University of Southampton

Faculty of Environmental and Life Sciences

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Dementia and the Intellectual Disability Population: Insights into the Early Presentation and Approaches to Assessment

by

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Thesis for the degree of Doctor of Clinical Psychology

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Abstract

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It is widely acknowledged that individuals with intellectual disability are more susceptible to dementia than the general population however the reason for this is unclear. Theories include variation in pre-existing levels of cognitive ability, genetic components, and the presence of additional comorbidities. Individuals with Down's Syndrome are also at increased risk due to a triplication of chromosome 21. Dementia is characterised by progressive decline however the early manifestation in this population is uncertain. As a result, a systematic review was conducted to explore the early symptoms associated with dementia in individuals with intellectual disability. 18 studies were included and analysed using narrative synthesis. The review found evidence that changes in multiple domains, including memory, executive function, attention, mobility, and behaviour were observed early in the course of dementia. The review emphasises the need for assessments to be broad in nature to encompass domains other than memory that may demonstrate early decline.

In light of this, there is a need for valid and reliable tools exploring a range of functions to be developed for the intellectual disability population. Although numerous assessment tools exist, only a small proportion were designed specifically for this population. The empirical study explored the psychometric properties of a newly developed battery aimed at assessing dementia in this population. 23 participants across different NHS settings completed the battery. The results suggest the battery demonstrates good internal consistency although the validity of the measure could not be established due to the small sample size. Floor and ceiling effects were present across several subtests. This battery appears to be a promising measure for use within the intellectual disability population however further amendments are required regarding the inclusion of certain subtests and scoring guidance. Further research should seek to verify the validity of the battery in comparison to other established measures.

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Research Thesis: Declaration of Authorship

Print name: Jade Dunning

Title of thesis: Dementia and the Intellectual Disability Population: Insights into the Early Presentation and Approaches to Assessment

I declare that this thesis and the work presented in it are my own and has been generated by me as the result of my own original research.

I confirm that:

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2. Where any part of this thesis has previously been submitted for a degree or any other qualification at this University or any other institution, this has been clearly stated;
3. Where I have consulted the published work of others, this is always clearly attributed;
4. Where I have quoted from the work of others, the source is always given. With the exception of such quotations, this thesis is entirely my own work;
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6. Where the thesis is based on work done by myself jointly with others, I have made clear exactly what was done by others and what I have contributed myself;
7. None of this work has been published before submission.

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Finally, I would like to dedicate this to my nan, for being such a wonderful human being. Your kindness knew no bounds. I hope I make you proud.

Abbreviations

AD.....Alzheimer's Disease

ADLs .....Activities of Daily Living

BPSD.....Behavioural and Psychological Symptoms of Dementia

BPVS-III.....British Picture Vocabulary Scale 3

CORE-LD .....Clinical Outcomes in Routine Evaluation – Learning Disability

DLD.....Dementia Questionnaire for People with Learning Disabilities

DS .....Down's Syndrome

DSM-5.....Diagnostic and Statistical Manual of Mental Disorders – Fifth Edition

EF.....Executive Function

ICD-10.....International Classification of Diseases – Tenth Revision

ID .....Intellectual Disability

NAID .....Neuropsychological Assessment of Dementia in Individuals with Intellectual Disabilities

NBID .....Neurocognitive Battery for Intellectual Disabilities

NHS.....National Health Service

Chapter 1 Early Markers of Dementia in the Intellectual Disability Population: A Systematic Literature Review

Journal choice: The '*Journal of Applied Research in Intellectual Disabilities'* has been used as a guide to prepare this review. The named journal specifies that review articles should not exceed 7000 words, with an abstract of maximum 150 words. Tables, figures, captions, and references are excluded from the word count.

Word count: 7,000 (including abstract, excluding tables, figures, and references)

1.1 Abstract

Adults with intellectual disability (ID) are at increased risk of dementia. Memory changes are often considered the earliest marker associated with dementia; however, this may not always be the case. Research indicates decline in language and executive function, as well as changes in non-cognitive domains may occur in the early stages, possibly before changes in memory. This systematic review evaluated recent literature on the early markers of dementia in ID. 18 peer-reviewed papers met the inclusion criteria. Across the studies, changes in multiple domains were found to occur early in the dementia course. Memory and executive function were frequently reported as sensitive to early decline, as well as changes in attention, mobility, and behaviour. These findings suggest symptoms other than memory decline also occur in the early stages of dementia in ID. This has implications for policy and guidelines regarding dementia assessment. Further research is required to address methodological limitations.

1.2 Introduction

1.2.1 Lifespan and Ageing

Historically, life expectancy for those with an intellectual disability (ID)[1] was significantly lower than the general population. In the 1930s life expectancy for this population was approximately 18.5 years, increasing to 59.1 years in the 1970s and to 66.2 years in the 1990s (Braddock, 1999). Similarly, individuals with Down's Syndrome (DS) experienced much shorter life expectancies, often not living beyond the age of 30 in the 1950s (Lott & Head, 2019). Despite advancements in medicine and specialist healthcare provisions, life expectancy continues to fall short of the general population (Coppus, 2013; Heslop et al., 2014; White et al., 2023). This can be understood to some extent by the continued presence of substantial health inequalities faced by the ID population (Emerson & Baines, 2011; McCarron et al., 2014), and high rates of avoidable deaths (White et al., 2023). Obtaining prevalence and life expectancy data for this population can be challenging. Many studies are methodologically flawed, use inconsistent definitions of ID, are selective regarding inclusion criteria, consist of small samples, and use a range of diagnostic criteria (McMahon & Hatton, 2021). In addition, there is likely a sub-section of individuals without a formal ID diagnosis, or don't interact with specialist services, resulting in underestimations of epidemiological data (Heslop & Glover, 2015).

1.2.2 Dementia Progression

It is estimated that 55 million people live with dementia globally (World Health Organization, 2023). Age is one of the main factors associated with dementia risk (Chen et al., 2009) due to brain volume reduction, disruption of neurotransmitter signals (Peters, 2006), and accumulation of amyloid-β (Power et al., 2018). A wealth of research suggests individuals with ID and DS are more susceptible to dementia than the general population (Coppus et al., 2006; Startin, Hamburg, et al., 2016; Strydom et al., 2013). The British Psychological Society (BPS; 2015) summarised dementia prevalence across the ID, DS, and general populations. An age-related

[1] The term 'intellectual disability' will be used throughout this paper, reflecting its more widely adopted use within policy and research (Cluley, 2018). Although services in the UK, such as community teams which have been involved with this research, typically use the term 'learning disability' and therefore many of the study materials also used this phrasing, it was felt the term 'intellectual disability' was more universally recognised (Fredman, 2006). 'Intellectual disability' will be the main term used throughout this paper however there may be instances where 'learning disability' is used instead. It should be assumed that these terms are interchangeable for the purposes of this paper.

trend was observed where individuals with ID showed an earlier onset and steeper increase in prevalence than the general population, with symptoms typically developing in their 50s and approximately 50% developing dementia. Those with DS demonstrate an even earlier onset, around age 30, and a more rapid increase in prevalence with approximately 75% of this population developing dementia. A review by Cipriani et al. (2018) concluded the majority of individuals with DS aged 35-40 years showed neuropathological changes related to Alzheimer's Disease (AD), the most common form of dementia, although clinical signs may not be evident until much later. Despite it being well accepted that individuals with ID and/or DS demonstrate cognitive changes indicative of dementia much earlier than in the general population (Strydom et al., 2010; Strydom et al., 2013; Takenoshita et al., 2020), the exact course of progression, particularly in the early stages, is less well understood.

DS is often considered a genetic form of dementia (Fortea et al., 2021) due to the presence of an additional chromosome 21 which is implicated in the amyloid cascade hypothesis (Hardy & Higgins, 1992). This theory suggests the protein amyloid-β is responsible for AD brain pathology, causing alterations in cell structure, atrophy, and vascular changes. The amyloid precursor protein which is responsible for the production of amyloid-β is situated on chromosome 21, causing an overexpression in those with DS, increasing their risk of dementia (Lott & Head, 2019). It is less clear why individuals with ID without DS are also at a greater risk of dementia, although hypotheses include pre-existing cognitive abilities, poorer physical health, and greater co-morbidities e.g., cardiovascular disease (Takenoshita et al., 2023).

Recent research has shifted focus to investigating the earliest stages of dementia in people with ID (Krinsky-McHale & Silverman, 2013). These stages occur prior to a clinical diagnosis and include preclinical and prodromal stages. In the preclinical stage, biological change may be present without observable symptoms (Sperling et al., 2013) whereas the prodromal stage is where the earliest symptomatic change is evident (Scharre, 2019). An understanding of these early stages is important to aid early diagnosis and is crucial when considering that time between diagnosis and death may be as little as three years (Strydom et al., 2010).

Research into both ID and DS suggest differences in the early stages, in language, functional skills, personality, and behaviour (Deb et al., 2007a; Strydom et al., 2010). Cosgrave et al. (2000) investigated dementia symptomology in DS longitudinally, enabling retrospective examination of symptom onset. Memory loss, spatial disorientation, and reduction in activities of daily living (ADLs) were recognised as the earliest features of dementia. However, this sample consisted of individuals with DS and moderate to severe ID and therefore may not represent early symptoms across the entire ID population. Devenny et al. (2000) attempted to sequence cognitive decline in

a DS sample, comparing individuals who remained cognitively 'healthy' to those considered to have 'questionable', 'early stage', and 'middle stage' dementia. They found that individuals considered to be in the early stages showed poorer performance on measures of visuospatial skills and working memory but showed preserved performance on short-term and semantic memory measures. However, the small sample and few participants in each dementia group limit the conclusions that can be drawn. Ball, Holland, Hon, et al. (2006) concluded that frontal lobe functions are implicated earliest, resulting in personality and behavioural changes which may be associated with executive dysfunction. Research in the general population has found associations between gait speed and global cognitive decline, including executive function (EF) and memory (Hughes et al., 2020; Mielke et al., 2013). It is possible that a similar pattern is also present within ID and DS. Anderson-Mooney et al. (2016) suggested that, although changes in gait occur throughout ageing in DS, a decline in higher-gait functioning may be more fully explained by pathology associated with dementia, such as reduced ability in adapting to environmental demands. In addition, they proposed that frontal pathology might also indicate reduced executive skills, however the evidence base regarding mobility changes in ID remains sparse.

Understanding the progression of dementia in this population is complicated by the heterogeneity within ID and abilities that might typically be assessed already being compromised (Elliott-King et al., 2016), as well as approaches to research. Often research has investigated dementia in ID and DS separately, and far less research is conducted in individuals with ID without DS (Takenoshita et al., 2020). Ethical issues surrounding consent and capacity create barriers to research and reduce the representativeness, as those with more severe and profound ID may be excluded due to being unable to give valid consent (Goldsmith & Skirton, 2015). Sub-sections of the ID population are often researched in silos of ability (i.e., mild-moderate ID, severe and profound ID) again limiting applicability of findings. Hence, there is a need to look beyond the results of individual studies to gain a wider perspective of the prodromal features of dementia in this population as a whole.

1.2.3 Aim of the review

Of note, this is not the first review of early markers of dementia within the ID/DS populations. Lautarescu et al. (2017) conducted a review exploring the early presentation in people with DS, highlighting cognitive and behavioural changes that are observed, namely EF and Behavioural and Psychological Symptoms of Dementia (BPSD) emerging prior to memory impairments. Similarly, Devshi et al. (2015) focused on non-cognitive aspects of functioning, specifically BPSD, comparing the prevalence of these symptoms to the general population. However, the papers within this review focused either on behavioural or psychological symptom

separately, rather than BPSD as a whole concept. Both reviews acknowledge the methodological shortcomings of included studies including the availability and use of appropriate measures and suggest factors such as dementia classification criteria and validity of measures as accounting for discrepancies between studies. Since these reviews were published, more research into the early markers of dementia in the ID population has been conducted, focusing on both cognitive and non-cognitive aspects of functioning, including mobility and functional skills.

To the best of the authors knowledge, a systematic review combining early cognitive and non-cognitive markers of dementia in ID has not been conducted, encompassing wider aspects of functioning such as mobility. Failure to fully understand the symptomology and early presentation of dementia has implications for diagnosis and management (Cipriani et al., 2018). Importantly, a holistic view is required to help identify clinical changes, rather than viewing symptoms in discreet silos as is often the case. Despite dementia being a national priority, individuals with ID are often neglected in policy (Burke et al., 2018) further reinforcing health inequalities they face. It is key for there to be a clearer understanding of the early presentation of dementia in the ID population to inform clinical practice. Therefore, this systematic review aimed to explore the early markers of dementia in ID, including both cognitive and non-cognitive symptoms. The results will be presented, critically evaluated and implications for clinical practice and future research discussed.

1.3 Method

A systematic review was conducted using the Preferred Reporting Items for Systematic Reviews and Meta-Analyses Statement (PRISMA; Page et al., 2021). The review was registered on PROSPERO in December 2023 (CRD42023480122).

1.3.1 Search Strategy

The online databases of PsychInfo, PubMed and Web of Science were searched for articles published between 1st January 2015 to 17th November 2023. Articles from 2015 onwards were included to avoid duplication of previous reviews, as Lautarescu et al. (2017) included articles up to 21st January 2015. Search terms and operators used are presented in Appendix A. Search terms were kept broad to encompass a wide range of literature which otherwise might not have been captured. In addition, a hand search of reference lists of included studies was undertaken.

The initial database searches yielded a total of 3821 papers which were screened for duplicates and papers which violated the limiters, reducing this to 2574 papers. A systematic selection process was employed which involved screening titles, abstracts, and full text articles.

An additional 11 papers were identified through hand searching and were screened using the same limiters and inclusion/exclusion criteria.

1.3.2 Inclusion and Exclusion Criteria

Inclusion criteria for the review included the following: papers were empirical, published in peer-reviewed journals, written in English and recruited a sample of individuals with ID and/or DS aged 18 years and older. The focus of the paper had to be on identifying early signs of dementia in this population, however participants were not required to have a formal diagnosis. The exclusion criteria included animal studies, studies exploring biological or genetic markers, and studies describing dementia symptoms generally (i.e., not early stages).

Article searches and initial screening was performed by the author (JD). The following two screening stages (titles/abstracts, and full texts) employed a second reviewer (AM) to independently review a sub-section of articles (20%). Any disagreements were resolved by a third reviewer (WD). At the title screening stage, inter-rater reliability between JD and AM was 56%. When a third reviewer was consulted, agreement between JD and WD was 90%. Full texts were then screened, with 80% agreement between JD and AM. Discrepancies were again reviewed by WD, with 100% agreement between JD and WD. Screening decisions were recorded and compared using the Rayyan software for systematic reviews (Ouzzani et al., 2016). Following the selection process, 18 papers were included in the review (see Figure 1).

1.3.3 Quality Assessment

A quality assessment was completed for the individual articles using the Standard Quality Assessment Criteria (QualSyst) tool (see Appendix B; Kmet et al., 2004). The QualSyst was chosen over other appraisal tools due to its applicability to both cross-sectional and longitudinal studies, as well as the incorporation of both quantitative and qualitative checklists. The QualSyst provides a checklist of criteria, with 'Yes', 'No', or 'Partial' responses indicating the degree to which each statement is satisfied and calculation of a summary score. Summary scores were interpreted using the following criteria: strong (≥0.80), good (0.70-0.79), adequate (0.50-0.69) and poor (<0.50). There is no set guidance on what score denotes 'good' quality, however a liberal cut-off of 0.55, as suggested by Kmet et al. (2004), was applied. Scores closer to one indicate higher quality studies. Quality assessments were completed by the author (JD), with a sub-section of 20% rated by a second reviewer (AM).

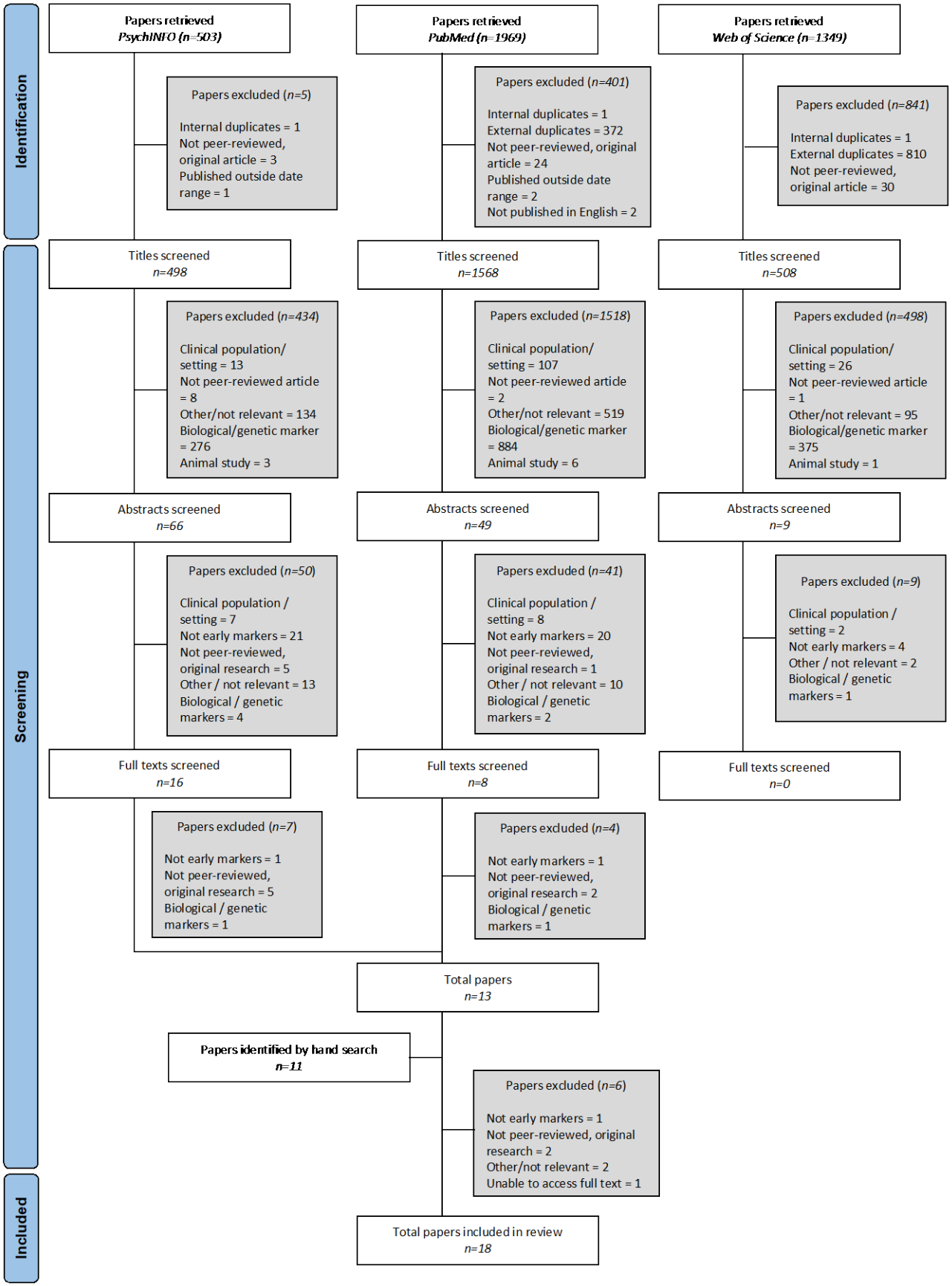
1.3.4 Data Extraction

The process of data extraction was completed by author, JD. For each included study, information regarding the following was extracted: study location, population, sample size, sample characteristics (age and gender), grouping of participants based on dementia status, methods of classifying dementia diagnosis, domains assessed, measures used and relevant findings relating to early symptoms.

1.3.5 Data Analysis

A narrative synthesis was conducted to explore and compare study findings. This approach was chosen instead of meta-analysis due to the heterogeneity of included studies specifically the measures used, and functions examined. The synthesis was based on methods proposed by Popay et al. (2006) which suggested there are four key elements to narrative synthesis that should be explored in an iterative manner: development of theory, preliminary synthesis of findings, exploration of relationships within the data, and assessing the robustness of the synthesis.

Figure 1. Flowchart of Screening Process for Selected Studies



1.4 Results

Descriptive statistics of the included papers (*n*=18) are summarised in Table 1, along with an overview of the key findings in Table 3.

1.4.1 Descriptive Statistics

Numerous studies exploring the early symptoms of dementia in the ID and DS populations have been conducted since 2015, focusing on cognitive and non-cognitive factors. Four studies were multi-national studies (2,7,9,11) including the United Kingdom, Spain, United States, Netherlands, Belgium, France, Italy, and Spain. Of the single nation studies, most were conducted in the United Kingdom (8,13,15, *n*=3), United States (12,14,16, *n*=3), the Netherlands (5,17,18, *n*=3), and Spain (3,4,10, *n*=3). Individual studies were conducted in Brazil (6) and Finland (1).

The majority of studies recruited adults with DS, however these varied regarding age. Several studies included adults with DS of any age (*n*=8)[2], adults over 20 years (*n*=1), over 25 years (*n*=2), over 30 years (*n*=2), over 35 years (*n*=1), and over 40 years (*n*=3). One study recruited individuals over 34 years with ID including DS (1), whilst another recruited care professionals and family members of individuals with ID (17). The reported mean age of participants ranged from 36.6 to 69.9 years with an overall mean of 46.6 years (*SD*=9.6). Two studies did not report mean age of participants and therefore were excluded from the summary statistics.

Sample sizes varied across studies, ranging from 28 to 524 participants (*M*=189.8, *SD*=131.0, median = 153). As might be expected, multi-site studies tended to have larger samples however many single site studies achieved samples greater than 200 (*n*=5). The majority of studies utilised a non-experimental cross-sectional design (*n*=14), whilst a small proportion adopted a longitudinal design (*n*=3) or mixed methods (*n*=1). Of the included studies, most conducted between-group comparisons (*n*=14) with a combination of comparisons to control groups, between dementia categories or both. Both cognitive and non-cognitive domains were explored in nine studies, only cognitive domains were explored in seven studies and only non-cognitive domains were explored in two studies. The majority of studies (*n*=16) reported information regarding participant's gender. Of these there was an approximately equal female to male ratio (female *M*=48.22%, *SD*=5.35, range 41.2-61%). Graphs displaying descriptive characteristics of the studies are displayed in Appendix C.

[2] This included studies that did not specify a specific age range of participants and those which considered participants as ≥16 years as adults.

Table 1. Descriptive Characteristics of Included Studies

Study No.	Authors & Publication Year	Country	Sample Size (*N*)	Dementia Groups Included	Mean Age (SD)	Female Number (%)	Participants	Design
1	Arvio & Bjelogrlic-Laakso (2021)	Finland	230	8 DS with AD	51 (NR)	102 (44.3)	Adults with ID, 34 years+	Cross-sectional, no comparator. Quantitative
2	Aschenbrenner et al. (2021)[1]		312	No diagnosis at baseline visit	44.5 (NR)	150 (48.1)	Adults with DS, 35 years+	Retrospective, longitudinal, no comparator
		UK (2 sites) Spain US (2 sites)	*n*=122 *n*=128 *n*=62		47.4 (6.6) / 44.1 (5.2) 43.8 (6.5) 42.8 (4.6) / 44.6 (7.0)			Quantitative
3	Benejam et al. (2015)	Spain	90	Absence of decline, *n*=75 DS-DAT group, *n*=15	36.1 (9.8) 51.1 (5.1)	33 (44) 9 (60)	Adults with DS	Single-centre, cross-sectional, dementia comparator group. Quantitative
4	Benejam et al. (2020)[2]	Spain	343	Asymptomatic (aDS) Prodromal AD (pDS) AD dementia (dDS)	41 (18.5)[3] aDS = 37 (15.0) pDS = 51 (4.5) dDS = 53 (8.5)	169 (49.0) aDS=132 (38.5) pDS=18 (5.2) dDS=19 (5.5)	Adults with DS	Single-centre, cross-sectional, between group comparison. Quantitative
5	Blok et al. (2017)	Netherlands	68	None	43.4 (9.5)	28 (41.2)	Adults with DS	Cross-sectional, no comparator. Quantitative

Study No.	Authors & Publication Year	Country	Sample Size (*N*)	Dementia Groups Included	Mean Age (SD)	Female Number (%)	Participants	Design
6	Conceição et al. (2023)	Brazil	66		40.9 (10.6)	28 (42.2)	Adults with DS, 20 years+	Single-centre, cross-sectional, between group comparison. Quantitative
				Stable, *n*=36	38.8 (10.4)	14 (38.6)		
				Prodromal, *n*=10	42.0 (12.1)	6 (60.0)		
				Dementia, *n*=20	44.3 (9.9)	8 (40.0)		
7	Dekker et al. (2021)[4]	Netherlands Belgium France Italy Spain	524	No dementia = 292 Questionable dementia (DS+Q) = 119 Clinically diagnosed dementia (DS+AD) = 113	DS = 47.4 (9.0) DS+Q = 53.8 (7.7) DS+AD = 57.3 (6.4)	DS = (48.3%) DS+Q = (51.3%) DS+AD = (38.9%)	Adults with phenotypical diagnosis of DS, over 30 years of age	Cross-sectional, between group comparisons. Quantitative
8	Firth et al. (2018)[5]	UK	283 (YA, *n*=119, OA, *n*=164)	Dementia and no dementia	YA = 25.2 (5.6) OA = 49.6 (7.5)	YA = 62 (52.1) OA = 77 (47.0)	Clinical diagnosis of DS aged 16 years+	Cross-sectional. YA group used as control group. Quantitative
9	Fonseca et al. (2020)	UK Brazil	162	AD, Prodromal dementia, stable	42.5 (8.2)	69 (42.6)	Adults with DS, over 30 years+	Cross-sectional, between group comparisons. Quantitative
10	Garcia-Alba et al. (2019)	Spain	41	CN-DS, MCI-DS, AD-DS	69.9 (0.6)	25 (61.0)	Adults with DS Control group: healthy adults without DS	Cross-sectional, between group comparisons and control group. Quantitative

Study No.	Authors & Publication Year	Country	Sample Size (*N*)	Dementia Groups Included	Mean Age (SD)	Female Number (%)	Participants	Design
11	Hartley et al. (2020)[6]	US UK	118	No clinical diagnosis at time point one Later categorised: CS, MCI-DS, AD, unable to determine	37.2 (7.7)	61 (52)	Adults with DS, aged 25+ with 2 or more data collection time points between 2010 - 2019	Longitudinal, data collected over 5 times points, dementia groups not used as comparator. Quantitative
12	Hom et al. (2021)[7]	US	144	CS, *n*=103 MCI-DS, *n*=41	CS = 48.7 (6.3) MCI-DS = 52.9 (6.7)	CS = 43.7% MCI-DS = 34.2%	Adults with DS, 40 years+	Cross-sectional, between group comparison. Quantitative
13	Mgaieth et al. (2023)[5]	UK	302 (YA, *n*=132, OA, *n*=170)	Dementia and no dementia	YA = 26.1 (5.4) OA = 49.6 (8.0)	YA = 73 (55.3) OA = 79 (46.5)	DS, 16 years+	Longitudinal, cohort study, between group comparison. Quantitative
14	Pulsifer et al. (2020)	US	168	CS (57.8%), MCI (22.6%), probable/definite dementia (19.6%)	51.4 (7.1) CS = 49.0 (6.6) MCI = 53.6 (6.9) Probable/definite = 55.6 (5.9)	72 (42.9) CS = 42 (43.3) MCI = 12 (31.6) Probable/definite = 18 (54.5)	Adults with DS above 40 years+	Cross-sectional, between group comparisons. Quantitative

Study No.	Authors & Publication Year	Country	Sample Size (*N*)	Dementia Groups Included	Mean Age (SD)	Female Number (%)	Participants	Design
15	Startin et al. (2019)[8]	UK	297	Preclinical, prodromal, clinical, missing	16-30 = 22.89 (4.11) 31-35 = 32.60 (1.30) 36-40 = 38.00 (1.44) 41-45 = 43.17 (1.31) 46-50 = 47.92 (1.31) 51-55 = 52.83 (1.46) 56-60 = 57.75 (1.40)	16-30 = 48 (51.1) 31-35 = 17 (56.7) 36-40 = 13 (48.1) 41-45 = 8 (33.3) 46-50 = 22 (42.3) 51-55 = 23 (54.8) 56-60 = 17 (60.7)	DS, 16 years+	Cross-sectional, between group comparisons. Quantitative
16	Van Pelt et al. (2020)	US	28	No dementia, Other/possible	36.6 (7.0)	13 (46.4)	Adults with DS, 25 years+	Cross-sectional, between group comparisons. Quantitative
17	Wissing et al. (2022)[9]	Netherlands	100 (survey respondents) 14 interviews	Suspected dementia	NR	NR	Care professionals and family members of people with ID and suspected dementia	Mixed methods, no comparator
18	Wissing et al. (2023)	Netherlands	141	No dementia, *n*=103 Questionable dementia, *n*=19 Dementia, *n*=19	No = 64.6 (1.1) Q = 61.0 (8.9) D = 65.6 (10.1)	NR	Adults with severe, severe to profound, or profound ID, aged 40+, with or without DS	Cross-sectional retrospective analysis of clinical records, between group comparisons. Quantitative

Notes. [1] This paper contains details of five study sites across three countries which each administered different test batteries across different cognitive domains. The combined study characteristics are summarised here. [2]This paper presented descriptive statistics as medians and interquartile ranges. [3]Sample descriptives are presented as median ages and interquartile ranges in parenthesis. [4]This study was conducted with informants rather than directly with people with DS. 954 informants completed the measures for 524 individuals with DS. As such, 186 interviews were conducted with one informant, 250 were conducted with two informants, and 88 were conducted with three informants. Demographics are reported for the individuals with DS. [5]This paper split the original sample into two age categories: younger adults (YA; 16-35) and older adults (OA; 36+). Age and gender descriptives are provided per group as this data was not available for the total sample. [6]This study was completed over 5 time points between 2009 and 2019. Data for time point 1 is presented. [7]Only data for the CS and MCI-DS groups were analysed in this study. [8]This study divided the original sample into 7 age groups. Descriptive statistics are reported for each age group. [9]This study reported descriptive statistics for informants completing the surveys and as such are not reported here.

Abbreviations: AD = Alzheimer's Disease, DS = Down's Syndrome, AD-DS = met criteria for AD, CN-DS = did not meet criteria for AD or MCI, CS = Cognitively Stable, DS-DAT – Down Syndrome – Dementia of the Alzheimer's Type, MCI = Mild Cognitive Impairment, MCI-DS = met criteria for MCI, NR = not reported.

1.4.2 Quality of Included Studies

Quality ratings are displayed in Table 2. Inter-rater agreement ranged from 71% to 86%, with ratings by JD ranging from 0.64 to 0.86 (*M* = 0.78, *SD* = 0.06) and ratings by AM ranging from 0.77 to 0.90 (*M* = 0.83, *SD* = 0.04). Most discrepancies reflected differences of opinion on the assignment of 'Yes' versus 'Partial' responses. All studies received ratings above the 0.55 cut-off. For quantitative papers, two were rated as adequate, seven were rated as good, and eight were rated as strong. The mixed methods paper was rated as good and above. All studies stated the objective, this was not always explicit but could be inferred and appeared to draw conclusions consistent with the reported results. Reasons for lower scores were related to study design, lack of comparator groups, and limited control of confounding variables, with only a selection of studies controlling for age and ID severity. Few studies provided clear estimates of variance, and the mixed methods paper failed to discuss researcher reflexivity.

1.4.3 Dementia Diagnosis

Many studies categorised participants based on already established dementia diagnosis, or retrospective categorisation using clinical judgement and/or neuropsychological evaluation. Although this enables between group comparisons (stable cognition, preclinical, prodromal, and clinical) which can be helpful in considering differences in functioning, the definitions of these categories varied between studies, or were ambiguous. Ten studies included groups which were categorised under three main domains: stable cognition (preclinical and participants without dementia broadly defined as the absence of decline, no decline beyond normal ageing, or asymptomatic), suspected dementia (prodromal and probable dementia defined as the presence of symptoms which do not meet diagnostic criteria), and clinical dementia (where diagnostic criteria are met). Three studies included an 'uncertain' or 'missing' group where dementia status could not be determined however these individuals were excluded from the analysis. One study included only suspected dementia. These variations complicate comparisons across studies.

Approaches to determining dementia status are included in Table 3 with varying degrees of detail provided. Several studies used clinician judgement of neurologists and neuropsychologists reviewing medical records whereas other studies included standardised assessments. The lack of inclusion of standardised tests increases the subjectivity associated with dementia groups and increases the risk of bias. Several studies failed to include any comparator group which limits the inferences that can be drawn regarding early symptoms.

Table 2. Quality Assessment for the Included Papers using the QualSyst Tool

Author and Title	Checklist item														Summary score (JD)	Second rater summary score (AM)	Inter-rater agreement
	1	2	3	4	5	6	7	8	9	10	11	12	13	14			
Arvio & Bjelogrlic-Laakso (2021)	1	2	2	1	n/a	n/a	n/a	1	2	1	1	0	1	2	0.64	-	-
Aschenbrenner et al. (2021)	2	1	2	2	n/a	n/a	n/a	2	2	2	1	0	2	2	0.77	0.82	86%
Benejam et al. (2015)	2	2	2	1	n/a	n/a	n/a	2	1	2	2	1	2	2	0.86	0.82	71%
Benejam et al. (2020)	2	2	2	2	n/a	n/a	n/a	1	2	1	2	1	2	2	0.86	-	-
Blok et al. (2017)	2	1	1	1	n/a	n/a	n/a	2	1	2	2	1	2	2	0.77	-	-
Conceição et al. (2023)	2	2	2	2	n/a	n/a	n/a	1	1	2	1	1	2	2	0.82	0.83	86%
Dekker et al. (2021)	2	1	1	2	n/a	n/a	n/a	2	2	2	2	2	2	1	0.86	-	-
Firth et al. **(**2018)	2	2	1	2	n/a	n/a	n/a	2	2	1	0	1	1	1	0.68	-	-
Fonseca et al. (2020)	2	2	1	2	n/a	n/a	n/a	1	2	2	1	0	2	1	0.73	-	-
Garcia-Alba et al. (2019)	2	2	1	1	n/a	n/a	n/a	1	1	2	1	1	2	2	0.73	-	-
Hartley et al. (2020)	2	2	2	2	n/a	n/a	n/a	1	2	1	1	0	2	1	0.73	-	-
Hom et al. (2021)	2	2	1	2	n/a	n/a	n/a	2	2	2	1	1	2	2	0.86	-	-
Mgaieth et al. (2023)	2	1	1	2	n/a	n/a	n/a	2	2	1	1	2	2	2	0.82	-	-
Pulsifer et al. (2020)	2	2	2	2	n/a	n/a	n/a	1	2	1	2	1	2	2	0.86	-	-
Startin et al. (2019)	1	2	1	2	n/a	n/a	n/a	1	2	2	1	2	2	2	0.82	-	-
Van Pelt et al. (2020)	2	1	1	2	n/a	n/a	n/a	1	2	2	0	2	2	2	0.77	-	-
Wissing et al. (2022)																	
Quantitative checklist:	1	2	2	1	n/a	n/a	n/a	1	n/a	2	0	n/a	2	2	0.72	0.77	71%
Qualitative checklist:	1	2	2	1	2	2	2	2	2	0	-	-	-	-	0.80	0.90	80%
Wissing et al. (2023)	2	1	2	2	n/a	n/a	n/a	1	1	2	1	1	2	2	0.77	-	-

Note. Responses scored as 0 = No, 1 = Partial, 2 = Yes, n/a = Not Applicable. Cell shading represent responses to statements: green = 'Yes', orange = 'Partial', red = 'No'.

Table 2. Summary of Key Findings of Included Studies

Study No.	Study	Method for classifying dementia diagnosis	Cognitive domains assessed	Non-cognitive domains assessed	Key Findings
1	Arvio & Bjelogrlic-Laakso (2021)	Individuals diagnosed prior to the study	Orientation, forgetfulness, speech	Mood, personality, functional skills, social interaction, behaviour	Common dementia symptoms: • Loss of energy, self-care skills, forgetfulness, and variable mood. No age-related differences
2	Aschenbrenner et al. (2021)	Not part of methodology	Language, visuospatial ability, memory, orientation, executive function, attention, praxis, perception	N/A	Cognitive domains sensitive to early decline: • Memory • Language • Selective attention • Praxis
3	Benejam et al. (2015)	Retrospective screening	Memory (free and cued recall)	N/A	Healthy DS group: • ↓ mCRT scores with age - poorer recall and greater number of errors. DS with dementia group: • Poorer performance across all mCRT domains. • DS with early-stage dementia showed little benefit from cueing, indicating difficulties with storage of information.
4	Benejam et al. (2020)	Classified by neurologists and neuropsychologists	Memory, attention, language, orientation, praxis, abstract thinking, perception	N/A	↓ in CAMCOG-DS and mCRT scores along the dementia continuum. Sig. differences between mild and moderate ID groups in asymptomatic DS but not prodromal DS.

Study No.	Study	Method for classifying dementia diagnosis	Cognitive domains assessed	Non-cognitive domains assessed	Key Findings
5	Blok et al. (2017)	Not part of methodology	Episodic memory	Temperament, adaptive functioning	Decline was observed across all domains. Earliest deterioration – episodic memory. No evidence changes in adaptative functioning or personality occur before changes in memory.
6	Conceição et al. (2023)	Classified by clinicians utilising neuropsychological assessments (CAMCOG-DS and CAMDEX-DS) using multidisciplinary approach	Orientation, language, memory, attention, praxis, abstract thinking, perception	Balance, gait	Poor gait performance a predictor of prodromal dementia and clinical dementia.
7	Dekker et al. (2021)	Based on clinical judgement using multidisciplinary evaluation, informant interview(s) and medical reports. Established prior to study	N/A	BPSD	Changes in frequency and severity of: • Anxious, sleep-related, restless and stereotypic, irritable, apathetic, depressive, and eating/drinking behaviour Higher frequency and severity of behaviours in the DS+AD group, lowest in DS group. ↑ observed in 1/3+ of DS+Q group in: • Anxious, sleep-related, irritable, apathetic, and depressive behaviour • Might be early signs of AD in DS
8	Firth et al. (2018)	Based on pre-existing clinical diagnosis after	Visuospatial memory, object memory, orientation, rule	Motor ability, co-ordination, adaptive skills, social skills	• Early decline in memory, sustained attention, motor co-ordination, and verbal fluency • Changes in executive abilities occur later

Study No.	Study	Method for classifying dementia diagnosis	Cognitive domains assessed	Non-cognitive domains assessed	Key Findings
		comprehensive clinical assessment	learning and set shifting, working memory, planning, semantic verbal fluency, attention		• Behavioural changes observed by informants occur last Test of memory and sustained attention most useful for tracking decline in preclinical/prodromal stages.
9	Fonseca et al. (2020)	Established using CAMDEX-DS, ICD-10 and DSM-IV criteria.	Executive dysfunction	Disinhibition, apathy	Prodromal group showed greater impairments in memory and executive skills than stable cognition group. • Deterioration begins with memory deficits and executive dysfunction (prodromal) • Disinhibition and apathy manifest later • More amnestic initial presentation in the presence of earlier frontal changes in DS compared to general population.
10	Garcia-Alba et al. (2019)	Categorised determined based on participants meeting criteria for MCI or AD, or not. Criteria described in paper	Memory, executive function, orientation	N/A	• MCI-DS group poorer performance on CAMCOG-DS, DVM, TO, WM, and ADVM measures compared to CN-DS – possible cognitive markers of MCI in DS • AD-DS group poorer performance on verbal and working memory compared to MCI-DS • Transition from MCI to AD characterised by worsening global cognition, an increase in temporal disorientation, and marked amnesic deficit.

Study No.	Study	Method for classifying dementia diagnosis	Cognitive domains assessed	Non-cognitive domains assessed	Key Findings
11	Hartley et al. (2020)	Diagnostic consensus including at least 3 clinicians, considering medical history, informant reports and ratings, evaluation of adaptive skills, direct assessment.	Episodic memory, visual attention, executive functioning	Motor planning, co-ordination	The Cued Recall Test emerged as a promising indicator of transition from preclinical to prodromal AD.
12	Hom et al. (2021)	Comprehensive assessment and consensus review by staff at study sites	Language, executive function, memory, visuomotor skills.	N/A	Language/executive abilities, memory and visuomotor skills predicted MCI-DS status. Differences based on modelling system used: • Path modelling – language/executive most affected by prodromal AD • Structural equation modelling – memory scores impacted first, followed by visuomotor scores, followed by language/executive scores.
13	Mgaieth et al. (2023)	Completed by specialists using comprehensive assessments and medical records. Assessments included cognition, and adaptive functioning	Verbal fluency	N/A	Semantic verbal fluency observed as an early indicator of cognitive deterioration. ↓ in words produced and increase in intrusion errors.

Study No.	Study	Method for classifying dementia diagnosis	Cognitive domains assessed	Non-cognitive domains assessed	Key Findings
14	Pulsifer et al. (2020)	Clinical status determined via consensus review by staff and researchers who had direct contact with individual, use of assessment results, and pre-existing level of cognitive functioning.	Language, verbal fluency	N/A	Assessment of language skills can aid early detection of cognitive decline • Receptive language a predictor of MCI-DS. • Semantic verbal fluency was identified as the strongest predictor of AD-DS. • Informant reports are able to identify early stages (CS to MCI-DS).
15	Startin et al. (2019)	For those 36+ years and no clinical diagnosis prior to the study, 2 psychiatrists reviewed CAMDEX-DS scores.	Memory, executive function	Motor co-ordination	Changes in memory and attention identified as domains most sensitive to progression from preclinical to prodromal dementia. • Prodromal group performed worse on several memory, executive and attentional measures than the preclinical group. • Memory and attention measures most sensitive to progression from pre-clinical to prodromal stage
16	Van Pelt et al. (2020)	NINCDS-ADRDA AD criteria used by panel of neurologists and neuropsychologists	N/A	Gait	Greater dual-task effects on gait velocity based on dementia diagnosis. No association between dementia and velocity, step length or base width. Dual-task gait may serve as an indicator of early-stage dementia in DS.

Study No.	Study	Method for classifying dementia diagnosis	Cognitive domains assessed	Non-cognitive domains assessed	Key Findings
		to reach consensus diagnosis.			
17	Wissing et al. (2022)	Not reported	Memory, planning, problem solving, orientation, visuospatial skills	BPSD, motor function	Most frequently observed symptoms included deterioration in: • ADL functions, BPSD (irritability, eating/drinking, anxious and apathetic behaviour). Cognitive and motor changes were observed to a lesser extent.
18	Wissing et al. (2023)	Data extracted from clinical records	Memory, orientation, language, object recognition, executive skills, visuospatial	ADLs, BPSD, motor function	Higher frequency of symptoms for those with diagnosed or questionable dementia. Most prevalent early symptoms: • Memory impairment, reduced mobility, increased anxious, apathetic and irritable behaviour.

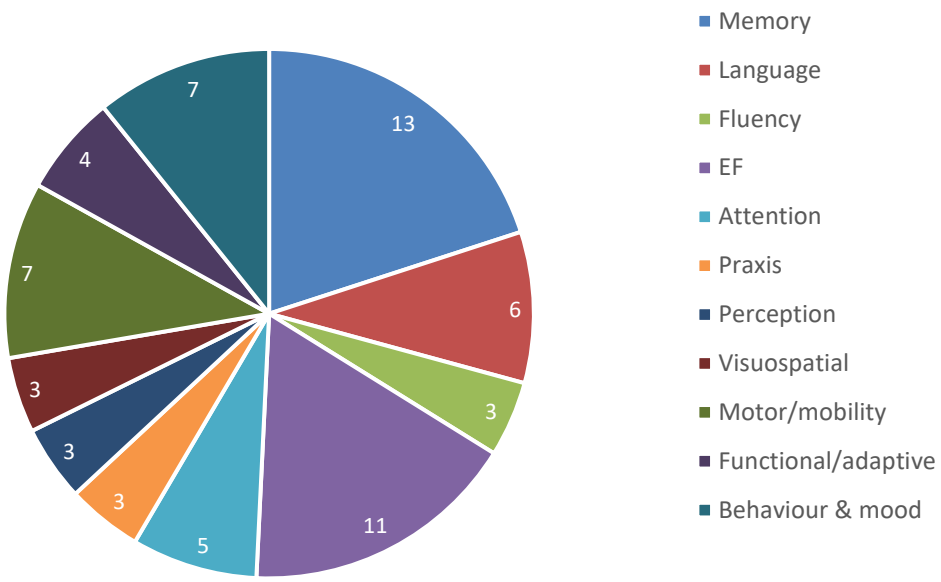
Abbreviations: ABS = Adaptive Behavior Scale, AD = Alzheimer's Disease, ADL = Activity of Daily Living, ADVM = Auditory Delayed Verbal Memory, BFT = Brief Praxis Test, BNT = Boston Naming Test, BPSD = Behavioral and Psychological Symptoms of Dementia, BPSD-DS II = Behavioral and Psychological Symptoms of Dementia in Down Syndrome II, BRIEF-A = Behavior Rating Inventory of Executive Function-Adult version, BRIEF-P = Behavior Rating Inventory of Executive Function-Parents, CAMCOG-DS = Cambridge Cognitive Examination for Older Adults with Down's Syndrome, CAMDEX-DS = Cambridge Examination for Mental Disorders of Older People with Down's Syndrome and Others with Intellectual Disabilities, CANTAB = Cambridge Neuropsychological Test Automated Battery, CEFA = Cambridge Executive Functioning Assessment, CN = did not meet criteria for AD or MCI, CS = Cognitive Stable, DLD = Dementia Questionnaire for People with Learning Disabilities, DMR = Dementia Questionnaire for Persons with Mental Retardation (now DLD), DS = Down Syndrome, DS+Q = Down Syndrome with Questionable Dementia, DSMSE = Down Syndrome

Mental Status Examination, DVM = Delayed Visual Memory, ID = intellectual disability, IED = Intra/Extra Dimensional Shift, KBIT = Kaufman Brief Intelligence Test, MCI = Mild Cognitive Impairment, mCRT = modified Cued Recall Test, MMSE = Mini-Mental State Examination, N/A = not applicable, NEPSY-II = A Developmental Neuropsychological Assessment 2, NINCDS-ADRDA = National Institute of Neurological and Communicative Diseases and Stroke/Alzheimer's Disease and Related Disorders Association, OMQ = Observer Memory Questionnaire, PAL = Paired Associates Learning, POMA = Performance-Oriented Mobility Assessment, PPS-LD = British Present Psychiatric State-Learning Disabilities Scale, PPT = Purdue Pegboard Test, PPVT = Peabody Picture Vocabulary Test, RADD-2 = The Rapid Assessment of Developmental Disabilities – Second Edition, RBMT = Rivermead Behavioural Memory Test, SIB = Severe Impairment Battery, SRT = Simple Reaction Time, SRZ/SRZ-i = Social Disability Scale for the Mentally Retarded, TO = Temporal Orientation test, TUG = Timed Up and Go test, TVZ = Temperament Scale for People with Intellectual Disability, VAT = Visual Association Test, VMI = Visuo-motor Integration, WM = Working Memory

1.4.4 Cognitive and Non-Cognitive Measures

A vast range of measures were used to assess cognitive and non-cognitive functioning. Appendix D displays the measures included and their frequency of use. Overall, 66 different measures were used, including direct assessment batteries (*n*=7), tests (*n*=11), tasks (*n*=37), and informant-based measures (*n*=10). Figure 2 shows the proportions of domains assessed across the studies. Generally, there was a bias towards assessing memory, with 13 studies (72%) exploring aspects of memory, closely followed by EF with 11 studies (61%) examining this. Far fewer studies explored BPSD, perception, visuospatial abilities, praxis, and fluency.

Figure 2. Frequency of Domains Assessed across Included Studies



1.4.4.1 Cognitive Domains

A range of cognitive domains were assessed across the included studies including language, memory, orientation, EF, visuospatial skills, praxis, perception, rule learning and verbal fluency.

1.4.4.1.1 Memory. 11 studies suggested that memory changes indicate early decline however, a selection of studies looked at memory as a broad construct whereas several studies looked at specific processes within memory, including episodic memory, recognition, cued recall, immediate memory, auditory memory, and visual memory. When considering memory broadly, the findings suggest memory performance declines across the AD continuum and that

changes occur early, distinguishing those with prodromal dementia from healthy/asymptomatic individuals. Benejam et al. (2015) concluded that memory changes were characterised by poorer recall, increased intrusion errors and a lack of benefit from cueing, indicative of a storage deficit. Specifically, episodic memory was cited as the earliest marker of decline in DS samples (Blok et al., 2017; Hartley et al., 2020), along with immediate memory (Blok et al., 2017), visual and verbal memory (García-Alba et al., 2019). One study found that adults with DS with higher amyloid-β burden, demonstrated poorer episodic memory and greater decline over time. Importantly, changes associated with memory were observed in individuals with DS regardless of the presence of dementia, concordant with previous research suggesting some degree of memory decline is expected with ageing (Nagdee, 2011).

The large variation in measures used is problematic. Several studies utilised the modified Cued Recall Test, which is specifically designed to assess verbal episodic memory in DS and demonstrates good validity and sensitivity to prodromal AD (Krinsky-McHale et al., 2022). Other studies used batteries designed for the ID/DS populations (e.g., CAMCOG-DS), or memory subtests from these (e.g., Cued Recall taken from the NEPSY-II). Measures developed for the general population were used in other studies, e.g., the Rivermead Behavioural Memory Test (RBMT). There are issues with using tests such as these which are not clearly validated for ID populations.

1.4.4.1.2 Executive Function. EF was the second most commonly assessed domain across the included studies. These studies found that EF difficulties might emerge earlier in the progression of dementia in ID than previously thought. Three studies commented that EF was impaired early in dementia, with participants in the prodromal groups demonstrating poorer performance on EF measures than those at preclinical stages. When considering specific constructs of EF, García-Alba et al. (2019) found working memory was poorer in the prodromal group compared to controls. However, this deterioration was less marked than delayed visual memory, suggesting deficits in storage and retrieval, rather than manipulation of information. A selection of other studies exploring EF found deterioration in these skills were implicated later, stating that constructs such as planning, set-shifting and disinhibition occur following memory changes (Firth et al., 2018; Fonseca et al., 2020). Using path modelling, Hom et al. (2021) demonstrated that EF (combined with language) was most impacted by prodromal status suggesting it is sensitive to early change. In contrast, when structural equation modelling was used, EF was impacted less than memory. The authors also found some EF measures had to be removed from the analysis due to high error rates, likely resulting from difficulties with comprehension of task instructions, as well as measures expected to load onto EF instead loading

on to language. Despite this, the findings still suggest significant deterioration of EF abilities in the prodromal stage.

Although a number of studies identified EF skills as early markers, Firth et al. (2018) suggested that even though executive dysfunction may be present within the prodromal stage, these are not the first changes observed. Fonseca et al. (2020) also argued that memory deficits better distinguished prodromal dementia from stable cognition than EF. They concluded that there is an initial amnestic presentation, however this occurs in the presence of earlier frontal changes than in the general population.

1.4.4.1.3 Attention. Two of the included studies reported attention to be an early indicator of decline. Aschenbrenner et al. (2021) suggested that selective attention was sensitive to early change, however they only used a single test (cancellation) to measure attentional abilities. Conversely, Firth et al. (2018) concluded that sustained attention was more useful in tracking decline. Through event sequencing models, they found multiple areas of cognition were implicated as being associated with early decline indicative of dementia pathology. Although, they noted that many of these domains were underpinned by attentional abilities. Rather than the results directly indicating a role of attention in the early stages of dementia, they deduced that these attentional difficulties may be an underlying factor in decline in other areas.

1.4.4.1.4 Language and Verbal Fluency. Verbal fluency is often considered an EF construct however, a selection of studies specifically evaluated verbal fluency across stages of dementia. Verbal fluency was reported by two studies as an early marker, and the strongest predictor of clinical dementia status in another study. Three studies reported language to be an early indicator, with one study specifying that receptive language was the best predictor (Pulsifer et al., 2020). Changes across language and verbal fluency were characterised by poorer recall and increased intrusions. Similar to memory and EF, modelling approaches differ in their outcomes regarding language with one approach suggesting language is more impacted than memory in the prodromal stage, whilst other approaches suggest the opposite. In later stages, semantic verbal fluency appears to be a strong predictor of clinical dementia (Pulsifer et al., 2020).

1.4.4.1.5 Other Cognitive Domains. Additionally, there were single instances of praxis, temporal orientation, and visuomotor skills being reported as early indicators. Hom et al. (2021) concluded that, although lower scores on measures of visuomotor skills are associated with prodromal dementia, these skills are affected to a lesser extent than memory, language, and EF. Similarly, García-Alba et al. (2019) concluded that although poorer temporal orientation was

observed in those with prodromal dementia and likely characterised transition into clinical dementia, verbal and working memory decline was more significant in the early stages.

1.4.4.2 Non-Cognitive Domains

The non-cognitive domains explored within the included studies can be divided into three groups: mobility/motor functions, behaviour/personality characteristics (including BPSD, and functional skills (including ADLs).

1.4.4.2.1 Mobility and Motor Skills. Three studies suggested that motor functions were impaired in the early stages, particularly reduced gait, mobility, and motor co-ordination. Although the majority of studies exploring mobility/motor symptoms also assessed cognitive abilities, these were often reported separately and therefore it is unclear whether mobility changes occurred alongside, before or after changes in cognition. Conceição et al. (2023) suggested that memory and scores on mobility measures were strongly correlated, such that memory deterioration was associated with declines in gait performance (initiation, symmetry, and continuity), with gait being a significant predictor of prodromal dementia. Wissing et al. (2023) concluded that both prodromal and clinical dementia groups demonstrated more motor symptoms than those without dementia, with the prodromal group being characterised by reduced walking skills, stiffness, and decreased balance. Decline in motor co-ordination was also considered in the early stages of dementia (Firth et al., 2018). Other papers suggested that motor changes are observed to a lesser extent than other non-cognitive symptoms (Wissing et al., 2022). However, these findings are based on informant reports for individuals with suspected dementia and may be affected by recollection bias. Praxis was also evaluated by several studies, although only one paper suggested this was sensitive to early decline (Aschenbrenner et al., 2021).

1.4.4.2.2 Behavioural and Psychological Symptoms. In total, seven papers included measures of BPSD and personality, however not all of these included measures of cognition. Common themes included the presence of variable mood and changes in BPSD symptoms, including increased anxious behaviour, irritability, and apathy. Most of the papers reporting these findings compared stable, prodromal and clinical groups and indicated these changes were present within the prodromal group, suggesting these may be early markers of decline. However, this was not a consistent finding across studies. Firth et al. (2018) concluded that behavioural changes reported by informants, occur last when compared to cognitive symptoms such as memory, attention, and EF. Fonseca et al. (2020) also concluded that changes in apathy manifest later than amnestic symptoms. Sleep-related changes were also indicated to

occur in the prodromal stage (Dekker et al., 2021) however this was the only study to draw conclusions regarding sleep.

1.4.4.2.3 Functional and Adaptive Skills. Four papers evaluated functional changes. Changes in ADLs were observed by Wissing et al. (2022) in a sample of individuals with severe/profound ID. They found the prodromal stage was characterised by changes in dressing and eating/drinking skills whereby there was a reduction in how independently individuals could complete these tasks. Similarly, Blok et al. (2017) found that decline in adaptive functioning was observed for individuals with DS. Both papers concluded that this decline was not present before changes in memory. Measures pertaining to functional skills and ADLs were informant-based, which are subject to response bias, including both recall and interpretation of symptoms. It is plausible that respondent's memory of symptom onset was inaccurate, and that symptoms were not conclusively due to dementia.

1.4.5 Determining the Trajectory of Decline

Although all the included studies were interested in changes indicative of early decline, it remains unclear what comes first. An added complication in determining trajectory of decline comes from the study methodologies. For example, a proportion of included papers did not employ between-group comparisons or compared those with clinical dementia to healthy controls. Therefore, it cannot be inferred at what stage symptoms first present. Nevertheless, a number of studies compared those considered as healthy, in the prodromal stage, and with a clinical diagnosis. When considering these papers, a range of conclusions are drawn.

In keeping with research in the general population, five papers concluded that changes in memory are one of the earliest indicators of decline. Several papers suggest this occurs first, whereas other papers argue memory decline occurs alongside changes in other domains such as attention, language, EF, and motor co-ordination. Several studies reported changes in EF and behaviour occur in the early stages, however Blok et al. (2017) found no evidence that these changes occur before memory decline. Firth et al. (2018) suggested a pattern of decline initiated by memory loss, reduced fluency, attention, and motor co-ordination, followed by changes in planning skills and cognitive flexibility, and finally behavioural symptoms. Fonseca et al. (2020) concluded that stable cognition and prodromal dementia are better distinguished by the presence of memory impairments than executive dysfunction, signifying a larger impact of memory decline in the early stages than EF. However, they acknowledged that executive dysfunction underpinned by frontal changes may occur earlier than observed in the general population.

Four papers suggest the presence of other symptoms early in the progression of dementia including BPSD, verbal fluency and gait performance however they do not directly compare this with the onset of memory decline. The few studies which do draw conclusions regarding the pattern of symptom onset did not use comparator groups and instead based their findings on age, inferring dementia pathology was likely in the older participants despite the lack of diagnostic support. Although these findings may be due to more than the ageing process, definitive conclusions from this cannot be drawn.

1.5 Discussion

1.5.1 Summary of Findings

This review aimed to explore symptom onset in dementia, particularly in the prodromal stage, and to consider the trajectory of decline, building upon previous research into the early symptoms of dementia within the ID/DS population. These findings provide further evidence for the presence of memory decline in the early stages of dementia in this population and add to a growing body of research which suggests that other abilities are also impacted early in the course of dementia.

Memory, attention, and EF were considered to be early symptoms across many of the included studies, while a selection of other studies suggested declines in language, fluency, motor ability, and BPSD also occur early in the disease process. Dementia is often associated with memory impairments, particularly episodic memory due to atrophy of the medial temporal lobes (Twamley et al., 2006). Memory decline is often considered to occur first in dementia in both the general and ID populations (Ball et al., 2008; Deb et al., 2007a) therefore, it is unsurprising that many studies reported memory as an early symptom. Hippocampal dysfunction is considered a mechanism in the onset of memory problems as this structure is involved in encoding and memory consolidation (Lott & Dierssen, 2010; Rao et al., 2022), consistent with storage deficits observed by Benejam et al. (2015). With respect to EF, a large proportion of the included studies placed emphasis on this also being an early feature, consistent with evidence that these difficulties might emerge earlier than previously thought (Deb et al., 2007a). Changes in attention were also observed although fewer studies explored this. It is somewhat surprising that few studies prioritised attention considering it is fundamental to many cognitive abilities (Lezak et al., 2012). Nevertheless, the presence of changes in selective attention is supported by previous research whereby a progressive pattern of decline in selective attention was observed in early AD (Krinsky-McHale et al., 2008). These individuals performed significantly worse than healthy peers on a cancellation task and demonstrated greater variability in performance over trials. This study

also used a cancellation task to measure selective attention therefore these findings may not be generalisable beyond this measure.

Regarding non-cognitive domains, the findings that mobility and BPSD changes might emerge as early indicators of decline are, in part supported by previous research, although views are mixed. Ball, Holland, Hon, et al. (2006) concluded personality and behavioural changes are prominent features of AD even in the absence of memory decline. Conversely, Adams and Oliver (2010) observed that behavioural changes in DS only occur in those also demonstrating cognitive decline. It is plausible that changes in non-cognitive abilities are underpinned by changes in cognition, rather than an independent feature. Hughes et al. (2020) found a relationship between EF and mobility in older adults without ID, suggesting that executive dysfunction might be linked with greater decline in mobility. Despite this, mobility decline may be more readily observed in the prodromal stage, compared to memory or EF particularly in the ID population.

Although this review adds to the understanding of symptoms thought to be sensitive to early decline in dementia, there continues to be a lack of consensus regarding the markers of prodromal dementia. It is unclear which domain is affected first with many studies drawing conflicting conclusions. Despite this, the current review supports the notion that the course of dementia in ID/DS differs to that of the general population. It is readily acknowledged that better understanding and detection of early dementia symptoms is necessary to provide the best support for individuals (Llewellyn, 2011). However, there are numerous challenges to the assessment of dementia, including individual heterogeneity, pre-existing cognitive deficits, and a lack of standardised approaches (Ballard et al., 2016). Exploring domains beyond memory may be a helpful next step in improving dementia diagnosis in this population.

1.5.2 Clinical Implications

Although previous research suggests dementia within ID and DS populations progresses systematically rather than manifesting globally (Devenny et al., 2000), this review describes in more detail areas that might be implicated early on and suggest memory, EF, language, attention, BPSD and motor/mobility should be included in routine dementia assessments. It is important that professionals and family members are aware of these factors, as they often raise concerns first. This may help individuals receive earlier assessments, tracking change from an earlier timepoint, leading to more timely diagnosis. Subsequently this can aid the implementation of appropriate post-diagnostic support. With a clearer definition of prodromal dementia in ID, this opens up scope to develop measures which are sensitive to this specific stage of decline (Krinsky-McHale et al., 2020).

In light of these findings, policies need to evolve (Burke et al., 2018). If non-cognitive symptoms do emerge prior to cognitive decline, guidance should be reviewed to encourage assessment of these domains and ensure baseline assessments are broad in nature (Prasher et al., 2015). Although informant-based assessments often incorporate social scales including measures of practical skills, mood, activity, interest, and behavioural disturbance, direct measures of these areas are less well utilised. Currently, the majority of tools used to assess dementia in ID rely heavily on memory measures, with limited inclusion of other domains. It is important that a well validated measure is established, encompassing a broader range of domains, including but not limited to EF, language, and mobility.

1.5.3 Methodological Considerations

Several methodological limitations need consideration when attempting to explore these early symptoms. Firstly, although many studies had large samples, the dementia groups tended to have few participants, reducing the generalisability of conclusions. Although between-group comparisons and longitudinal methods allowed exploration of group differences and change over time, the limited number of participants within the groups reduces the statistical power. Additionally, the differences between studies in their approach to group classifications is problematic. Four papers did not use a comparator group, three papers compared healthy controls and those with dementia, one paper compared healthy controls with possible dementia, whilst ten papers used multiple groups (healthy, prodromal, clinical). The absence of groups, particularly the prodromal group, makes it difficult to isolate symptoms that develop prior to a clinical diagnosis. Consequently, despite some studies reporting memory, language, and praxis as being sensitive to early decline (Aschenbrenner et al., 2021), clear conclusions cannot be drawn. Only three studies were longitudinal. These studies have the benefit of tracking change over time, rather than comparing heterogeneous groups as in cross-sectional studies (Wang & Cheng, 2020). The longitudinal studies could also review scores retrospectively, in light of later diagnoses. All except one study recruited only individuals with DS, rather than ID therefore, although this review provides evidence regarding early markers for this population, it does not add to the evidence base for ID more broadly.

A further limitation relates to the conceptualisation of cognitive functions. For example, a proportion of studies explored verbal fluency as a separate construct however other studies combined it with language or EF. This lack of consistency has implications for whether verbal fluency is considered an early marker. Additionally, there are issues related to the measures used and their lack of validity in ID populations. Martin et al. (2000) found that although individuals with ID could complete the RBMT, its appropriateness for determining memory impairments is

unclear. It is possible that the findings of this review reflect differences in methodologies and the range of measures used, rather than true group differences.

The impact of normal ageing and premorbid abilities cannot be discounted. The majority of studies explored dementia in those with mild and/or moderate ID. In previous research exploring assessment tools for dementia in ID, individuals with severe/profound ID have been screened out (Crayton et al., 1998). Therefore, it is difficult to determine trajectories across the ID population, and whether variation in symptomology is based on pre-existing cognitive abilities. Aschenbrenner et al. (2021) highlighted that premorbid abilities influenced effect sizes, suggesting those with severe ID demonstrated poorer performance and subsequently had a lower baseline than other participants. This limited the degree of decline that could be measured across time points. Similarly, to previous reviews in this area, many included studies only provided details of clinical characteristics rather than comparisons between domains and the course of decline (Lautarescu et al., 2017). Subsequently, it remains unclear how symptoms develop over time in this population.

1.5.4 Strengths and Limitations of the Review

A strength of this review is its attempt to combine research across multiple domains, rather than looking at abilities in isolation. The scope of this review was initially broad, to reduce the likelihood of not capturing relevant articles. However, a limitation is that the final iteration of studies used samples primarily of individuals with DS, therefore these findings cannot be applied to the ID population. It is likely that this is more a reflection of the evidence base lacking studies exploring dementia in ID, rather than the review methodology. Additionally, memory was the most studied aspect of cognition which could bias the conclusions, suggesting it is more common as an early symptom than it truly is.

Another limitation is the lack of consistency in how studies categorised dementia groups, and the inclusion of studies where longitudinal or between-group comparisons were not adopted. This resulted in difficulties synthesising and comparing data across studies. Additionally, the vast heterogeneity of measures used further complicates this. A smaller, more homogeneous selection of measures would be of benefit in synthesising the findings and in providing recommendations of suitable measures for use in detecting dementia in this population. Previous reviews have encountered similar challenges, with minimal overlap in measures used and suggest this is a reflection of the lack of standardised measures available (Paiva et al., 2020). The quality assessment indicated the majority of studies were of good or strong quality. Objectives and samples were sufficiently described however, methods of comparison and the control of

confounding variables were lacking, which are important factors for determining differences in prodromal dementia. There was variability in the detail provided regarding participant recruitment and selection, and the robustness of outcome measurement therefore it is unclear whether lower scores are due to methodological issues or a lack of clear reporting. Although the assessment tool used encouraged reviewers to consider sources of bias and their impact, interpretation was largely subjective. The use of a higher cut-off threshold (e.g., 0.75) may have been helpful in dealing with methodological issues such as the lack of comparator groups.

1.5.5 Recommendations for Future Research

Future research should seek to further verify the trajectory of decline in dementia for this population, focusing on prodromal features. Much of the evidence base continues to utilise samples consisting predominantly of individuals with DS. As a result, our understanding of early dementia in ID across the range of cognitive abilities remains limited. This review has established further evidence for a differing picture of dementia progression than previously assumed. It is imperative that additional, methodologically sound research is conducted to provide clarity regarding early symptoms and progression, with clear criteria for classifying prodromal dementia and adopting validated measures for this population. Research should seek to use a more focused range of measures, encompassing important domains including EF, attention, language, mobility, and BPSD. Furthermore, much of the current research has focused on Western samples. Future research should consider dementia in different ethnicities, ensuring the individuals with ID and DS from ethnic minorities are also represented and their needs considered.

1.5.6 Conclusion

In conclusion, the narrative synthesis presented provides insight into the early symptomology of dementia in ID and DS, pulling together a far wider range of domains than previous reviews. Memory, EF, language, attention, behaviour, and motor skills/mobility have been proposed as early indicators of prodromal dementia in this population and provides support for the need of broader assessments when concerns regarding dementia are raised. There still remains a lack of agreement as to the onset of these symptoms chronologically. Further methodologically sound studies are needed to explore this further.

Chapter 2 The Development and Validation of a New Battery for Dementia Assessment in the Intellectual Disability Population: The Neurocognitive Battery for Intellectual Disabilities

Journal choice: The journal '*Aging, Neuropsychology, and Cognition'* has been used as a guide to prepare this paper. The named journal specifies that manuscripts can be any length. There are no restrictions on word count, number of figures, or amount of supporting information. Therefore, a word limit of 10,000 words has been used.

Word count: 9,999 (including abstract, excluding tables, figures, and references)

2.1 Abstract

Individuals with intellectual disability (ID) are at greater risk of dementia than the general population. Although national guidance has been published detailing approaches to the assessment, diagnosis, and care of people with ID and dementia, there is a lack of clear recommendations regarding assessment tools. Several assessment tools are already available however they are limited by their psychometric properties, lack of validation within this population, and tendency to focus on memory decline. A new battery, the Neurocognitive Battery for Intellectual Disabilities (NBID), has been developed to address such limitations, incorporating measures of a broader range of functions, including executive skills, language, and mobility. This paper details the continued development and validation of the NBID. Participants (n=23) recruited from NHS sites completed the NBID to establish its psychometric properties. Results suggest the NBID is a reliable measure for individuals with ID, demonstrating good internal consistency (α=.829). Sub-groups based on age and pre-existing cognitive ability were derived. Significant differences were observed between age groups and ability groups, suggesting the presence of distinct groups and feasibility of developing statistically based change cut-offs. The majority of cognitive domains demonstrated significant correlations to one another. The mobility domain was not significantly correlated with any other domain. Floor and ceiling effects were present across a number of subtests within the battery. Although the sample was small, this new battery appears to show promise as a tool for assessing dementia in the ID population. Suggestions for further refinement of the battery are considered.

2.2 Introduction

There is vast heterogeneity in cognitive and functional abilities amongst individuals with intellectual disability (ID; British Psychological Society [BPS], 2015). However diagnostic classification systems, such as the Diagnostic and Statistical Manual of Mental Disorders (DSM-5; American Psychiatric Association, 2013) and the International Classification of Diseases (ICD-11; World Health Organization, 2019) are guided by three core principles:

- Presence of significant impairment in intellectual functioning (typically IQ<70)
- Presence of significant impairment in adaptive functioning
- Onset before the age of 18

It is estimated that there are 1.3 million people with an ID in England (Public Health England, 2023). Down's Syndrome (DS) is one of the most common genetic causes of ID and is associated with early onset of cognitive decline linked with dementia and shorter lifespan (Ballard et al., 2016). Improvements in medicine and social care have increased life expectancy for people with ID and DS (Coppus, 2013; Department of Health, 2001) subsequently increasing the chances of these individuals' developing dementia. However, our understanding of the ageing process and its consequences, such as health issues and the trajectory of cognitive decline, remain less well understood than in the general population (Walker, 2015). Typically, individuals with ID face increased health and social inequality (Emerson & Baines, 2011; McMahon & Hatton, 2021), which is further compounded by the presence of comorbidities such as dementia, adding further complexities to the care and support individuals with ID may require (Llewellyn, 2011).

2.2.1 Dementia

Dementia is a neurodegenerative condition typically associated with changes in memory, however changes in other domains, such as language and executive skills, are also important clinical features (National Institute for Health and Care Excellence [NICE]; 2018b). Typically, a diagnosis of dementia is one of exclusion (Lezak et al., 2012), pulling information from multiple sources regarding current level of functioning and changes in abilities, as well as the use of standardised screening measures. Screening measures are based upon data collected via large scale studies exploring performance of normative samples (i.e., of healthy functioning individuals) and determining cut-off scores where dementia is suggested if performance falls below this (Strauss et al., 2006). This approach is based on the assumption that the majority of individuals premorbid performance would be within the average range in relation to intelligence (Lezak et al., 2012). As individuals with ID do not fall within the average range regarding cognitive abilities, and

there is vast heterogeneity in their abilities, the same approach cannot be adopted for this population.

2.2.2 Dementia in the ID Population

Despite it being well documented that people with ID and DS are at greater risk of dementia than the general population (Startin, Hamburg, et al., 2016; Strydom et al., 2013; Wu & Morris, 2013) prevalence estimates vary due to methodological challenges including diagnostic barriers and ambiguity (BPS, 2015). Distinguishing symptoms of dementia from an individuals' cognitive profile related to ID presents a barrier in providing accurate and timely diagnosis, particularly for those with moderate/severe ID where their functioning may be more difficult to assess due to pre-existing cognitive and communication difficulties (NICEa, 2018). Additionally, research indicates an age-related trend in dementia onset, with those with ID developing dementia earlier than the general population, and those with DS developing dementia much earlier than both the ID and general population (BPS, 2015).

Research often investigates dementia prevalence in individuals with ID with and without DS separately. Strydom et al. (2007) conducted an epidemiological study exploring dementia prevalence in a sample of 222 individuals with ID without DS aged 60 years and above, applying both DSM-IV and ICD-10 diagnostic criteria. They found that, when using DSM-IV criteria, 13.1% of participants 60 years and above and 18.3% of participants 65 year and above met diagnostic criteria for dementia. Rates were higher when using the DSM-IV compared to ICD-10 criteria which often excluded those with mild and moderate dementia, conceivably related to differing sensitivity and specificity of the diagnostic tools.

Most epidemiological research has focused on the prevalence of dementia in DS. Holland et al. (1998) found a sharp increase in prevalence rates across the lifespan, with estimates of 3.4% for those aged 30-39, 10.3% for those aged 40-49 and 40% for those aged 50-59 years. Coppus et al. (2006) conducted a large-scale prevalence study in the Netherlands and found overall prevalence rates were 16.8% in their sample of 506 people with DS aged 45 years and over. When grouped by age, they found prevalence's of 8.9%, 17.7%, 32.1% and 25.6% for up to 49 years, 50-54 years, 55-59 years, and 60 years and over respectively. Likely explanations of the decrease in prevalence after 60 years is due to increased mortality. However, estimates for this older age bracket are inconsistent, with Tyrrell et al. (2001) reporting 41.7% prevalence of dementia for those 60-69 years and 50% for those above 70 years, despite a similar overall prevalence (13.3%). Although these studies are helpful in considering the extent and impact of dementia in the ID and DS populations, they are not without limitations. Much of the research demonstrates inconsistent

findings, with some describing rates of dementia in ID populations to be in line with that of the general population (Zigman et al., 2004). This may reflect methodological differences i.e., cross-sectional vs. longitudinal, sampling methods and representativeness, and the diagnostic criteria adopted (Strydom et al., 2010). The exact reasons for higher prevalence rates and earlier onset of dementia in this population remains unclear however there are a number of hypotheses, including pre-existing levels of cognitive ability, genetic components, and additional comorbidities.

Variability in onset and degree of cognitive change is hypothesised to be linked with degree of intellectual disability prior to the onset of change. This theory, based on the reserve capacity model (Stern, 2002), suggests that a higher degree of premorbid difficulties reflects an earlier onset and faster deterioration in cognitive abilities, due to increased neuropathological susceptibility (Bush & Beail, 2004; Strydom et al., 2009). Although a plausible theory in understanding the vast differences in onset of cognitive deterioration in these populations, agreement of an operationalised concept remains unclear (Stern, 2002) with 'reserve' encompassing constructs such as cognitive reserve, brain reserve and brain maintenance (Stern et al., 2020). Anderson et al. (2020) reviewed the relationship between ID severity and dementia, finding a sparsity of evidence for how ID severity is associated with dementia prevalence and presentation. Margallo-Lana et al. (2007) found those with more severe ID showed less decline on cognitive assessment, however another study found more rapid decline in cognitive performance (Oliver et al., 1998). A number of other studies found no association between severity of ID and the development of dementia (McCarron et al., 2014; Strydom et al., 2009).

Models of Alzheimer's Disease (AD) suggest the neuropathology for this sub-classification of dementia is an overabundance of amyloid-β in the brain, leading to the development of plaques and neurofibrillary tangles which disrupt the transmission of nerve impulses (Hardy & Selkoe, 2002). The gene responsible for amyloid-β production, the amyloid precursor protein (APP), is located on chromosome 21, where there is a triplication in DS resulting in over-expression of APP, therefore making individuals with DS more susceptible to AD (Lott & Head, 2019). Other genetic factors thought to be associated with the development of dementia in DS are additional genes found on chromosome 21 (such as the *DYRK1A* gene), and maternal linkage. These will not be discussed within the scope of this paper; however, an overview is provided by Evans et al. (2013).

Individuals with ID are generally found to have poorer physical health and multiple comorbidities due to continued health inequalities (McMahon & Hatton, 2021), including higher risk of cardiovascular disease and epilepsy (Evans et al., 2013). Evidence from the general population suggests a relationship exists between epilepsy and poor cognitive performance across

a range of domains including memory, EF, attention, and processing speed, and subsequently an increased risk of dementia (Sen et al., 2018). However, there is a lack of large, population-based research to draw firm conclusions about the association between dementia and epilepsy (Subota et al., 2017).

2.2.3 Dementia Assessment in the ID Population

Ideally, a similar process to detecting dementia in the general population would be available for individuals with ID. However, the large degree of variability in cognitive profiles for this population possess a challenge as baseline levels of functioning cannot be assumed, and comparisons to normative samples is not possible. Floor effects (clustering at the lowest possible sore) are also common, particularly where tools have been adapted from the general population (Elliott-King et al., 2016). Even measures developed or adapted specifically for the ID population demonstrate the presence of floor effects (O'Caoimh et al., 2013) impacting the measures sensitivity. Many existing assessment tools demonstrate high floor effects when assessing those with severe ID, despite the absence of floor effects for those with mild ID (Paiva et al., 2020). Ceiling effects (clustering at the highest possible value) are also problematic. Burt et al. (1998) suggested it may not be appropriate to apply ICD-10 diagnostic criteria for dementia to the ID population. For example, if individuals are not first assessed until they are in the later stages of dementia, they may become untestable before a repeat assessment can be completed and therefore not demonstrate sufficient decline on formal measures to meet diagnostic thresholds. This issue could be overcome through routine baseline assessments as proposed in professional guidelines (BPS, 2015; NICE, 2016). Research has previously addressed the bias introduced by floor effects by utilising IQ cut-offs to exclude individuals with more severe ID who would likely perform largely at floor level (Fleming et al., 2022).

An additional difficulty of assessing dementia in this population is the assumption that the disease progression mirrors that of dementia in the general population. Strydom et al. (2010) argued the presentation of dementia in individuals with ID, with and without DS, is dissimilar to the presentation of dementia in the general population. They highlighted changes in speech, personality and behaviour, and functional decline appear to be an earlier feature. Deb et al. (2007a) conducted a series of focus groups and interviews with carers of individuals with DS and dementia to explore their perspectives on early changes. They found that although there were similarities in presentations between individuals with DS and the general population who developed dementia, several symptoms associated with frontal lobe functioning, such as loss of interest in activities, emotional and behavioural changes, motor slowness, and reduction in

functional skills were evident much earlier in the course of dementia for those with DS. Consequently, alternative approaches to diagnosis are required.

Approaches to dementia assessment in this population involve baseline assessment, prospective monitoring, and reactive screening. Due to available resources, many services within the UK adopt a reactive approach (Jethwa & Cassidy, 2010), whereby an assessment of functioning is completed once concerns are raised. However, this can create delays in diagnosis as follow-up assessments are required to confirm the presence of decline. This is overcome to some extent by the use of routine baseline assessments, so that services have information on an individual's baseline level of functioning prior to the onset of symptoms. Both the BPS (2015) and NICE (2016) recommend baseline assessments for individuals with DS should be completed at the age of 30, due to their increased risk of dementia. Once a baseline is established, comparisons can be made at a later time point to determine change in functioning. Prospective screening is another approach involving repeating assessment at regular intervals, with increased frequency of assessments with age, however there is often a compromise between depth of assessment and frequency (Gardner & Slater, 2021). Beresford-Webb et al. (2021) argue that there remains a need for single use assessments, which are not reliant on repeat administration due to the limited availability of baselines, particularly for individuals with ID where this is less common practice.

2.2.4 Assessment Tools and Psychometric Properties

Although guidance exists for the assessment of suspected dementia in ID (BPS, 2015; NICE, 2016) and a variety of assessment tools have been developed, there remains a lack of specific recommendations or a standardised approach to such assessments. Auty and Scior (2008) noted marked variability in dementia pathways in UK services, with individual services selecting what processes and assessments to adopt. However, response rates in their study were low (26%) which may reflect a bias in those completing the survey, such as those with a research interest being more inclined to respond than those who feel dementia cases represent a small proportion of their clinical roles. Despite the introduction of national guidance, this challenge still persists to some extent. Within the UK, dementia assessment for people with ID is typically completed by specialist community ID services, utilising both proactive and reactive assessment processes. Gardner and Slater (2021) reviewed three services within England to compare their assessment pathways. They found all three services offered proactive screening for those with DS, but not for those without DS, instead offering reactive assessments once concerns had been raised. The services all varied in the age at which baselines were offered (although all later than recommended in national guidance), frequency of follow-ups and tools used. They highlighted the lack of consistency of dementia assessments for this population across the country, even within

specialist services, and suggest little has changed despite readily accessible guidance. However, the findings from these services, which were close geographically, may not be generalisable to all ID services across England.

Several systematic reviews have been conducted exploring the available assessment tools and their psychometric properties. Zeilinger et al. (2013) reviewed 97 studies, primarily exploring the purpose of the tool (was it designed/intended for use to assess dementia), and the intended population (was it designed for use with people with ID). They identified 114 tools, including 79 direct measures and 35 informant-based measures, and four batteries. All the batteries identified had been developed specifically to assess dementia in the ID population however, this was the case for only 36 of the 114 tools. McKenzie et al. (2018) reviewed 81 papers and identified 22 tools used for assessing dementia in this population, categorising 12 of these as cognitive and 10 as behavioural tools. Although a range of assessment tools exists, they highlighted that many tools had originally been developed and utilised in the general population and then adapted for people with ID, with only a small proportion being specifically designed for the ID population. There was also a lack of clarity regarding the validity of tools. Similarly, Paiva et al. (2020) identified 39 assessment tools, including self-report and informant questionnaires, scales and inventories along with 13 test batteries to explore the presence of cognitive and behavioural change in people with ID. Despite the large numbers of measures encompassed in the review, there were a lack of available psychometrics which limit the possibility of providing further clinical recommendations.

Although there is no agreed assessment battery for this population, national guidance (BPS, 2015) recommends that any battery should assess a variety of memory domains, including prospective, short-term, and long-term visual and verbal memory, orientation, language including expressive and receptive, EF, and learning. These guidelines summarise the utility of several widely used tools, namely the Neuropsychological Assessment of Dementia in Adults with Intellectual Disabilities (NAID; Crayton et al., 1998), and the Cambridge Examination for Mental Disorders of Older People with Down's Syndrome and Others with Intellectual Disabilities (CAMDEX-DS; Ball, Holland, Huppert, et al., 2006). An overview of commonly used measures and their psychometric properties is presented in Table 4.

Table 3. Psychometric Properties of Measures Commonly Used to Assess Dementia in the ID Population

Measure	Authors & Publication Year	Reliability	Validity	Comments
Neuropsychological Assessment of Dementia in Adults with Intellectual Disabilities (NAID)[1,2]	Crayton et al. (1998)	Split-half reliability: All items Spearman-Brown Formula >.8 Ranged from .82 (picture naming) to .96 (picture identification)	Internal consistency: Cronbach's alpha ranged from .74 (picture memory) to .95 (picture identification) across subtests Concurrent validity: Ranged from .64 (acceptable) to .77 (good)	• Direct assessment • Developed specifically for ID population.
Cambridge Examination or Mental Disorders of Older People with Down's Syndrome and Others with Intellectual Disabilities (CAMDEX-DS)[3,4]	Ball et al. (2004)	Limited reliability information available Inter-rater reliability: for 91% of items, Kappa fell in the almost perfect range κ >.8. The additional items fell within the substantial agreement range κ >.6	Face validity: Considered to have good face validity Predictive validity: High specificity (.88, [95% confidence interval = .80 - .98]) and sensitivity (.94 [95% confidence interval = .53 - .98]) Concurrent validity: Good concurrent validity when scores compared to the presence of formal AD diagnosis at a later time point	• Direct assessment • Modified from CAMDEX and standardised for ID population. • Decline measured as score reduction of 1 standard deviation. • Sensitivity statistics based on 8 participants showing cognitive decline.
Cambridge Cognitive Examination Modified for use in a group with Down Syndrome (CAMCOG-DS)[4]	Ball, Holland, Huppert, et al. (2006)	NR	Predictive validity: Able to discriminate between diagnostic groups and those without dementia. Concurrent validity: High correlation with Mini-Mental State Examination (MMSE)	• Direct assessment • Standardised for use with people with ID.

Measure	Authors & Publication Year	Reliability	Validity	Comments
Severe Impairment Battery (SIB)[4,5]	Saxton et al. (1993)	<u>Test-retest reliability:</u> High when sample of individuals with ID without dementia	<u>Concurrent validity:</u> Good concurrent validity with DLD in sample of people with DS without dementia Some evidence to suggest unable to distinguish differences between groups of varying ID severity	• Direct assessment • Not specifically designed for individuals with ID or DS. Has been used with people with ID but not standardised. • Cut-off scores only provided for general population. • Research suggests not appropriate for those with severe ID.
Test of Severe Impairment (TSI)[4]	Albert and Cohen (1992)	NR	NR	• Direct assessment • Not designed for individuals with ID or specifically for detection of dementia. • Limited psychometrics in ID population. Some evidence of good validity and reliability however research has been conducted with small samples and many years ago.
Down Syndrome Mental Status Examination (DSMSE)[4]	Haxby (1989)	No available reliability information	Some evidence that DSMSE is able to discriminate between those with and without dementia for those with moderate ID.	• Direct assessment • Designed to assess age-related differences in people with DS. • Unsuitable for use with severe ID.
Cognitive Scale for Down Syndrome (CS-DS)[4,6]	Startin, Rodger, et al. (2016)	<u>Test-retest:</u> Good (.95) <u>Internal consistency</u> Good internal consistency (.96)	<u>Discriminant validity:</u> Significant differences observed between individuals with and without cognitive decline.	• Informant-based questionnaire • Designed specifically for DS.

Measure	Authors & Publication Year	Reliability	Validity	Comments
			Concurrent validity: Positive correlations found between CS-DS and KBIT-2 (*r*=.56) and short ABS (*r*=.76)	
Dementia Rating Scale (DRS)[4]	Mattis (1988)	NR	NR	• Informant-based • Not developed specifically for use in ID population. • Limited research into psychometrics for ID population. When used in the general population, found to have high test re-test reliability, and good sensitivity and specificity. The DRS demonstrated moderate-high correlation with MMSE.
Dementia Screening Questionnaire for Individuals with Intellectual Disabilities (DSQIID)[7]	Deb et al. (2007b)	Inter-rater reliability = .9 Test-retest reliability = .95	Internal consistency: Average across all 53 items α=.91 Sensitivity & specificity: When screening cut-off of 20 used, sensitivity = .92 and specificity = .97	• Informant-based
Dementia Questionnaire for People with Learning Disabilities (DLD)[4,8]	Evenhuis et al. (2007)	Inter-rater reliability: Poor inter-rater reliability. Ranged from .44 to .94 across subtests. All correlations satisfactory except behavioural disturbance.	Predictive validity: Some evidence to suggest DLD is able to discriminate between groups of individuals with and without dementia (higher scores obtained by those without dementia). Concurrent validity:	• Informant-based • Designed specifically to detect dementia in ID population. • Validation primarily completed with individuals with AD. • Less evidence for its accuracy in detecting dementia in severe ID.

Measure	Authors & Publication Year	Reliability	Validity	Comments
			Correlated highly with several other measures of cognitive decline in the ID/DS population. Sensitivity: Varies between those with ID and those with DS. ID and dementia, values between 39-85% DS and AD, values between 80-81% Specificity: Varies between those with ID and those with DS. ID and dementia, values between 57-100% DS and AD, values between 83-100%	
Dementia Scale for Down Syndrome (DSDS)[4,9]	Gedye (1995)	Inter-rater reliability = high (.91)	Specificity = 89% Sensitivity = 85% Good correlation with the DLD	• Informant-based • Developed for those with DS mostly with severe or profound ID.

Note. Data obtained from the following sources: [1]Oliver and Adams (2007),[2]Oliver et al. (2022), [3]Ball et al. (2004), [4]McKenzie et al. (2018), [5]Witts and Elders (1998), [6]Startin, Rodger, et al. (2016), [7]Deb et al. (2007b), [8]Evenhuis (2018), [9]Jozsvai et al. (2018)

Abbreviations: NR = Not reported

In clinical practice, the NAID has typically been the most widely used instrument, reportedly adopted by over 30 ID services (BPS, 2015). The NAID is a battery designed to assess cognitive domains associated with decline indicative of dementia, including memory, orientation, praxis, and language and has demonstrated acceptable levels of internal consistency and reliability at sub-scale level, with reliability ranging from .82 to .96 and internal consistency ranging from .74 to .95 (Oliver & Adams, 2007). The original development papers (Crayton et al., 1998; Oliver et al., 1998) only included participants with DS who passed a brief screening test to exclude those with significant cognitive impairments regardless of cause, therefore despite the large-scale validation of this tool, it continues to lack applicability to those without DS. Additionally, floor effects were still observed despite screening for pre-existing cognitive impairments (Crayton et al., 1998).

Until recently, robust administration and scoring guidance for the NAID has not been easily available, impacting how reliably services use this tool (Gleave & Westbrook, 2023). Another limitation is its reliance on memory measures, and lack of inclusion of measures exploring changes in EF, visuospatial skills, and language abilities, as well as changes in non-cognitive domains (e.g., behaviour, mobility, and functional skills) which have been suggested to be sensitive to early decline (Blok et al., 2017; Firth et al., 2018; Wissing et al., 2023). Oliver et al. (2022) acknowledge the need for a broad assessment, particularly in the early stages of dementia. A revised NAID (NAID-R) has been published (Gleave et al., 2023) however this failed to incorporate further measures of EF and other important domains, such as mobility. Additionally, it is unclear how readily adopted this revision has been within clinical practice at this early stage especially as the authors advised the original NAID should continue to be used for repeat assessments.

Despite the limitations, the NAID provided two options for interpretation of scores (Oliver & Adams, 2007). Either using single point score classifications similar to the approach adopted in the general population where one score is compared to what is typically expected of others of a similar ability and using Reliable Change Index to determine difference between scores at two time points (Jacobson & Truax, 1991). In order to utilise reliable change, estimates of cognitive ability were derived to create ability bands within the normative data. Adams and Oliver (2010) demonstrated the use of this approach further, using z-scores to determine change in scores and suggest this statistical approach may be more sensitive to cognitive decline than informant-based observations.

Other batteries which are frequently adopted are the CAMCOG-DS-II (Beresford-Webb & Zaman, 2021) and LonDownS cognitive test battery (Startin, Hamburg, et al., 2016). Limited information for the CAMCOG-DS-II is available regarding its structure, utility, and psychometrics,

although floor and ceiling effects appear to be low, and domains correlated well with already established measures (Baksh et al., 2023). For this reason, this battery is not described in detail here. The LonDownS battery is considered to be a valid tool (Tristão et al., 2023) however it takes a substantial amount of time to administer and consists of both computer-based and pencil-based tests which present challenges for use within clinical services. Startin, Hamburg, et al. (2016) found lower completion rates for the computer-based tasks, partly due to technical issues. Those with dementia had far lower rates of completion, with half of these individuals being unable to complete any computer-based items suggesting these items might not be suitable for those at risk of dementia.

2.2.5 The Neurocognitive Battery for Intellectual Disabilities

As discussed above, the course of dementia in individuals with ID does not necessarily follow the same course observed in the general population (Deb et al., 2007a). This raises questions regarding the use of tools which were originally developed for use within the general population, and their sensitivity to detecting dementia in people with ID and/or DS. Recent research indicates the need for common assessment tools developed with this population in mind, exploring areas including memory, EF, language, behaviour (Lautarescu et al., 2017), mobility, and functional skills (Conceição et al., 2023; Wissing et al., 2022).

Subsequently, a new battery, the Neurocognitive battery for Intellectual Disabilities (NBID), was developed in an attempt to overcome these challenges, address the significant floor and ceiling effects of currently available tools, and to offer a more valid and reliable approach to the assessment of cognitive decline in ID (Campbell et al., 2023). The NBID is an assessment battery specifically developed for the ID population using updated DSM-5 criteria for dementia and encompassing a wider range of skills than previously assessed. The NBID assesses eight domains including orientation, attention, EF, learning and memory, language, perceptual motor skills, social cognition, and mobility. A key feature of the NBID is the incorporation of additional measures of EF in recognition of this being an early indicator of cognitive decline in this population (Deb et al., 2007a; Fonseca et al., 2020). Campbell et al. (2023) provides a full overview of the development of the NBID and qualitative feedback from participants.

2.2.6 Aims and Rationale

The present study aims to refine the test selection, order and administration of the NBID based on preliminary findings and qualitative data from the pilot study (Campbell et al., 2023). The main scope of the current study was to explore the psychometric properties of the battery,

particularly its internal consistency. Additionally, it was hoped concurrent validity could be determined based on comparisons to validated direct and indirect measures, as well as evaluation of the factor structure to determine whether the current domains are the most accurate representation. Therefore, the present study aimed to address the following questions:

1. What are the psychometric properties of the NBID?
    a. Does the NBID have adequate internal consistency?
    b. What is the concurrent validity of the NBID when compared to existing measures?
    c. What is the factor structure of the NBID?
2. Can sub-group classifications be made using estimates of overall cognitive ability for calculating reliable change for NBID total scores?
3. When grouped by age, does performance on the NBID significantly differ between groups?
4. Is performance on the NBID influenced by psychological wellbeing?

2.3 Method

2.3.1 Design

To evaluate the psychometric properties of the NBID, a cross-sectional design was used where all participants completed the NBID alongside a selection of other measures assessing psychological wellbeing and cognitive ability (unless already available). In order to explore concurrent validity, results from previously completed assessments were requested from individuals' healthcare records. Further context of this study is provided in Appendix E.

2.3.2 Participants

A-priori power calculations were performed using G*Power 3.1 (Faul et al., 2007) to determine the minimum sample size required to test a two-tailed correlational hypothesis. Using a correlation coefficient of 0.3, estimates indicated a minimum sample size of 84 participants was required to achieve 80% power for detecting a medium effect, with a significant criterion of $\alpha = .05$. However, similar studies exploring the development and psychometrics of the NAID used samples of 70 and 57 (Crayton et al., 1998; Oliver et al., 1998). The LonDownS was validated with a sample of 305 (Startin, Hamburg, et al., 2016), Jozsvai et al. (2002)'s unnamed battery was validated with a sample of 35 while Johansson and Terenius (2002)'s instrument was validated with just nine participants.

Six NHS Trusts were contacted regarding study recruitment. In order to meet inclusion criteria, participants were required to have a confirmed ID with or without DS, be over 18 years of age, be English speaking, be open to their local community ID team, be able to provide informed consent to participate, and to be able to engage and tolerate neuropsychological testing of up to two hours. Participants were excluded if they were non-verbal or had significantly limited verbal skills which would impact their ability to complete neuropsychological testing, or experiencing any of the following: current acute distress (e.g., psychosis), temporary changes in cognitive function (e.g., related to delirium), current acute physical complaints (e.g., UTI), significant pain, or recent significant life events (e.g. bereavement, placement breakdown/changes to their living situation) that may impact their cognitive functioning.

2.3.3 Measures

2.3.3.1 Clinical Outcomes in Routine Evaluation – Learning Disability (CORE-LD; Brooks et al., 2013; Appendix F)

The CORE-LD was used to assess participants psychological wellbeing prior to completing the NBID to ensure participants were not experiencing severe psychological distress. The CORE-LD is a 14-item self-report measure widely used in clinical settings, specifically developed for individuals with ID. Participants rated questions relating to how they have felt over the previous week, including three risk questions. The CORE-LD demonstrates good internal consistency (α=.80; Brooks et al., 2013). Higher scores indicate lower levels of psychological wellbeing. As there are no clinical cut-offs, responses were screened on a case-by-case basis to consider whether participants were suitable to complete the NBID, particularly considering responses to risk items. If concern was raised regarding participants psychological wellbeing, the assessment was abandoned, and if consent was provided, contact was made with their GP outlining the findings. It was made explicit no interpretation of scores could be offered.

2.3.3.2 *Dementia Questionnaire for People with Learning Disabilities (DLD; Evenhuis et al., 2007; Copyright)*

The DLD is a 50-item informant-based screening tool which assesses change in cognitive and social functioning. The cognitive domain comprises of subscales related to short-term memory, long-term memory and orientation whilst the social domain comprises of subscales for speech, practical skills, mood, activity and interest, and behavioural disturbance. Scores for the relevant subscales are totalled to give overall domain scores. Although the psychometric properties are variable, ranging from excellent to modest (see Table 3; Zeilinger et al., 2022), the

DLD is widely used in both clinical practice and within research to aid validation studies therefore it was adopted here in order to aid comparisons to other studies.

2.3.3.3 *British Picture Vocabulary Scale 3 (BPVS-III; Dunn et al., 2009; Copyright)*

The BPVS-III is a measure of receptive vocabulary in which individuals are shown four images and asked which image best illustrates a word given by the examiner. The BPVS-III was designed for individuals aged 3-16 but is also suitable for adults, including those with ID. Ezard et al. (2022) suggest that the BPVS-III is an appropriate tool for estimating premorbid functioning in those with acquired cognitive impairment, particularly in the presence of reading or communication difficulties and can be considered a relatively stable indicator of ID severity for healthy individuals (Ball et al., 2008). As such, it is widely used as an estimate for general intellectual functioning in the ID population (Bevins & Hurse, 2016). For this study, BPVS-III scores were used as a proxy for ID severity.

2.3.3.4 Neurocognitive Battery for Intellectual Disabilities (NBID; Campbell et al., 2023; see Appendix G)

The NBID is an assessment battery developed specifically for individuals with ID, assessing eight domains of functioning. In order to increase accessibility and feasibility of the measure, several adaptations were made through co-production with pilot study participants. Changes included adding coloured images and amending scoring criteria for several subtests, particularly to give credit to recognition items when responses are freely recalled. Specific changes were made to the Adapted Trails subtest which included changes to the shape outline contrasts to aid perception, order of sequence (now required to order from biggest to smallest), and re-organisation of shapes on the page to avoid connection lines crossing. The Finger Counting task was removed due to its high ceiling. The Coding subtest was altered so that participants were required to fill in the missing numbers, rather than symbols. This would provide the option of participants providing an oral response if they experienced motor difficulties. Despite the significant ceiling effects observed on the Cats & Dogs subtest during the pilot, this task was retained for the purposes of this study. Table 5 details the domains and subtests used following the revisions made to the NBID. A more detailed description of amendments is presented in Appendix H.

Table 4. Overview of the Neurocognitive Battery Domains

Cognitive Domain	Process		Subtest
Orientation	-	1	Autobiographical Memory
Attention	Sustained Attention	8a	Digit Span – Forwards
		8b	Digit Span – Backwards
	Selective Attention	12	Coding
		15	Cancellation
Executive Functioning	Planning	6a	Adapted Trails – Sequencing
	Decision making / reasoning	6a	Adapted Trails – Sequencing
		13a	Cats & Dogs - Naming
	Working memory	6a	Adapted Trails – Sequencing
	Inhibition	13b	Cats & Dogs – Inhibition
		13c	Cats & Dogs – Switching
	Cognitive Flexibility	6b	Adapted Trails – Switching
		13c	Cats & Dogs - Switching
		13d	Cats & Dogs – Sorting
Memory & Learning	Immediate memory span	5	Immediate Recall & Learning
	Recent memory (free recall)	2	Remote Memory
		7b	Figure Drawing – Immediate Recall
		14	Delayed Recall & Recognition
		16a	Figure Drawing – Delayed Recall
	Recent memory (recognition)	16b	Figure Drawing – Visual Recognition
Language	Expressive language	3	Picture Naming
		9a	Verbal Fluency (Semantic)
		9b	Verbal Fluency (Phonemic)
	Receptive language	4	Picture Identification
		17	Language Reasoning
Perceptual Motor	Visuo-constructional ability	7a	Figure Drawing – Copy
		18	Construction
	Praxis	11a	Action of Request
Social Cognition	Emotion recognition	10	Emotion Recognition
Mobility	-	11b	Action on Request – 360° Turn

2.3.4 Procedure

Six NHS Trusts were initially approached and expressed interest in collaboration. Clinicians within these trusts were asked to share study information with individuals they identified as meeting the inclusion criteria and whom they thought would be interested in taking part. A summary flowchart of the consent and recruitment process is presented in Appendix I. Clinicians discussed the study with potential participants who then provided consent for their information to be shared with the research team in order to provide further information about the study, answer any questions, and arrange the assessment. A research team was assembled to aid the recruitment and data collection process. In addition to the author (JD) and research supervisors, the research team consisted of a Clinical Psychologist (MR), four Assistant Psychologist, and two

Trainee Clinical Psychologists within one of the recruitment sites. Participants were allocated to one of the members of the research team to conduct the assessments. Two training workshops were held (one ran by JD and the other by the site Principal Investigator, MR) regarding the administration procedure and scoring of the NBID to improve inter-rater reliability within the research team.

The full range of measures were expected to be administered in one testing session, lasting approximately 90-120 minutes, allowing time to review study documents, complete the measures and allow for any necessary breaks. However, an additional testing session could be facilitated if required. Participants were offered to complete the assessment either at their home, in a clinical setting, or day centre depending on their preference. Participants were given the option of being supported during the assessment. If a DLD had not been completed within the last three months, an informant was asked to complete this if available.

During the testing session, the information sheet (Appendix J) was reviewed, and a consent form signed (Appendix K). The CORE-LD was completed to establish participants psychological wellbeing and to open up conversations relating to factors which may impact study eligibility. Following this, the BPVS-III was administered followed by the NBID. Prior to the end of the session, participants were provided with debrief information (Appendix L) and were offered the choice of a monetary voucher for taking part. All easy read documents were co-produced. The author (JD) met with an individual with ID to appraise the study documents for their accessibility, use of language, and appropriateness of images. An overview of the study was provided, and each document was reviewed, with any language considered vague or ambiguous amended based on suggestions provided by the individual with ID.

2.3.5 Ethics

Due to the nature of the study population, a number of ethical considerations were required in the design and implementation of the study. Individuals with ID are considered vulnerable adults and may lack the ability to give consent to taking part in research activities. Additionally, it is possible that an individual's capacity may fluctuate across time points. Initial proposals included individuals who may lack capacity to consent for themselves and would therefore require consent by proxy. This was with the view to widening the participant pool and increasing inclusivity. The study aimed to collect representative normative data and therefore including individuals across a wide range of abilities in the ID population was paramount. However, NHS ethics did not approve this initial proposal and, due to time constraints, it was

decided that only individuals who are able to provide consent for themselves would be included in the recruitment strategy.

Ethical approval was obtained from the University of Southampton Research and Governance committee and NHS Health Research Authority (Appendix M). Although six NHS Trusts were approached and were included in the NHS REC approvals following expressions of interest, trust level Research and Development processes were only completed and approved for three sites (three were not followed to completion due to time constraints). Appendix N shows trust approvals for the included sites. Participants provided informed consent using easy read documents, reminded of their right to withdraw, and provided a full debrief upon completion of the assessment.

2.3.6 Data Analysis

Assessments were scored manually by the test administrator (member of the research team) following the scoring guidance provided. All assessments were checked by the lead researcher (JD) for errors and inconsistencies in scoring. Raw scores and domain scores were entered into IBM SPSS Statistics 29 for the purposes of data analysis.

Descriptive statistics were calculated for each subtest, domain and total NBID score. Normality tests were conducted to determine data distribution, as well as skewness and kurtosis. The presence of floor and ceiling effects was investigated, as well as the frequency of errors across subtests. Group comparisons were conducted via independent t-tests to explore any differences between NBID scores depending on age or cognitive ability. Median split of BPVS-III was used to determine the cognitive ability group using BPVS-III raw scores. In grouping by age, a similar approach was adopted to previous studies in this area, using the age of 35 to group participants due to research suggesting AD neuropathology (Firth et al., 2018; Mgaieth et al., 2023). To determine the internal consistency, Cronbach's alpha (α) was conducted at both subtest and domain levels. Finally, further reliability analysis was conducted as well as comparisons between NBID scores and mood measures. Factor analysis was not conducted due to the sample size. Concurrent validity using established measures of cognition was explored.

2.4 Results

2.4.1 Descriptive Statistics

Overall, 25 participants were recruited for the study. One participant withdrew on the day of the assessment due to unrelated psychological distress, while another participant's assessment

was discontinued following completion of mood measures due to risk issues. Of the remaining 23 participants, eight were female (35%) and 15 were male (65%), with an age range of 21-75 years (*M*=42.91, *SD*=15.07). In terms of general intellectual ability, BPVS-III data were available for 12 participants (52%) as well as Weschler IQ estimates for a further eight participants (30%). 11 participants were considered to have mild ID, two participants demonstrated moderate ID whilst seven participants appeared to be in the borderline ID range.

Table 6 reports the outcomes of mood and preliminary measures. CORE-LD scores ranged from 0-16 (*M*=5.39, *SD*=4.74) suggesting a degree of psychological distress for the majority of participants. However, given the nature of psychological difficulties amongst individuals with ID (Cooper et al., 2007), this finding is to be expected although it is important to consider the potential impacts on assessment scores. DLD data was obtained for four participants (17%) therefore there were limitations to how this data could be used in further analysis. NAID data was available for only one participant therefore this data could not be used in the analysis.

Table 5. Descriptive Statistics for Mood and Preliminary Measures

Measure	Subscale	*M*	*SD*	Minimum Score	Maximum Score
CORE-LD	*Total*	5.39	4.74	0	16
BPVS-III	*Raw Score*	113.50	29.13	63	148
DLD	*SCS*	3.50	1.73	2	6
	SSS	11.25	7.23	2	17

Abbreviations: SCS = Sum of Cognitive Scores, SSS = Sum of Social Scores

2.4.2 Missing Data

Items which were unable to be administered in light of participants characteristics i.e., being unable to complete a 360° turn due to mobility difficulties, were given a score of 0 to differentiate from truly missing data. Missing data were characterised by either item non-response (participant failing to provide a response to an item) or administration factors. Three individuals did not complete the Adapted Trails subtest. For two of these cases, printing errors resulted in the subtest being unable to be administered, and one participant declined to attempt. One participant did not complete Action on Request, while a further two did not complete the 360° Turn. Two participants did not complete Coding due to printing errors, one participant declined to complete Cats & Dogs Switching and Inhibition, whilst administration error for another participant during Cats & Dogs Inhibition resulted in this item not being scored. One

participant declined to complete Cancellation, Figure Delay Recall and Recognition, Language Reasoning and Construction subtests.

Missing data analysis was conducted to evaluate the impact of missing values. 12 items demonstrated missing values (42.9%), which included Trails (Sequencing and Switching), Action on Request, 360° Turn, Cats & Dogs (Switching and Sorting), Coding, Cancellation, Language Reasoning, Delayed Figure Recall (free recall and recognition), and Construction. Missing data was present for six participants (26.1%), impacting 19 values across the data set (3.0%). Appendix O shows the proportions and patterns of missing value across the data. Based on observations, data appeared to be missing at random (MAR).

Cases with missing values were not automatically removed from the analysis due to the sample size and wanting to avoid reducing this further. Methods of imputation (including mean substitution and multiple imputation) were considered to replace missing values with estimates in keeping with the pattern of data available however, due to the nature of the data, it was decided that missing values would not be replaced. Whitaker (2012) noted that cognition for those with IQ below 70 does not follow a normal distribution, as such using estimates of individuals' abilities could be considered inaccurate (Spreat, 2017). Therefore, not all analyses were conducted with the full sample size (*n*=23).

2.4.3 Data Normality

Parametric assumptions were analysed for all measures via exploration of histograms and using Shapiro-Wilk tests which are considered more appropriate for smaller samples (Mishra et al., 2019). Box plots and normal Q-Q plots were also inspected. BPVS-III raw scores were found to be reasonably normally distributed, $W(12)=.898$, $p=.151$, however CORE-LD scores were not normally distributed, $W(23)=.886$, $p=.013$. Three outliers were observed for the CORE-LD however these scores were within the scale range and considered representative of normal variations within the data and therefore were not removed.

For the NBID data, parametric assumptions were explored at both subtest level, domain level, and for total scores. When considered as a whole sample, reasonable normal distribution was found for the total NBID score, $W(23)=.948$, $p=.269$, as well as for both the Memory domain, $W(23)=.954$, $p=.357$, and the Attention domain, $W(23)=.938$, $P=.160$, and five subtests (Trails Sequencing and Switching, Figure Drawing – Immediate Recall, Coding, and Cancellation). The remaining subtests and domains demonstrated non-normal distributions and tended to be negatively skewed (see Table 6 and Table 7 respectively). When non-normal distributions were

apparent, logarithmic transformations were performed to address this however this did not result in more normally distributed data therefore analysis was conducted on untransformed variables.

2.4.4 NBID Score Distribution

Distributions for the NBID data were analysed at subtest level for the entire sample (Table 7). The percentage of participants unable to complete a subtest was calculated, along with the percentage of participants performing at floor and ceiling. For subtests where not all participants completed the task, the percentages were calculated based on the total participants to complete the subtest. Outliers were also screened. For many items where outliers were present, this was due to a large proportion of participants performing at either floor or ceiling for the subtest and therefore were not excluded due to being considered in keeping with the overall pattern of data. An overview of the total and domain scores are presented in Table 8. The 360° Turn subtest time was analysed as opposed to the number of steps, due to inconsistency in scoring.

Table 6. Distribution of Data for Each Battery Item

Subtest / Item	*n*	% not completed	% floor	% ceiling	*M*	*SD*	Median	Range	Skewness	Kurtosis
Orientation	23	0.00	0.00	13.04	17.17	2.79	18.00	9-20	-1.61	2.71
Remote Memory	23	0.00	8.70	73.91	1.65	0.65	2.00	0-2	-1.73	1.95
Picture Naming	23	0.00	0.00	39.13	10.87	1.33	11.00	8-12	-1.28	0.81
Picture Identification	23	0.00	0.00	91.30	11.87	0.46	12.00	10-12	-3.71	13.96
Memory – Immediate Recall	23	0.00	0.00	30.43	16.74	4.45	19.00	3-20	-1.87	3.32
Trails – Sequencing	20	13.04	10.00	20.00	3.80	2.40	4.00	0-7	-0.15	-1.21
Trails - Switching	20	13.04	5.00	10.00	4.15	2.43	4.00	0-9	0.42	0.02
Figure Drawing - Copy	23	0.00	4.35	13.04	8.17	3.47	9.00	0-12	-1.29	0.94
Figure Drawing – Immediate Recall	23	0.00	0.00	17.39	7.35	3.43	8.00	2-12	-0.24	-1.11
Digit Span Forwards	23	0.00	0.00	30.43	8.04	1.64	8.00	5-10	-0.08	-1.37
Digit Span Backwards	23	0.00	30.43	4.35	2.22	2.09	2.00	0-8	1.03	1.34
Verbal Fluency - Category	23	0.00	0.00	21.74	2.78	0.80	3.00	2-4	0.43	-1.25
Verbal Fluency - Letter	23	0.00	21.74	0.00	1.04	0.83	1.00	0-3	0.98	1.31
Emotion Recognition	23	0.00	0.00	0.00	3.22	1.24	3.00	1-5	0.02	-0.72
Action on Request	22	4.35	0.00	95.45	29.95	0.21	30.00	29-30	-4.69	22.00
360° Turn Time	21	8.70	13.04	-	3.43	2.27	3.00	0-10	0.83	2.42
Coding	21	8.70	9.52	4.76	23.00	15.55	19.00	0-50	0.14	-1.01
Cats & Dogs - Naming	23	0.00	0.00	91.03	15.91	0.29	16.00	15-16	-3.14	8.61
Cats & Dogs – Inhibition	23	0.00	0.00	78.26	14.35	3.76	16.00	2-16	-2.31	4.74
Cats & Dogs – Switching	21	8.70	9.52	47.62	12.00	5.33	15.00	0-16	-1.16	0.34
Cats & Dogs – Sorting	22	4.35	0.00	95.45	15.95	0.21	16.00	15-16	-4.69	22.00
Delayed Recall & Recognition	23	0.00	4.34	13.04	6.39	2.86	7.00	0-10	-0.79	-0.29
Cancellation	22	4.35	9.09	4.55	9.55	6.58	9.00	0-28	0.92	1.62
Figure Drawing – Delayed Recall	22	4.35	13.64	13.64	6.27	4.14	6.00	0-12	-0.10	-1.31
Figure Drawing – Recognition	22	4.35	13.64	18.18	1.55	0.96	1.50	0-3	0.04	-0.82
Language Reasoning	22	4.35	0.00	31.82	6.36	1.56	7.00	3-8	-0.59	-0.75
Construction	22	4.35	0.00	27.27	5.91	0.97	6.00	4-7	-0.83	0.06

Table 7. Descriptive Statistics for the Overall NBID and Domain Scores

Domain	*n*	*M*	*SD*	Median	Range	Skewness	Kurtosis
Total	23	243.48	54.90	237.00	132-318	-0.31	-.69
Orientation	23	17.13	2.93	18.00	8-20	-1.80	3.63
Attention	23	40.39	23.25	34.00	5-80	0.27	-1.18
Executive	23	63.30	13.70	67.00	20-78	-1.62	3.44
Memory	23	39.65	13.02	40.00	6-61	-0.63	0.91
Language	23	32.65	4.09	34.00	25-38	-0.69	0.91
Perceptual Motor	23	42.48	6.97	45.00	16-49	-2.71	9.30
Social	23	3.22	1.24	3.00	1-5	0.02	-0.72
Mobility	18	3.43	2.27	3.00	0-10	0.83	2.24

2.4.4.1 Floor and Ceiling Effects

The proportion of individuals who performed at floor and ceiling levels of each subtest was examined (Figure 3). For this analysis, floor effects were considered as scores of zero on any subtest whilst ceiling effects were considered as achieving the maximum possible score. Of the 27 subtests in the NBID, 13 subtests (48%) demonstrated floor effects (*M* = 5.67%, *SD* = 7.64%) whilst 24 subtests (89%) demonstrated ceiling effects (*M* = 34.07%, *SD* = 31.64%). Additionally, the impact of age on floor performance was examined (see Figure 4). Participants were grouped based on the following age ranges: 20-29 (*n*=4), 30-39 (*n*=6), 40-49 (*n*=6), 50-59 (*n*=5), and 70-79 years (*n*=2). There was no signficant correlation between floor frequency and age (r_s=.27, *p*=.301). Figure 4 shows the greatest proprortion of floor effects were present for the 40-49 age group. Comparisons for ceiling effects were not calculated due to the intended nature of several tests being for screening purpises therefore anticipating participants to score at the ceiling. Within health research, floor or ceiling effects are considered present when 15% or more responses are at the lowest or highest score respectively (McHorney & Tarlov, 1995; Terwee et al., 2007). When considering individual subtests, significant floor effects were present on Digit Span – Backwards and Verbal Fluency (Letter) whilst 16 subtests demonstrated significant ceiling effects.

Figure 3. Floor and Ceiling Effects by Subtest

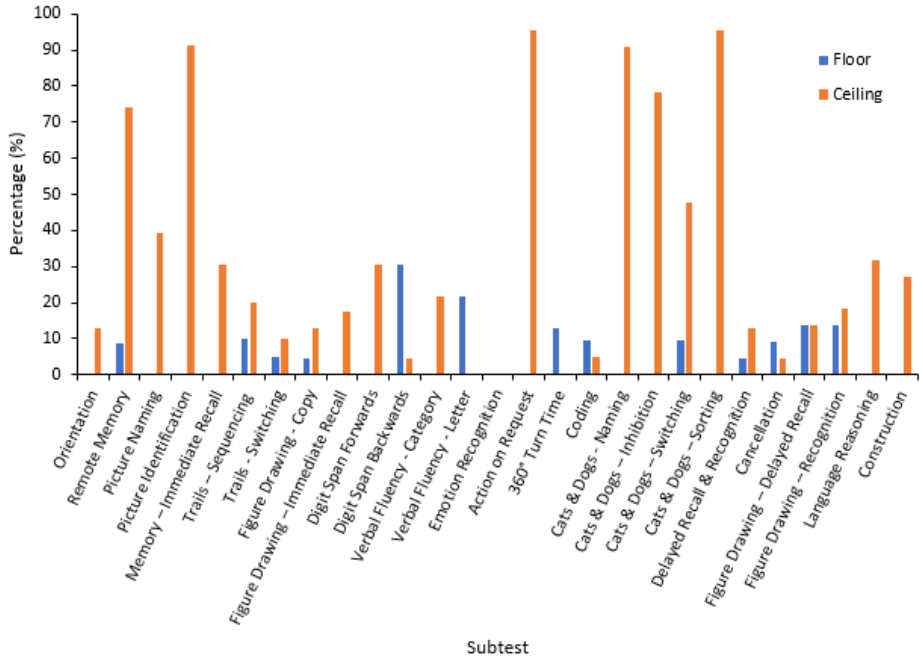
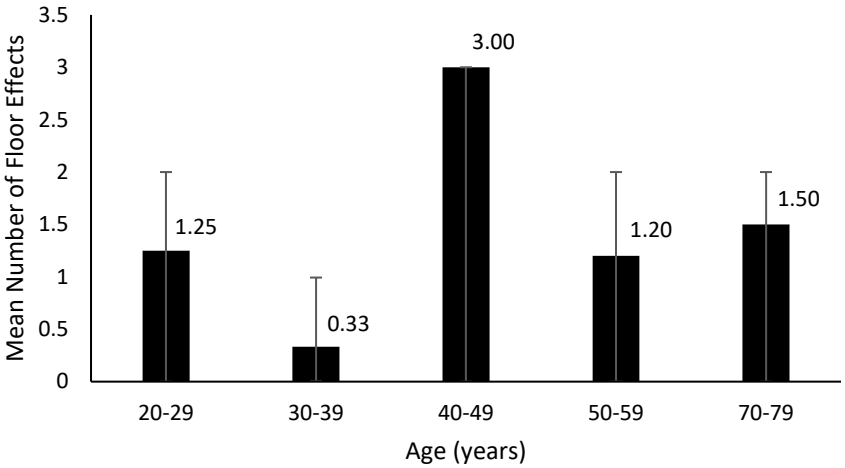


Figure 4. Mean Number of Floor Effects by Age Group



Note. The sample did not include any participants in the 60-69 age bracket. Subsequently this age bracket has been omitted.

2.4.4.2 Error Types and Frequencies

The frequency of response errors was also examined to provide further context to the NBID subtest scores (see Table 9). Several subtests provided measures of errors within the individual items. Errors were observed across other subtests, although these are not reported in detail here. Coding demonstrated the highest frequency of errors, with a total of 63 across all participants (*M*=3.00, *SD*=9.47), with 43 errors being attributed to one participant, suggesting either difficulty

in comprehension of the instructions or poorer attentional skills. Upon further inspection, this participant demonstrated high levels of errors across all subtests with error measures. Cancellation also produced a high frequency of errors, with a total of 59 errors across the sample, whilst Figure Recall – Recognition totalled 47 errors. The presence of set-loss errors (considered to be rule violations) and perseveration errors (repetition of previous response/strategy) were common for both semantic and phonemic verbal fluency, with 29 and 24 errors respectively.

Table 8. Descriptive Statistics for Error Measures

Subtest / Item	Total	*M*	*SD*	Range
Verbal Fluency – Category	29	1.26	2.40	0-11
Verbal Fluency – Letter	24	1.04	2.10	0-9
Coding	63	3.00	9.41	0-43
Cancellation	59	2.68	7.47	0-34
Figure Drawing - Recognition	47	2.14	1.76	0-6

2.4.4.3 NBID Performance by Age

NBID total score was compared by age groups of ≤35 years and ≥36 years. As scores for the ≤35 age group were not normally distributed, a Mann-Whitney U test was performed. The ≤35 age group scored higher on the NBID overall (*Mdn=288*, *n*=9) than the ≥36 age group (*Mdn*=221.5, *n*=14). This was found to be significantly different, *U*=28.00, *z*=-2.21, *p*=.027.

2.4.4.4 NBID Performance by Estimated Ability

Overall performance on the NBID was also explored based on estimates of pre-existing ability. The median score on the BPVS-III was 115. Those with scores ≥116 became a distinct group to those with scores of ≤115. A Mann-Whitney U test was performed to explore group differences. The group which scored ≥116 on the BPVS-III scored higher on the NBID overall (*Mdn*=277, *n*=6) than the group with BPVS-III scores of ≤115 (*Mdn*=217.5, *n*=6). This was found to be significantly different, *U*=3.00, *z*=-2.40, *p*=.016.

2.4.5 Internal Consistency

Cronbach's alpha (α) was computed for the total NBID score (Table 10). Internal consistency for the NBID when including all subtest scores was considered to be good (α=.829). When determined using the eight domains, consistency was acceptable (α=.772). When considering the

reliability of each domain, internal consistency ranged from unacceptable (Perceptual Motor, α=.196) to acceptable (Memory, α=.789).

Non-parametric (Spearman's Rho) correlations between the eight domains are shown in Table 9. Due to the number of comparisons made, an adjusted *p* value was used in an attempt to reduce the risk of Type 1 errors. At this adjusted level, 14 of the domain correlations reached significance. All domains showed positive correlations except for Mobility, which was negatively correlated with the seven other domains, although this was not significant. The strongest correlations were observed between Memory and Perceptual Motor (r_s=.868), Attention and Perceptual Motor (r_s=.784), and Attention and Language (r_s=.783). Except for Mobility, the Attention domain showed significant positive correlations with all other domains, although with Orientation this was at the *p*=.05 level. This suggests higher performance on attentional measures is associated with higher scores across other domains except Mobility. As expected, cognitive domains were significantly correlated, whereas the correlations between cognitive and non-cognitive domains were not significant.

Table 9. Correlation Matrix of Internal Consistency for NBID Total and Domain Scores

Domain	1	2	3	4	5	6	7	8	α
1. Orientation	-								-
2. Attention	.475*	-							.552
3. Executive	.569**	.750**	-						.475
4. Memory	.493*	.730**	.511*	-					.789
5. Language	.596**	.783**	.741**	.672**	-				.773
6. Perceputal Motor	.582*	.784**	.658**	.868**	.768**	-			.196
7. Social	.302	.668**	.635**	.464*	.554**	.423*	-		-
8. Mobility	-.223	-.143	-.107	-.189	-.234	-.328	-.258	-	-

Note. Cronbach's alpha is not reported for domains consisting of a single subtest. * denotes correlation is significant at the *p*<.05 level. ** denotes correlation is significant at the *p*<.01 level.

Upon inspection of the correlation matrix and the construction of individual items, it was decided to remove the Remote Memory subtest from data analysis. This was due to its limited scoring system (comprising of just two items), high rates of ceiling performance, and suggestion that its removal would increase Cronbach's α. Upon removal, Cronbach's α increased to .809 for the Memory domain and to .840 for the total NBID score. For the Perceptual Motor domain, item deletion calculations suggested that removal of items in this domain did not increase α to above .5 therefore items were retained.

2.4.5.1 Internal Consistency by Age

Cronbach's alpha was conducted for NBID total scores for each group to determine whether internal consistency would increase if groups were more homogenous. Internal consistency for the NBID was considered acceptable for the ≤35 group (α=.768) and good for the ≥36 group (α=.857). Internal consistency was not significantly improved by removal of items.

2.4.5.2 Internal Consistency by Estimated Ability

Internal consistency was also assessed for BPVS-III scores to explore the presence of sub-group variability in order to inform the use of reliable change statistics as demonstrated by Adams and Oliver (2010) for individuals with differing levels of cognitive ability. Again, groups were based on median split of BPVS-III raw scores. Internal consistency of the NBID for the lower BPVS-III group (≤115), was considered questionable (α=.649) whereas for the higher BPVS-III group (≥116), internal consistency was considered acceptable (α=.782). Internal consistency was not significantly improved by removal of items.

2.4.6 Correlation Between Mood and NBID

The relationship between CORE-LD score and NBID total score was investigated to determine whether psychological wellbeing impacted performance on the cognitive battery. Bivariate Pearson's correlation was used as variables were linear and there were no outliers. The correlation between CORE-LD scores and NBID total score was not statistically significant, $r(21)=-.15$, $p=.48$, 95% CI [-.53, .28]. The effect size was small, and CORE-LD accounted for 2.3% of variance in the NBID total score. As CORE-LD data were not normally distributed, bootstrapping was used to obtain bias corrected accelerated 95% CI [-.61, .34].

2.4.7 Concurrent Validity

Due to the limited data available, correlational analysis to establish concurrent validity could not be performed. NBID total scores and DLD domain scores were plotted against each other however did not provide any meaningful conclusions.

2.5 Discussion

This study aimed to further evaluate the psychometric properties of a newly developed battery for detecting dementia in the ID population, as well as to further establish its practicality and feasibility in clinical practice. Building on work by Campbell et al. (2023), amendments to the NBID were made utilising both participant and researcher feedback. This paper also aimed to

explore whether the BPVS-III could be used as an estimate of cognitive ability to determine sub-groups within the sample.

2.5.1 Summary of Findings

The present study found that the NBID is a promising assessment tool for use within the ID population. Despite the continued presence of floor and ceiling effects across several subtests, the NBID appears to be reliable, demonstrating good internal consistency. Age groups within the sample were compared against total NBID scores and showed those ≤35 years achieved a significantly higher total score than those ≥36 years. It is possible this reflects the nature of cognitive change over time for people with ID, as it is not uncommon for cognitive decline to be present regardless of dementia (Nagdee, 2011). When comparing groups based on estimated cognitive ability, those with higher scores on the BPVS-III achieved significantly higher total NBID scores than those with lower BPVS-III scores. This suggests it could be possible to develop reliable change cut-offs based on estimates of cognitive ability in order to support diagnosis. Being able to statistically determine whether change has occurred would increase the robustness of the NBID, rather than relying solely on clinician judgement to determine whether the extent of change is large enough to indicate decline consistent with dementia. Furthermore, reliable change methods could be adopted to determine the difference between scores at different time points, and whether this is greater than would be expected due to measurement error (Jacobson & Truax, 1991). It is reasonable to assume that most ID services would have a sense of an individual's cognitive ability as part of informing a diagnosis of ID. This may not be the case for older individuals who may not have a formal diagnosis of ID but are supported nevertheless by the ID service. Nevertheless, the BPVS-III appears to be an appropriate alternative method (Ezard et al., 2022; Startin et al., 2019). Oliver and Adams (2007) adopted a similar approach in validation of the NAID however it is unclear how readily adopted this was in clinical practice.

Regarding internal consistency of the NBID, the overall measure was determined to have good reliability when calculated at subtest level and acceptable reliability when calculated based on domains. It is encouraging that in both respects, the overall NBID measure appears to be reliable. This suggests that NBID subtests provided a good degree of homogeneity in measuring overall cognition for an ID population. The differences in reliability between subtest and domain calculations may be explained by how well individual subtests load onto each domain they have been assigned to, and that investigating internal consistency at subtest level partially overcomes this. The Perceptual Motor domain showed the weakest internal consistency suggesting this is not an overly reliable measure and therefore, interpreting data related to this domain should be done with caution. On review of this subtest, it consists of measures of visuoconstructional ability and

praxis which may be too distinct to be considered one domain. However, reliability analysis did not suggest that removal of items from this domain would substantially improve the reliability of the domain. Factor analysis may provide useful insights into the construction of the domains for the NBID using a larger sample.

The majority of cognitive domains appeared to be significantly positively correlated with each other, suggesting that higher performance in one domain is associated with higher performance in another. Previous research concluded that cognitive measures for individuals with lower abilities are more highly correlated than those with higher abilities (Detterman & Daniel, 1989). Non-cognitive domains were not significantly correlated with cognitive domains. Surprisingly, the Perceptual Motor domain showed the strongest correlations, particularly with Attention, Memory, and Language. Although not significant, negative correlations were found between Mobility and the other domains, suggesting that an increase in time taken to perform the 360° Turn is associated with lower scores on other measures which supports the hypothesis that poorer mobility skills are thought to be associated with cognitive decline (Conceição et al., 2023). It is possible that some correlations were found to be non-significant due to statistical power and the number of items comprising of domains, for instance, the Orientation and Mobility comprise of one subtest score, whereas other domains have several scores contributing to the domain.

Unfortunately, due to limited data, validity analysis was not conducted therefore the concurrent validity of the NBID remains unknown. Based on the above findings, and those demonstrated by Campbell et al. (2023), the measure appears to have good face validity. The NBID also appears to be robust to differences in mood and psychological wellbeing, as no correlation observed between NBID and CORE-LD scores, therefore within this sample, mood does not appear to be a confounding factor. However, it is acknowledged that high rates of psychological distress, including anxiety and low mood are common in ID (Cooper et al., 2007). The presence of floor and ceiling effects continues be of concern and indicate the need for further amendments to address this. It is possible the inclusion of individuals with borderline ID impacted the ceiling of the NBID in this study, as this rate was far greater than observed by Campbell et al. (2023) although was in keeping with the pattern of greater ceiling effects compared to floor effects. Floor effects are not uncommon and exist within other measures such as the CAMDEX-DS (O'Caoimh et al., 2013). Although ceiling effects are problematic, decline in functioning can still be measured, however the measure may be less sensitive to subtle changes.

2.5.2 Clinical Implications

The results from this study have important implications for clinical practice, particularly regarding further development of the NBID. There was evidence to suggest the presence of age differences in cognitive ability, which highlights the importance of early screening and is consistent with previous research (Firth et al., 2018; Mgaieth et al., 2023). However, this study utilised a sample of individuals with ID, which sets this study apart from much of the previous research where samples have focused on DS only.

2.5.2.1 Further NBID Developments

The findings suggest the NBID continues to demonstrate both floor and ceiling effects, although the proportion of ceiling effects is much higher. To some extent, this is expected as several subtests are intended as screening measures, where it would be expected that most healthy individuals would achieve the maximum score, for example, the Picture Naming and Identification tests. However, for other subtests which are not intended as such, e.g., the Cats & Dogs test, we must question how sensitive the measure is to EF in individual with ID. Although designed to measure cognitive flexibility in terms of response inhibition, rule maintenance and set shifting, the high rate of ceiling responses across all trials provides little meaning by way of interpretation. Previous research has indicated that the Cats & Dogs measure correlates moderately well with object memory and other measures including the DLD cognitive scale (Bevins & Hurse, 2016). Therefore, further consideration needs to be given to whether this item is retained or removed. Regardless, Ball et al. (2008) argued for the inclusion of EF measures when assessing for dementia. Further EF measures should be explored, such as the coloured Stroop, for inclusion in the NBID. Additionally, factor analysis may demonstrate verbal fluency loads more accurately onto the EF construct than language, as it is often considered an EF task (Whiteside et al., 2016).

Additionally, it will be important to extend the floor and ceiling of the NBID. Following this, consideration should be given to the inclusion of start points and discontinuation rules, similar to the structure of the Wechsler Adult Intelligence Scales (Wechsler, 2008), to reduce the overall administration time of the battery. Further scoring considerations are needed, particularly relating to acceptable responses to Picture Naming items and scoring of the 360° Turn. Despite providing training on the administration and scoring of the assessment to the research team, inconsistency was still observed therefore more thorough review of this is required.

2.5.3 Methodological Considerations

There were a range of methodological limitations to the present study. Firstly, the sample size is significantly lower than the recruitment target and therefore the study was underpowered. This is partly due to challenges faced regarding research timeframes and recruiting from only two of the approached six NHS Trusts, limiting the participant pool. However, small sample sizes are not uncommon within ID research, and previous measures have attempted to be validated with fewer participants (Johansson & Terenius, 2002). Barriers to participation exist such as ethical issues, practicalities, and lack of support (Crook et al., 2016). Additionally, due to delays with approvals from NHS trust Research and Development departments, the data collection period spanned just five weeks. The sample size likely resulted in the assessment scores not fully reflecting the distribution of scores across the ID population and therefore inferences regarding the psychometric properties must be cautious. This is further complicated by the presence of missing data. Although imputation of missing values is often suggested for MAR data, given the context of this study, it was decided that accepting missing values was more reflective of real-world performance particularly in this population, and any estimates of ability would not reflect true performance phenomenon (Spreat, 2017; Whitaker, 2012). Secondly, a large proportion of participants did not have available BPVS-III scores which limits the utility of sub-group classifications and comparisons made according to ability groups, and a number of participants were considered to be in the borderline ID range, which likely inflated subtest means and score distributions. Thirdly, the presence of floor effects on certain subtests such as mobility, may not be pure floor effects. For example, individuals who were unable to complete this item scored zero due to physical limitations, rather than attempting and performing poorly. However, as mentioned, floor effects are not uncommon particularly when screening for dementia in ID (Paiva et al., 2020; Tyrrell et al., 2001), as well as when ensuring assessments are applicable to those with moderate/severe ID (Kalsy & Oliver, 2005). Fourthly, the lack of previous assessments (NAID and DLD) available for the sample meant examination of concurrent validity could not be performed and remains unknown. Fifthly, due to the presence of screening items, such as Picture Naming, many individuals performed at ceiling, skewing the distribution of these subtests. Although distributions at total and domain level appeared more normally distributed, the same could not be said for individual subtests. This is not surprising given the nature of psychometric properties for neuropsychological constructs, particularly as they are often more unstable when broken down to item level (Taber, 2018). Transformations did not improve the approximate normality of the distributions, and there is conflicting evidence as to the robustness of the α coefficient with non-normal data (Headrick & Sheng, 2013; Sheng & Sheng, 2012).

Despite this, the current study also has several strengths including the adoption of co-production and feedback from previous participants in developing the measure. The inclusion of multiple EF measures as well as measures for motor and mobility skills is a considerable strength of the NBID, particularly as the focus of research has recently shifted to that of early symptoms of dementia and ID, highlighting that domains other than memory might be implicated early in the progression of dementia (Krinsky-McHale & Silverman, 2013). Including a wider range of measures suggested to be sensitive to early decline will future proof the measure and aid earlier and more timely diagnosis.

2.5.4 Future Research

To further investigate the psychometric properties of the NBID, future studies should be conducted, increasing the sample size, and exploring the validity of the NBID in comparison to validated measures such as the NAID and DLD. Further considerations are needed regarding the structure of the battery, including the usefulness of the Remote Memory subtest, as removal of this was indicted through reliability analysis. Additionally, further thought needs to be given to the inclusion of the Cats & Dogs subtest due to its high ceiling rates and lack of sensitivity to EF across ID abilities (Willner et al., 2010). Despite this, Bevins and Hurse (2016) argued for the continued use of this subtest within the ID population. The Cats & Dogs subtest makes up a substantial proportion of the EF domain within the NBID and its complete removal would have a large impact on this domain. It would be beneficial to explore whether other measures of EF are more suitable to be included, such as coloured Stroop or Weigl test.

Importantly, future studies evaluating the NBID should seek to include a wider pool of participants, such as those who lack the capacity to consent for themselves, as this may capture a wider spread of abilities within the ID population, and thus increase the generalisability of the findings.

2.5.5 Conclusion

The present study adds to the emerging evidence base regarding the psychometric properties of this new measure. The NBID has been shown to be easily administered with individuals generally tolerating it well. The measure has also been shown to be reliable albeit with a small sample, and that inclusion of measures other than memory can help to obtain a clearer picture of individuals abilities in order to establish the presence of change in the future.

Appendix A Search Terms and Databases

Database	Search Terms				
PsychInfo	Intellectual Disability Terms		Dementia Terms		Symptoms Terms
	(intellectual* OR learning* OR mental* OR developmental*)N1(disab* OR retard* OR handicap)) OR "Down* Syndrome*" OR trisomy* 21	AND	Dementi* OR Alzheimer*	AND	Symptom* OR marker* OR sign* OR feature* OR presentation* OR change*
PubMed	(intellectual*) OR (learning*) OR (mental*) OR (developmental*) AND (disab*) OR (retard*) OR (handicap*) OR ("Down* syndrome") OR (trisomy* 21)	AND	(dementia*) OR (Alzheimer*)	AND	Symptom* OR marker* OR sign* OR feature* OR presentation* OR change*
Web of Science	(intellectual*) OR (learning*) OR (mental*) OR (developmental*) AND (disab*) OR (retard*) OR (handicap*) OR ("Down* syndrome") OR (trisom* 21)	AND	(dementia*) OR (Alzheimer*)	AND	(symptom* OR marker* OR sign* OR feature* OR presentation* OR change*)

Appendix B Quality Assessment Checklist

Checklist for assessing the quality of quantitative studies

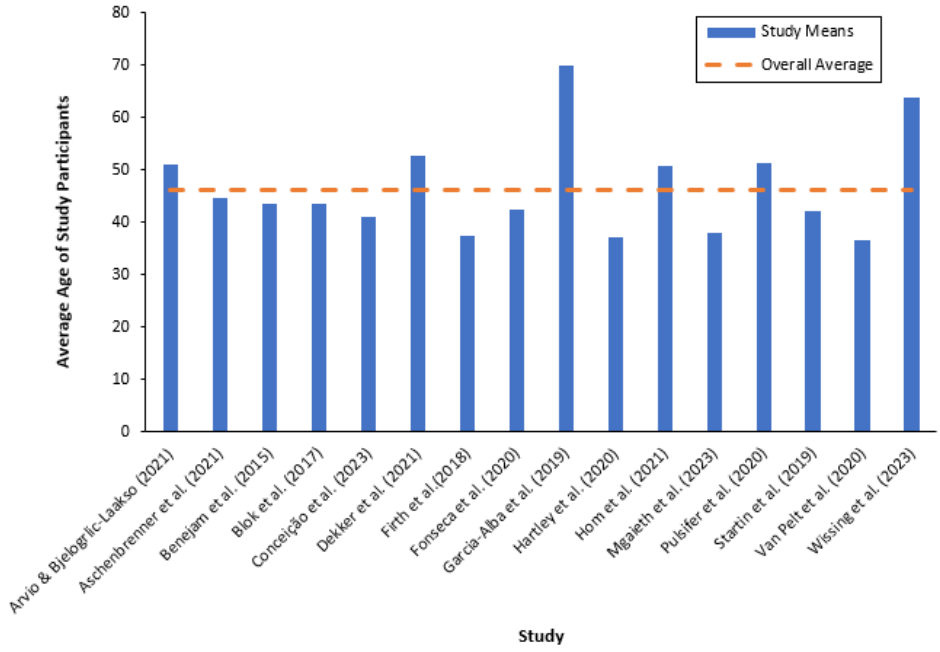
	Criteria	Yes (2)	Partial (1)	No (0)	N/A
1	Question / objective sufficiently described?				
2	Study design evident and appropriate?				
3	Method of subject / comparison group selection or source of information / input variables described and appropriate?				
4	Subject (and comparison group, if applicable) characteristics sufficiently described?				
5	If interventional and random allocation was possible, was it described?				
6	If interventional and blinding of investigators was possible, was it reported?				
7	If interventional and blinding of subjects was possible, was it reported?				
8	Outcome and (if applicable) exposure measures(s) well defined and robust to measurement / misclassification bias? Means of assessments reported?				
9	Sample size appropriate?				
10	Analytic method described / justified and appropriate?				
11	Some estimate of variance is reported for the main results?				
12	Controlled for confounding?				
13	Results reported in sufficient detail?				
14	Conclusions supported by the results?				

Checklist for assessing the quality of qualitative studies

	Criteria	Yes (2)	Partial (1)	No (0)
1	Question / objective sufficiently described?			
2	Study design evident and appropriate?			
3	Context for the study clear?			
4	Connection to a theoretical framework / wider body of knowledge?			
5	Sampling strategy described, relevant and justified?			
6	Data collection methods clearly described and systematic?			
7	Data analysis clearly described and systematic?			
8	Use of verification procedure(s) to establish credibility?			
9	Conclusions supported by the results?			
10	Reflexivity of the account?			

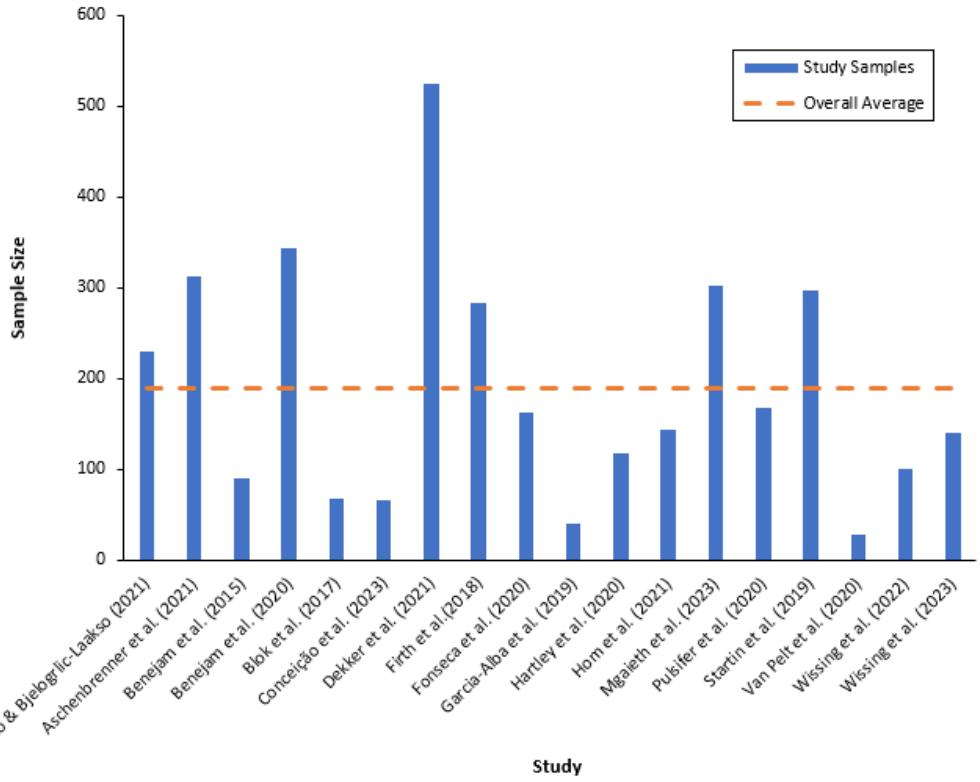
Appendix C Figures Displaying Descriptive Statistics for the Included Studies

C.1 Mean Ages of Participants per Study

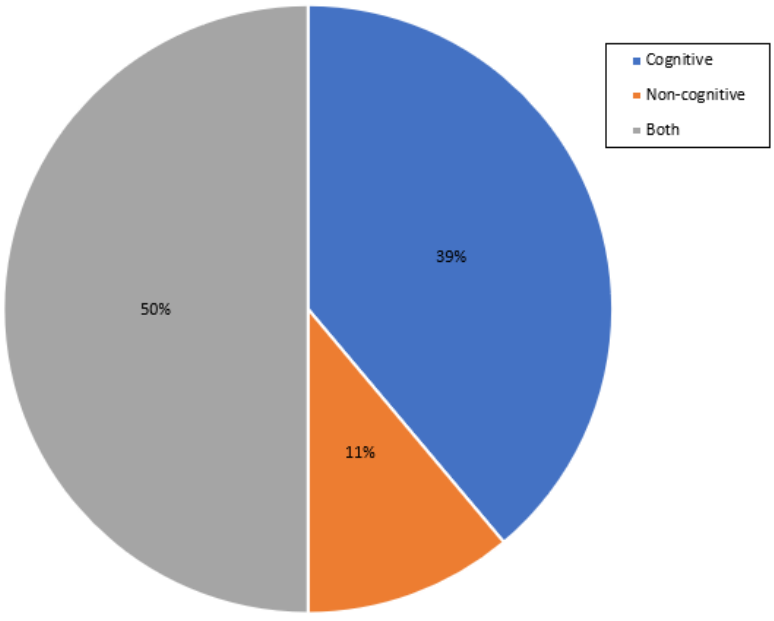


Note. Benejam et al. (2020) and Wissing et al. (2022) omitted from figure above as mean ages of individuals with ID not reported.

C.2 Sample Size Reported by Each Study



C.3 Domains Assessed in Included Studies



Appendix D Measures Used in Included Studies and the Frequency of Use

Measure	Frequency
Direct Tools:	
Scales and Questionnaires:	
British Present Psychiatric State-Learning Disabilities Scale (PPS-LD)	1
Batteries:	
NEPSY 2nd edition: A Developmental Neuropsychological Assessment (NEPSY-II)	1
Cambridge Cognitive Examination for Older Adults with Down's Syndrome (CAMCOG-DS)	4
Cambridge Examination for Mental Disorders of Older People with Down's Syndrome and Others with Intellectual Disabilities (CAMDEX-DS)	4
Severe Impairment Battery (SIB)	2
Kaufman Brief Intelligence Test (KBIT)	2
Kaufman Brief Intelligence Test 2nd edition (KBIT-2)	4
The Rapid Assessment of Developmental Disabilities – Second Edition (RADD-2)	1
Tests:	
Boston Naming Test (BNT)	3
Peabody Picture Vocabulary Test-4th Edition (PPVT)	1
Rivermead Behavioural Memory Test (RBMT)	1
Brief Praxis Test (BPT)	2
Purdue Pegboard Test (PPT)	3
Timed Up and Go (TUG)	2
Modified Cued Recall Test (mCRT)	2
Visual Association Test (VAT)	1
Performance-Oriented Mobility Assessment (POMA)	1
The Beery Buktenica Developmental Test of Visual-Motor Integration	1
Tinetti Balance and Gait Assessment Tool	1
Tasks:	
Verbal Fluency	3
McCarthy Verbal Fluency	1
Semantic Verbal Fluency	1
Spatial Reversal, taken from Cambridge Executive Functioning Assessment (CEFA)	1
Scrambled Boxes, taken from CEFA	1
Cats & Dogs, taken from CEFA	2
Tower of London, taken from CEFA	2
Finger-nose pointing	3
Paired Associates Learning (PAL), taken from the Cambridge Neuropsychological Test Automated Battery (CANTAB)	3
Intra/extra Dimensional Set Shift (IED) task, taken from CANTAB	3
Simple Reaction Time (SRT) task, taken from CANTAB	4
Digit Span	1
Corsi Block-Tapping Test (CBT) Forward	1

Measure	Frequency
Corsi Block-Tapping Test (CBT) Backward	1
Object Memory	2
Cancellation Task	1
4-Meter Walk	1
Fuld Object Memory Test	1
Orientation	2
Car and Motorbike Score, taken from NEPSY-II	1
Cued Recall Test, taken from NEPSY-II	3
Visuomotor Perception, taken from NEPSY-II	1
Visuospatial Precision, taken from NEPSY-II	1
Temporal Orientation Test (TO)	1
Delayed Visual Memory (DVM)	1
Auditory Delayed Verbal Memory (ADVM)	1
Cat and Dog Modified Stroop Task	1
Block Design	3
GAITRite™	1
Riddles, taken from the KBIT	1
Visuo-motor Integration (VMI)	1
Story Recall	1
Ideomotor Praxis	1
Adapted version of Tower of London	1
Adapted Category Fluency Test	1
Down Syndrome Mental Status Examination (DSMSE)	1
Modified MMSE	1
Informant-based Questionnaires:	
Dementia Questionnaire for People with Learning Disabilities (DLD) *Previously called* Dementia Questionnaire for Persons with Mental (DMR)	6
Temperament Scale for People with Intellectual Disability (TVZ)	1
Social Disability Scale for the Mentally Retarded (SRZ/SRZ-i)	1
Short Adaptive Behavior Scale (Short ABS)	2
Observer Memory Questionnaire (OMQ)	2
Vineland Adapted Behavior Scale, taken from Vineland 2nd edition (Vineland-II)	1
Behavior Rating Inventory of Executive Function – Adult version (BRIEF-A)	1
Working Memory (WM), taken from Behavior Rating Inventory of Executive Function-Parents (BRIEF-P)	1
Communication Domain, taken from Vineland-3	1
Behavioral and Psychological Symptoms of Dementia in Down Syndrome II (BPSD-DS II)	1
	Total = 66

Appendix E Context of the Present Study

This appendix provides additional details pertinent to the current study. The study was originally conceptualised following recognition within a local ID service, that the measures used for assessing dementia in ID do not live up to this 'gold standard' badge given to them. The lack of standardisation and robust psychometrics, coupled with the primary focus on memory change led to the development of a new measure. This project was an extension of a previous study, exploring the feasibility of the new measure however this did not utilise a clinical sample. Therefore, this project was planned to be undertaken within the NHS using a clinical sample, capturing a range of abilities within the ID population to establish the measures psychometric properties.

Initially six NHS trusts were approached and expressed interest in supporting with recruitment and were included in the IRAS application. The initial ethics application aimed to recruit individuals across these six trusts and recruit both individuals who had capacity to consent to take part, as well as those who lacked capacity to consent and where proxy consent would be required, in keeping with the HRA e-learning module titled 'Research Involving Participants Lacking Mental Capacity'. However, when the project was reviewed by a Research Ethics Committee specialising in issues of capacity, the project was denied on the grounds participant who lacked capacity could not be included. This resulted in delays as protocols were amended and revisions submitted to the Committee.

Following receiving these approvals, each trust was approached to complete their site level approvals through the Research & Development teams. These processes varied from trust to trust and pushed the recruitment start date back significantly. Due to time constraints, approvals were not followed through to completion with three trusts. The recruitment window was just five weeks, far shorter than had originally been anticipated which resulted in a far smaller sample.

Due to the large undertaking of this project, a research team was assembled to aid in identification of participants as well as administering assessments. The majority of the research team were situated in one of the research sites. The investment from the main site and Principal Investigator there was invaluable in generating awareness and support for the study. However due to the scale of the project there were likely oversights and issues which were compounded by delays in obtaining approvals, e.g., the time needed to distribute resources, gathering assessment materials and managing data handling across multiple sites.

Appendix F CORE-LD

LD-CORE

Site ID

letters only numbers only

Client ID

Therapist ID numbers only (1) numbers only (2)

Sub codes

D D / M M / Y Y Y Y

Date form given

Age Male Female

Stage Completed
S Screening
R Referral
A Assessment
F First Therapy Session
P Pre-therapy (unspecified)
D During Therapy
L Last Therapy Session
X Follow up 1
Y Follow up 2

Stage

Episode

Total Score

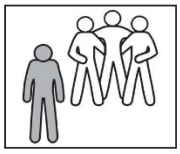
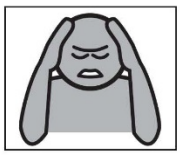
Risk Score

IMPORTANT – PLEASE READ THIS FIRST

HOW DO YOU FEEL?

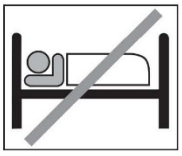
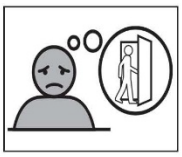
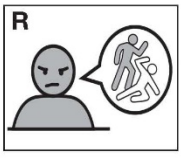
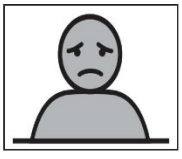
This form has 14 questions about how you have been OVER THE LAST WEEK.
People with a learning disability helped make these questions.
Please tick the box that fits how you feel.

Over the last week

		Not at all	Sometimes	A lot
1	 **Have you felt very very lonely?** Have you felt really alone?			
2	 **Have you felt confused?** Has it been hard to think straight?			
3	 **Have you felt happy with the things you have done?**			

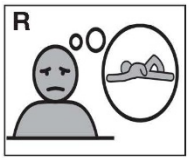
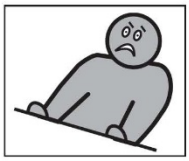
Please turn over

Over the last week

		Not at all	Sometimes	A lot
4	 **Have you found it hard to say how you feel?**			
5	 **Have you had difficulty getting to sleep or staying asleep?**			
6	 **Have you felt frustrated or upset with your learning disability?**			
7	 **Have you felt sad about people you have lost?** For example family, staff, friends			
8	 **Have you threatened or shouted at someone?**			
9	 **Have you felt unhappy?**			

Please turn over

Over the last week

		Not at all	Sometimes	A lot
10	 **Have you felt people are getting at you?** Have you felt people were picking on you?			
11	 **Have you thought about ending your life?** Have you wanted to be dead?			
12	 **Have you bottled up angry feelings?** Have you felt ready to blow inside?			
13	 **Have you hurt yourself on purpose?** eg. cutting, picking, hitting yourself, not taking tablets, drinking lots of alcohol			
14	 **Have you felt really scared or frightened?**			

THANK YOU FOR DOING THIS QUESTIONNAIRE

Scoring – all questions except question 3:
Not at all = 0 Sometimes = 1 A lot = 2

Question 3 only
Not at all = 2 Sometimes = 1 A lot = 0

Add together the item scores for the Total Score.
Divide by the number of questions completed to get the Mean Score, multiply by 10 to get the Total Clinical Score

Total Score ☐ **Mean** ☐ **Total Clinical Score** ☐

CORE - LD
Learning Disabilities

INSTRUCTIONS FOR USE

CORE-LD is a validated self report outcome measure for use with clients with a learning disability receiving any form of psychological therapy.

It is designed to be used as part of the therapeutic process and completed with the therapist ideally between the first and third session. Services may well repeat the questionnaire at intervals during therapy and always at the end of therapy.

INTRODUCING CORE-LD TO YOUR CLIENT

You and your client will both need a copy of CORE-LD.

It is expected that your client will mark their answers on their copy.

Tell the client this is a special questionnaire because it was made together with people with a learning disability who understand what it's like living with a learning disability and the problems people face.

Emphasise there are NO right or wrong answers the questions are simply about how they feel.

Use the first question to model the process, show how the picture supports the question and how the scoring beakers range from 'not at all' (empty) to 'sometimes' (half full) to 'a lot' (full).

Encourage the client to answer all the questions but where this is not possible reassure the client and go onto the next question.

On completion of the questionnaire explain you will ask the client to do the questionnaire again to help see if things have changed. It is helpful to discuss these changes within the therapy relating back to the relevant items.

USING THE MEASURE

1. clinically on a single score for one person: don't base any major clinical decisions on scores alone, i.e. don't say "low score, doesn't need anything"

As yet there are:

- NO clinical/non-clinical cutting points
- NO "clinically improved" criteria
- NO "reliably improved/deteriorated" criteria

However with increased data collection it is hoped these will be available within the next two years. It should be noted there will probably be greater variability with CORE-LD and people with a learning disability than with CORE-OM and the non LD population. Therefore services should always use data with greater caution.

2. clinically looking at change: it is safer to use changes in scores within an individual alongside other clinical indicators to decide things like changing therapy, adding other interventions, positively connoting and supporting progress. Ideally, discuss with client.

3. data collection: always attempt to contribute data, ideally item data, with age, gender, location, date and where in services, to some national database to improve on the guides the data can provide.

4. for research: use cautiously to explore both the measure but also substantive questions about things that affect wellness in people with a learning disability.

5. for service management and reporting: aggregate data and report to practitioners first and foremost: encourage them to own and think about the data; then share with commissioners, referrers and above all, user & carer groups. See (3): always share anonymised data including item data.

6. for commissioning: too early to use as evidence for reliable commissioning. In a few years however, when there are some good sized clinical and non-clinical datasets, it should become a part of good commissioning.

Appendix G Neurocognitive Battery for Intellectual Disabilities (NBID)

G.1 NBID Record Form

Neurocognitive Battery Intellectual Disability (NBID)
Record Form

Prior to testing: Check that the environment is suitable for testing- a table to work at and that the examinee has space to stand and turn 360°. You will need to have pens/pencils and a stopwatch. A clipboard may be helpful.

Instructions: For each subtest, **Bold text** indicates the verbal instructions to be read aloud to the examinee. *Italic text* indicates pointers on what to consider during administration and/or scoring. Record verbal responses verbatim.

Client Sticker:	
Date:	
Assessor Name & Job Title:	
People Present:	
Location:	

Additional needs:

Does the person use glasses?	
If so, are they wearing them during this assessment?	
Does the person use a hearing aid?	
If so, is the hearing aid fitted and switched on during this assessment?	
Does this person need someone to sign for them?	
If so, is a signer present?	
Is the person left-handed or right-handed?	

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Administration

Were there any deviations from standard test administration procedures during this assessment?

Yes / No

If yes, please describe these below with sufficient detail to allow repeat assessments to follow an identical administration process for this person.

Score Summary

Domain	Raw Score
Orientation	
1. Total	/20
Attention	
8a. Digit Span - Forwards	/10
8b. Digit Span - Backwards	/8
12. Coding	/50
15. Cancellation	/28
Total:	**/96**
Executive Functioning	
6a. Trails Shapes- Sequencing	Time in seconds:
6a. Trails Shapes- Sequencing	/7
6b. Trails Shapes- Switching	Time in seconds:
6b. Trails Shapes- Switching	/9
13a. Cats & Dogs – Naming	/16
13b. Cats & Dogs – Inhibition	/16
13c. Cats & Dogs - Switching	/16
13d. Cats & Dogs - Sorting	/16
13d. Cats & Dogs - Sorting	Time in seconds:
Total:	**/80**
Learning & Memory	
2. Remote Memory	/2
5. Immediate Recall & 3 Trial Learning (verbal)	/20
7b. Figure Drawing – Immediate Recall (visual)	/12
14. Delayed Recall & Recognition (verbal)	/10
16a. Figure Drawing – Delayed Recall (visual)	/12
16b. Figure Drawing – Visual Recognition (visual)	/3
Total:	**/59**
Language	
3. Picture Naming	/12
4. Picture Identification	/12
9a. Verbal Fluency – Category	/4
9b. Verbal Fluency – Letter	/4
17. Reasoning	/8
Total:	**/40**
Perceptual Motor	
7a. Figure Drawing – Copy	/12
11a. Action on Request	/30
18. Construction	/7
Total:	**/49**
Social Cognition	
10. Emotion Recognition	/6
Total:	**/6**
Mobility	
11b. Action on Request- 360° turn	Number of steps:
	Time in seconds:

1. Orientation

	Response	Score
What is your name? *1 point if first name only*		0 1 2
How old are you?		0 1
When is your birthday? *1 point for each element (day, month, year)*		0 1 2 3
Where do you live? *2 points for full address, 1 point only for town or name of home*		0 1 2
What day is it today?		0 2
If incorrect/no answer, administer this item. If correct, skip and award 1 point **Is it? _____, _____ or _____?** (correct answer) (2 days ago) (tomorrow)		0 1
What month is it now?		0 2
If incorrect/no answer, administer this item. If correct, skip and award 1 point **Is it? _____, _____ or _____?** (2 months ago) (correct answer) (next month)		0 1
What year is it now?		0 2
If incorrect/no answer, administer this item. If correct, skip and award 1 point **Is it? _____, _____ or _____?** (next year) (2 years ago) (correct answer)		0 1
What is the name of this town?		0 2
If incorrect/no answer, administer this item. If correct, skip and award 1 point **Is it? _____, _____ or _____?** (nearby town) (correct answer) (far away town)		0 1
Total Score (max 20)		

2. Remote Memory

Use page 1 of Stimulus Book for item 2.

	Response	Score
What town were you born in?		0 1
Who was Elvis Presley? *Point to picture in Stimulus Book* *Accept "singer", "entertainer" or similar*		0 1
	Total Score (max 2)	

Scoring
1 point if correct

3. Picture Naming

Start from page 2 of Stimulus Book. Present each picture and say…
What is this?

Picture	Response	Score
(Training) **Glass**		
(Training) **Sock**		
Football		0 1
Chicken		0 1
Hand		0 1
Bus		0 1
Guitar		0 1
Hive		0 1
Pond		0 1
Button		0 1
Bed		0 1
Window		0 1
Letter		0 1
Dress		0 1
	Total Score (max 12)	

Scoring
1 point if correct

4. Picture Identification

Start from page 16 of Stimulus Book. Present each item and say…

Show me the [item]. Where is the [item]?

Picture	Response	Score
(Training) **Glass**		
(Training) **Sock**		
Football		0 1
Chicken		0 1
Hand		0 1
Bus		0 1
Guitar		0 1
Hive		0 1
Pond		0 1
Button		0 1
Bed		0 1
Window		0 1
Letter		0 1
Dress		0 1
	Total Score (max 12)	

Scoring
1 point if correct

5. **Memory– Immediate Recall & 3-Trial Learning: Name and address**

Show picture of Andy Smith page 30 from Stimulus Book. **This is Andy Smith. Try to remember his name**. *Leave picture out for whole subtest.*

	Response	Score
What is his name?		0 1

Say **Andy Smith lives at 35 North Street, London**.

	Response	Score
Where does he live?		0 1

I'm going to tell you his name and address again. I would like you to repeat it after me. We will do it three times so you can learn it.
If participant tries to repeat whilst you're still talking, tell them to wait until you have finished. Always do all 3 trials even if correct.

Scoring- 1 point per correct element

	1st trial	*2nd trial*	*3rd trial*
Andy Smith **35 North Street** **London**	____ _____ __ _____ _____ _____	____ _____ __ _____ _____ _____	____ _____ __ _____ _____ _____
Score			

Please try to remember his name and address as I will ask you about it later.
Turn page of Stimulus Book to remove picture of Andy Smith from view.

Total Score (max 20)	

Scoring
1 point if correct

6. Adapted Trails Shapes

Part A - Sequencing

Show Trials A Demonstration in Response Booklet

Here are some circles. Some are bigger than others. Without taking your pencil off the paper, draw a line joining the circles together from the biggest to the smallest.

I'll show you *(complete Demonstration)*
(Pass a pen to the person, point to the Practice)
Now you do these. Start here *(Point to start position, complete Practice)*

If correct say: **Well done, you joined them from biggest to smallest.**

If the person makes a mistake say: **That's not quite right, you need to join them from the biggest to the smallest like this…** *(use example on page 31 of stimulus book to demonstrate rule, reiterate using Demonstration and then draw out correct order in the Practice).*

Now do these. Start here *(point to start position)* **Ready? Go** *(begin timing)*

	Shapes Sequencing
Completion Time	
Accuracy Score (max 7)	
Comments	

Scoring errors for Trails Shapes- Sequencing (max score 7)
There are 8 circles to be sequenced. If there are no errors, this means the participant has correctly sequenced all the circles in the correct order and they would score "7". An error is when a line is drawn to a circle that is out of the correct sequence. To calculate if errors have occurred, count the number of times the participant draws a line to an out-of-sequence circle. Each out-of-sequence line is classed as one error. Total the number of errors and subtract this from 7 to obtain the final score.

Part B - Switching

Show Trials B Demonstration on Response Booklet

In this one, there are triangles and circles. Some are bigger than others. Without taking your pencil off the paper, draw a line joining the shapes together from biggest to smallest, swapping between triangles and circles.

I'll show you *(complete Demonstration)* **Start at the biggest triangle** *[point]*, **then go to the biggest circle** *[point]*, **then go to the next smallest triangle** *[point]*, **and then to the next smallest circle** *[point]*. *(Pass a pen to the person, point to the Practice)*
Now you do these. Start here *(Point to start position, complete Practice)*

If correct say: **Well done, you joined them triangle, circle, triangle, circle.**

If the person makes a mistake say: **That's not quite right, you need to go from the biggest Triangle** *[point]*, **then go to the biggest circle** *[point]*, **then go to the next smallest triangle** *[point]*, **and then to the next smallest circle** *[point] (use example on page 32 of stimulus book to demonstrate rule, reiterate using Demonstration and then draw out correct order in the Practice).*

Now do these. Start here *(point to start position)* **Ready? Go** *(begin timing)*

	Shapes Switching
Completion Time	
Accuracy Score (max 9)	
Comments	

There are a total of 9 switches to be made. If there are no errors, this means the participant has correctly alternated between connecting circles and triangles in descending size order and they would score "9". To calculate if errors have occurred, from the start position, following their line, count the number of times the participant draws a line to an out-of-sequence shape and/or size. Each out-of-sequence line is classed as one error. Total the number of errors and subtract this from 9.

Qualitative analyse: Note the point at which errors occur. Is an error due to failing to correctly alternate between shapes (indicating a set-loss type error) or failing to correctly connect to the next ascending size (indicating a sequencing type error).

7. Figure Drawing

Open Stimulus Book to page 33 to show Figure Drawing.
Pull out and provide Figure Drawing (Copy) page from Response Booklet, ensure it is placed in landscape orientation (so that it is consistent with the Stimulus Book)

Part A - Copy

Have a look at this picture. I would like you to copy it for me, here.
[point to Copy page of Response Booklet]

Accuracy Score (max 6)	
Location Score (max 6)	
Copy Total (max 12):	

Once examinee has finished, turn page of Stimulus Book to hide the Figure Drawing. Also remove examinee's copy of Figure Drawing from view.

Scoring *- See next page for full scoring criteria.*

1 point if each correct element and 1 point for proper placement.
Accuracy - 1 point for each feature accurately drawn (regardless of location).
Location - 1 point for each feature in the correct location

Item	*Accuracy (0 or 1)*	*Location (0 or 1)*	*Criteria*
1. Rectangle			**Drawing:** lines unbroken and straight, angels approx. 90 degrees **Placement:** placed so proportionate to top two-thirds of overall figure
2. Triangle			**Drawing:** lines unbroken and straight, all sides meet at a point **Placement:** placed approximately in middle of bottom side of rectangle
3. Circle			**Drawing:** round, unbroken and closed **Placement:** placed in top right quadrant of rectangle
4. Diagonal line			**Drawing:** line moderately straight between rectangle corners **Placement:** end of lines should meet (or be close to) corners of square
5. 3 parallel lines			**Drawing:** lines are straight and not crossing each other, approximately parallel **Placement:** in top left section of rectangle, crossing diagonal line
6. Filled oval			**Drawing:** unbroken and filled **Placement:** in approximately bottom left area of triangle

Part B - Immediate Recall

Pull out and provide Figure Drawing (Immediate Recall) page from Response Booklet, ensure it is placed in landscape orientation.

Now try drawing it again here from memory

[point to Immediate Recall page of Response Booklet]

Accuracy Score (max 6)	
Location Score (max 6)	
Immediate Recall Total (max 12):	

Once examinee has finished, remove Figure Drawing Immediate Recall from view.

Scoring

1 point if each correct element and 1 point for proper placement.
Accuracy - 1 point for each feature accurately drawn (regardless of location).
Location - 1 point for each feature in the correct location

Item	*Accuracy (0 or 1)*	*Location (0 or 1)*	*Criteria*
1. Rectangle			**Drawing:** lines unbroken and straight, angels approx. 90 degrees **Placement:** placed so proportionate to top two-thirds of overall figure
2. Triangle			**Drawing:** lines unbroken and straight, all sides meet at a point **Placement:** placed approximately in middle of bottom side of rectangle
3. Circle			**Drawing:** round, unbroken and closed **Placement:** placed in top right quadrant of rectangle
4. Diagonal line			**Drawing:** line moderately straight between rectangle corners **Placement:** end of lines should meet (or be close to) corners of square
5. 3 parallel lines			**Drawing:** lines are straight and not crossing each other, approximately parallel **Placement:** in top left section of rectangle, crossing diagonal line
6. Filled oval			**Drawing:** unbroken and filled **Placement:** in approximately bottom left area of triangle

8. Digit Span

Part A - Forwards

I'm going to say some numbers and I'd like you to repeat them after me

Discontinue after two failed trials of the same length.

	Response	Score
2		0 1
5		0 1
8-7		0 1
4-1		0 1
5-8-2		0 1
6-9-4		0 1
9-4-3-9		0 1
7-2-8-6		0 1
4-2-7-3-1		0 1
7-5-8-3-6		0 1
	Total Score (max 10):	

Scoring
1 point if correct

Part B - Backwards

Now I'm going to say some more numbers but this time I want you to say them backwards. So if I said, 3-6 you would say_____ *(6-3).*

If correct, say **Yes that's right, 6-3.**

If incorrect or no response, say **You would say 6-3. You say them backwards.**

Discontinue after two failed trials of the same length.

	Response	Score
2-5		0 1
6-9		0 1
5-1-3		0 1
8-4-6		0 1
2-9-5-3		0 1
4-7-9-4		0 1
5-1-5-1-2		0 1
7-8-6-4-5		0 1
	Total Score (max 8):	

Scoring
1 point if correct

9. Verbal Fluency

Part A – Category

I would like you to tell me as many different animals as you can. See how many you can say in one minute.

Ready? Go. *Time 60 seconds. Record responses verbatim in 15 second interval boxes.*

0 – 15s	16s – 30s	31s – 45s	46s – 60s
		Total	
		Errors made	

Scoring *– 1 point for each correct response.*
Scoring: 0 = 0, 1-4 = 1, 5-9 = 2, 10-14 = 3, 15+ = 4
Errors include repetitions, perseveration, deviations from category.

Part B - Letter

I would like you to tell me as many different things as you can beginning with the letter "F". You cannot give me names, like Frank, or names of places, like France. See how many you can say in one minute.

Ready? Go. *Time 60 seconds. Record responses verbatim in 15 second interval boxes.*

0 – 15s	16s – 30s	31s – 45s	46s – 60s
		Total	
		Errors made	

Scoring *– 1 point for each correct response.*
Scoring: 0 = 0, 1-4 = 1, 5-9 = 2, 10-14 = 3, 15+ = 4
Errors include repetitions, perseveration, deviations from category.

10. Emotion Recognition

Open Stimulus Book to page 34 to Emotion Recognition Faces

Point to each picture and ask **What emotion is this?**

If they state a behaviour, prompt for an emotion/feeling.

	Response	Score
Picture 1 – scared		0 1
Picture 2 – surprise		0 1
Picture 3 – angry		0 1
Picture 4 – happy		0 1
Picture 5 – sad		0 1
Picture 6 - worried		0 1
	Total score (max 6)	

Scoring
1 point if correct

11. Action on Request

Part A - Action on Request

I am going to ask you to do some strange actions. Some things you may think are a bit odd or silly. Clap your hands.

If the participant does not perform the action, repeat the instruction again whilst being careful not to give visual prompts with your hands.
If no response is given, say, **Do this** *(model the action).*

	Action	Unable to Complete	Model	Verbal Prompt	Verbal Instruction
1	**Clap your hands**	0	1	2	3
2	**Stick out your tongue**	0	1	2	3
3	**Wave good-bye**	0	1	2	3
4	**Scratch your head**	0	1	2	3
5	**Put your hands in the air**	0	1	2	3
6	**Close your eyes**	0	1	2	3
7	**Show me how you would blow out a candle**	0	1	2	3
8	**Show me how you would comb your hair**	0	1	2	3
9	**Show me how you would clean your teeth**	0	1	2	3
10	**Show me how you would use a spoon**	0	1	2	3
				Total Score (max 30)	

Comments:

Scoring
3 points for correct answer on the first verbal instruction, 2 points for the verbal prompt, and 1 point for the model.

Part B – Mobility
(Only administer if examinee is deemed to have sufficient mobility to complete this task)

Ask the participant to stand up and to move away from the table/chair. You want to be able to observe their whole body so position yourself appropriately. Examiner to place item such as a pen on the floor in front of the participant to indicate start/stop position.

I would like you to do a complete turn on the spot. I would like you to stop when the pen is in front you again, like it is now. Ready? Go.

Begin timing as soon as you say 'Go'.

Count the number of steps taken to do a complete 360° turn and time taken in seconds. Note whether the turn was completed clockwise or anti-clockwise. Comments about their mobility can be added to the box below (e.g. leaning to one side, shuffling, pace, balance).

Clockwise or anti-clockwise?	Number of steps	Time to complete

If they did not perform the action, repeat the instruction again whilst being careful not to give visual prompts with your hands. Begin timing again when you say 'go'.

If no response is given, say, **Do this** *(model the action).* **Now you try. Go** *(begin timing again).*

Action was completed with (✓ below):

Verbal instruction only	Verbal instruction with prompt	Model
Comments:		

12. Coding

Open Response Booklet to the Coding Page.

Look at these boxes *(point to the boxes at the top)***. Each box has a shape in the top part and a number in the bottom part. Down here** *(point to incomplete boxes)* **the boxes have shapes in the top part but are empty in the bottom part.**

You must draw the number that goes with this shape. *Point to the top line and show how you are completing the sample.*

Now I would like you to try doing these ones here. *Ask the person to complete sample items. Provide correct feedback as required to help understand the task.*

Now you know what to do, I would like you to try these *(point to remaining items)***. Work through them in order as quickly as you can**. **Ready? Go** *Begin timing.*

The time limit is 120 seconds. Ask examinee to stop when this is reached.

Total Correct:		**Total Errors:**	
		Total Score *(max 50)*	

Scoring
Total the number of correct responses. Then total number of errors. Total score is Number correct minus Number errors.

13. **Cats & Dogs** – *Use laminated picture cards of cat and dog. Ensure that for each sub-trial, cards are in the following order (numbered on the back for ease): dog, dog, cat, dog, cat, cat, dog, dog, cat, dog, cat, cat, dog, cat, dog, cat.*

For trials (a), (b) and (c), present cards at a rate of 1 per second. Make sure they are in number order. For trial (d) prompt person to sort only one at a time if needed.

a. Naming: **I am going to show you some pictures of a cat and a dog. When you see the cat, say cat. When you see the dog, say dog.**

b. Inhibition: **I'm going to show you the pictures again. This time when you see the dog, say dog. When you see the cat, do not say anything.**

c. Switching: **I'm going to show you the pictures again. This time when you see the dog, say cat. When you see the cat, do not say anything.**

d. *Show page 35 of Stimulus Book- starting places for sorting cats and dogs.* **Now sort the pictures into two piles. Put Cats here** *[point to cat]***. Put Dogs here** *[point to dog]***.** *(Time this block)*

Mark errors in table below.

	No. of errors (tally)	Score
(a) Naming	dog, dog, cat, dog, cat, cat, dog, dog, cat, dog, cat, cat, dog, cat, dog, cat	
(b) Inhibition	dog, dog, X, dog, X, X, dog, dog, X, dog, X, X, dog, X, dog, X	
(c) Switching	cat, cat, X, cat, X, X, cat, cat, X, cat, X, X, cat, X, cat, X	
(d) Sorting		Score: Time:

Total Score (max 64):	

Scoring
Total score for each block is 16. For each block, subtract number of errors from 16.

Comments:

14. Delayed Recall & Recognition - Name and Address

Show the picture of Andy Smith from Stimulus Book, page 36.

	Response	Score
What was this man's name? *2 points for full name, 1 point for first or last name only*		0 1 2
If incorrect/no answer, administer this item. If correct, skip and award 1 point **Was it…** **Andy Smith*** **Thomas Richards** **Kevin Jones?**		0 1
What was his address? *1 point for each element: 35, North, Street, London*		0 1 2 3 4
If incorrect/no answer, administer this item. If correct, skip and award 1 point **Was it…**		
Number 52 **Number 35*** **Number 81**		0 1
North Street* **South Street** **West Street**		0 1
Portsmouth **Brighton** **London***		0 1
Free Recall Score (max 6)		
Recognition Score (max 4)		
Total Score (max 10):		

**Correct answer*

15. Cancellation

Open Response Booklet to Cancellation Page

Look at these shapes. This shape is a blue cross and this shape is a green heart. Now look at this row. It has crosses and hearts but some of them are a different colour. I am going to look at the shapes in the row and draw a line through each BLUE CROSS and GREEN HEART. I am not going to draw a line through anything else. *(Demo- draw a single line through each of the target shapes).*

Point to practice items. **Now you do these here. Draw a line through each BLUE CROSS and GREEN HEART. Do not draw a line through anything else. When you finish the first row, go to the beginning of the next row. Work as fast as you can without making any mistakes. You cannot go back to mark any shapes that you miss. Are you ready? Go."** *(Complete practice items)*

Turn to next page. **When I say go, draw a line through each BLUE CROSS and GREEN HEART. Start here and go in order. When you finish a row go to the next row. If you get to here** *[point to blue heart in bottom right corner],* **there are more on the next page. Are you ready? Go.** *Begin timing.*

The time limit is 45 seconds, ask examinee to stop when this is reached.

Total Correct		**Total Errors**	
		Total Score (max 28)	

Scoring
Marks on correct targets are scored as correct. Marks on non-targets are incorrect. Total up the correct and incorrect responses. Total score is number correct minus number incorrect.
Use guidance as per WAIS-IV manual, Cancellation sub-test (p. 191) if there are any marked adjacent shapes, marks through multiple shapes or other marks.

16. Delayed Recall and Recognition - Figure Drawing

Part A - Delayed recall

Open page in Response Booklet Figure Drawing Delayed Recall. Position in landscape orientation in front of examinee.

Earlier, I asked you to copy and remember a picture. Please draw what you can remember of the picture.

Delayed Accuracy Score (max 6)	
Delayed Location Score (max 6)	
Delayed Recall Total (max 12):	

Scoring *- See below for full scoring criteria.*

1 point if each correct element and 1 point for proper placement.
Accuracy - 1 point for each feature accurately drawn (regardless of location).
Location - 1 point for each feature in the correct location

Item	*Accuracy (0 or 1)*	*Location (0 or 1)*	*Criteria*
1. Rectangle			**Drawing:** lines unbroken and straight, angels approx. 90 degrees **Placement:** placed so proportionate to top two-thirds of overall figure
2. Triangle			**Drawing:** lines unbroken and straight, all sides meet at a point **Placement:** placed approximately in middle of bottom side of rectangle
3. Circle			**Drawing:** round, unbroken and closed **Placement:** placed in top right quadrant of rectangle
4. Diagonal line			**Drawing:** line moderately straight between rectangle corners **Placement:** end of lines should meet (or be close to) corners of square
5. 3 parallel lines			**Drawing:** lines are straight and not crossing each other, approximately parallel **Placement:** in top left section of rectangle, crossing diagonal line
6. Filled oval			**Drawing:** unbroken and filled **Placement:** in approximately bottom left area of triangle

Part B – Recognition

Show page 37 of the Stimulus Book.

Look at all of these shapes *[indicate all 12 shapes]***. Can you point out which of these were in the picture I asked you to remember?**

Enter the shape number of all shapes endorsed			
Correct responses		**Incorrect responses**	
		Total Score (max 3)	

Scoring
1 point each for correctly identifying shapes 3, 7 and 10.

17. Language - Reasoning

Show picture of couple speaking on the phone in Stimulus Book page 38.

This picture shows a couple talking on the phone about something they would like to do.

Ask questions "1 to 10" below.

*Lvl**	Question	Response	Score
1	**1) Point to the Boy** *1 point if correctly points to boy*		0 1
	2) Point to a phone *1 point if correctly points to either phone*		0 1
2	*Provide the following information* – **The girl is asking the boy to go to the cinema to watch a film about Barbie and romance** *[point to speech bubble above girl's head].* **The boy does not want to go.**		
	3) Who is on the phone? *1 point if correctly states either the boy or the girl*		0 1
	4) What kind of film does the girl want to see? *1 point if state's a film about Barbie, romance, love, relationships or any description that would convey this*		0 1
3	**5) What is the girl saying?** *1 point if examinee able to say/express the girl wishes to watch the film at the cinema*		0 1

	6) What might the boy be thinking? *1 point if examinee able to say/express any thoughts or feelings that would indicate not wishing to see the film*		0 1
4	*Provide the following information* – **I think he's worried** *[point to boy's face]*		
	7) Why might the boy be feeling worried? *1 point if able to give a reasonable explanation of why the boy could be worried, e.g., does not like Barbie, does not want to upset the girl*		0 1
	8) If the boy does not want to go, what should he do? *1 point if able to give a reasonable solution/show some degree of problem solving, e.g., boy can make up an excuse he is busy, boy could be honest and say he does not like Barbie.*		0 1
		Total Score (max 8)	

** Scoring is informed by the Test of Abstract Language Comprehension (TALC 2) and levels that corresponds to the cognitive load of the question as per Blanks language scheme (1978). 1 point is assigned to each question if the examinee's answer demonstrates sufficient evidence of that level: Lvl 1 refers to the ability to name items/objects; Lvl 2 refers to the ability to classify, describe and show understanding of the function; Lvl 3 indicates ability to refer to own knowledge in order to make predictions and generalisations; Lvl 4 indicates ability to identify causality, problem solve and create solutions.*

Scoring
1 point for a correct answer to each question

18. Construction

Present person with Construction page from Response Booklet.

Please draw a circle here *[point to top left section of Construction page]*
*If examinee unable to draw, show circle from page 39 Stimulus Book and say '***copy this***'*

Please draw a square here *[point to top right section of Construction page]*
*If examinee unable to draw, show square from page 40 Stimulus Book and say '***copy this***'*

Please draw a tree here *[point to bottom half of Construction page]*

Circle	0 1 2
Square	0 1 2
Tree	0 1 2 3
Total score (max 7):	

Scoring for circle & square
A closed circular/four-sided shape is required.
0 if does not represent the desired shape.
1 point if copied.
2 points if drawn independently.

Scoring for tree
0 if not drawn or the image does not represent a tree.
1 point for each element drawn:
-Tree top (scores only 1 point if any or all of the following are drawn- branches, leaves, buds, fruit)
-Tree trunk (scores only 1 point if present)
-Ground (scores only 1 point if any or all of the following are drawn- roots, ground, soil, grass, pot)

G.2 NBID Response Booklet

Neurocognitive Battery Intellectual Disability (NBID)

Response Booklet

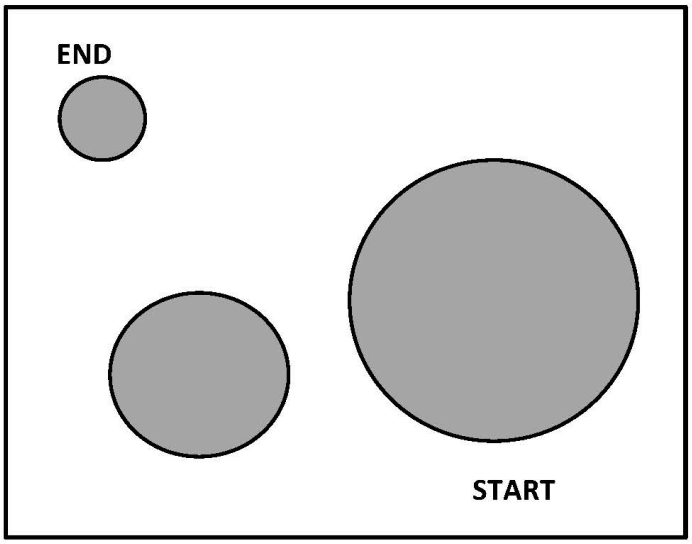
Print single-sided and in colour

Client Sticker	
Date	
Assessor	
Designation	
People Present	
Location	

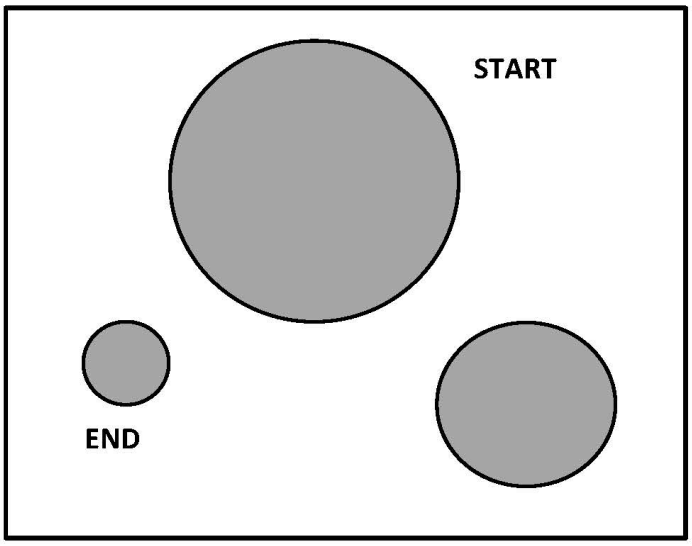
Adapted Trails Shapes

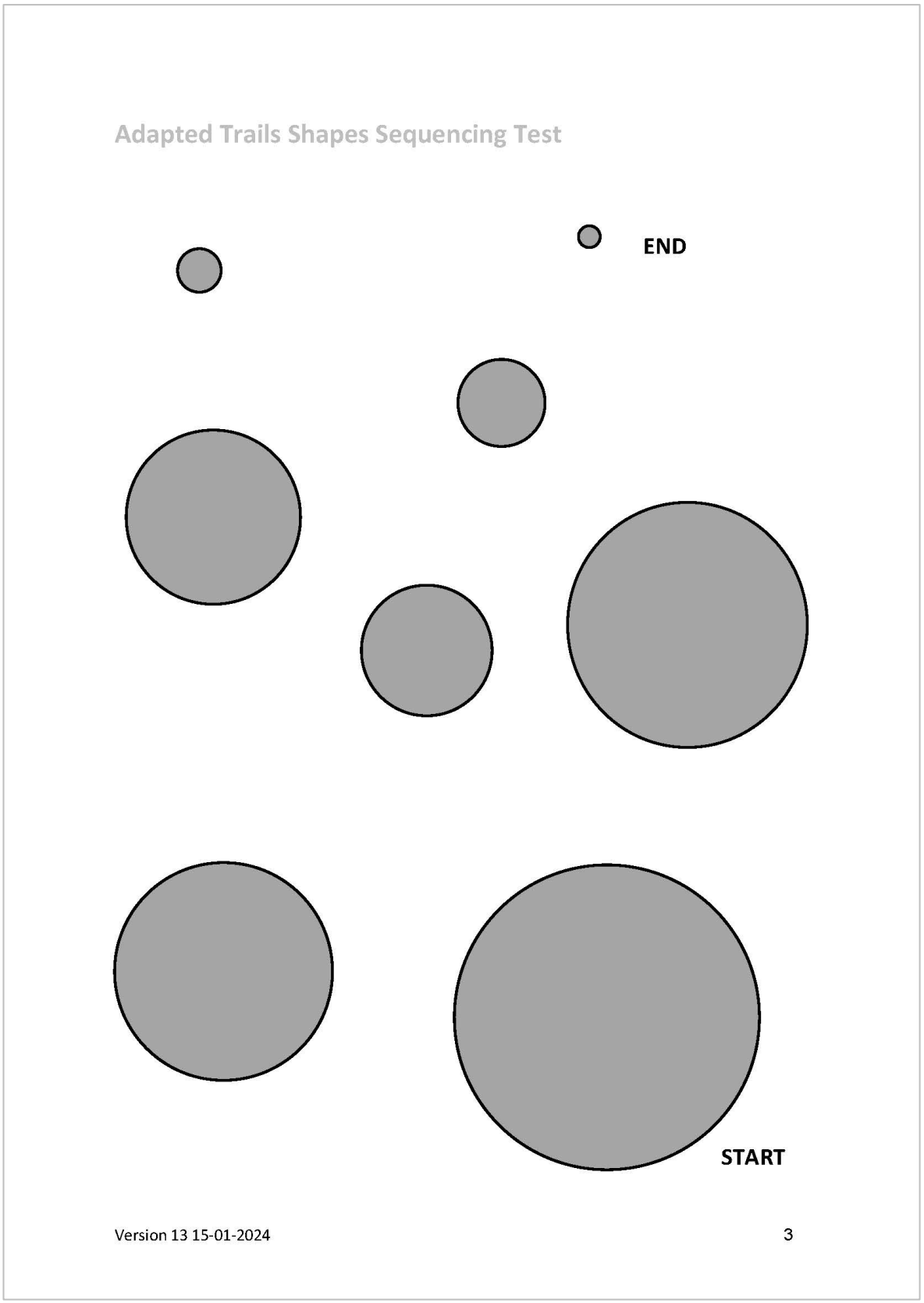
A) Sequencing

Demonstration



Practice

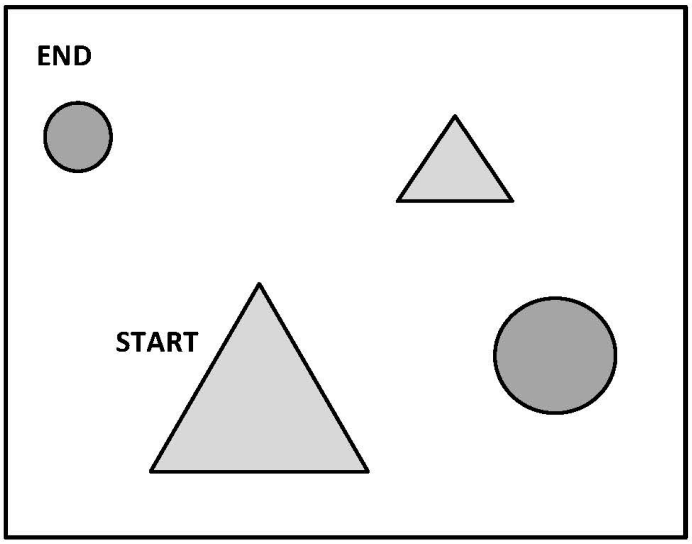



Adapted Trails Shapes Sequencing Test
END
START
Version 13 15-01-2024
3

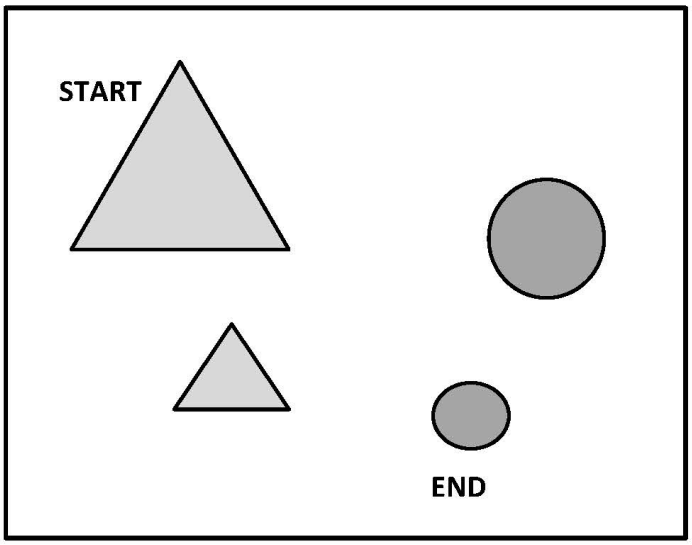
Adapted Trails Shapes

b) Switching

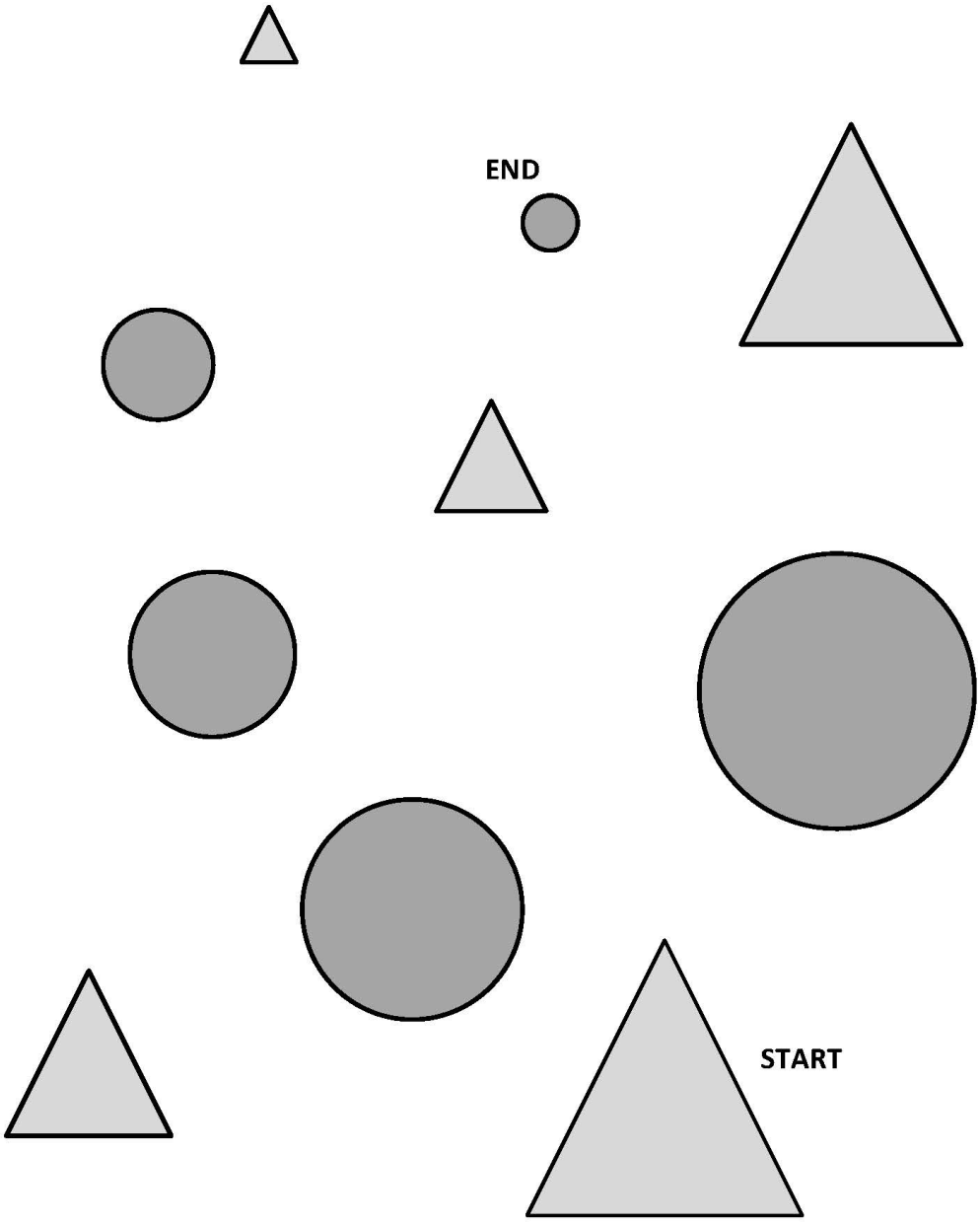
Demonstration



Practice



Adapted Trials Shapes Switching Test



Version 13 15-01-2024 5

Figure Drawing (Copy)

Draw it here

Figure Drawing (Immediate Recall)

Draw it here

Coding

─	L	Ɔ	>	⊥	+	∧	‖	Π
1	2	3	4	5	6	7	8	9

Demo

+	‖	Ɔ

Sample

Π	⊥	>	─	∧	L

─	>	‖	L	∧	+	Π	Ɔ	⊥	‖

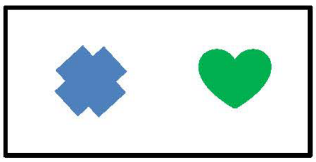
Ɔ	─	Π	L	⊥	+	>	Ɔ	∧	L

Π	‖	─	>	∧	+	⊥	Π	─	L

>	∧	L	⊥	+	Π	⊥	‖	+	>

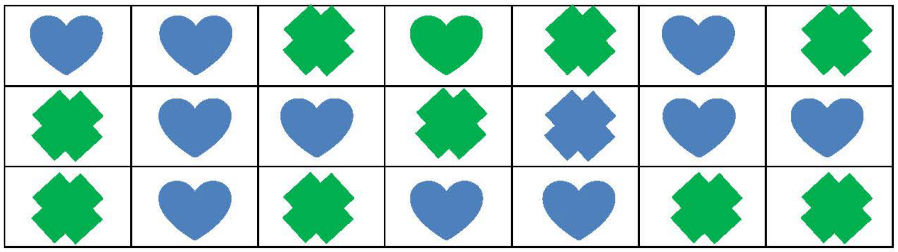
Ɔ	─	∧	‖	Ɔ	─	Ɔ	Π	+	Ɔ

Cancellation

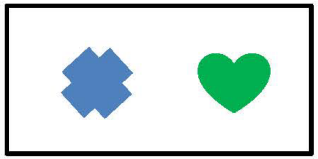


Demonstration

Practise



Version 13 15-01-2024 9



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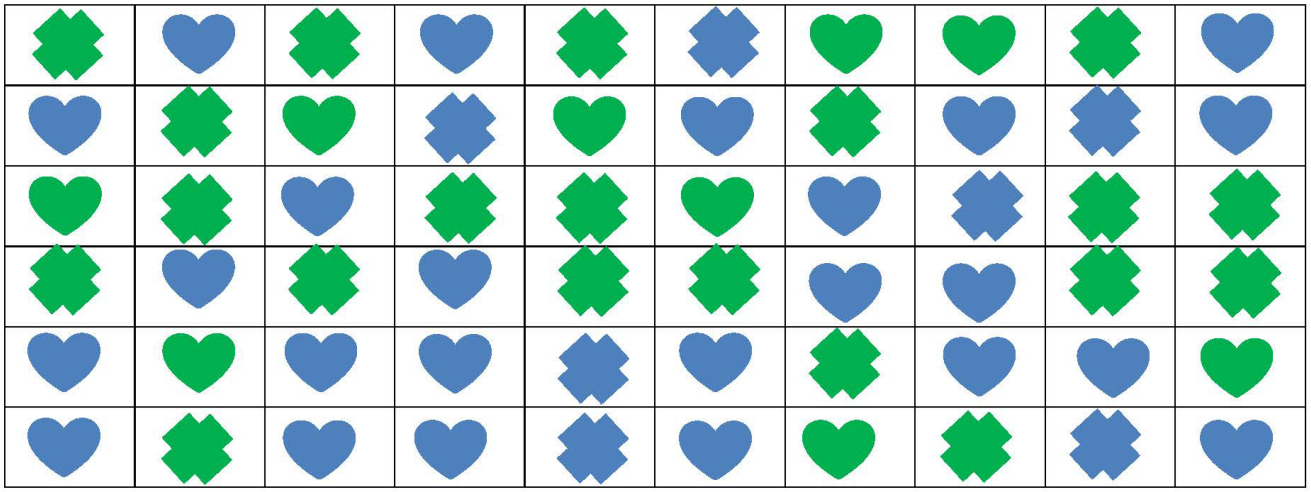
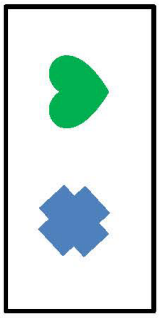
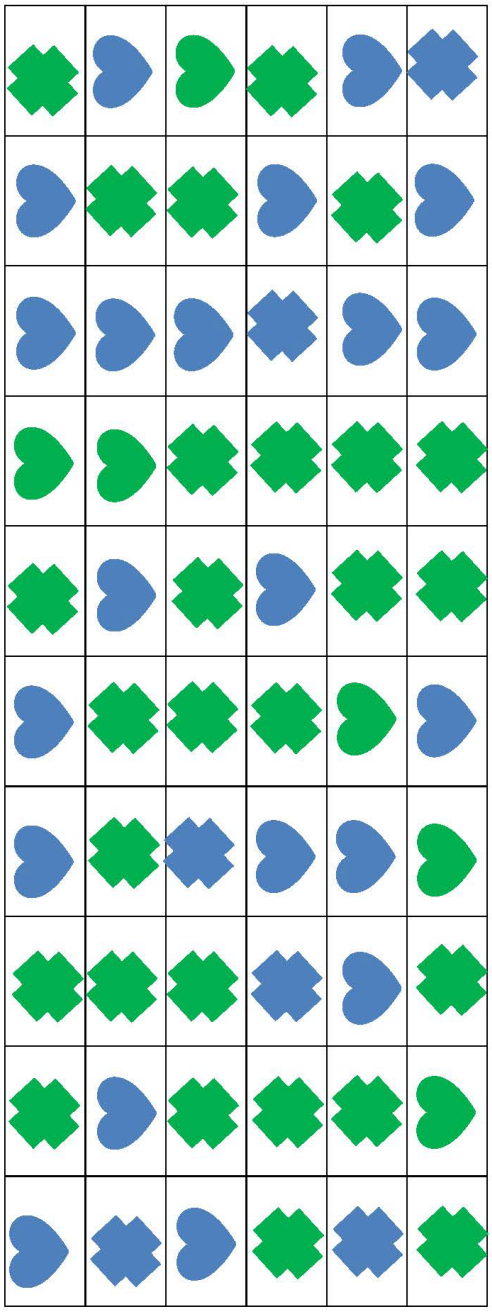






Task items (2)

11

Version 13 15-01-2024

Figure Drawing (Delayed Recall)

Draw it here

Construction

Circle

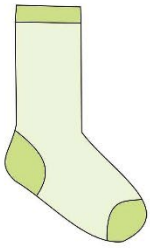
Square

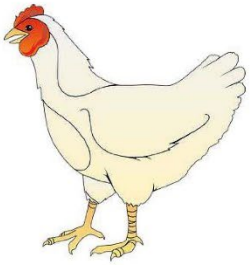
Tree

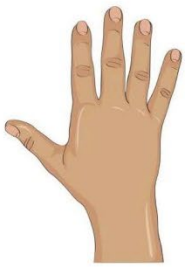
G.3 NBID Stimuli

Neurocognitive Battery
Intellectual Disability (NBID)

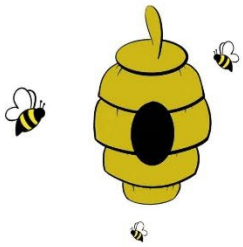
Stimulus Book

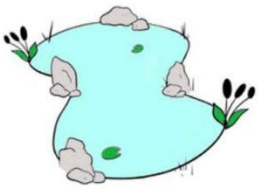


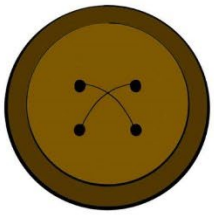


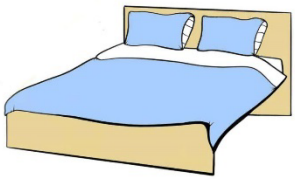



123
CITY









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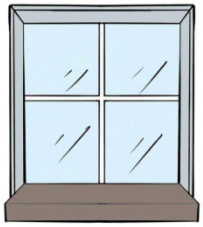

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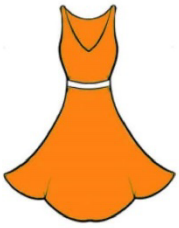

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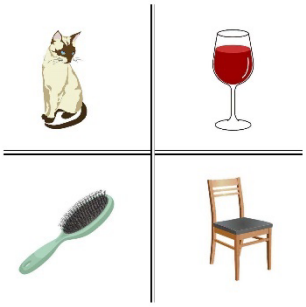
Picture Identification (training)



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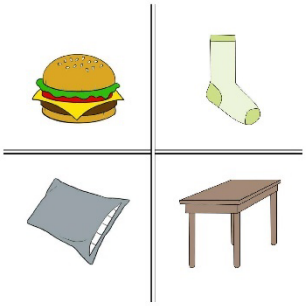
Picture Identification (training)



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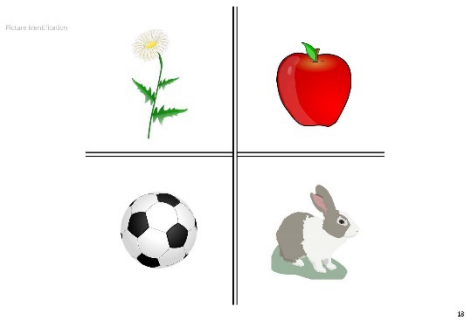

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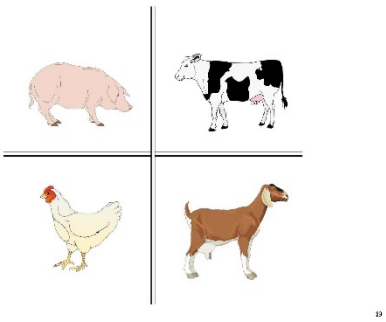

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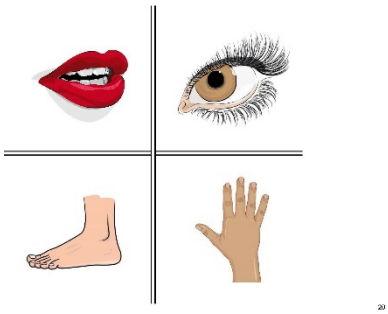

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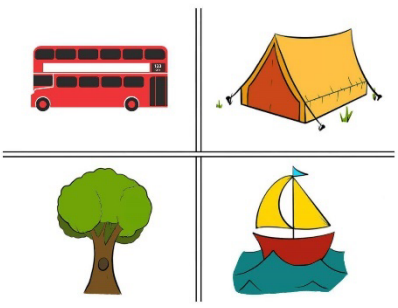

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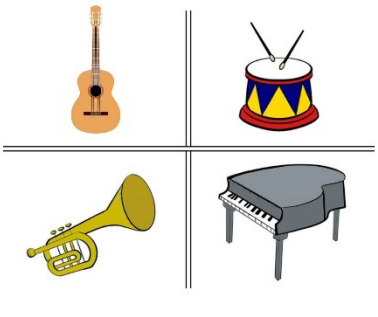

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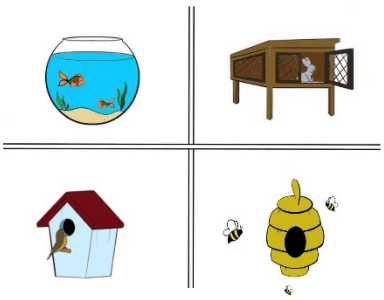

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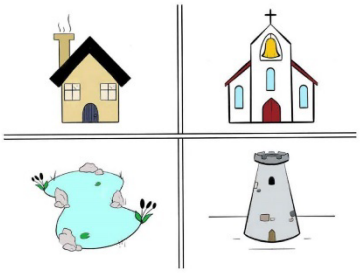

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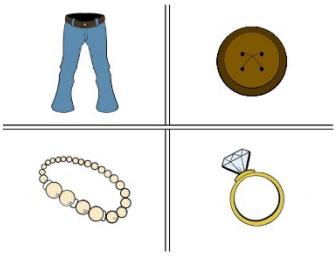

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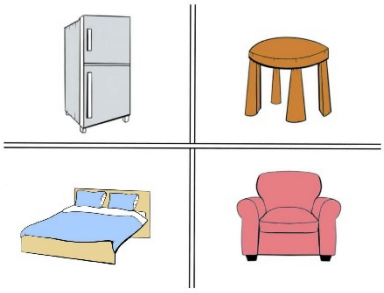

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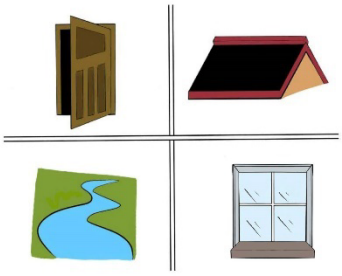

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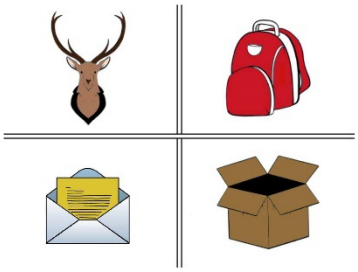

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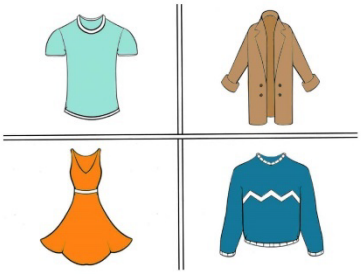

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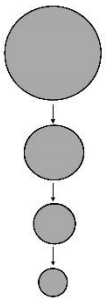
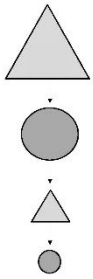
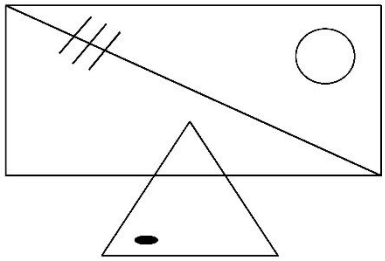
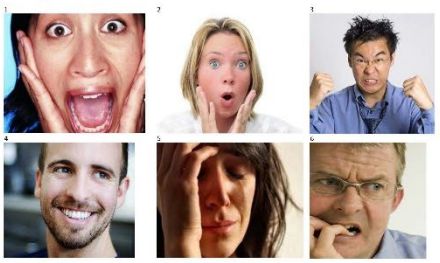
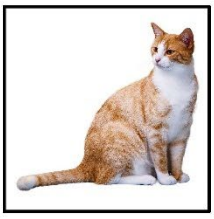
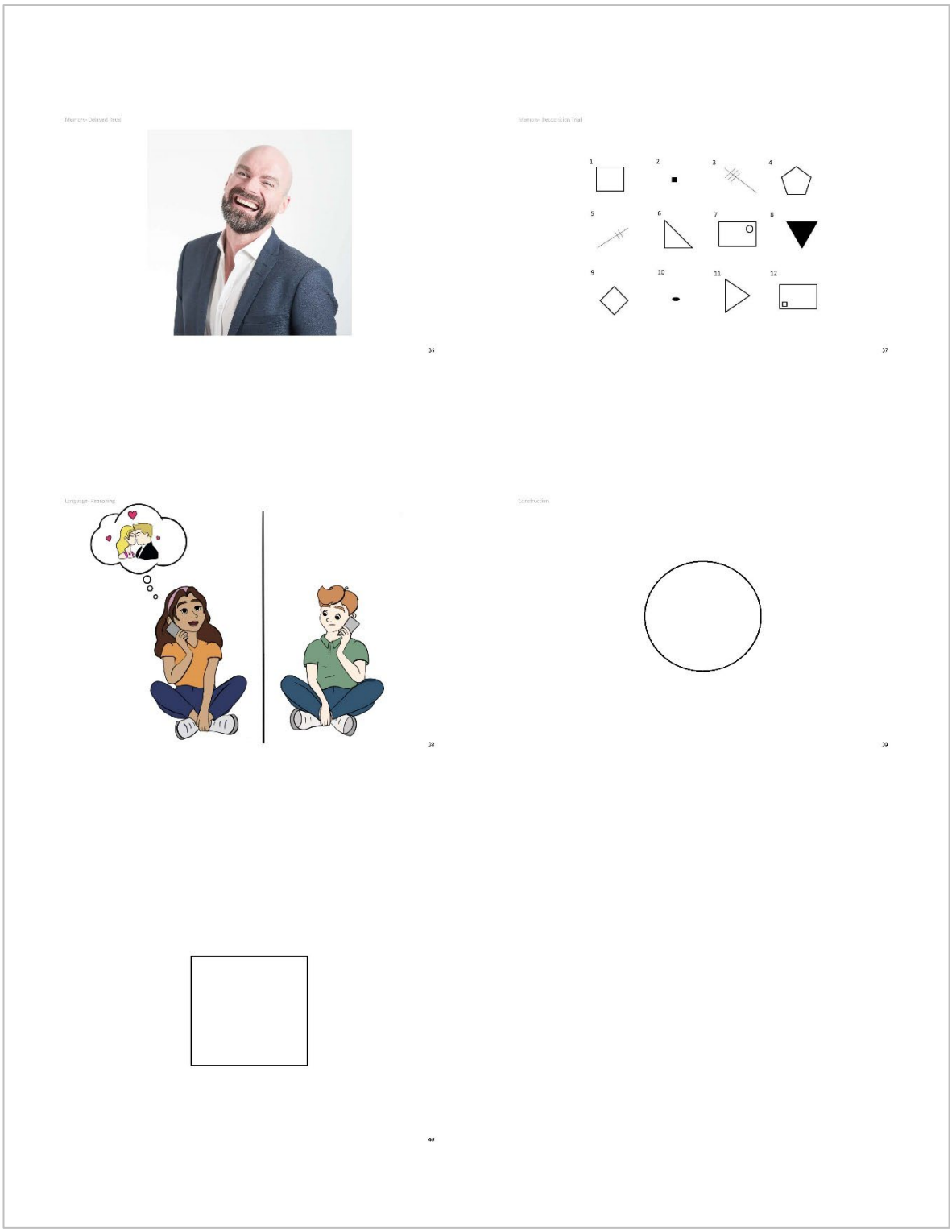












Note. For ease of inclusion, nine pages on the stimulus book are presented on one appendix page.

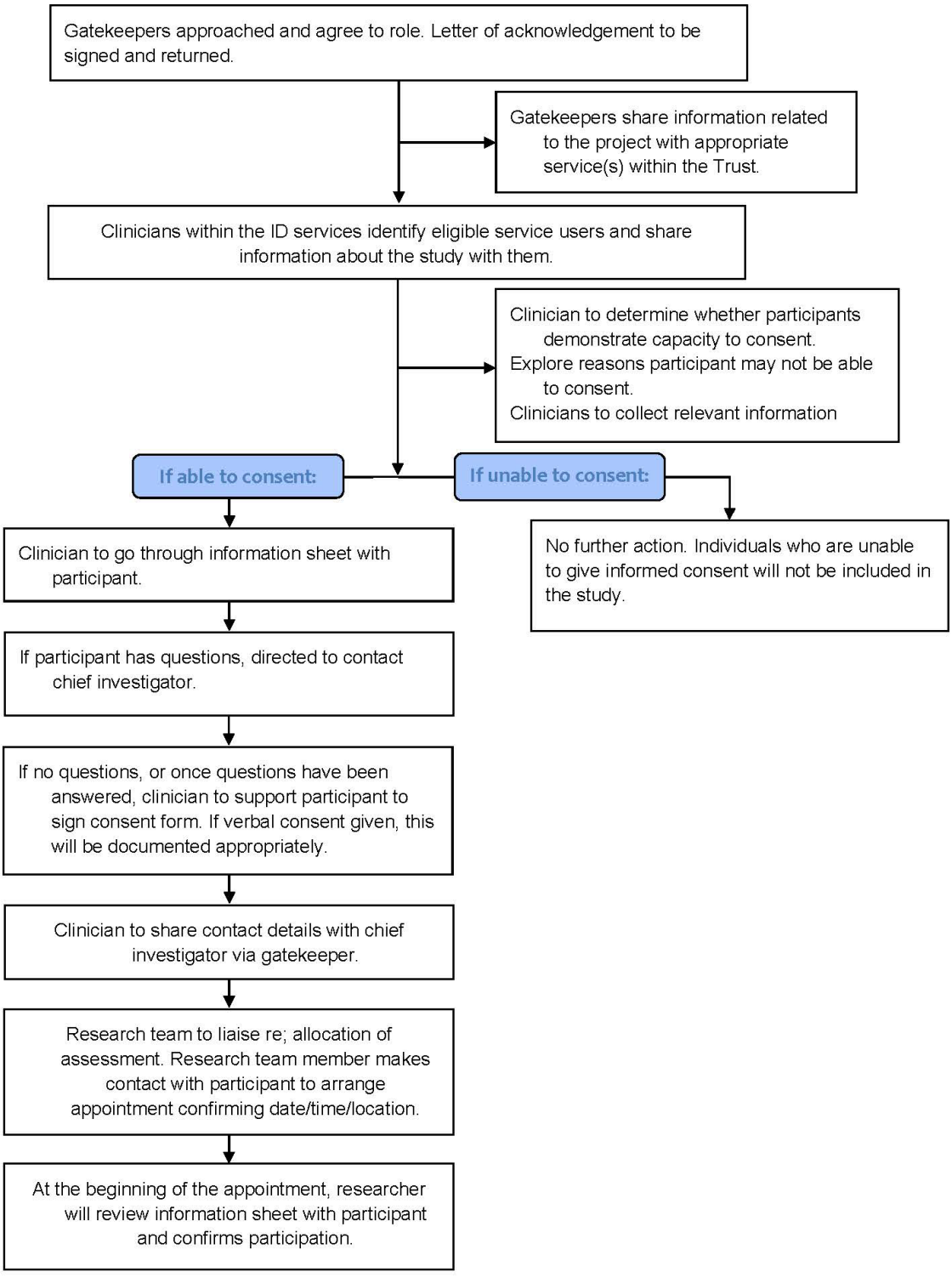
Appendix H NBID Amendments Based on Co-Production

Subtest	Suggestions for change from pilot study	Considerations and amendments
Orientation	• Analogue clock – change to digital as less familiarity with analogue clocks	• Time question removed • Scoring amendments: o Credit given for recognition items if response freely recalled o Additional credit given for elements of birthday, address etc (scoring 0,1,2) • Max score increased from 16 to 20
Remote Memory	N/A	N/A
Picture Naming	• May benefit from increasing difficulty • Coloured pictures	• Colour images included
Picture Identification	• May benefit from increasing difficulty • Coloured pictures	• Colour images included
Immediate Recall & 3 Trial Learning	N/A	N/A
Finger counting	N/A	• Subtest removed
Adapted Trails	• Holding the pen on the paper was detrimental to actually seeing the stimuli for most participants • Coloured shapes	<u>Sequencing:</u> • Stimuli changed – increased contrast between outline and shape fill • Switched order from smallest to biggest to biggest to smallest • Scoring refined – correct connections not correct circles to be more consistent with switching condition • Reordered items so no crossing lines when drawing connections <u>Switching:</u> In addition to amendments made for sequencing condition: • Changed order from circles first to triangles first as to differentiate from previous task.
Figure Drawing (Copy, Immediate Recall, Delayed Recall)	N/A	• Scoring revisions – inclusion of accuracy and location scores (similar to RBANS scoring criteria) with clearer guidance provided for each
Digit Span	N/A	N/A
Verbal Fluency	N/A	• Included recording of errors

Subtest	Suggestions for change from pilot study	Considerations and amendments
Emotion Recognition	N/A	• Letter changed from P to F. FAS used regularly, availability of norms for these letters • Numbers added to pictures • Considered using images of people with ID and different cultures – unable to implement for the present study
Action on Request	• May benefit from increasing difficulty	• Revision of scoring – column for not being able to complete (score 0) • 'Match' changed to 'candle' • Mobility subtest added
Coding	• Clearer instructions	• Instructions modified to be clearer • Format changes – individuals required to write numbers instead of symbols. Introduces possibility of providing oral responses if motor difficulties present.
Cats & Dogs	• Strong evidence of ceiling effects across all conditions indicates that the item needs to be made more difficult removed or replaced. • Stimuli need to be in colour.	• Colour stimuli <u>Inhibition & Switching:</u> • Administration order changed so inhibition/switch response not the first item
Delayed Recall & Recognition – Name and Address	N/A	• Scoring amended – free recall and recognition scores
Cancellation	N/A	N/A
Language reasoning	N/A	• Additional guidance given for scoring
Construction	N/A	• Clock drawing changed to tree to remove requirement of telling time

Appendix I Consent and Recruitment Process

Document: Recruitment process and obtaining consent/consent by proxy (v1.2, 17.07.23)
Study Title: The Development and Validation of a New Battery for Dementia Assessment in the Intellectual Disability Population



Appendix J Participant Information Sheet

Document: Participant Information Sheet (v1.2; 17.07.2023)
Study Title: The Development and Validation of a New Battery for Dementia Assessment in the Learning Disability Population
Researcher: Jade Dunning
ERGO Number: 79119
IRAS ID: 324094

Making a new thinking skills test for people with learning disabilities

Information Sheet

If you need help with this letter, you can ask someone to read it with you.

My name is Jade Dunning.

I am a Trainee Clinical Psychologist.

I am doing some research on memory and people with learning disabilities.

I am writing to ask if you are happy to take part in my research.

What is the research about?

Sometimes when people get older, they have problems with their thinking skills, like finding it harder to remember things.

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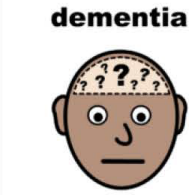
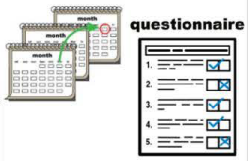
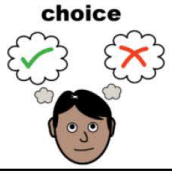
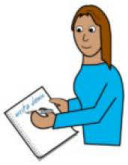
	Sometimes when people have problems with their thinking skills and remembering things it can be caused by dementia. Dementia changes how our brain works.
	It can be hard to know if someone has dementia. I want to help make it easier for people who struggle with their thinking skills to find out if they have dementia or not.
	We have made a new test to try and do this. This is a test of your thinking skills like how well you can remember things, solve problems and what you know. The test is made up of games and puzzles that help us to find out more about someone's thinking skills.
	We want to find out how good the puzzles and games are.
Why have I been asked to take part?	
	I would like to do the test with people who don't have problems with their thinking skills, like you. Someone who works with you from the learning disability team thought you might like to help with my research.
	I would also like to see the results of tests you have done before to see if they are similar or different.

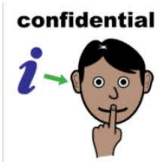
2

	We hope this research will help people with a learning disability find out if they have dementia and get the right support.
What will happen to me if I take part?	
	If you say yes to taking part (consent):
	• We will do some games and puzzles.
	• This will tell us what you are good at.
	• This will tell us what you might find more difficult.
	The test will take between 1 hour and 30 minutes and 2 hours.
	You can choose if you want to do the test.

3

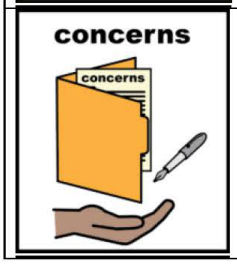
	If you say no, it will not change the care you get.
	If you say yes you will be helping other people with learning disabilities.
	If you feel upset or distressed at any time, you can ask to take a break.
	We will speak to you about where you would like the appointment to take place. It could be at home, an NHS building or a day centre.
What information will be collected?	
	I will write down some of the things you tell us. This will include:
	• Your name • Your age • Your birthday • Your gender • Your doctor's contact details • How you feel (if you are happy, sad or worried). • Things you are good at • Things you are not so good at

4

	Everything you tell me is confidential. That means it is private and I cannot tell anyone else. I can only tell someone if you, or someone else is at risk of harm. This is to keep everyone safe!
	If you say yes, I will share some of the information with your doctor.
	All information I write down will be kept safe on a computer protected by the University of Southampton or in a locked cabinet.
	Only people working with me will be able to see it.
Do I have to take part?	
	You can choose if you want to take part. You can say yes or no.
	This decision will not affect the care you get!
What happens if I change my mind?	

	If you change your mind, you can ask to stop at any time.
What will happen to the results?	
	We will use this research to help other people with learning disabilities.
	I will write about what I find in a report. This report will help to share knowledge and ways of supporting people with a learning disability.
	I won't use your name or write anything that would let people know who you are.
	The report will be seen by University students, scientists, NHS workers or others who support people with a learning disability.
	If you would like to see the results of the research, we can show you.
Where can I get more information?	

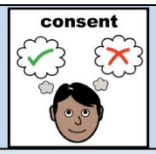
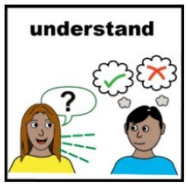
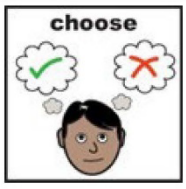
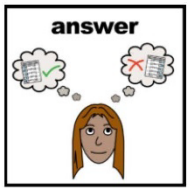
6

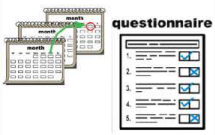
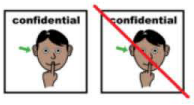
	If you have any questions you can, or ask someone to email me at jd16g12@soton.ac.uk
	If you have any concerns or complaints, you can contact 023 8059 5058 or rgoinfo@soton.ac.uk

7

Appendix K Participant Consent Form

Document: Participant Consent Form (v1.2; 17.07.2023)
Study Title: The Development and Validation of a New Battery for Dementia Assessment in the Intellectual Disability Population
Researcher: Jade Dunning
ERGO Number: 79119
IRAS ID: 324094

	**Making a new thinking skills test for people with learning disabilities**	
	Consent form Please tick the box next to each statement when you are sure you understand and consent to it. If you need help with this form, you can ask someone to read it with you.	
	I have read and understood the information sheet (v1.2; 17.07.2023):	☐
	**I understand and agree that:** It is my choice to take part	☐
	**I understand and agree that:** If I do not want to take part it will not affect the care I get	☐
	**I understand and agree that:** If I take part, I do not have to answer all the questions	☐

	**I understand and agree that:** It is ok to change my mind about taking part. Changing my mind will not affect the care I get.	☐
	I have been able to ask questions and they have been answered.	☐
	I am happy for my details to be shared with the research team so I can be contacted about the study.	☐
	I am happy for the research team to use the information and answers I give. This information will be kept safe in a locked cabinet and password protected computer.	☐
	I am happy for the research team to see my most recent memory test.	☐
	If I tell the research team something that makes them worry that me or someone else might get hurt, they will need to tell other people. This is to keep everyone safe.	☐

2

Images from www.easyonthei-leeds.nhs.uk (*symbols copyright © LYPFT*)

	I am happy for my results to be shared with my doctor	☐
	The research team will write the results up in a report. The report might include some of the things I say.	☐
	This report will be seen by University students, scientists, NHS workers or others who support people with a learning disability.	☐
	My name or personal information will not be included in the report.	☐

Please tick the box to show if you would like to take part in this project:

I consent to taking part in this project. ☐

Please sign your name below:

Participant name: ……..

Name of the person supporting you / healthcare professional:

……

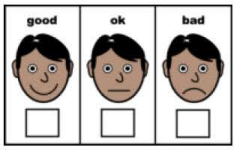
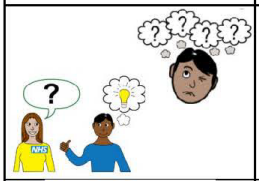
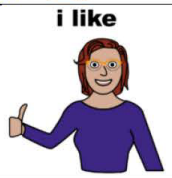
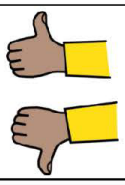
3

Images from www.easyonthei-leeds.nhs.uk (*symbols copyright © LYPFT*)

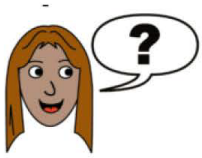
Date:.....
Name of researcher: .....
Date:.....

Appendix L Participant Debrief Form

Document: Debrief Form (v1.2; 17.07.2023)
Study Title: The Development and Validation of a New Battery for Dementia Assessment in the Learning Disability Population
Researcher: Jade Dunning
ERGO Number: 79119
IRAS ID: 324094

Making a new thinking skills test for people with learning disabilities	
	**Debrief Form** If you need help with this letter, you can ask someone to read it with you.
	The aim of this research was to check the following:
	How good our puzzles and games were
	If the puzzles and games were too easy or too hard
	If you enjoyed the puzzles and games
	By taking part, you have helped us:
	Build a new test that tells us what you are good at and what things you find more difficult.

Images from www.easyonthei-leeds.nhs.uk (*symbols copyright © LYPFT*)

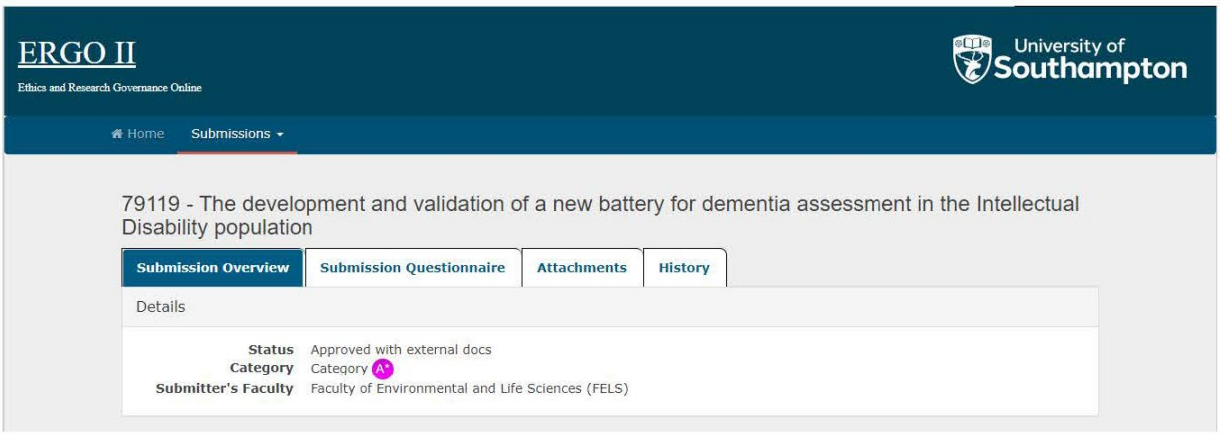
	And helps us understand how everyone has different skills!
	I will write up these findings in a report. The report will not include your name or anything that would let people know who you are.
	I will share the results with your doctor if you are happy for me to do this.
	If you want to see the final report or if you have any questions you can ask me. My email is jd16g12@soton.ac.uk
	Thank you for taking part and helping my research!

2

Images from www.easyonthei-leeds.nhs.uk (*symbols copyright © LYPFT*)

Appendix M Ethics Approvals

M.1 University of Southampton Ethics Approval



M.2 Health Research Authority (HRA) Letter of Approval

Ymchwil Iechyd a Gofal Cymru
Health and Care Research Wales

NHS
Health Research Authority

Miss Jade Dunning
Chestnut Drive
Shave Lane
Horton, Ilminster, Somerset
TA19 9QP

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

18 August 2023

Dear Miss Dunning

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

Study title:	**The Development and Validation of a New Battery for Dementia Assessment in the Intellectual Disability Population**
IRAS project ID:	**324094**
Protocol number:	**N/A**
REC reference:	**23/SC/0229**
Sponsor	**University of Southampton**

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.

Please see IRAS Help for information on working with NHS/HSC organisations in Northern Ireland and Scotland.

How should I work with participating non-NHS organisations?
HRA and HCRW Approval does not apply to non-NHS organisations. You should work with your non-NHS organisations to obtain local agreement in accordance with their procedures.

What are my notification responsibilities during the study?

The standard conditions document "*After Ethical Review – guidance for sponsors and investigators*", issued with your REC favourable opinion, gives detailed guidance on reporting expectations for studies, including:

- Registration of research
- Notifying amendments
- Notifying the end of the study

The HRA website also provides guidance on these topics, and is updated in the light of changes in reporting expectations or procedures.

Who should I contact for further information?
Please do not hesitate to contact me for assistance with this application. My contact details are below.

Your IRAS project ID is **324094**. Please quote this on all correspondence.

Yours sincerely,
Rekha Keshvara

Approvals Manager

Email: approvals@hra.nhs.uk

Copy to: *Mrs Linda Hammond*

Appendix N NHS Trusts Letters of Access

N.1 Dorset Healthcare University NHS Foundation Trust Site Letter of Access

NHS
Dorset HealthCare University
NHS Foundation Trust

PRIVATE & CONFIDENTIAL
Miss Jade Dunning
University of Southampton
School of Psychology
Faculty of Environmental & Life Sciences
Building 44, Highfield Campus
Southampton
SO17 1BJ

Research & Development
11 Shelley Road
Boscombe
Bournemouth
Dorset
BH1 4JQ

Tel: 01202 443024
Web: www.dorsethealthcare.nhs.uk
Email:
dhc.researchdevelopment@nhs.net

10TH November, 2023

Dear Jade

Letter of Access for Research

IRAS 324094 – The Development & Validatin of a New Battery for Dementia Assessment in the Intellectual Disability

This letter should be presented to each participating organisation] before you commence your research at that site. The participating organisation(s) is/are:

Dorset HealthCare University NHS Foundation Trust

In accepting this letter, each participating organisation confirms your right of access to conduct research through their organisation for the purpose and on the terms and conditions set out below. This right of access commences on **10/11/2023** and ends on **31/07/2024** unless terminated earlier in accordance with the clauses below.

You have a right of access to conduct such research as confirmed in writing in the confirmation of capacity and capability for research from Dorset HealthCare University NHS Foundation Trust. Please note that you cannot start the research until the Principal Investigator for the research project has received notification from us giving confirmation from the individual organisation(s) of their agreement to conduct the research.

The information supplied about your role in research at the organisation(s) has been reviewed and you do not require an honorary research contract with the organisation(s). We are satisfied that such pre-engagement checks as we consider necessary have been carried out. Evidence of checks should be available on request to the organisation(s).

You are considered to be a legal visitor to the organisations premises. You are not entitled

to any form of payment or access to other benefits provided by the organisation(s) or this organisation to employees and this letter does not give rise to any other relationship between you and the organisation(s), in particular that of an employee.

While undertaking research through the organisation(s) you will remain accountable to your substantive employer but you are required to follow the reasonable instructions of the organisation(s) or those instructions given on their behalf in relation to the terms of this right of access.

Where any third party claim is made, whether or not legal proceedings are issued, arising out of or in connection with your right of access, you are required to co-operate fully with any investigation by the organisation(s) in connection with any such claim and to give all such assistance as may reasonably be required regarding the conduct of any legal proceedings.

You must act in accordance with the organisations policies and procedures, which are available to you upon request, and the UK Policy Framework for Health and Social Care Research.

You are required to co-operate with the organisation(s) in discharging its/their duties under the Health and Safety at Work Act 1974 and other health and safety legislation and to take reasonable care for the health and safety of yourself and others while on the organisations premises. You must observe the same standards of care and propriety in dealing with patients, staff, visitors, equipment and premises as is expected of any other contract holder and you must act appropriately, responsibly and professionally at all times.

If you have a physical or mental health condition or disability which may affect your research role and which might require special adjustments to your role, if you have not already done so, you must notify your employer and each organisation prior to commencing your research role at that organisation.

You are required to ensure that all information regarding patients or staff remains secure and *strictly confidential* at all times. You must ensure that you understand and comply with the requirements of the NHS Confidentiality Code of Practice and the Data Protection Act 2018. Furthermore you should be aware that under the Act, unauthorised disclosure of information is an offence and such disclosures may lead to prosecution.

You should ensure that, where you are issued with an identity or security card, a bleep number, email or library account, keys or protective clothing, these are returned upon termination of this arrangement. Please also ensure that while on the organisations premises you wear your ID badge at all times, or are able to prove your identity if challenged. Please note that the organisation(s) do not accept responsibility for damage to or loss of personal property.

This organisation may revoke this letter and any organisation(s) may terminate your right to attend at any time either by giving seven days' written notice to you or immediately without any notice if you are in breach of any of the terms or conditions described in this letter or if you commit any act that we reasonably consider to amount to serious misconduct or to be disruptive and/or prejudicial to the interests and/or business of the organisation(s) or if you are convicted of any criminal offence. You must not undertake regulated activity if you are barred from such work. If you are barred from working with adults or children this letter of access is immediately terminated. Your employer will immediately withdraw you from undertaking this or any other regulated activity and you MUST stop undertaking any regulated activity immediately.

Your substantive employer is responsible for your conduct during this research project and may in the circumstances described above instigate disciplinary action against you.

No organisation will indemnify you against any liability incurred as a result of any breach of confidentiality or breach of the Data Protection Act 2018. Any breach of the Data Protection Act 2018 may result in legal action against you and/or your substantive employer.

If your current role or involvement in research changes, or any of the information provided in your Research Passport changes, you must inform your employer through their normal procedures. You must also inform your nominated manager in each participating organisation and the R&D office in this organisation.

Yours sincerely

Paul Walters
Associate Medical Director for Research & Development

cc: Dr Alethea Charlton, University of Southampton

Dorset HealthCare

Our vision is to lead and inspire through excellence, compassion and expertise in all we do

N.2 Somerset NHS Foundation Trust Site Letter of Access

Somerset NHS Foundation Trust

Research & Development Operational Group
Department of Clinical Research
Musgrove Park Hospital
TAUNTON
TA1 5DA

E-mail: research@somersetft.nhs.uk

Miss J Dunning
Trainee Clinical Psychologist
University of Southampton
School of Psychology
Faculty of Environment and Life Sciences
Building 44, Highfield Campus
SOUTHAMPTON SO17 1BJ

13 November 2023

Dear Miss Dunning

LETTER OF ACCESS FOR RESEARCH – THE DEVELOPMENT AND VALIDATION OF A NEW BATTERY FOR DEMENTIA ASSESSMENT IN THE INTELLECTUAL DISABILITY POPULATION (R&D ref: 3180)

The participating organisation is: Somerset NHS Foundation Trust, Musgrove Park Hospital.

In accepting this letter, each participating organisation confirms your right of access to conduct research through their organisation for the purpose and on the terms and conditions set out below. This right of access commences on 14 November 2023 and ends on 31 July 2024 unless terminated earlier in accordance with the clauses below.

You have a right of access to conduct such research as confirmed in writing in the letter of permission for research from Somerset NHS Foundation Trust. Please note that you cannot start the research until the Principal Investigator for the research project has received a letter from us giving confirmation from the individual organisation of their agreement to conduct the research.

The information supplied about your role in research at the organisation has been reviewed and you do not require an honorary research contract with the organisation. We are satisfied that such pre-engagement checks as we consider necessary have been carried out. Evidence of checks should be available on request to the organisation.

You are considered to be a legal visitor to the organisations premises. You are not entitled to any form of payment or access to other benefits provided by the organisation to employees and this letter does not give rise to any other relationship between you and the organisation, in particular that of an employee.

While undertaking research through the organisation you will remain accountable to your substantive employer but you are required to follow the reasonable instructions of the organisation or those instructions given on their behalf in relation to the terms of this right of access.

Where any third party claim is made, whether or not legal proceedings are issued, arising out of or in connection with your right of access, you are required to co-operate fully with any investigation by the organisation in connection with any such claim and to give all such assistance as may reasonably be required regarding the conduct of any legal proceedings.

You must act in accordance with the organisations policies and procedures, which are available to you upon request, and the Research Governance Framework.

You are required to co-operate with the organisation in discharging its duties under the Health and Safety at Work etc Act 1974 and other health and safety legislation and to take reasonable care for the health and safety of yourself and others while on the organisations premises. You must observe the same standards of care and propriety in dealing with patients, staff, visitors, equipment and premises as is expected of any other contract holder and you must act appropriately, responsibly and professionally at all times.

If you have a physical or mental health condition or disability which may affect your research role and which might require special adjustments to your role, if you have not already done so, you must notify your employer and each organisation prior to commencing your research role at that organisation.

You are required to ensure that all information regarding patients or staff remains secure and *strictly confidential* at all times. You must ensure that you understand and comply with the requirements of the NHS Confidentiality Code of Practice and the Data Protection Act 2018. Furthermore you should be aware that under the Act, unauthorised disclosure of information is an offence and such disclosures may lead to prosecution.

You should ensure that, where you are issued with an identity or security card, a bleep number, email or library account, keys or protective clothing, these are returned upon termination of this arrangement. Please also ensure that while on the organisations premises you wear your ID badge at all times, or are able to prove your identity if challenged. Please note that the organisation does not accept responsibility for damage to or loss of personal property.

This organisation may revoke this letter and any organisation may terminate your right to attend at any time either by giving seven days' written notice to you or immediately without any notice if you are in breach of any of the terms or conditions described in this letter or if you commit any act that we reasonably consider to amount to serious misconduct or to be disruptive and/or prejudicial to the interests and/or business of the organisation or if you are convicted of any criminal offence. You must not undertake regulated activity if you are barred from such work. If you are barred from working with adults or children this letter of access is immediately terminated. Your employer will immediately withdraw you from undertaking this or any other regulated activity and you MUST stop undertaking any regulated activity immediately.

Your substantive employer is responsible for your conduct during this research project and may in the circumstances described above instigate disciplinary action against you.

No organisation will indemnify you against any liability incurred as a result of any breach of confidentiality or breach of the Data Protection Act 2018. Any breach of the Data Protection Act 2018 may result in legal action against you and/or your substantive employer.

If your current role or involvement in research changes, or any of the information provided in your Research Passport changes, you must inform your employer through their normal procedures. You must also inform your nominated manager in this participating organisation and the R&D office in this organisation.

Yours sincerely

Dr R INNES
Chair
Research & Development Operational Group

cc. Dr L Robbins, Consultant Clinical Psychologist, Somerset NHS Foundation Trust, Learning Disabilities East, Summerlands Hospital Site, Preston Road, YEOVIL BA20 2BX
Dr A Charlton, Deputy Programme Director/Docorate in Clinical Psychology, University of Southampton, Department of Psychology, University Road, SOUTHAMPTON SO17 1BJ

N.3 Southern Health NHS Foundation Trust Site Letter of Access

NHS
Southern Health
NHS Foundation Trust

Research & Innovation
Tom Rudd Unit
Moorgreen Hospital
Botley Road
West End
Southampton
SO30 3JB

Tel: 023 8047 5160

www.southernhealth.nhs.uk

LOA/JP

1 December 2023

Jade Dunning
School of Psychology
Faculty of Environment and Life Sciences
Building 44
Highfield Campus
Southampton
SO17 1BJ

Jade.Dunning@soton.ac.uk

Dear Jade

Letter of access for research – The Development and Validation of a New Battery for Dementia Assessment in the Intellectual Disability Population

I am pleased to offer you a Letter of Access to conduct research within Southern Health NHS Foundation Trust for the purpose and on the terms and conditions set out below.

Please note that you cannot start the research at this organisation until the Investigator for the research project has received a letter from the Southern Health Research & Innovation Department giving Southern Health NHS Foundation Trust permission to conduct the project.

This right of access commences on 01/12/2023 and ends on 31/07/2024 unless terminated earlier in accordance with the clauses below. Upon expiry of the contract, you will not be able to undertake any research activity which requires access to SHFT patients or SHFT patient data.

The information supplied about your role in research at Southern Health NHS Foundation Trust has been reviewed and you do not require an honorary research contract with the organisation. Your employer has confirmed in writing to Southern Health NHS Foundation Trust that the necessary pre-engagement checks are in place in accordance with the role you plan to carry out in this Trust. We are satisfied that such pre-engagement checks as we consider necessary have been carried out.

You are considered to be a legal visitor to Southern Health NHS Foundation Trust premises. You are not entitled to any form of payment or access to other benefits

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provided by Southern Health NHS Foundation Trust to employees and this letter does not give rise to any other relationship between you and Southern Health NHS Foundation Trust, in particular that of an employee.

While undertaking research through Southern Health NHS Foundation Trust, you will remain accountable to Psychology Department University of Southampton but you are required to follow the reasonable instructions of your nominated manager Dr Alda Almaro at Southern Health NHS Foundation Trust or those given on her/his behalf in relation to the terms of this right of access.

Where any third party claim is made, whether or not legal proceedings are issued, arising out of or in connection with your right of access, you are required to co-operate fully with any investigation by University of Southampton or this organisation in connection with any such claim and to give all such assistance as may reasonably be required regarding the conduct of any legal proceedings.

You must act in accordance with Southern Health NHS Foundation Trust policies and procedures, which are available via the Trust website, and the UK Policy Framework for Health and Social Care Research.

You are required to co-operate with Southern Health NHS Foundation Trust in discharging its duties under the Health and Safety at Work etc Act 1974 and other health and safety legislation and to take reasonable care for the health and safety of yourself and others while on Southern Health NHS Foundation Trust premises. Although you are not a contract holder, you must observe the same standards of care and propriety in dealing with patients, staff, visitors, equipment and premises as is expected of a contract holder and you must act appropriately, responsibly and professionally at all times.

If you have a physical or mental health condition or disability which may affect your research role and which might require special adjustments to your role, if you have not already done so, you must notify your employer and Southern Health NHS Foundation Trust prior to commencing your research role at each site.

You are required to ensure that all information regarding patients or staff remains secure and strictly confidential at all times. You must ensure that you understand and comply with the requirements of the NHS Confidentiality Policy and the Data Protection Act 2018. Furthermore you should be aware that under the Act, unauthorised disclosure of information is an offence and such disclosures may lead to prosecution. Southern Health NHS Foundation Trust will not indemnify you against any liability incurred as a result of any breach of confidentiality or breach of the Data Protection Act 2018. Any breach of the Data Protection Act 2018 may result in legal action against you and/or your substantive employer.

You should ensure that, where you are issued with an identity or security card, a bleep number, email or library account, keys or protective clothing, these are returned upon termination of this arrangement. Please also ensure that while on the premises you wear

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your ID badge at all times, or are able to prove your identity if challenged. Please note that Southern Health NHS Foundation Trust accept no responsibility for damage to or loss of personal property.

This letter may be revoked and your right to attend Southern Health NHS Foundation Trust terminated at any time either by giving seven days' written notice to you or immediately without any notice if you are in breach of any of the terms or conditions described in this letter or if you commit any act that we reasonably consider to amount to serious misconduct or to be disruptive and/or prejudicial to the interests and/or business of the organisation or if you are convicted of any criminal offence. You must not undertake regulated activity if you are barred from such work. If you are barred from working with adults or children this letter of access is immediately terminated. Your employer will immediately withdraw you from undertaking this or any other regulated activity and you MUST stop undertaking any regulated activity immediately.

Your substantive employer is responsible for your conduct during this research project and may in the circumstances described above instigate disciplinary action against you.

If your circumstances change in relation to your health, criminal record, professional registration or suitability to work with adults or children, or any other aspect that may impact on your suitability to conduct research, or your role in research changes, you must inform the organisation that employs you through its normal procedures. You must also inform your nominated manager at Southern Health NHS Foundation Trust.

It is a condition of your Letter of Access that you have had recent Information Governance (Data Security) training (this must be refreshed every 12 months) and that you understand that you are responsible for your conduct whilst contacting patients and collecting data within the Trust.

Yours sincerely

Research & Innovation
Southern Health NHS Foundation Trust

cc: **HR Department**, Southern Health NHS Foundation Trust
HR department of the substantive employer

I confirm that I understand and accept the terms of this letter:

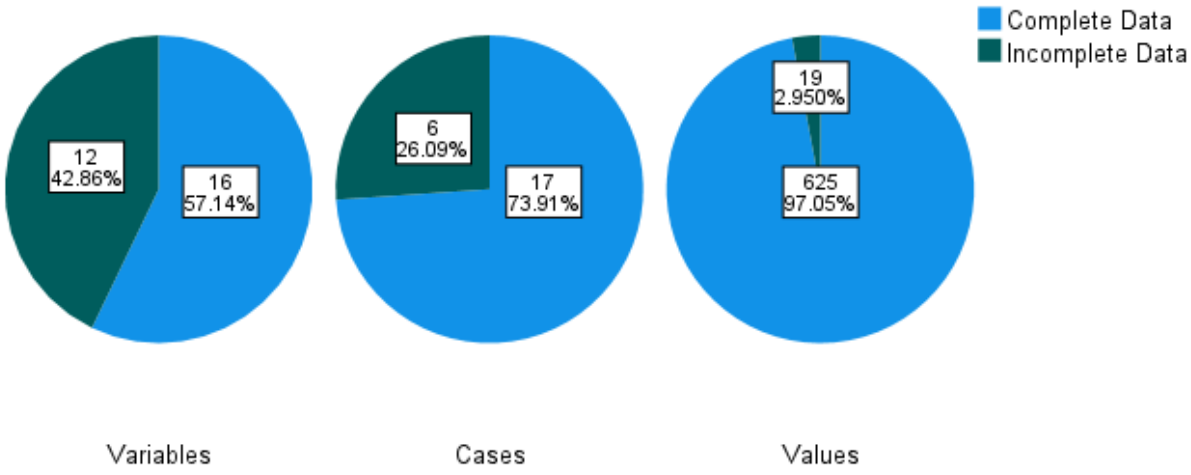
Name: Jade Dunning.....

Signed:  .....

Date: 01/12/2023.....

Appendix O Patterns of Missing Values

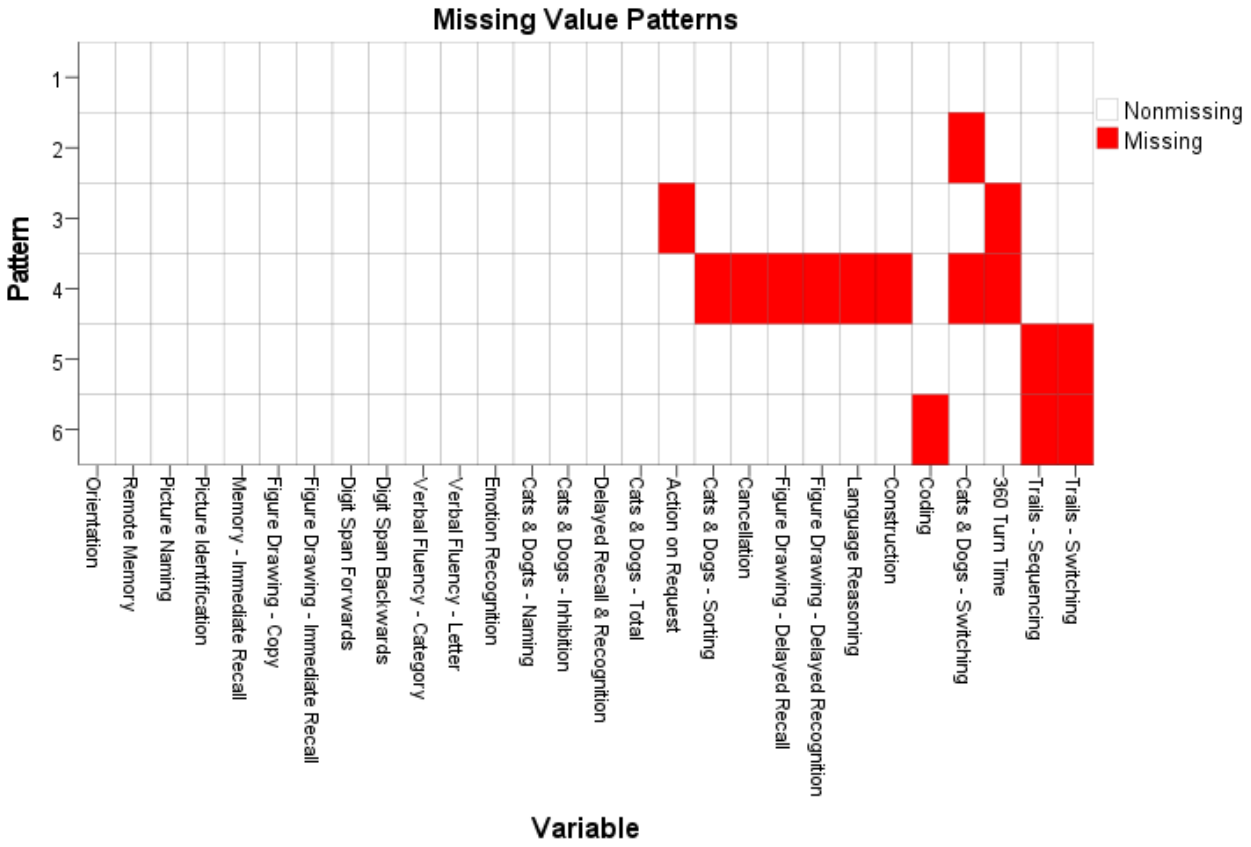
O.1 Overall Summary of Missing Values



O.2 Missing Variables Summary Statistics

Variable	Missing		Valid *N*
	N	Percent (%)	
6a – Trails Shapes – Switching	3	13.0	20
6a – Trails Shapes - Sequencing	3	13.0	20
11b - 360° Turn Time	2	8.7	21
13c – Cats & Dogs – Switching	2	8.7	21
12 – Coding	2	8.7	21
18 – Construction	1	4.3	22
17 – Language Reasoning	1	4.3	22
16 – Figure Drawing – Delayed Recognition	1	4.3	22
16 – Figure Drawing – Delayed Recall	1	4.3	22
15 – Cancellation	1	4.3	22
13d – Cats & Dogs – Sorting	1	4.3	22
11a – Action on Request	1	4.3	22

O.3 Missing Values Patterns Output



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