between cognitive (non-informant) measures, or reported acceptability, feasibility, validity, reliability of tests with PWID.

Exclusion Criteria

1) Conference presentations, case studies, protocols, book chapters, reviews/ commentaries, unpublished theses
2) Sample of adults without an intellectual disability
3) Included participants under 18 years of age in the sample;
4) Papers not written in English;
5) Includes informant-report measures only

6) Studies describing interventions for people with intellectual disability and/or dementia.

Summary of Included Studies

38 cross-sectional studies, 25 longitudinal studies, two studies which used both cross-sectional and longitudinal methods, and one longitudinal randomised control trial were identified. Longitudinal follow-up period ranged from two weeks (Manning et al., 1998) to 50 years (Carr & Collins, 2018) Sample size ranged from 14 (Margallo-Lana et al., 2003) to 561 (Krinsky-McHale et al., 2020). All studies were published between 1984 and 2021; 21 were conducted in the UK, 28 in the USA, six in Spain, four in Brazil, three in Canada, two in Ireland, one in Italy and one between Ireland and the USA.

One study reported a sample entirely of non-DS ID (Webb et al., 2020), two reported a 'mixed' sample of ID aetiologies (Masson et al., 2010; Willner et al., 2010), five included samples of PWDS and people with ID of other aetiologies, and the remaining studies reported samples of only PWDS. Most studies included a range of ID severity, though only eight studies included people with profound intellectual disabilities. ID severity was not reported in 15 studies. Assessment of ID severity was mostly through previous assessments, with some using general IQ tests such as the WASI, or DSM/ICD criteria.

Of studies including participants with dementia, or concerned with dementia-related outcomes, 27 studies reported AD as the primary dementia subtype of interest, though in 20 studies this was not specified. Most studies used ICD or DSM criteria, clinical judgement, or a combination of these to ascertain dementia status. The proportion of female participants ranged from 16.7% (Cosgrave et al., 1998) to 100& (McCarron et al., 2014, 2017). Many studies did not report on the verbal ability of participants, though some stated non-verbal participants were excluded from the sample.

Appendix E: Study and Sample Characteristics

Table E 1

Study Characteristics

Author, Year	Country	Type of study	Follow-up	Sample size (N)	Tests included	General findings	Psychometric/ acceptability findings
Adams & Oliver, 2010	UK	Longitudinal	8- and 16-months	DS= 30	NAID; BPVS; VABS; AADS; BPV; CEFA= TOL; WST; CaD; SB	Significant decline in EF from baseline to 16 month follow-up and between 8month and16 month follow-up in participants with cognitive deterioration. Not seen between baseline and 8 month follow up. May not indicate early symptoms. No indication of clinical presentation of effects.	Floor effects found for some participants.
Alexander et al., 1997	USA	Cross-sectional	N/A	old-DS = 17 young-DS = 24	HOM; BD; PPVT-R; SBIS; BP; DSpan-F; BTS; OI, and SRep; ME and GC subtests of the ITPA; DSMSE	After controlling for ID severity, older participants with DS (41–61 years) showed poorer scores on BD than younger participants (22–38 years), but not on other measures.	N/A
Ball et al., 2004	UK	Longitudinal	6 years	DS Time Point One: 74 (AD=9) DS Time Point Two: 56 (AD= 11)	CAMDEX-DS; CAMCOG-DS	N/A	CAMCOG-DS (Direct component): 14 participants scored at floor at baseline CAMDEX-DS (Informant component) = good predictor of later dementia diagnosis. Good concurrent validity and inter-rater reliability (Kappa >0.8 for 91% of items and >0.6 for all items.
Ball et al., 2008	UK	Cross-sectional	NA	103 (25= DS-AD; 78= DS-no-AD)	CEFA; CAMDEX-DS; BPVS II; CAMCOG-DS; RaB (RBMT-C); CODB	Supports frontal lobe impairment as preclinical indicator of AD in DS. DS-AD group showed poorer performance on all measures than DS-no-AD. Memory informant report only related to delayed memory scores. CAMDEX-DS personality/ behaviour changes predicted performance on EF and executive memory CEFA tests for DS-no-AD, but not episodic memory score.	Some floor and ceiling effects. Only spatial reversal not affected by ID severity. TOL and delayed recall affected by ID severity and increasing age. Floor effects seen for ID-AD group on TOL and delayed recall. CaD less sensitive to dementia status.

Author, Year	Country	Type of study	Follow-up	Sample size (N)	Tests included	General findings	Psychometric/ acceptability findings
Ball et al., 2010	UK	Cross-sectional	N/A	DS= 78	CAMDEX-DS; CAMCOG-DS; SIB; RaB (RBMT-C); CODB; CEFA	Decline in informant-related memory score significantly associated with lower delayed memory score, but not other memory measures. Adaptive behaviour scores and apathy scores both significantly predicted lower scores on EF tests in CEFA and CAMCOG-DS. But WM scores significantly associated with antidepressant use.	N/A
Benejam et al., 2015	Spain	Cross-sectional	N/A	DS-no-AD = 75 DS-AD = 15	CRT-M (Spanish); K-BIT	DS-no-AD scored better in free recall with fewer intrusion errors. CRT-M scores may discriminate between AD and no-AD groups. However age most associated with decline in CRT-M score.	DS-no-AD scored higher in CRT-M if semantic cue given. CRT-M instructions not understood by those with severe ID or late-stage AD.
Benejam et al., 2020	Spain	Cross-sectional	N/A	Completed CAMCOG-DS = 343 Completed CRT-M = 271	CAMDEX-DS; CAMCOG-DS; CRT-M (all in Spanish)	CAMCOG-DS and m-CRT scores show progressive decline after age 40, especially for moderate ID.	Completion rates lower in MCI and AD. Floor/not able to complete for many with severe and all with profound ID. CAMCOG-DS and m-CRT were able to detect MCI and AD with high accuracy in mild and moderate ID. Could predict AD onset.
Bevins & Hurse, 2014	UK	Cross-sectional	N/A	24 DS 4= oID	DLD ; CaD, WST, and VF from CEFA; BVPS-II; Object memory (NAID)	N/A	WST too complex, showed floor effects and was removed. CaD showed narrow scores and ceiling effects. VF did not correlate with other measures. CaD did not correlate with VC, suggesting little reliance on verbal ability. CaD correlated with object memory, suggests response inhibition and WM are related to visuospatial memory skills. CaD negatively correlated with informant-reported cognitive decline on the DLD.
Burt et al., 2005	USA	Cross-Sectional	N/A	DS= 78 oID= 90	BSRT; EOWPVT-R; MfO; TSI; PPT; BBDT-VMI; BD; ScIB; DSAD; FPT; m-CF PPVT-R; A-SICD; LIPS; BSID; Shoebox task and Shoebox delayed; M-DR; M-SR; DMR; RSMB; PIMRA; Memory Problems Checklist; RSMB	Scores on PPT for using both hands related to executive functioning difficulty. Low scored on cognitive tests associated with dementia diagnosis (clinician judgement) if slope scores are used.	Grooved pegboard could not be administered due to floor effects.

Author, Year	Country	Type of study	Follow-up	Sample size (N)	Tests included	General findings	Psychometric/ acceptability findings
Carr & Collins, 2018	UK	Longitudinal	Cognitive change over 50 years	DS= 22	LIPS; BPVS; WPPSI; RBMT-C; NAID	NAID test scores reduced over time even for those without dementia diagnoses. Isolated dementia-related change from ageing-related change. Isolated differences between age-related and dementia-related changes in DS, finding verbal skills to be relatively unchanged	Participants with dementia by age 50 and/or profound ID unable to complete NAID.
Carvalho et al., 2018	Brazil	Cross-sectional	N/A	30	MoCA; ABCD (Portuguese); IQCODE; L-IADL; FAQ; Katz-IADL	DS performed similarly to people with AD in the TD population, although DS scored higher on episodic IM tests. Significant positive correlation between scores on the Lawton-IADL (functioning) and scores on Mental State, Episodic Memory, Linguistic Comprehension and Total ABCD.	Performance on ABCD correlates with indices of functioning. Performance on the MoCA was variable - highest scores falling far below the cut-off score for cognitive impairment in the TD population.
Cooper et al., 2016	UK	Longitudinal comparison RCT	1 year	21 at baseline, 13 completed (mild-severe ID)	MfO (NAID); CT; ABS ; PR memory (CANTAB); CaD; TOL; CRT; CFT; StoryRT (adapted from RBMT-C)	N/A	CANTAB PR showed floor effects at baseline, Participants showed difficulty completing CaD and StoryRT. MfO, CFT and CRT easy to complete and sensitive to change over time. TOL showed no floor effects but less sensitive to change. Cognitive testing more sensitive than informant-rated adaptive behaviour score.
Cosgrave et al., 1998	Ireland	Cross-sectional	N/A	Moderate ID: DS-Dementia = 19 DS-no-Dementia = 29 Severe ID: DS-Dementia = 11 DS-no-Dementia = 11	TSI; DSMSE	Moderate ID DS-dementia group scored significantly lower on TSI and DSMSE, compared to moderate ID DS-no dementia. Moderate ID, Severe ID and no dementia groups showed significant differences between each other on TSI and DSMSE scores.	TSI-Reliability 0.89. TSI showed sensitivity to change over time in severe ID, and wider score range than DSMSE, suggesting more appropriately scaled. Severe dementia and moderate ID were unable to score on TSI or DSMSE. 91% of moderate ID with no dementia unable to score on delayed memory task of TSI. Suggests short-term memory decline as early indicator.
Crayton et al., 1998	UK	Cross-sectional	N/A	70 (younger group and older group. Older group= 40+)	BPVS; VABS; CAMDEX; CODB	Younger group performed significantly higher on memory tests than older group.	Younger (under 40 yrs) participants showed significant negative correlations between VABS and all cognitive tests, especially orientation and memory, suggesting effect of pre-existing global cognitive impairment. Floor effects found for most tests with some participants. 18.6% of participants could not complete baseline.

Author, Year	Country	Type of study	Follow-up	Sample size (N)	Tests included	General findings	Psychometric/ acceptability findings
Das et al., 1995	Canada	Cross-sectional	NA	young-oID= 16 young-DS = 16 old-DS = 16 old-oID= 15	WAIS/WAIS-R; CAS battery; SBIS; MAT; DRS; PPVT-R-Form M;	Faster cognitive decline in DS group than oID. High correlation between IQ and DRS score in older groups only. old-DS performed worse on all tasks, particularly low scores on tasks requiring planning and attention. DS groups scored poorly in verbal expression tasks.	Floor effects on FM for most participants. High skew in MAT scores indicating inappropriately high difficulty. CAS subtests showed floor effects particularly for older participants. PPVT-R scores effectively discriminated between older and younger groups, but no indication of whether this indicated dementia-related or age-related decline sensitivity.
Das & Mishra, 1995	Canada	Cross-sectional	N/A	DS = 31 oID= 41	CAS	PWDS 40+ show difficulties in articulation, PWDS 50+ show difficulty in task planning and attention. Generally, PWDS show difficulty in phonological encoding and verbal short-term memory, perhaps related to 'phonological loop' impairment.	N/A
De Vreese et al., 2021	Italy	Cross-sectional	N/A	DS= 46 oID= 165	s-PCFT (Italian); VABS; AFAST; DLD	Significantly lower scores on orientation and memory subtests of s-PCFT.	No significant difference in s-PCFT scores by age, but significantly lower scores for participants with cognitive decline vs. without. s-PCFT showed wide range of scores. No floor effects reported (especially language and praxis) and minimal ceiling effects for all tests. High internal consistency, good inter-rater reliability and test re-test reliability (intraclass correlation coefficients of 0.85 and 0.90]. Acceptable concurrent validity between s-PCFT and DLD.
Devenny et al., 1996	USA	Longitudinal	3-5 years	DS = 28 oID = 18	EMS; BSRT; VisMT	No functional deterioration or age-related memory decline in adults with DS.No participant groups showed significant changes in test scores between baseline and follow up across 5 years. All groups showed higher scores in tasks from first to second testing period.	All participants could answer most questions on the EMS. High test-retest reliability for BSRT.

Author, Year	Country	Type of study	Follow-up	Sample size (N)	Tests included	General findings	Psychometric/ acceptability findings
Devenny et al., 2000	USA	Longitudinal	4+ years apart	noAD-DS = 44 possible-AD-DS = 10 Early-AD-DS = 5 Middle-Stage AD-DS = 7 oID= 40	WISC-R; CRT; BSRT	Pattern of decline beginning with memory and scores on Coding, BD, Object Assembly, Arithmetic, Picture Completion from healthy to middle-stage-AD in DS. Later stages, decline on vocabulary, digit span and information subtests (WISC-R) seen.	Picture Arrangement and Similarities subtests on the WISC-R showed floor effects at baseline.
Devenny et al., 2002	USA	Longitudinal	2 years+	oID= 66 DS-no AD = 75 DS-AD = 19	CRT-M	DS-AD significantly lower scores than DS-no-AD group. Scores negatively related to IQ and age. Poor performance on CRT-M associated with early-stage-AD.	Cut-off of ≤23 on total score gave sensitivity of 94.7% and specificity of 93.9% with a positive predictive value of 81.9% when comparing DS-AD with oID-no-AD. DS-AD group could name a non-test item within same category when given a semantic cue, indicating preserved semantic knowledge.
De Sola et al.2015	Spain	Cross-sectional	N/A	89 DS	K-BIT; TESTAD: (CANTAB-MOT; CANTAB-PAL; CANTAB-PRM; CANTAB-SSP;DSF;CRT-M; WST; TT; D-span backward; BNT; CaD; TOL^{DX})	Language impairment may be pre-existing in PWDS (Receptive more preserved). Higher scores in visuo-spatial CANTAB tasks.	Floor effects seen for WST, SSP, visual backward and digit span backward. Ceiling effects seen in CaD and CRT.
Doty et al., 1984	USA	Longitudinal	6 months	1,600 TD	UPSIT, WMS	Age-related changes in olfactory function found. UPSIT scores distinguished between participants with olfactory disorders and controls.	Test-retest reliability established. UPSIT did not correlate with WMS scores.
Doty et al., 1996	USA	Cross-sectional	N/A	198 TD	UPSIT, CC-SIT	12 odour items from the UPSIT were used to develop the CC-SIT. Norm data established.	Scores did not differ between participants of north American, European, South American or Asian cultures. Indicates cross-cultural acceptability.
Fonseca et al., 2014	Brazil	Longitudinal	14-22 months	18	IQCODE; NI; CAMCOG	87% probability of cognitive decline when accompanied by experienced bereavement.	Both IQCODE and CAMCOG can support assessment of cognitive decline in DS. Floor effects seen in CAMCOG and some IQCODE items. Some IQCODE items not relevant/ not ecologically valid to PWID (e.g. paying bills)
Fonseca et al., 2019a	Brazil	Cross-sectional and longitudinal	N/A	DS= 70 DS-AD = 11 DS-MCI = 18	CAMCOG-DS; CAMDEX-DS (in Brazillian)	N/A	CAMDEX-DS comparable to 'gold standard'- diagnostic accuracy of 96.7%. Shows good inter-rater reliability (kappa of >0.8 for 93% of items). CAMDEX-DS consistent with CAMCOG

Author, Year	Country	Type of study	Follow-up	Sample size (N)	Tests included	General findings	Psychometric/ acceptability findings
							(probability of a participant with dementia showing cognitive decline of 83%).
Fonseca et al., 2019b	Brazil	Cross-sectional	N/A	DS= 70 DS-AD = 11 DS-MCI = 18	CAMDEX-DS; CAMCOG-DS; WASI; CEFA; FSBS	FSBS Informant ratings of disinhibition and executive dysfunction associated with stage of dementia. Negative association between direct EF test scores and informant-rated executive dysfunction scores. Significantly higher odds ratio of AD with higher FSBS score. Apathy may also be important early indicator of AD for PWDS.	Non-verbal participants and those with advanced dementia could not complete the CAMCOG-DS.
Fortea et al., 2020	Spain	Cross-sectional	N/A	Healthy DS = 257 DS-MCI = 48 DS- AD = 83 TD controls = 242	CAMDEX-DS; CAMCOG-DS (both Spanish)	Decline in CAMCOG-DS were found in ages 50+ starting age 40+. Can detect MCI (prodromal) diagnosed at median age of 50.2 years (IQR 47.5–54.1) and AD at median 53.7 years (49.5–57.2).	N/A
García-Alba et al., 2017	Spain	Cross-sectional	N/A	DS= 63	TOL^{DX} ; K-BIT-2; ABS-RC:2; CAMDEX-DS; BT-intellectual disability; WST; BRIEF-P	N/A	Satisfactory (sensitivity = 0.76 and specificity = 0.81) psychometric properties of TOL^{DX} for ID shown, no floor effects. TOL^{DX} highly associated with other EF measures and an distinguish between mild and moderate ID participants. Scores imply inappropriate scaling of test items in TOL^{DX}
García-Alba et al., 2019	Spain	Longitudinal	3 times over 3 years	DS = 41 DS-AD = 13 DS-MCI = 14 DS-Control = 14	CAMCOG-DS; ADVM; WM; DVM; TO	DS-AD significantly poorer scores on all tests, especially in delayed visual memory and WM compared to DS-controls.DS-AD showed poorer scores vs. DS-MCI group in WM and verbal memory. DS-MCI scored poorly vs. DS-controls on CAMCOG-DS and DVM. Global deterioration (overall decline) may characterise progression from MCI to AD in DS.	Floor effects found for severe-profound ID.
Hartley et al., 2020	USA	Longitudinal	4-5 time points across 1-8 years	(Drop-out across time points) Time 1: Healthy DS= 109, MCI-DS= 9	CRT-M; CaD, SCT; BD; PP	CRT-M associated with increased neocortical APP, able to identify transition from preclinical to MCI AD (detected elevated APP as measured by Pittsburgh Compound-B).	CRT-M sensitive to preclinical and prodromal AD decline in DS.

Author, Year	Country	Type of study	Follow-up	Sample size (N)	Tests included	General findings	Psychometric/ acceptability findings
				Time 2: Healthy DS= 101 MCI-DS= 8, DS-DAT= 6; Time 3: Healthy DS= 53, MCI-DS= 7, DS-AD= 3, Time 4: Healthy DS= 37, MCI-DS= 7, DS-AD= 2, Time 5: Healthy DS= 12, MCI-DS= 4, AD-DS= 1			
Head et al., 2011	USA	Cross-sectional	N/A	Study 1: DS-no-AD = 17 DS-AD = 17 TD control= 11 AD controls = 12 Study 2: DS-AD = 52 DS-no-AD = 78	BPT; SIB; DMR	No association between scores on SIB and DMR, nor with blood plasma amyloid levels.	Lack of sensitivity of SIB and DMR to detect dementia or cognitive decline in DS.
Hon et al., 1999	UK	Cross-sectional	N/A	DS aged 30–44 = 45 DS 45+ = 29	CAMDEX; CAMCOG; MMSE	45+ group scored more poorly than other groups on all tests except attention and calculation.	CAMCOG showed floor effects for 11% of participants due to severe ID, sensory impairments, and/ or severe dementia. MMSE had narrow range of scores, higher floor effects, than CAMCOG.
Hutchinson & Oakes, 2011	UK	Cross-sectional	N/A	DS= 37	SIB; DLD	N/A	Good concurrent criterion validity with cognitive component of DLD (-0.73). SIB showed few floor effects and some ceiling effects in healthy PWDS.
Jozsvai et al. 2002	Canada	Cross-sectional	N/A	DS-AD= 12 DS-no-AD=23 (age >40= 9, age 40+= 14)	MMSE-O; PPVT-R; DSDS; GL; BNT; PX; FOME; Information and Orientation tasks	BNT and BD scores most affected by age (increasing age= lower scores). These tests unlikely to be sensitive to dementia decline as similar scores between older participants regardless of dementia status.	MMSE-O and FOME scores lower in DS-AD 40+ group. No difference in older group between BNT and BD scores; low clinical applicability in dementia diagnosis. FOME identified as most sensitive (and orientation).
Kay et al., 2003	UK	Cross-sectional	N/A	85 (PWID mild, moderate and severe).	PCFT & ABS	N/A	Participants with severe ID scored at floor on PFCT. Range of scores seen across ID severity groups. High correlation between PCFT and ABS scores (0.87). All PCFT tests valid for mild-severe ID except PCFT recall.

Author, Year	Country	Type of study	Follow-up	Sample size (N)	Tests included	General findings	Psychometric/ acceptability findings
Krinsky-McHale, Devenny & Silverman, 2002	USA	Longitudinal	3 or 3+ over 3 years	DS-AD = 14 DS-no-AD = 71	BSRT;MMSE-O; BD; FOME; DSDS;	Early-stage-AD showed significantly lower LTM and retrieval performance prior to other symptoms 3 years before diagnosis.	Lower scores in BSRT could distinguish between AD and no-AD groups. BSRT sensitive to age-associated decline in verbal explicit memory related to DS.
Krinsky-McHale et al., 2008)	USA	Longitudinal	At least 3 across 3 years	Total = 30 DS-AD = 5, Non-AD DS = 25	DSDS; mSRT; SCT	Progressive decline in selective attention up to 2 years prior to diagnosis of MCI in non-AD DS.	SCT was easy to administer and showed good sensitivity and specificity. Differences in SCT performance related to dementia severity.
Krinsky-McHale et al., 2020	USA	Longitudinal	14- to 22-month intervals	DS= 561	mSRT; mMMSE-DS; TSI; M-CFT; WISC-R-blocks tests; DSMSE; DSMSE (BLOCK-T); DLD; ABSI; RSMB; VMI; NI; CUSPAD; BBDT-VMI	Several measures showed ability to distinguish between MCI and dementia (diagnosed at follow up intervals).	Floor effects seen for majority of tasks in sample (severe ID).
Masson et al. 2010	UK	Cross-Sectional	N/A	43 PWID of mixed aetiology	WASI; adapted TOL; DEX-IR; ABS-RC:2	TOL correlated negatively on DEX-IR and positively with ABS-RC:2 scores, indicating good clinical utility and validity in detecting EF dysfunction.	TOL showed good scale of difficulty (all participants scored on item one, 9 on final item).
Manning et al., 1998	USA	Longitudinal	2 time points (2 weeks between)	DS= 21	PPVT-R; DSMSE; RAVLT; DSpan; OPS	Scores declined with age on all tests, except apraxia subtest of DSMSE. PWDS given glucose before DSMSE completion showed significantly higher scores in most tests of verbal ability and memory and overall score than control group.	N/A
Margallo-Lana et al., 2003	UK	Longitudinal	4 weeks	DS= 14	PCFT	Non-specialists able to administer.	Extremely high inter-rater reliability for detecting cognitive deterioration and very high test-re-test reliability (both 0.99, $p < 0.01$). High reliability and temporal stability. Floor effects for BD which was removed from the PCFT.
Margallo-Lana et al., 2007	UK	Longitudinal	15 years	92	PCFT; ABS	Participants who scored lower on the PCFT later received a dementia diagnosis. Indicates clinical utility. This increased with age.	PCFT has utility for those with mild to moderate ID, but not severe, due to floor effects at baseline. Score decline in all PCFT subtests associated with diagnosis of dementia.
McCarron et al., 2014	Ireland, USA	Longitudinal	Every year for 14 years	DS= 77	DSMSE; DLSQ; DMR; TSI	Mean age of dementia diagnosis = 55.41 years. Median survival rate after diagnosis= 7 years.	Decline on TSI scores and DSMSE seen one year before diagnosis. Rate of decline on TSI and DSMSE more gradual after diagnosis given. DMR most sensitive to change over time, up to 5 years before diagnosis.

Author, Year	Country	Type of study	Follow-up	Sample size (N)	Tests included	General findings	Psychometric/ acceptability findings
McCarron et al., 2017	Ireland	Longitudinal	20 years (linked to McCarron et al. 2014)	DS= 77	DLSQ; DQPID; DSMSE; TSI; DLD	97.4% of participants diagnosed with dementia at 20 year follow up (mean diagnosis age 55 y/o). Dementia risk not related to ID severity. Dementia associated with cognitive and functional decline and seizures. AD risk 23% risk at age 50 years, 45% at age 55 years and 88% risk at age 65 years.	Several instruments (especially DQPID), showed a gradual decline in scores 1 year before dementia onset. Informant based measures seem more sensitive than direct measures. Direct measures show less sensitivity (change over time) after dementia diagnosis. Difficult to profile further decline.
Nelson et al., 2001	USA	Longitudinal	1 year	DS= 26	NBAP; DSDS; PPVT-III	Atrophy/dysfunction of frontal lobe may indicate early AD in DS. Pragmatic language decline shown after symptoms of depression/apathy (indifference). Participants with abnormal physical findings (atrophy and ventricular enlargement of prefrontal lobe on MRI and pathological reflexes during neurological examination) scored significantly lower on DSDS memory and PPVT receptive language.	NBAP pragmatic communication scale reliably detected early signs of probable AD (MCI).
Nelson et al., 2005	USA	Cross-sectional	NA	DS= 20	WAIS-III; DMR; WGTA; ODL; RL; DNMP; DNMS	Object memory scores predicted by FSIQ. Scores on DMR strongest predictor of reversal learning errors. Age associated with learning and memory scores.	Many could not complete DNMP spatial task- possible floor effects.
Nelson et al., 2007	USA	Longitudinal	1 year	DS=34 at baseline; 19 at follow up	WAIS-III; NBAP; DMR; SVDL; RL; DNMS; ASL or landmark	NBAP reported as the strongest predictor of dementia status.	DMNS may show floor effects- only one participant could score. Strong correlation between DMR and pragnosia (defined by Nelson as 'communication style deficits) scores. All tests showed high reliability and validity.
Oliver et al., 1998	UK	Longitudinal	At 6, 13, 20, 25 and 50 months	DS= 57	BVPS; VABS; VMT adapted from MST; MfO; MfPT; CODB	After age 30, 28.3% of participants showed cognitive deterioration; increased with age and ID severity. Orientation, learning and memory deficits first to appear, then in apraxia, agnosia and aphasia.	N/A
Oliver et al., 2005	UK	Longitudinal	0, 6, 13, 20, 25 and 50 months	DS-CognitiveDeterioration = 12 DS-no-CognitiveDeterioration <40 = 19 DS-no-	BPVS; VABS; Orientation task; Picture naming task; Action on Request; CRT; CANTAB	Dementia status associated with decline in delayed response and associative (conditioned) learning tasks.	Floor effects found for tasks at early follow up stages.

Author, Year	Country	Type of study	Follow-up	Sample size (N)	Tests included	General findings	Psychometric/ acceptability findings
				CognitiveDeterioration >40 = 21			
Palmer, 2006	USA	Cross-sectional	N/A	Dementia = 10 (DS = 6; oID= 4) No-dementia = 12 (DS = 4; oID= 8)	CTT; BNT; COWAT; FOME; ESDCL;	Low scores on ESDCL in dementia group. Scores were poorer on tasks of memory and learning in the dementia group. Similar to areas seen in AD for TD individuals. BNT scores, animal naming and FOME memory scores lower in dementia group. Suggests tests associated with dementia-related decline.	Floor effects for CTT noted for participant with dementia (could not complete). Cut-off scores identified.
Powell et al., 2014	USA	Cross-sectional	N/A	DS-no-dementia = 10 DS-dementia = 10 oID= 10	BPT & SIB	Low scores on BPT related to frontotemporal atrophy, reduced white matter integrity (myleniation).	BPT sensitive to functional decline (through BPT scores) in DS individuals.
Poveda & Broxholme, 2016	UK	Longitudinal	5-40 months	55 (DS, old) 15 participants had 'probable' dementia, 31 no dementia, 9 unsure.	LDDB; VABS; DMR; BPVS	After follow up, 'probable' group performed worse (not significant), 'no dementia' group improved significantly in LDDB score at follow up. Language subtest may distinguish between no and probable dementia.	Change over time on LDDB shown only for PWDS. Decline in LDDB scores associated with DMR increase score (showing decline)
Pulsifer et al. 2020	USA	Cross-Sectional	N/A	DS= 168	BNT; M-CFT; VABS	Decline in language scores (VABS) related to AD dementia status in early stages (MCI).	BNT scores significantly decrease with age, but not significant predictor of dementia status via logistic regression. M-CFT strong indicator of MCI.
Pyo et al., 2007	USA	Cross-sectional	N/A	DS-AD = 13 Healthy-DS= 31	WG - MTB (mFOME; TSI; AMT; Orientation); PPVT-III; PRMT;Comprehension Test (NEPSY); TO	AD-DS group scored lower than healthy-DS on WG-AMT and WG-Orientation. No differences between groups found on TSI total score or immediate and delayed memory subtest scores.	WG-AMT and O tasks useful for studying ageing in moderate- severe ID, but not sensitive enough to distinguish (significant group overlap). mFOME too difficult for moderate to severe ID. Free recall test too difficult for all participants with low verbal comprehension skills. PRMT could distinguish DS-AD from healthy Ds in most cases. No significant difference in TSI scores between groups. May not be sensitive enough to detect early AD.

Author, Year	Country	Type of study	Follow-up	Sample size (N)	Tests included	General findings	Psychometric/ acceptability findings
Pyo et al., 2010	USA	Cross-sectional	N/A	AD = 26 No-AD = 33 healthy DS = 9 healthy oID= 24 DS-AD = 15 oID-AD = 11	r-PRMT; mFOME; TSI; NEPSY	N/A	Healthy DS scored higher than DS-AD on r-PRMT, with no overlap. Healthy oID scored much lower than oID-AD with significant overlap. r-PRMT scores could discriminate between healthy DS and DS-AD, but not in other ID groups. TSI could not discriminate between AD and no AD groups.
Pyo et al., 2011	USA	Longitudinal	12 months	DS and oID-AD = 21 DS and oID-no-AD = 42	WG-AMT	AD group scored more poorly than control group at baseline. Scores for people with DS-no-AD decreased significantly at follow up.	Mny participants could not answer their age correctly. Working Group's AMT may be useful as dementia screening tool in moderate to severe ID and DS, but needs further validation. Unclear whether reliable dementia screening tool for moderate to severe oID. Limited score variability – requires modification.
Sano et al., 2005	USA	Cross-sectional and longitudinal	Multiple time points	316	MMSE-O; VT adapted from EOWPVT-R BPT; NDT; mFOME	Decline in memory and verbal learning were highly associated with dementia status.	mFOME useful regardless of ID severity. Orientation and visual memory showed floor effects in moderate-severe ID. High accessibility of BPT, minimal floor/ceiling effects, BPT sensitive to change over time. Vocabulary score sensitive to dementia status but not ID severity.
Schmitt et al., 2010	USA	Cross-Sectional	N/A	TD= 103	UPSIT; RBANS	UPSIT may be unaffected by premorbid functioning.	Significant moderate correlation for UPSIT scores and RBANS total, delayed memory index and language index. UPSIT scores not correlated to IQ scores.
Shultz et al., 2004	USA	Cross-sectional	NA	oID = 38 DS = 26 oID+DS-dementia = 19 oID+DS-no-dementia = 19	DSDS; DMR; RSMB; S-MMSE; DHQ, PAL	Dementia groups showed poorer scores than no dementia groups.	MMSE and paired-associate learning task related to IQ and dementia, but not age or gender. DSDS and DMR seem sensitive to dementia-related decline; unrelated to ID severity, age or gender. Both scores could differentiate between groups.
Sinai et al., 2016	UK	Cross-sectional	N/A	49 PWID (aged 45+). Dementia and no-dementia groups.	ACTB= TOL; object memory (NAID); Modified Dots; VF; F-NT; GA; BRIEF; K-BIT-2; NEPSY Visuomotor Precision; DLD; CANTAB Intra-Extra Dimensional shift and Paired Associates	N/A	Participants could attempt most ACTB tasks. Significant differences between dementia and no-dementia groups on CANTAB Simple Reaction Time median latency, NEPSY Visuomotor Precision and CANTAB Paired Associates Learning. Floor effects shown for CANTAB Intra-Extra Dimensional shift

Author, Year	Country	Type of study	Follow-up	Sample size (N)	Tests included	General findings	Psychometric/ acceptability findings
					Learning tasks		stages completed and Modified Dots Task. No significant difference on TOL between groups.
Tabert et al. 2005	USA	Longitudinal	Mean= 42 months	TD control= 63 TD-MCI= 147 TD-AD= 100	MMSE; UPSIT; BSIT	Both UPSIT and BSIT showed discriminative ability between MCI and AD groups. Incorrect responses on BSIT related to AD risk. UPSIT and BSIT both significantly predicted progression from MCI to AD.	10 most sensitive items on the UPSIT identified as valid for inclusion into the BSIT.
Tyrer et al. 2010	UK	Cross-sectional	N/A	ID(undefined)=168	PFCT; s-PCFT; K-BIT	N/A	PFCT reliable for people with severe ID. Scores lower for PWDS who later receive a diagnosis of dementia. Not as sensitive to dementia-related decline in severe ID. Long administration time. Extremely high validity of both short and long versions (high correlation with K-BIT)
Walsh et al., 2015	USA	Cross-sectional	N/A	114 PWDS (62% with dementia)	RADD; DMR; BADLS; SIB; BPT	Participants with dementia scored poorly on all measures in comparison to participants without dementia.	Dementia and non-dementia participants with profound ID performed at floor level. High test-retest reliability (0.95, $p<0.001$) of RADD. Good criterion validity (0.67, $p=0.001$). High sensitivity (0.87) and specificity (0.81) in differentiating dementia vs. no dementia. Sensitivity and specificity lower for for those diagnosed over 2 years ago (0.73).
Webb et al. 2020	UK	Cross-sectional	N/A	101 (no DS)	BADS-ID & BADS-C; both with differing adaptations of tests in the BADS: 1) Rule Shift Card Test, 2) Key Search Test, 3) Temporal Judgement Test, 4) Zoo Map Test, 5) Action Program Test and 6) Modified Six Elements Test. (Comparison with CEFA data from Willner et al. 2010)	N/A	Simplified instructions for rule shift and action program tests were better understood by participants in BADS-ID compared to BADS-C. Reflected in reduced floor scored for this task in BADS-ID. Supermarket task raised floor effects from 87.5% to 2.8%, and significantly increased the proportion of ceiling effects from 2.5% to 59.2% for the second trial in comparison to BADS-C. The temporal judgement test was considered to assess cognitive estimation, rather than time factored into planning everyday tasks related to EF and was removed in BADS-

Author, Year	Country	Type of study	Follow-up	Sample size (N)	Tests included	General findings	Psychometric/ acceptability findings
							IDBADS-ID internal consistency, face validity and inter-rater reliability comparable to the CEFA.
Willner et al. 2010	USA	Cross-Sectional	N/A	40	BADS-C; CEFA;BPVS; WASI	Overall scores on CEFA and BADS-C very weakly related to IQ and verbal ability (BPVS). Factor structure of EF may be tripartite.	Many participants scored at floor for 3 of 6 BADS-C subtests, including zoo map and WST. CaD not related to BPVS, TOL related to PBVS. CaD and VF easy to administer.
Witts & Elders, 1998	UK	Cross-sectional	NA	33	SIB; VABS	N/ A	SIB shows high test-retest reliability and criterion validity of SIB and no floor effects. SIB may be good for longitudinal use for PWDS of varying ID severity.

Note. < = less than; > = more than; AADS = Assessment for Adults with Developmental Disabilities; ABCD battery = Arizona Battery for Communication Disorders of Dementia; 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= Alzheimer's Disease Dementia; ADVM = Auditory delayed verbal memory; AFAST = Alzheimer's Functional Assessment Tool Scale for informants; AMT = Autobiographical Memory Test; ApoE = Apolipoprotein E; A-SICD= Sequenced Inventory of Communication Development for Adolescents and Adults with Severe Handicaps; ASL or landmark = Allocentric spatial learning or landmark; ASM = Auditory sequential memory; 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; BSID = Bayley Scales of Infant Development; BSIT= Brief Smell Identification Test; BSRT = Busckke selective reminding test and modified versions; BT-ID = Barcelona Test-ID; BTS = Block tapping span; CaD = Cats and Dogs; CAL = Conditioned associative learning; 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; CAS = Das–Naglieri Cognitive Assessment System; CEFA = Cambridge Executive Functioning Assessment; CFT = Category Fluency Test; CLD = Checklist with Symptoms of Dementia; CO = Colour Ordering; CoD = Copy of drawings; COWAT = The Controlled Oral Word Association Test; CRT = The Cued Recall Test; CRT-M = Cued Recall Test-Modified; CTT = The Colour Trails Test; CUSPAD = Columbia University Scale to Assess Psychopathology in Alzheimer's Disease; DHQ = Demographic health questionnaire; DLD/ DMR = Dementia Questionnaire for People with Learning Disabilities; DLSQ = Daily Living Skills Questionnaire; DM = Delayed memory; DMTS = Delayed match-to-sample; DNMP = Spatial delayed non-match-to-position; DNMS = Object delayed non-match-to-sample; DQPID = Dementia Questionnaire for People with Intellectual Disabilities; Dresponse = Delayed response; DRS = Dementia Rating Scale; DS = Down syndrome; DSDS = Dementia scale for Down Syndrome; DSMSE = Down Syndrome Mental State Examination; DSpan = Digit span; DSpan-B = Digit Span – backwards; DSpan-F = Digit Span – forwards; DSQID = Dementia Screening Questionnaire for Individuals with Intellectual Disabilities; DVM = Delayed visual memory; DSAD= Dyspraxia Scale for Adults with Down Syndrome; ECTs = Experimental Computerised Tasks; 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-NT = Finger-Nose Test; FAQ = Pfeffer Functional Activities Questionnaire; FM = Figure Memory; FOME = The Fuld Object-Memory Evaluation; FPT= Fragmented Picture Test; FS = Finger Sequencing; FSBS = Frontal Systems Behavior Scale; FSBS = Frontal Systems Behavior Scale; FSIQ = Full-Scale IQ; GA = Gait Assessment; GC = Grammatic Closure; GL= Grocery List task; H-NTLA = Hiskey-Nebraska Test of Learning Aptitude; HADS = Hospital Anxiety and Depression Scale; HC = Healthy controls; HOM = Hidden Object Memory Test; HSSA = Hampshire Social Services Assessment; IBR-MSE = Mental State Examination from the New York Institute for Basic Research; IM = Immediate memory; IQ = Intelligent quotient; IQCODE = Informant Questionnaire on Cognition Decline in the Elderly; IQR = Interquartile range; ITPA = Illinois Tests of Psycholinguistic Ability; Katz-IADL = Katz Index of Independence in Activities of Daily Living; K-BIT = Kaufman Brief Intelligence Test ;LDDB; Learning Disabilities Dementia Battery; L-IADL = Lawton Instrumental Activities of Daily Living; LIPS = Leiter International Performance Scale; LT = Long-term; LTM = Long-term memory; M-CFT = McCarthy Category Fluency Test; M-DR= McCarthy Digit Recall; M-SR= McCarthy Sentence Recall; MAT = Matrix Analogies Test-Expanded Form; MCI = Mild cognitive impairment;; ME = Manual Expression; MEAMS= Middlesex Elderly Assessment of Mental State; MfO = Memory for objects; MfPT = Memory for pictures; MfS = Memory for sentences; MSE= Mental State Examination (traditional); mMMSE-DS = Modified Mini Mental Status Evaluation— Down Syndrome; MMSE = Mini Mental Status Evaluation/Modified Mini Mental Status Evaluation; MN = Matching Numbers; MoCA = The Montreal Cognitive Assessment; mOMT = Modified Objective Memory Test; MSCA = McCarthy Scales of Children's Abilities; MtS = Matching-to-Sample; N/A = Not applicable; NBAP; Neuropsychology behavior and affect profile; NEPSY = A Developmental NEuroPSYchological Assessment; oID = Intellectual disability from other causes than DS; PIMRA= Psychopathology Inventory for Mentally Retarded Adults; PPVT/ PPVT-R = Peabody Picture Vocabulary Test/ Peabody Picture Vocabulary Test-Revised; PR = Pattern recognition; PRMT/r-PRMT = Picture Recognition Memory Test/ revised Picture Recognition Memory Test; PWDS= People with Down Syndrome; PWID= People with an intellectual disability; QoL = Quality of Life; RA = Receptive Attention;

RADD/RADD-2 = The Rapid Assessment of Developmental Disabilities/Second Edition; RAVLT = Rey Auditory Verbal Learning Test; PX= Test of Apraxia; RBMT-C = Rivermead Behavioural Memory Test for Children; RCPM = Raven Coloured Progressive Matrices; RL = Reversal learning; RSMB = Reiss Screen for Maladaptive Behaviour; S-MMSE = Shultz Mini Mental State Exam; SAE = Selective Attention-Expressive; ScIB= Scales of Independent Behaviour; SB = Scrambled Boxes; SBIS = Stanford-Binet Intelligence Scales; SCT = Symbol Cancellation task; SIB = Severe Impairment Battery; SR = Sentence Recall; Srecog = Spatial recognition; Srep = Sentence repetition; Sreversal = Spatial Reversal; SRT/mSRT = The Selective Reminding Test/ Modified - The Selective Reminding Test; STM = Short-term memory; SVDL = Simple visual discrimination learning; TACL-III = Test of Auditory Comprehension of Language-3; TBGAT = Tinetti Balance and Gait Assessment Tool; TD = Typically Developing; TO = Temporal Orientation; ToL = Tower of London; TOL^{DX} = Tower of London-Drexel University: 2nd Edition; TSI = Test for Severe Impairment; TT = Token Test; UPSIT= University of Pennsylvania Smell Identification Test; VABS = Vineland Adaptive Behaviour Scales; VAT = Visual Association Test; VF = Verbal Fluency; VisMT = Visual Memory Test; VMT = Verbal Memory Test; VS = Visual Search; VSM = Visual sequential memory; VT = Vocabulary Test; WAIS/WAIS-R = Wechsler Adult Intelligence Scale/Revised; WASI = Wechsler Abbreviated Scale of Intelligence; WST = Weigl Colour-Form Sort Test; WG-AMT= Working Group's Autobiographical Memory Test; WGTA = Wisconsin General Testing Apparatus; WISC/WISC-R = Wechsler Intelligence Scale for Children/Revised; WM = Working Memory; WMS= Wechsler Memory Scale; WPPSI/WPPSI-R = Wechsler Preschool and Primary Scale of Intelligence/Revised; WR = Word Recall

Table E 2

Sample Characteristics

Author, Year	Subtype of ID	ID Severity	Criteria/ method used for assessing level of ID	Comparator / control group	Dementia subtypes (if included)	Diagnostic criteria for dementia	Mean age (years, *SD*) and/ or range (in years)	% female	Verbal ability (for inclusion in sample)
Adams & Oliver, 2010	DS	NR	NA	Longitudinal	N/A	N/A	44.5 (7.5) 34-64	50.0%	All verbal (at least single word responses)
Alexander et al., 1997	DS	NR	NR	Between age groups	N/A	N/A	22-61	46.3%	NR
Ball et al., 2004	DS	NR	NR	Longitudinal	AD	CAMDEX	NR	41.9% at Time 1	NR
Ball et al., 2008	DS	Mild = 35.2% Moderate = 49.2%	ICD-10 and BPVS-II	Between groups (AD and no-AD)	AD	CAMDEX-DS	49 (36-72)	42%	NR
Ball et al., 2010	DS	Mild = 40% Moderate = 60%	ICD-10	Between test methods (cognitive and behavioural)	AD	CAMDEX-DS	46.7 (36-72)	41%	Excluded severe-profound ID
Benejam et al., 2015	DS	DS-noAD= 33% mild, 67% moderate DS-AD= 93% moderate, 7% severe	K-BIT and informant report for DS-AD group	Between groups (AD and no-AD)	AD	NR	DS = 36.1 (9.8) DS-AD = 51.1 (5.1)	DS = 44% DS-AD = 60%	NR
Benejam et al., 2020	DS	CAMCOG-DS group = 91 mild, 205 moderate 47 severe CRT-M group= 85 mild, 161 moderate, 25 severe	DSM-V and K-BIT	Between groups (No-AD, MCI, and AD)	AD	CAMDEX-DS	CAMCOG-DS group = 41 (18.5) mCRT group = 39 (18.0)	CAMCOG-DS group = 49.1% mCRT group = 47.2%	NR
Bevins & Hurse, 2014	DS + oID	mild-moderate ID	BVPS-II	Between instruments	AD	NR	49.5 (9.32) 21–66	57.1%	Participants scored 13–120 on BVPS-II
Burt et al., 2005	DS +oID	Mild= 16 Moderate= 51 Severe = 23	LIPS; ICD-10	Between methods of testing (cognitive and clinical judgement)	Range of dementia (not specified)	ICD-10	30-69	NR	Non-verbal participants included- verbal tests omitted for

Author, Year	Subtype of ID	ID Severity	Criteria/ method used for assessing level of ID	Comparator / control group	Dementia subtypes (if included)	Diagnostic criteria for dementia	Mean age (years, *SD*) and/ or range (in years)	% female	Verbal ability (for inclusion in sample)
									these individuals.
Carr & Collins, 2018	DS	NR	NR	Longitudinal	N/A	N/A	N/A	40.9%	NR
Carvalho et al., 2018	DS	NR	NA	Between groups (DS and older-AD no-DS)	NR	N/A	47.8 (6.7)	40%	Sample had various levels of literacy (not specified)
Cooper et al., 2016	DS	Mild = 36% Moderate = 33% Severe = 43% Profound = 5%	NR	Longitudinal	N/A	N/A	54.15 (3.10)	48%	NR
Cosgrave et al., 1998	DS	Moderate = 80% Severe = 36.7%	Psychiatrist evaluation based on prior neuropsychological testing, caregiver reports, interviews	Between instruments and between groups (ID severity and dementia status)	NR	ICD-10	51.9 (8.7) 35-75	16.7%	NR
Crayton et al., 1998	DS	NR	NR	Between groups (age)	N/A	NR	42.8 (7.38) 28-58	55.7%	Individuals with simple word responses/ who were unable to perform simple motor commands excluded
Das et al., 1995	DS + oID	Mild-severe	Historical WAIS, WAIS-R, or Stanford-Binet scores.	Between groups (age and ID aetiology)	NR	NR	Young subgroups = 43.7 (2.9) 40-49 old DS subgroup = 55.2 (3.9) 50-62 old non-DS subgroup = 56.7 (2.9)	NR	NR
Das & Mishra, 1995	DS	NR	NR	Between groups (age and ID aetiology)	NR	NR	26-60	NR	NR
De Vreese et al., 2021	DS (46) + oID (165)	DSM-IV-TR: Mild= 26.1%	DSM-IV-TR + VABS	Comparison of instruments	NR	NR	Median (IQR) = 52 (10.0)	40.8%	NR

Author, Year	Subtype of ID	ID Severity	Criteria/ method used for assessing level of ID	Comparator / control group	Dementia subtypes (if included)	Diagnostic criteria for dementia	Mean age (years, *SD*) and/ or range (in years)	% female	Verbal ability (for inclusion in sample)
		Moderate= 46.4% Severe= 24.2% VABS: Mild= 35.5.% Moderate= 39.8% Severe= 21.3%					40-84		
Devenny et al., 1992	DS + oID	Mild-moderate	NR	Longitudinal and between groups (DS and. no-DS)	NA	NA	DS <35 = 31.7 DS >35 = 41.8 27-55	NR	NR
Devenny et al., 2000	DS	Mild-moderate	NR	Longitudinal and between groups (dementia groups)	NR	ICD-10	ID-no-DS = 53.68 ± 11.03 DS = no overall given	NR	NR
Devenny et al., 2002	DS	Mild-moderate	NR	Between groups (DS-AD, 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
De Sola et al., 2015	DS	Mild/moderate= 58.1% Severe= 41.9%	DSM-4	Between groups (gender, age, IQ)	N/A	N/A	23.3 (4.3) 16-34	48.8%	NR
Doty et al., 1984	N/A	N/A	N/A	Psychometric properties	N/A	N/A	10-99	NR	N/A
Doty et al., 1996	N/A	N/A	N/A	Psychometric properties	N/A	N/A	5-96	NR	N/A
Fonseca et al., 2014	DS	NR	NA	Longitudinal	NR	ICD-10, DSM-IV	42.44 (6.11) 35-55 (at baseline)	33.3%	14 participants were illiterate
Fonseca et al., 2019a	DS	Mild = 37.8% Moderate = 37.8% Severe = 24.4%	NR	Psychometric properties	AD	CAMDEX-DS, DSM-5, ICD-10	42.45 (8.51)	35.9%	NR
Fonseca et al., 2019b	DS	Mild = 37.8% Moderate = 37.8% Severe = 24.4%	AAIDD, WASI, ICD-10	Between groups (dementia severity)	AD	CAMDEX-DS, DSM-5 and ICD-10	42.45 (8.51)	35.9%	2 participants were non-verbal and could not complete the tests
Fortea et al., 2020	DS	Mild = 19% Moderate = 45%	DSM-V	Between groups (dementia severity)	AD	Clinician consensus	Median (IQR) Asymptomatic DS = 38.7(31.1-48.2)	DS = 45% Controls = 67%	NR

Author, Year	Subtype of ID	ID Severity	Criteria/ method used for assessing level of ID	Comparator / control group	Dementia subtypes (if included)	Diagnostic criteria for dementia	Mean age (years, *SD*) and/ or range (in years)	% female	Verbal ability (for inclusion in sample)
		Severe/ Profound = 25%					DS-MCI = 50.2 (47.5-54.1) DS-AD = 53.7 (49.5-57.2) Healthy DS = 56.6 (50.4-63.8)		
García-Alba et al., 2017	DS	Mild = 62.9% Moderate = 37.1%	DSM-V	Psychometric properties and between groups (ID severity)	N/A	CAMDEX-DS (includes criteria from the DSM-IV and ICD-10) dementia status was an exclusion criterion	All aged ≥ 39	47.6%	NR
García-Alba et al., 2019	DS	Mild-moderate	DSM-V, K-BIT, Vineland II	Between groups (between DS and no-DS & dementia status)	MCI, AD	CAMDEX-DS, adaptive skills, and clinician judgement	DS-controls = 44.64 (3.30) DS-MCI = 51.64 (3.95) DS-AD = 53.54 (6.58) Controls = 45.21 (4.39)	DS = 61.0% Controls= 71.4% DS-Control = 71.4% DS-MCI = 42.9% DS-AD = 69.2%	NR
Hartley et al., 2020	DS	NR	NR	Longitudinal	MCI, AD	Clinician consensus	Time 1= 37.24 (7.7); Time 2= 38.89 (8.09), Time 3= 42.18 (7.04); Time 4= (44.11 (7.02); Time 5= 45.77 (6.62)	Time 1, 2= 52. % Time 3, 4= 48%;Time 5= 53%	NR
Head et al., 2011	DS	Mild- Profound	Historical FSIQ scores	Between groups (dementia status and ID severity)	AD	DSM-IV	Study 1: DS-no-AD = 44.1 (1.4) 37-54 Controls = 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)	Study 1: DS-no-AD = 47.1% Controls= 54.5% DS-AD = 11.8% AD-controls = 41.7% Study 2: DS-AD = 50% DS-no-AD = 34.6%	NR

Author, Year	Subtype of ID	ID Severity	Criteria/ method used for assessing level of ID	Comparator / control group	Dementia subtypes (if included)	Diagnostic criteria for dementia	Mean age (years, *SD*) and/ or range (in years)	% female	Verbal ability (for inclusion in sample)
							41-63 DS-no-AD = 45.1 (1.9 26-60		
Hon et al., 1999	DS	Mild = 18% Moderate = 57% Severe = 20% Profound = 5%	ICD-10	Between instruments and between groups (age)	AD	ICD-10, DSM IV, CAMDEX	42.6 (8.2) 30 - 65	41.9%	NR
Hutchinson & Oakes, 2011	DS	NR	NR	Psychometric properties of instruments	N/A	N/A	38.97 (9.18) 20–58	43.2%	NR
Jozsvai et al. 2002	DS	None below moderate ID (otherwise NR)	NR	Psychometric properties between instruments and between groups (dementia status)	AD	DSDS score (compatible with DSM-IV)	DS-AD= 50.9 (5.6) Young DS-no-AD= 33.4 (3.4) Old DS-no-AD= 46.2 (5.6)	NR	All participants fell within the same general range of verbal ability.
Kay et al., 2003	DS	Mild-profound and untestable	SBIS scores in medical records	Between instruments	AD (excluded from sample)	Clinical assessment	38.2	34.1%	NR
Krinsky-McHale, Devenny & Silverman, 2002	DS	Mild-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
Krinsky-McHale et al., 2008	DS	Mild-moderate	Historical WAIS-R or Stanford-Binet or LIPS scores	Longitudinal and psychometric properties	MCI, AD	Test performance and clinical judgement	DS-AD = 51.44 (5.20) 45-58 DS-no-AD = 49.40 (4.57) 44-62 (at baseline)	NR	NR
Krinsky-McHale et al., 2020	DS	Mean FSIQ= 33.3 (severe)	Working Group Manualized comprehensive evaluation	Longitudinal and between instruments and groups	MCI, AD	Test performance and clinical judgement	51.6 (9.1) (at baseline)	NR	Could verbally assent to participate
Masson et al. 2010	Mixed	Mean FSIQ= 58.28 (4.2) (moderate)	WASI	Psychometric properties of instruments	N/A	N/A	40.58 (11.34) 19-61	30.2%	Only participants able to provide consent included.

Author, Year	Subtype of ID	ID Severity	Criteria/ method used for assessing level of ID	Comparator / control group	Dementia subtypes (if included)	Diagnostic criteria for dementia	Mean age (years, *SD*) and/ or range (in years)	% female	Verbal ability (for inclusion in sample)
Manning et al., 1998	DS	NR	NR	Longitudinal	N/A	N/A	35 (9.2) 18-55	71.4%	NR
Margallo-Lana et al., 2003	DS	Mild = 42% Moderate = 28.5% Severe = 14.2% Profound = 14.2%	Historic IQ test scores	Psychometric properties of instruments	N/A	N/A	Males =44.1 (6.7) 33-55 Females = 38.6 (4.7) 35-44	22%	NR
Margallo-Lana et al., 2007	DS	NR	NR	Longitudinal	NR	Clinical judgement, ICD-10 criteria, record reviews, neuropathology examinations	39.1 (10.7) 20-72	31.5%	NR
McCarron et al., 2014	DS	Moderate = 88.4% Severe = 21.7%	Medical records, ICD-10	Longitudinal	NR	ICD-10	NR	100%	NR
McCarron et al., 2017	DS	Moderate = 88.4% Severe = 21.7%	Medical records, ICD-10	Longitudinal	NR	ICD-10	NR	100%	NR
Nelson et al., 2001	DS	NR	NR	Between groups (normal and abnormal physical findings)	NR	NR	40.03 (11.8)	61.5%	NR
Nelson et al., 2005	DS	Mean FSIQ= 51.31 (moderate)	WAIS-III	Between instruments and between groups (age)	NR	NR	37.2 (9.5) 22–58	60%	NR
Nelson et al., 2007	DS	Mean baseline FSIQ= 51.31 (moderate)	WAIS-III	Psychometric properties of instruments	NR	NR	40.45 (8.67) 24-55	52.9%	NR
Oliver et al., 1998	DS	NR	NR	Between groups (cognitive deterioration)	NR	NR	42.34 (7.26)	59.6%	Participants excluded if unable to say single words or execute simple motor functions
Oliver et al., 2005	DS	NR	BPVS, VABS	Between groups (cognitive deterioration; age)	NR	NR	41.82 (7.37)	59.6%	NR

Author, Year	Subtype of ID	ID Severity	Criteria/ method used for assessing level of ID	Comparator / control group	Dementia subtypes (if included)	Diagnostic criteria for dementia	Mean age (years, *SD*) and/ or range (in years)	% female	Verbal ability (for inclusion in sample)
Palmer, 2006	DS + oID	Mild-moderate	NR	Between groups (DS-AD, DS-no-AD, oID)	AD	DSM-IV-TR	AD = 50.50 (6.77) 36–62 Controls = 44.50 (9.07) 33–66	AD = 60% Controls= 66.7%	NR
Powell et al., 2014	DS	"low, medium or high-functioning"	Medical records	Between groups (DS-AD, DS-no-AD, controls)	AD	NINCDS-ADRDA and clinical consensus	DS = 51.38 (6.48) DS-no-AD = 50.61 (5.53) DS-AD = 52.16 (7.54) Controls= 51.07 (2.14)	70%	NR
Poveda & Broxholme, 2016	DS+oID	Moderate (Mean IQ= 45.8- 58.57) IQ score unavailable= 37	NR	Longitudinal and Psychometric properties (identifying dementia status)	NR	LDDB, VABS, DMR	50.88 (9.82) 29-71	DS= 24:14 oID= 7:10	NR
Pulsifer et al. 2020	DS	Mild-severe	NR	Psychometric properties, between measures and between groups (dementia status)	AD	Clinical consensus	Controls= 49 (6.59) DS-MCI= 53.63 (6.94) DS-AD= 55.64 (5.87)	42.9%	NR
Pyo et al., 2007	DS + oID	Moderate-severe	Medical records	Between groups (AD, no-AD)	AD	DSM-IV-TR	AD = 53.13 (10.56) 43-74 Controls = 49.95 (5.13) 40-59	AD = 15.4% Controls = 24.4%	NR
Pyo et al., 2009	DS + oID	Moderate-severe	Medical records	Between groups (AD, no-AD)	AD	DSM-IV-TR	AD = 53.99 (10.20) Controls = 50.76 (5.76)	AD = 31.3% Controls = 2.9%	NR
Pyo et al., 2010	DS + oID	Moderate-severe	Medical records	Between groups (AD, no-AD)	AD	DSM-IV-TR	DS = 49.21 (4.41) oID =52.87 (5.25) DS-AD = 47.89 (4.18) oID-AD = 57.13 (10.52)	DS= 0% oID = 8.3% DS-AD = 13.3% oID-AD = 36.4%	NR
Pyo et al., 2011	DS + oID	Moderate-severe	Medical records	Between groups (AD, no-AD)	AD	DSM-IV-TR	DS-controls = 47.71 (5.21) oID-controls = 51.93 (7.05) DS-AD = 48.26 (2.43)	DS-controls = 0% oID-controls = 6.9% DS-AD = 20%	NR

Author, Year	Subtype of ID	ID Severity	Criteria/ method used for assessing level of ID	Comparator / control group	Dementia subtypes (if included)	Diagnostic criteria for dementia	Mean age (years, *SD*) and/ or range (in years)	% female	Verbal ability (for inclusion in sample)
							oID-AD = 57.99 (11.14)	oID-AD = 45.5%	
Sano et al., 2005	DS	Mild= 15% Moderate= 52% Severe= 29% Profound= 4%	Medical records	Between instruments	NR	DSM-IV	48.7 (6.2) 33-77	51.3%	NR
Schmitt et al., 2010	N/A	N/A	WTAR	Psychometric properties between instruments	NR	RBANS	NR	NR	NR
Shultz et al., 2004	DS (68%), oID	Mean FSIQ= 41.1	Prior assessment	Psychometric properties of instruments and between groups (dementia, no-dementia)	NR	DSM-IV or ICD-10	56 (45-74)	45%	NR
Sinai et al., 2016	DS	Mild = 37.1% Moderate/Severe = 62.9%	Informant report, case notes	Between groups (dementia, no-dementia)	NR	Informant report, clinical consensus	52.7 (6.06) 45-64	53.1%	NR
Tabert et al. 2005	N/A	N/A	N/A	Between groups (dementia status)	MCI, AD	DSM-4, clinical consensus	Controls= 65.71 (9.38) MCI= 67.63 (9.85) AD= 71.72 (9.54)	Controls= 54% MCI= 55.1% AD= 63.8	NR
Walsh et al., 2015	DS	Mild = 35% Moderate = 39% Severe = 23% Profound = 3%	Unknown (already diagnosed)	Psychometric properties of instruments	NR	ICD-10 and DSM-IV	49.8 (8.9)	45%	NR
Webb et al. 2020	oID	Mean IQ= 60.35 (5.39)	WASI	Between instruments, psychometric properties of instruments	N/A	N/A	42.05 (12.76)	38.2%	NR
Willner et al. 2010	Mixed	Mean FSIQ= 59	NR	Between measures comparison, psychometric properties	N/A	N/A	Mean 40.1 (10.8)	47.5%	NR
Witts & Elders, 1998	DS	NR	NR	Psychometric properties of instruments	NA	NA	36 (8.9) 22-53	45.5%	NR

Note. AAIDD = The American Association on Intellectual and Developmental Disabilities; AD= Alzheimer's Disease Dementia; BPVS/ BPVS-II = British Picture Vocabulary Scale/ British Picture Vocabulary Scale-2nd Edition; BSID = Bayley Scales of Infant Development; 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; DLD = Dementia Questionnaire for People with Learning Disabilities; DM-ID = Diagnostic Manual — Intellectual Disability; 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;; ICD-10/ ICD-11 = International Classification of Diseases-10th Edition/ 11th Edition; ID = Intellectual disability; IQ = Intelligent Quotient; IQR = Interquartile range; KBIT = Kaufman Brief Intelligence Test; LIPS = Leiter International Performance Scale; MCI = Mild Cognitive Impairment; mCRT = Modified Cued Recall Test; N/A = 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; VABS = Vineland Adaptive Behavior Scales; WAIS/ WAIS-R/ WAIS-III/ WAIS-IV = Wechsler Adult Intelligence Scale/ Revised/ 3rd Edition/ 4th Edition; WASI = Wechsler Abbreviated Scale of Intelligence; WTAR= Wechsler Test of Adult Reading

Appendix F: HRA Approval Letter

Ymchwil Iechyd a Gofal Cymru
Health and Care Research Wales

NHS
Health Research Authority

Dr Matthew Jones-Chesters
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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 organisations.	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.

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 G: UEL EISC Ethics Approval Letter

University of East London

School of Psychology Ethics Committee

NOTICE OF ETHICS REVIEW DECISION LETTER

For research involving human participants

BSc/MSc/MA/Professional Doctorates in Clinical, Counselling and Educational Psychology

Reviewer: Please complete sections in blue | **Student:** Please complete/read sections in orange

Details	
Reviewer:	Please type your full name **Fevronia Christodoulidi**
Supervisor:	Please type supervisor's full name **Matthew Jones Chesters**
Student:	Please type student's full name **Elicia McGregor and Zakiya Reid-Wisdom**
Course:	Please type course name **Prof Doc Clinical in Psychology**
Title of proposed study:	Assessment of cognition in people with intellectual disabilities using a novel set of neuropsychological tests

Checklist (Optional)	YES	NO	N/A
Concerns regarding study aims (e.g., ethically/morally questionable, unsuitable topic area for level of study, etc.)	☐	☐	☐
Detailed account of participants, including inclusion and exclusion criteria	☐	☐	☐
Concerns regarding participants/target sample	☐	☐	☐
Detailed account of recruitment strategy	☐	☐	☐
Concerns regarding recruitment strategy	☐	☐	☐
All relevant study materials attached (e.g., freely available questionnaires, interview schedules, tests, etc.)	☐	☐	☐
Study materials (e.g., questionnaires, tests, etc.) are appropriate for target sample	☐	☐	☐

Clear and detailed outline of data collection	☐	☐	☐
Data collection appropriate for target sample	☐	☐	☐
If deception being used, rationale provided, and appropriate steps followed to communicate study aims at a later point	☐	☐	☐
If data collection is not anonymous, appropriate steps taken at later stages to ensure participant anonymity (e.g., data analysis, dissemination, etc.) – anonymisation, pseudonymisation	☐	☐	☐
Concerns regarding data storage (e.g., location, type of data, etc.)	☐	☐	☐
Concerns regarding data sharing (e.g., who will have access and how)	☐	☐	☐
Concerns regarding data retention (e.g., unspecified length of time, unclear why data will be retained/who will have access/where stored)	☐	☐	☐
If required, General Risk Assessment form attached	☐	☐	☐
Any physical/psychological risks/burdens to participants have been sufficiently considered and appropriate attempts will be made to minimise	☐	☐	☐
Any physical/psychological risks to the researcher have been sufficiently considered and appropriate attempts will be made to minimise	☐	☐	☐
If required, Country-Specific Risk Assessment form attached	☐	☐	☐
If required, a DBS or equivalent certificate number/information provided	☐	☐	☐
If required, permissions from recruiting organisations attached (e.g., school, charity organisation, etc.)	☐	☐	☐
All relevant information included in the participant information sheet (PIS)	☐	☐	☐
Information in the PIS is study specific	☐	☐	☐
Language used in the PIS is appropriate for the target audience	☐	☐	☐
All issues specific to the study are covered in the consent form	☐	☐	☐
Language used in the consent form is appropriate for the target audience	☐	☐	☐
All necessary information included in the participant debrief sheet	☐	☐	☐
Language used in the debrief sheet is appropriate for the target audience	☐	☐	☐
Study advertisement included	☐	☐	☐
Content of study advertisement is appropriate (e.g., researcher's personal contact details are not shared, appropriate language/visual material used, etc.)	☐	☐	☐

Decision options	
APPROVED	Ethics approval for the above-named research study has been granted from the date of approval (see end of this notice), to the date it is submitted for assessment.
APPROVED - BUT MINOR AMENDMENTS ARE REQUIRED BEFORE THE RESEARCH COMMENCES	In this circumstance, the student must confirm with their supervisor that all minor amendments have been made before the research commences. Students are to do this by filling in the confirmation box at the end of this form once all amendments have been attended to and emailing a copy of

	this decision notice to the supervisor. The supervisor will then forward the student's confirmation to the School for its records. **Minor amendments guidance:** typically involve clarifying/amending information presented to participants (e.g., in the PIS, instructions), further detailing of how data will be securely handled/stored, and/or ensuring consistency in information presented across materials.
NOT APPROVED - MAJOR AMENDMENTS AND RE-SUBMISSION REQUIRED	In this circumstance, a revised ethics application **must** be submitted and approved **before** any research takes place. The revised application will be reviewed by the same reviewer. If in doubt, students should ask their supervisor for support in revising their ethics application. **Major amendments guidance:** typically insufficient information has been provided, insufficient consideration given to several key aspects, there are serious concerns regarding any aspect of the project, and/or serious concerns in the candidate's ability to ethically, safely and sensitively execute the study.

Decision on the above-named proposed research study	
Please indicate the decision:	APPROVED

Minor amendments
Please clearly detail the amendments the student is required to make

Major amendments
Please clearly detail the amendments the student is required to make

Assessment of risk to researcher		
Has an adequate risk assessment been offered in the application form?	**YES** ☒	**NO** ☐
	If no, please request resubmission with an **adequate risk assessment.**	
If the proposed research could expose the researcher to any kind of emotional, physical or health and safety hazard, please rate the degree of risk:		
HIGH	Please do not approve a high-risk application. Travel to countries/provinces/areas deemed to be high risk should not be permitted and an application not be approved on this basis. If unsure, please refer to the Chair of Ethics.	☐
MEDIUM	Approve but include appropriate recommendations in the below box.	☐
LOW	Approve and if necessary, include any recommendations in the below box.	☒
Reviewer recommendations in relation to risk (if any):	Please insert any recommendations	

Reviewer's signature	
Reviewer: (Typed name to act as signature)	**Dr Fevronia Christodoulidi**
Date:	**01/08/2023**
This reviewer has assessed the ethics application for the named research study on behalf of the School of Psychology Ethics Committee	
RESEARCHER PLEASE NOTE For the researcher and participants involved in the above-named study to be covered by UEL's insurance, prior ethics approval from the School of Psychology (acting on behalf of the UEL Ethics Committee), and	

Appendix H: Participant Invitation Letter

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 XXXX, 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.

Why have you been asked to participate?

Participant Identification Number:

You have been invited to participate in my research
as I am looking to involve people who have Down Syndrome (Trisomy 21), 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 XXXXXXXXXX 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 affects 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.

What are the potential benefits of taking part?

Participant Identification Number:

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 u1945505@uel.ac.uk, or
- by ringing us on [redacted]

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
  Phone: [redacted]

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.

Participant Identification Number:

Email: t.patel@uel.ac.uk

Appendix I: Carer Invitation Letter

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 Elicia, 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 Down Syndrome (Trisomy 21), 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 [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 affects 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.

What are the potential benefits of taking part?

Participant Identification Number:

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 xxxxxxxx, or
- by ringing us on xxxxxxxxxx

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: xxxxxxxxxxxxxx

Phone: xxxxxxxxxx

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: xxxxxxxxxxxxx

Appendix I: Easy-Read 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 Elicia, 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 Down Syndrome, 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 (XXXXXXXXX) 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 [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: XXXXXX, 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 K: Data Management Plan

University of East London
Pioneering Futures Since 1898

UEL Data Management Plan

Completed plans must be sent to researchdata@uel.ac.uk for review

If you are bidding for funding from an external body, complete the Data Management Plan required by the funder (if specified).

Research data is defined as information or material captured or created during the course of research, and which underpins, tests, or validates the content of the final research output. The nature of it can vary greatly according to discipline. It is often empirical or statistical, but also includes material such as drafts, prototypes, and multimedia objects that underpin creative or 'non-traditional' outputs. Research data is often digital, but includes a wide range of paper-based and other physical objects.

Administrative Data	
PI/Researcher	**Dr. Matthew Jones-Chesters** **Supervising Elicia McGregor (Trainee Clinical Psychologist)**
PI/Researcher ID (e.g. ORCiD)	Dr. Matthew Jones Chesters ORCiD: 0000-0001-8146-7873 Elicia McGregor ORCiD: 0000-0002-3447-0081
PI/Researcher email	Dr. Matthew Jones-Chesters: m.h.jones-chesters@uel.ac.uk Elicia McGregor: u1945505@uel.ac.uk
Research Title	CREATING AND PILOTING A DIAGNOSTIC TOOL FOR THE DEMENTIAS FOR PEOPLE WITH LEARNING DISABILITIES
Project ID	IRAS Project ID: 295654
Research start date and duration	Start date: September 2020
Research Description	Many individuals with a learning disability are living longer due to better healthcare and are therefore more likely to develop dementia. There are many different types of dementia, each with their own unique profiles. Dementia may look different in people with a learning disability, as these individuals have pre-existing differences in the brain compared to 'typically developing' people. There have been tests created to screen for dementia in the learning disability population, but these tools do not accurately assess all areas of cognition, often overlooking executive function. Many existing tests are also made for use with a carer or parent rather than the person with a learning disability. Therefore, this research aims to create and pilot a diagnostic tool to identify the dementias

	in people with learning disabilities which examines all cognitive areas and is appropriate for use within this population. This study aims to create a novel scalable diagnostic measure of dementia for the Learning Disability population which is feasible, acceptable and accessible. It will be made with accessible, low cost materials for ease of use within the NHS. Feasibility and acceptability studies work best with a narrow scope (Bowen et al., 2009), therefore this tool will be piloted within the Down Syndrome population to gather preliminary norm data. This tool will aim to 1) assess all cognitive domains to aid in differential dementia diagnosis, 2) be accessible and acceptable to people with Learning Disabilities and 3) be administrable with low cost to NHS services.
Funder	The University of East London
Grant Reference Number (Post-award)	
Date of first version (of DMP)	31.01.2021
Date of last update (of DMP)	26.02.2021
Related Policies	e.g. Research Data Management Policy UK Research and Innovation Guidelines: https://www.ukri.org/about-us/policies-standards-and-data/good-research-resource-hub/ General Data Protection Regulation: https://www.ukri.org/about-us/policies-standards-and-data/gdpr-and-research-an-overview-for-researchers/
Does this research follow on from previous research? If so, provide details	N/A
Data Collection	

What data will you collect or create?	Qualitative data (feedback on improvements/revisions to be made to the novel battery) collected via questionnaire, and quantitative data (scores on novel created neuropsychological battery tasks). Video recordings in .mp4 format Questionnaire responses in print/ paper format. Scores on battery tasks will be in .sav format (for processing and analysis using SPSS). We will have 6 participants, therefore 6 .mp4 files. As these files are identifiable, they will be scored immediately after collection and safely destroyed after they have been analysed for scoring. No other personal or sensitive data will be collected. There will be 6 participants, and from each we will collect a video of the diagnostic process, consent form and questionnaire responses.
How will the data be collected or created?	Participant performance will be video-recorded and stored as .mp4 files. Participant testing will be video-recorded to ensure accurate scoring and interpretations of test accessibility. This will be immediately uploaded to OneDrive for Business after collection through a UEL computer, using a USB cable link. The video will then be deleted from the video camera device. Paper data will be immediately entered into a .sav SPSS file, kept within OneDrive for Business. All information provided and recorded will be kept strictly confidential. Data will be uploaded to the UEL OneDrive, which is a secure, encrypted online service. After uploading, all paper information will be safely destroyed, alongside data on the video camera. All data will be anonymised by assigning a numerical code instead of participant names. For up to 3 weeks after participation, a separate document will be kept which links names to their numerical code, in case participants decide to withdraw from the study during this period. After 3 weeks, names will be deleted from our records. As above, through paper responses and video-recorded performance of tests. Performance data will be quantitative and data on improvement or refinement of tests will be qualitative. Participants and guardians will be invited to read the information sheet and ask questions, before signing the consent form if they agree to participate. An easy-read information sheet and assent form will also be provided. After testing, participants will be given a debrief letter and easy-read debrief letter. Therefore, consent will be collected through a written/paper medium. A copy of the consent form will be given to participants and their carers each for their reference.

Documentation and Metadata	
What documentation and metadata will accompany the data?	A spreadsheet (.csv) file containing locations of all data available. This spreadsheet will be encrypted (password-protected). Only the researcher and P.I will have access to this password. Locations for a sample of the completed questionnaire, blank consent forms, participant information sheets and scoring guides will also be included in this spreadsheet.
Ethics and Intellectual Property	
Identify any ethical issues and how these will be managed	Importantly, participants with Learning Disabilities may be socially naïve and vulnerable to coercion (Khemka, Hickson, Casella, Accetturi & Rooney, 2009). Further, power relations between the participant and the researcher considering social naiveté can make it difficult for participants to communicate discomfort (Spears & Smith, 2001; Khemka et al., 2009). Therefore, participants will be accompanied by an advocate, carer or guardian. Additionally, to facilitate informed consent, an easy-read information sheet will be given alongside an information sheet with denser detail. Further, a video of the researcher explaining the purpose and process of the research will be sent to participants before they decide whether to consent to take part. Data will be anonymised by assigning each participant a unique numerical code which will be stored separately to their identifiable information. Identifiable information will be safely destroyed after 3 weeks, kept only for this time in case participants wish to withdraw. Access to this information will require a password known only by the researcher and the P.I.
Identify any copyright and Intellectual Property Rights issues and how these will be managed	No intellectual property will be created in this project.

Storage and Backup	
How will the data be stored and backed up during the research?	Data will be stored and backed up to the UEL OneDrive, a secure and encrypted service. Once uploaded here, all records of data in paper or video camera format will be destroyed. The storage will be on the UEL OneDrive account and then backed up to the researcher's H: Drive. Completed consent forms will be stored in a locked file folder drawer within UEL, under the care of the P.I. Pseudoanonymised data will be kept in a spreadsheet (.csv) within a folder separate to the identifiable data spreadsheet (.csv). We will back data up to the UEL H: drive, managed by logging in to a UEL managed computer. Identifiable data will be destroyed after 3 weeks of collection, retained only in the case that participants wish to withdraw in this time.
How will you manage access and security?	Links to the folder will be password-protected. The only people with access to this folder will be the researcher and the PI. The video camera will be kept in the locked office of the P.I, and will never have recordings of participants kept overnight from the day of collection. The camera will have the recording uploaded to the aforementioned UEL OneDrive and will then be wiped before being locked in the P.I's office at the end of each testing day. Paper format data will also be stored in the office of the P.I in a locked file folder drawer.
Data Sharing	
How will you share the data?	Only anonymised data will be shared. The diagnostic tool created will be made publicly and freely available to NHS services. Anonymised data will be shared in the academic thesis produced and in the published thesis produced. This will be shared openly via UEL's Research Repository as per Selection/Preservation below, and this decision will eb detailed in the information and consent sheets presented to potential participants.
Are any restrictions on data sharing required?	N/A
Selection and Preservation	

Which data are of long-term value and should be retained, shared, and/or preserved?	No personal/pseudonymised data of long-term value will be collected, as this is a pilot study for a diagnostic tool. Data kept will be on tool administration and anonymised data for prospective norms. These will be kept in the UEL OneDrive during analysis and write-up, and in the UEL data repository after analysis (see below).
What is the long-term preservation plan for the data?	Data will be preserved in UEL's data repository (https://repository.uel.ac.uk). After the study has been completed, data will continue to be stored in this secure location, only accessible by the research team and a limited number of library staff for 10 years, as recommended by the UK Research and Innovation (UKRI) guidelines. After this, all data will be destroyed. This data will not contain sensitive information and so will be suitable for sharing via the repository. Data will be deposited and shareable in .sav (spss dataset, anonymised) and .pdf (completed written up thesis, anonymised) formats. [Your Thesis will remain on the repository PJ and just to add that for the dataset UEL's Research Data Management Policy is to review at the end of the project and every 5 years until data are destroyed or transferred PJ]
Responsibilities and Resources	
Who will be responsible for data management?	Elicia McGregor and Dr. Matthew Jones-Chesters
What resources will you require to deliver your plan?	Internet/computer access, UEL OneDrive Access. After the researcher leaves UEL, responsibility or data will remain with Principal Investigator Dr. Matthew Jones-Chesters.
Review	
	Review after feedback from Ethics and regularly thereafter. Please send any amendments, as necessary to: researchdata@uel.ac.uk

Date: 26/02/2021	Reviewer name: Penny Jackson Research Data Management Officer

Guidance

Brief information to help answer each section is below. Aim to be specific and concise.

For assistance in writing your data management plan, or with research data management more generally, please contact: **researchdata@uel.ac.uk**

Administrative Data

Related Policies

List any other relevant funder, institutional, departmental or group policies on data management, data sharing and data security. Some of the information you give in the remainder of the DMP will be determined by the content of other policies. If so, point/link to them here.

Data collection

Describe the data aspects of your research, how you will capture/generate them, the file formats you are using and why. Mention your reasons for choosing particular data standards and approaches. Note the likely volume of data to be created.

Documentation and Metadata

What metadata will be created to describe the data? Consider what other documentation is needed to enable reuse. This may include information on the methodology used to collect the data, analytical and procedural information, definitions of variables, the format and file type of the data and software used to collect and/or process the data. How will this be captured and recorded?

Ethics and Intellectual Property

Detail any ethical and privacy issues, including the consent of participants. Explain the copyright/IPR and whether there are any data licensing issues – either for data you are reusing, or your data which you will make available to others.

Storage and Backup

Give a rough idea of data volume. Say where and on what media you will store data, and how they will be backed-up. Mention security measures to protect data which are sensitive or valuable. Who will have access to the data during the project and how will this be controlled?

Data Sharing

Note who would be interested in your data, and describe how you will make them available (with any restrictions). Detail any reasons not to share, as well as embargo periods or if you want time to exploit your data for publishing.

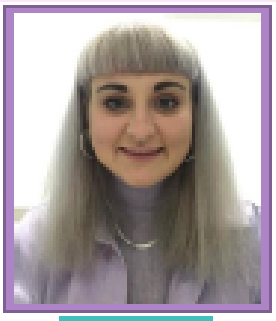
Appendix L: Study Recruitment Poster

IRAS ID:295654

Are you a person with Down Syndrome aged between 30 and 55 years old?

Are you currently receiving care from [SERVICE NAME] ?

Hello! My name is Elicia. 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 Down Syndrome who are aged between 30 and 55 years old, who do not have a diagnosis of dementia, and who come to [SERVICE], to take part in my study.



University of East London

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 Down Syndrome that accurately look at all the different things that the brain can do accurately. I would like to make a test for people with Down Syndrome that feels engaging and considers what people with Down Syndrome 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 Down Syndrome and other learning disabilities 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 [CONTACT] at [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 M: 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.	

FOR **PARTICIPANT INVITATION LETTER v1.03 (07.08.2022)** ONE COPY FOR THE FILE,
ONE COPY FOR PARTICIPANT

Appendix N: 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.	

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 O: Participant Easy-Read Debrief Letter

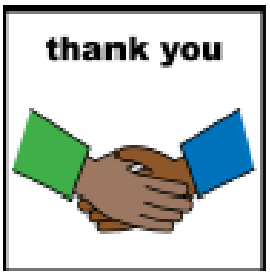
Participant Identification Number:

University of East London

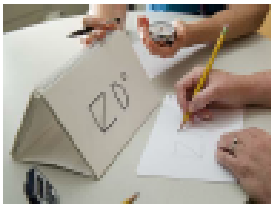
School of Psychology

Assessment of Cognition in People with Intellectual Disabilities – Participant Debrief Letter (Easy-Read)

V19.01.2023



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?

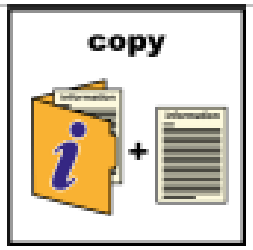
All the information you provided will be kept strictly confidential. This means that nobody apart from the research team will be able to see it. Your data will be stored on a safe

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.

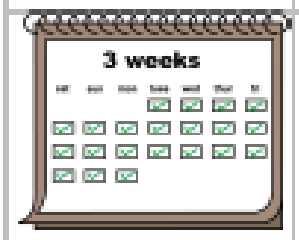
Your anonymised data will be seen by the researchers (XXXXXXX) and our supervisor (Dr. Matthew Jones-Chesters). Group data will be included in our research report, which will be read by examiners, and will be in a paper that other people can see. In this paper, you will only be known as a number, so 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.



Can I have a copy?

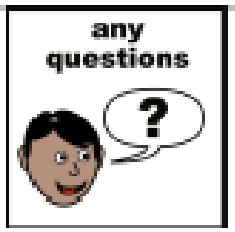
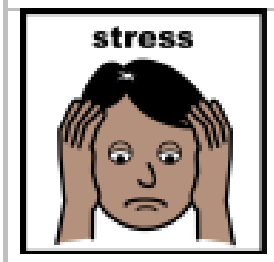
If you want, we can give you a copy of the results of this study once it is finished.



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 [SERVICE] for support.
	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: XXXXXX, email: XXXXXXXX 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: XXXXXXXXXX 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: XXXXXXXXXXX You can also visit this website to learn more about how your information is used: www.hra.nhs.uk/information-about-patients/

Appendix P: 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 (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 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:

- 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 Q: 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 R: 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 not 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 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 S: Data Distribution Table for Motor and Language Functions

Though the sample size was too small to formally analyse data as planned, a data distribution table and boxplot for 'Motor and Language Functions' subtests (where $n = 4$) is presented in this appendix as an example of planned analyses.

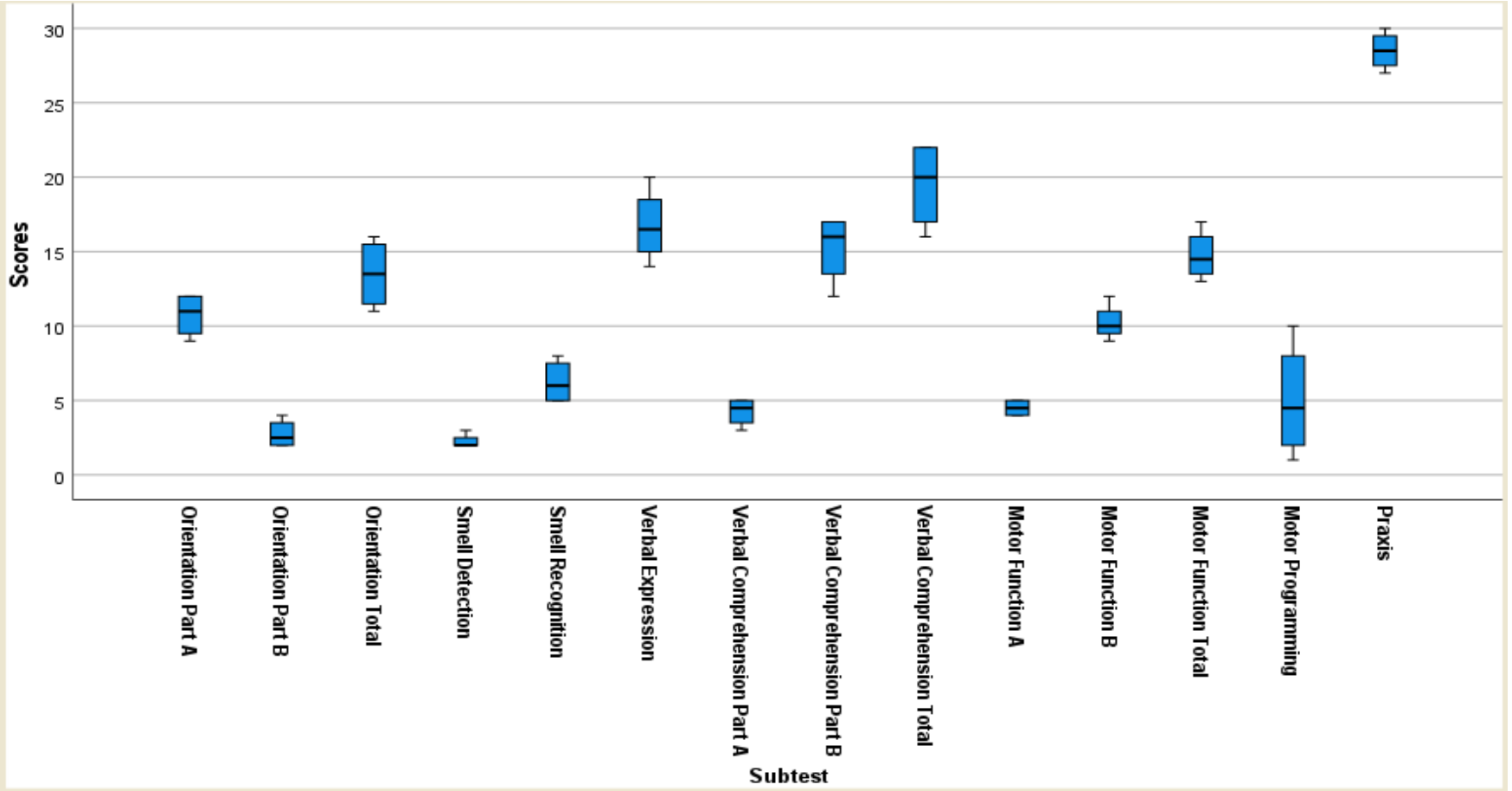
Table S. *Data Distributions - Motor & Language Functions*

Motor & Language Functions							
Subtest	**Skewness**			**Kurtosis**			***Normality***
	Value	**SE**	***Z***	**Value**	**SE**	***Z***	**Shapiro-Wilk test *p***
Orientation Subtotal A	-0.37	1.01	-0.37	**-3.90**	2.62	-1.49	.224
Orientation Subtotal B	0.86	1.01	0.85	**-1.29**	2.62	-0.49	.272
Orientation Total (A + B)	0.00	1.01	0.00	**-4.34**	2.62	-1.66	.488
Smell Detection	**2.00**	1.01	**1.98**	**4.00**	2.62	1.53	**.001**
Smell Recognition Total	0.37	1.01	0.36	**-3.90**	2.62	-1.49	.224
Verbal Expression	0.56	1.01	0.55	0.93	2.62	0.35	.911
Verbal Comprehension A	-0.86	1.01	-0.85	**-1.29**	2.62	-0.49	.272
Verbal Comprehension B	**-1.19**	1.01	-1.18	0.44	2.62	0.17	.220
Verbal Comprehension Total (A + B)	**-0.37**	1.01	-0.36	**-3.90**	2.62	-1.49	.224

Motor Function Subtotal A	0.00	1.01	0.00	**-6.00**	2.62	**-2.30**	**.024**
Motor Function Subtotal B	**1.13**	1.01	1.12	**2.23**	2.62	0.85	.406
Motor Function Total (A + B)	0.75	1.01	0.74	0.34	2.62	0.13	.850
Motor Programming Total	0.60	1.01	0.59	-0.77	2.62	-0.29	.850
Praxis Total	0.00	1.01	0	**-1.20**	2.62	-0.46	.972

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

Figure S. *Boxplot - Motor & Language Functions*



Appendix T: 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 noting this can be 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 autistic individuals) • 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