Neuropsychological Assessments of Dementia in Down Syndrome and Intellectual Disabilities

Second Edition

Vee P. Prasher
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To
Claire Adams and Ravinder Mann.
My professional colleagues whose support is very much appreciated and without whom I could not continue my ongoing academic duties.

Preface

Neuropsychological testing in the field of psychiatry is now an established part of the mental health assessment process, arguably more so for the non-intellectually disabled population than for persons with intellectual disability (ID). In the past neuropsychological assessments of persons with ID usually meant an assessment for global developmental delay, of intelligence (intelligence quotient testing) or of level of adaptive behavior. Popular tests included the Stanford-Binet, Wechsler Intelligence Scales, Bayley Scales of Infant Development, the Griffin Mental Developmental Scales, and the Vineland Social Maturity Scale. These were assessments of the "overall" level of ability. Arthur Dalton in New York was one of a few pioneering clinicians who, in the late 1970s and onwards, focused on the development of tests for specific areas of cognition in persons with ID. Following his work, subsequent researchers, in the latter part of the twentieth century, have proposed and developed a number of measures not only to detect the level of cognitive abilities but also to measure cognitive decline, a perquisite to the diagnosis of dementia.

At the beginning of the twenty-first century, as highlighted in this book *Neuropsychological Assessments of Dementia in Down Syndrome and Intellectual Disabilities* (*2nd edition*), several neuropsychological measures have been developed with some widely tested, to aid the clinical diagnosis of dementia or specifically dementia in Alzheimer's disease (DAD). Neuropsychological assessments no longer remain the sole responsibility of psychologists, as psychiatrists, researchers, ID nurses, neuroscientists, and even family members all now play a part in the development and administration of neuropsychological tests.

As a consequence of the development in neuropsychological tests of older persons with ID, there has been a steady growth in the publication of research reports, case studies, reviews, and drug trials, using such instruments. It is now standard practice for at least one neuropsychological measure to be used in standard clinical practice, and indeed internationally recognized diagnostic criteria for the diagnosis of dementia often require that one or more of these measures are used as part of the diagnostic pathway.

As in the first edition of *Neuropsychological Assessments of Dementia in Down Syndrome and Intellectual Disabilities*, it would continue to be an injustice to the

researchers and clinicians who developed these tests for their tests to be further appraised by myself in this second edition of the book. This book contains an up-to-date review of the most important neuropsychological measures used in the assessment of dementia by the researchers who developed or who are the principal researchers associated with the tests. A number of neuropsychological tests not previously reviewed have now been added to this second edition of *Neuropsychological Assessments of Dementia in Down Syndrome and Intellectual Disabilities*. I am extremely grateful to all the contributors who continue to feel that neuropsychological assessments of dementia in persons with ID remain an important area of clinical care and have kindly continued to contribute to this second edition.

As for the first edition of *Neuropsychological Assessments of Dementia in Down Syndrome and Intellectual Disabilities*, the overall organization of this second edition is that the most popular and most widely used tests have been given precedence in chapter order as compared to the newer, less well-developed tests. A number of measures not reviewed for the first edition have now been added as later chapters. Test researchers were asked to comment on new areas of development since the publication of the first edition. Personal views on the test continued to be encouraged. As for the first edition, where possible, to aid readers, a sample page of each test has been included in the "Appendix" section. This gives the readers a chance to catch sight of the layout of at least a few of the test questions.

As for the first edition of *Neuropsychological Assessments of Dementia in Down Syndrome and Intellectual Disabilities*, a few comments will be made on the terminology adopted in this text. The term "Alzheimer's disease" has been used to denote the neuropathological disease process, while "Dementia in Alzheimer's disease" (DAD) has been used to refer to the clinical aspects of the neurodegenerative condition. Dementia in Alzheimer's type (DAT) is used where it is specifically used as a diagnostic term in the test measure. It is accepted that such terms have not as yet gained universal acceptance. Further the term "intellectual disability(ies)" is used in this text to be synonymous with "mental retardation," "learning disabilities," "mental handicap," and "intellectual handicap."

Birmingham, UK Vee P. Prasher

Acknowledgment

I continue to be indebted to the scholarly clinicians and researchers who have contributed so benevolently and whose names are listed at the beginning of this book. Without their ongoing support and contribution, *Neuropsychological Assessments of Down Syndrome and Intellectual Disabilities* (*Second edition*) would not exist.

Contents

Abbreviations

ABDQ	Adaptive Behavior Dementia Questionnaire
ABS	Adaptive Behavior Scale
AD	Alzheimer's disease
CAMDEX-DS	Cambridge Examination for Mental Disorders of Older People with Down's Syndrome and Others with Intellectual Disabilities
DAD	Dementia in Alzheimer's disease
DAT	Dementia of Alzheimer's type
DLD	Dementia Questionnaire for people with Learning Disabilities
DMR	Dementia Questionnaire for Mentally Retarded Persons
DS	Down syndrome
DSDS	Down Syndrome Dementia Scale
DSM-IV	Diagnostic and Statistical Manual of Mental Disorders—Fourth Edition
DSQIID	Dementia Screening Questionnaire for Individuals with Intellectual Disabilities
ICD-10	International Classification of Diseases and Related Health Problems—Tenth Revision
ID	Intellectual Disability
IQ	Intelligence Quotient
MCI	Mild Cognitive Impairment
MMSE	Mini-Mental State Examination
NTG-EDSD	National Task Group-Early Detection Screen for Dementia
RADD	Rapid Assessment for Developmental Disabilities
SIB	Severe Impairment Battery
TSI	Test for Severe Impairment

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Chapter 1
Overview of the Diagnostic Instruments for Dementia in People with Intellectual Disability

Maria Lusia Margallo-Lana, Stephen P. Tyrer, and Peter B. Moore

Introduction

According to the ICD-10 diagnostic guidelines [1], dementia is a disease of the brain, usually of a chronic or progressive nature, in which there is disturbance of multiple higher cortical functions, including memory, thinking, orientation, comprehension, calculation, learning capacity, language and judgment. Consciousness is not clouded. Impairments of cognitive function in dementia are commonly accompanied, and occasionally preceded, by deterioration in emotional control, social behaviour, or motivation. Dementia is a clinical diagnosis that requires evidence of cognitive decline sufficient to impair function in daily life over a period of at least 6 months [2].

When diagnosing dementia in adults with intellectual disability (ID), the focus should be to recognise decline in relation to premorbid level of functioning. In those whose cognitive function is already affected by ID, assessment of change is difficult, particularly if there is no measure of earlier functioning. There are no agreed reliable diagnostic instruments to detect dementia in people with ID but there are valid and reliable tools that aid diagnosis, which are either administered to informants or, less commonly, rely on direct assessment of the subject.

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When diagnosing dementia in adults with ID, the most important fact to understand is that the diagnosis requires a change in status from baseline functioning. Baseline functioning is often unknown as this information is normally obtained by reports from carers who often have not known the person with ID for long enough to give an accurate picture of premorbid level of functioning. Ideally, longitudinal assessments that document both baseline and present cognitive functioning as well as behavioural functioning over a period of at least 6 months is necessary before sufficient information can be obtained to make a confident diagnosis of dementia [3]. Another difficulty in diagnosing dementia in this population is that there will be varying baseline profiles of abilities and disabilities and varying sensory impairment [4, 5]. It has been suggested that behavioural problems are early signs of dementia in people with DS. People with ID have a wide range of behavioural problems but in the absence of changes in level of functioning and lack of cognitive decline it is difficult to say if they are part of a dementing illness or not [4].

The perception of cognitive decline in this population will also depend upon the premorbid level of intellectual functioning [4]. In individuals with mild ID, the symptoms and signs of dementia can be very similar to that seen in the general population, whereas in individuals with more severe ID, dementia may present with neurological symptoms and maybe associated with diagnostic uncertainty. In order to be meaningful, changes in performance on cognitive testing must be accompanied by changes in everyday independent functioning. To be indicative of dementia, any changes over time must also be greater than those related to normal ageing in adults with ID [4, 6].

Three types of tests are used to help with the diagnosis of dementia in people within this group: (1) those that rely on direct assessment of the individual, (2) those administered to informants and (3) those that combine direct cognitive tests and informant reports in the form of a test battery. Among these, some are tests used in the general population with or without modification for the use in the ID population and some are developed specifically to be used exclusively within this population. Within these separate formats instruments vary according to whether they aim to give a global assessment of cognitive performance or to assess cognitive functions known to deteriorate earlier in dementia, such as recent memory, attention or executive function. In the same way some questionnaires that assess global independent level of functioning are used as benchmarks for future comparison as opposed to those that assess abilities that may decline early in the disease.

Informant-Based Tests

Informant-based reporting has been shown to be effective in facilitating dementia diagnosis [7]. In studies comparing informant reports to direct cognitive tests, informant report have been shown to be more effective than cognitive assessments [8–10]. The informant-based tests that have been most widely-used include the Dementia Questionnaire for Persons with Learning Disability (DLD) [11, 12], the Dementia Screening Questionnaire for Individuals with Intellectual Disabilities (DSQIID) [13], the Dementia Scale for Down Syndrome (DSDS) [5], the Early Signs of Dementia Checklist (ESDC) [14] and the The Short Informant Questionnaire on Cognitive Decline in the Elderly (IQCODE) [15].

Dementia Questionnaire for Persons with Learning Disabilities (DLD) [12]

The DLD (Chap. 3) (originally termed the DMR) was developed by Evenhuis and colleagues [16] as an aid to the diagnosis of dementia in people with ID. The publication in 1992 was an English translation of the original test published in Dutch in 1990 [12]. The 50 items are grouped in eight sub-scales divided in two subcategories: (1) cognitive scores: short-term memory; long term-memory; spatial and temporal orientation; and (2) social scores: speech; practical skills; mood; activity and interest; behaviour and disturbance. A family member or staff who knows the patient well scores his or her behaviour over the previous 2 months according to a three response categories: 0 points = no deficit, 1 point = moderate deficit, 2 points = severe deficit. The questionnaire does not require previous training but includes simple instructions. It takes 15–20 min to complete.

The DLD has been evaluated in a number of studies [17–19]. Inter-rater reliability, internal consistency of items, relationship between intellectual level and scores, influence of some physical handicaps on the scores, relationship between diagnosis of dementia and scores, and the relationship between the diagnosis of dementia and scores was investigated in two cross sectional studies among older residents of three Dutch institutions [17, 20].

This test was specifically designed for use with people with ID and when used together with standardised tools, it has been useful in providing important information for clinicians assessing dementia in people with ID. The test, however, has some drawbacks. The instrument is less sensitive for assessing individuals with dementia in the severe and profound ranges of ID who may never have been able to perform many of the skills assessed in the questionnaire [16]. Thompson [21] and Evenhuis [17] have also pointed out that it is difficult to discern sensitivity of the DLD when used with depressed individuals and they recommend the use of additional tools. Elliott-King and colleagues [7] state that it is necessary to use a scale that rates behaviour when using the DLD as the DLD itself does not cover the full range of factors affected by the dementing process. Although the author reported results for single cross-sectional scores, Evenhuis [19] recommended that score changes over time should be the most valid criterion, as single assessment cut-off scores could be inaccurate [22, 23]. Cut-off scores for dementia should be used cautiously and in conjunction with information gathered from other neuropsychological instruments [10, 19].

Dementia Screening Questionnaire for Individuals with Intellectual Disabilities (DSQIID) [13]

The DSQIID [13] (Chap. 11) is an observer-rated questionnaire, which is completed by carers of people with Down syndrome (DS) who have known the individual for some time. It has three parts. The first asks about the ‘best’ ability the person has or has had. The second contains 43 questions about behaviour or symptoms that are usually associated with dementia in adults with DS. Each item is scored on a four-point scale: ‘always has been the case’; ‘always, but worse’; ‘new symptoms’; and ‘does not

apply'. Items with a response of 'always been the case' or 'does not apply' are scored 0, those with 'always but worse' or 'new symptom' are scored 1. Part 3 of the DSQIID contains ten questions, all of which are comparative; for example, 'speaks (signs) less' and 'seems generally more tired'. A response of 'yes' is scored 1 and a response of 'no' is scored 0. Scores from parts 2 and 3 are added to provide a total score.

The 53 items of the (DSQIID) [13] cover areas such as loss of memory, confusion, loss of skills, social withdrawal, behavioural changes, psychological symptoms, physical symptoms, sleep disturbance and speech abnormalities. The questionnaire has three parts related to the person's 'best' ability, behaviour or symptoms and comparative questions (such as 'speaks (signs) less' and 'seems generally more tired'). Scores from part two and three are added to provide a total score. The questionnaire takes 10–15 min to administer and has a fixed cut off of 20. It has excellent internal consistency (average $\alpha = 0.91$), inter-rater (ICC = 0.90) and test-retest reliability (ICC = 0.95). However, its fixed cut-off may compromise its usefulness in people with more severe ID or advanced dementia [24].

Deb and colleagues [13] indicate that to avoid the floor effect, the DSQIID has a scoring system by which only recent changes in behaviour are scored rather than all behaviours, which allows the use of the DSQIID in a cross-sectional context. However, it is probably best used at regular intervals over a period of time to identify the change in score. The DSQIID has been administered to over 800 patients in numerous studies and all found it to be informative [7].

Dementia Scale for Down Syndrome (DSDS)

The DSDS scale (Chap. 4) is an informant scored questionnaire that was designed to detect cognitive decline, especially at the lower range of functioning The DSDS items were developed for adults with DS mostly with severe or profound ID. Informants are asked to rate subjects on up to 60 items, 20 of which may indicate early stages of dementia, 20 middle stages, and 20 late stages of dementia. In addition, informants are asked to report whether behaviours are typical of the individual during earlier adulthood, whether behaviours are present or absent, and whether or not the date of onset for the behaviours is known [25]. The scale also includes questions that allow the differentiation between dementia, depression, hearing and vision loss, problems with pain, medication- induced cognitive decline and hypothyroidism [5]. Although the manual requires a chartered psychologist to gather information on changes in behaviour from two informants, it has been used in clinical practice and in screening by other mental health professionals [10, 26].

The DSDS appears to be a good screening tool for dementia in people with ID. The DSDS has a specificity of 89% and a sensitivity of 85% in identifying dementia in people with ID. There is a good correlation between this schedule and the DLD [10]. This scale only takes into account new behaviours which have appeared recently and have lasted for at least 6 months. By using this criteria, the impairments in cognitive and daily living skills which have pre-existed the index

illness can be excluded to avoid a floor effect. It appears that repeated assessments using the DSDS can improve accuracy of diagnosis when compared to a single assessment when the dementia process has progressed further. A high correlation between the diagnosis of DAD and the DSDS findings has been reported in subjects in the middle or late stages of dementia. Disparity in diagnosis has been found between DSDS and the clinical diagnosis when subjects with mild or moderate ID have presented with symptoms of early stage dementia.

Early Signs of Dementia Checklist (ESDC)

The ESDC [14] is a list of 37 questions with binary scores. It is a checklist that scores clinical signs of mental deterioration and was found to have very good internal consistency and inter-rater reliability [14]. There appears to be a more comprehensive version consisting of 64 questions, which was used by Hoekman and Maaskant [27] in a current validity and sensitivity study. They found poor agreement with other instruments, but reasonable sensitivity and specificity when compared with expert opinion. Strydom and Hassiotis [22] pointed out the methodological problems of the study such as using a consensus diagnosis of dementia rather than a clinical assessment of mental state and cognition, a small number of participants with dementia and the exclusion of those with severe dementia.

The Short Informant Questionnaire on Cognitive Decline in the Elderly (IQCODE)

The Short Informant Questionnaire on Cognitive Decline in the Elderly (IQCODE) [15] has been evaluated by Schultz and colleagues [28] for use in people with ID. In this population, mediocre test re-test reliability and poor correlation with current mental status has been reported [29]. This is usually measured by informant report and a number of instruments are available. However, none have been adapted for people with ID to screen for dementia.

Subject-Based Assessments

The opinions and views of informants and family members sometimes affect the ratings that are used in informant-based schedules. The advantage of testing subjects directly is that assessments are more accurate and not influenced by others' memories and perceptions. For an accurate assessment the subject being tested needs to be in a cooperative mood and able to engage with the assessor.

The Prudhoe Cognitive Function Test (PCFT)

The PCFT was designed by Kay and colleagues in 1985 [9], but was not published until 2003 [30]. It was originally designed for the direct assessment of cognitive abilities in people with DS but it can be of value in assessing cognitive abilities in ID generally. The original aim was to use the test serially to detect cognitive decline over time. Administration of the PCFT takes no more than 45 min but there are two shorter versions that only take 10 min to administer and have extremely good correlations (more than 0.97) with the longer version, so the shorter versions are preferred [31]. The instrument covers the major domains of cognitive functioning including orientation, recall, language, praxis and calculation.

The PCFT is a highly reliable instrument with excellent temporal stability [30]. Inter-rater and test-retest reliability are both excellent (ICC = 0.99 for both) [30], but there is a floor effect in the more intellectually disabled. It is recommended that a behavioural assessment, using the Adaptive Behaviour Scale or similar instrument, should be used in conjunction with this test, particularly in those subjects with severe and profound impairment [7]. The PCFT has been validated against The Kaufman Brief Intelligence Test (K-BIT) [32], an instrument that has been utilised widely to assess cognitive abilities in people with intellectual disabilities. Comparison of performance on the long PCFT in 167 subjects with equal representation of mild, moderate and severe ID showed high correlations between the verbal and performance sections of the Kaufman Brief Intelligence Scale (K-BIT) with correlation coefficients of 0.85 and 0.78 respectively [31]. The PCFT has face validity in terms of the acceptability of test items to both user and subjects. It also has content validity as the test questions are representative of the skills in the specified domains of the test.

The Test for Severe Impairment (TSI)

The Test for Severe Impairment (TSI) [33] (Chap. 8) was originally designed to assess people with severe dementia in the general population. However, it has particular value for those with more severe levels of ID. The test takes approximately 10 min and has no significant ceiling or floor effect [34]. It includes assessment of motor performance, language production and comprehension, memory, conceptualisation and general knowledge. Only 8 out of the 24 items available require the person to answer a question verbally. The test has good internal consistency ($\alpha = 0.89$) and excellent inter-rater and test-retest reliability (rho = 0.97 and rho = 0.98 respectively). Inter-rater and test-retest reliability are also good among those with severe ID (rho = 0.81 and rho = 0.84 respectively) [35].

Severe Impairment Battery (SIB)

The Severe Impairment Battery (SIB) [36] (Chap. 13) was developed to assess a range of cognitive functioning in patients in the general population with severe dementia who are unable to complete standard neuropsychological tests. There have been three versions of the test and a recently published short version. The test has been translated and validated in Korean, Italian, Spanish and French populations [37–39]. It relies on direct assessment of the individual and takes into account the specific behavioural and cognitive deficits associated with severe dementia, allowing for non-verbal or partially correct response such as matching. It is brief, taking approximately 30 min to administer (15 min for the short version). It is composed of simple one-step commands which are presented in conjunction with gestured commands. The SIB is divided into six subscales: attention, orientation, language, memory, visuospatial ability and construction. There are also brief evaluations of praxis and orientation to name.

The psychometric properties of this scale have been assessed in people with DS with and without dementia [40, 41]. Witts and Elders [42] concluded that the SIB has good test re-test reliability and criterion validity and that in general terms it is suitable for the neuropsychological cognitive assessment of adults with DS. McKenzie and colleagues [40] in their study concluded that the orientation domain of the SIB may be a discriminant subtest as an early indicator of cognitive decline related to Alzheimer's dementia in people with DS.

Although the SIB was not specifically designed for people with ID, preliminary studies in this population appear to show that it is a potentially useful instrument to assess cognitive decline in conditions such as DS. Like many other neuropsychometric assessments however, it is necessary that the participant retains some degree of cognitive abilities to complete its performance. The encouraging performance of the SIB in Witts and Elders study [42] was probably due to the relatively high functioning sample. The test is unlikely to be useful for people with profound ID or severe dementia.

The Adaptive Behaviour Dementia Questionnaire (ABDQ)

The ABDQ was developed by Prasher and colleagues in 2004 [43] (Chap. 10) as a screening questionnaire for Dementia in Alzheimer's Disease (DAD) in adults with DS. It was based on the analysis of 5-year consecutive data on changes on part I of the Adapive Behaviour Scale [44] as part of an ongoing yearly thorough assessment of adults with DS. For the development of the ABDQ, 150 adults with DS (mean age 44.0 ± 1.46, range 16–76) were assessed on baseline by review of previously reported intelligence tests, previous level of functioning as determined by review of medical notes, from carer interview and from the mental state examination; severity of the ID was classified using ICD-10 criteria [1]. All persons were followed up on an annual

basis as part of ongoing clinical care with detailed reassessments of their physical and mental health, adaptive behaviour and social needs. Findings for the absence or presence of DAD were compared to changes in the ABS measurements over the 5 years follow up to determine which items of the ABS best correlated with deterioration in intellectual functioning and could be subsequently used to develop a screening questionnaire.

The ABDQ is a brief questionnaire with good validity and inter-rater reliability that screens for DAD not just dementia *per se*. The ABDQ has been developed from over 10 years of research investigating changes in adaptive behaviour in adults with DS. It can be used for all adults with ID irrespectively of ID and severity of DAD. Once the baseline level of independent functioning of a particular person has been established with the full ABS, the ABDQ appears to be a useful instrument for the ongoing assessment of people with DS. The instrument may prove to be of value as a tool to assess treatment response in drug trials (ongoing evaluation being carried out at the present) and to monitor changes over time without having to repeat the full ABS which is rather lengthy and time consuming.

However given the heterogeneity on presentation on the early stages of dementia and the relative sparse information about behavioural changes associated with the early stages of DAD in this population it is difficult to assume that a individual patient will fit on any of the behavioural categories included in the ABDQ. Individual clinicians may need to rely on changes identified in the full ABS questionnaire to identify decline and monitor illness progression rather than the ABDQ. In any case, although part of the presentation of dementia, changes on behaviour on its own cannot be used to screen for dementia (whether in people with ID on in the general population). Diagnosis of dementia requires a fuller clinical assessment together with a mixture of direct assessments of cognitive abilities, informant based history and exclusion of other causes of behavioural changes.

Cambridge Cognitive Examination (CAMCOG)

The CAMCOG is part of the Cambridge Examination for Mental Disorders in the Elderly-Revised (CAMDEX-R) [45, 46] (Chap. 7). The CAMDEX is a diagnostic assessment that provides a means to identify and differentiate it from other common disorders and the normal process of ageing developed for the general population [45]. The CAMDEX-R is the revised and up-dated version of the CAMDEX [46] enabling a clinical diagnosis of dementia to be made on the basis of international agreed criteria (e.g. DSM-IV, ICD-10). The CAMCOG is a concise group of neuropsychological tests covering all areas of cognitive function that characteristically decline with the onset of dementia. The MMSE [47] is also contained in the CAMCOG and can be used to obtain a global estimate of ability.

Data are collected within the CAMDEX-R through structured clinical interview of an informant supplying systematic information about the presenting disorder, past and family history, present state and history. The CAMCOG is administered by a qualified clinician such as a psychiatrist, psychologist, geriatrician, epidemiologist or other mental health professional working within psychiatry for the elderly.

The evaluator works directly with the person being assessed using verbal and visual stimulus items. These items relate to subscales for orientation, language, memory, praxis, attention/calculation, abstract thinking and perception thus giving subscale scores and a total score.

The CAMDEX-R and CAMCOG have been used with some modifications for adults with DS [48–51]. These authors concluded that the modified CAMCOG was useful to assess areas of cognitive function known to decline with dementia in persons with DS. However, person with pre-existing severe ID, severe sensory impairments and/or already advanced dementia may not be able to score above the "floor level" of the test [49].

Although the modified CAMCOG has shown promising results in high functioning people with DS, giving its length and level of complexity, it is unlikely to be able to be used successfully in the majority of people with DS. The CAMDEX and CAMCOG have not yet been validated to permit a clinical differentiation of the various types of dementia [48]. Additionally, studies examining the early detection of dementia in persons in the general population have suggested that CAMCOG scores are affected by age, hearing and visual defects (e.g. decreased visual acuity and contrast sensitivity due to cataracts) [52, 53].

The Dyspraxia Scale

The Dyspraxia scale was developed by Dalton and Fedor in 1998 [54] (Chap. 5). Dyspraxia is a partial loss of ability to perform purposeful or skilled motor actions in the absence of paralysis, sensory loss, abnormal posture or tone, abnormal involuntary movements, lack of coordination, poor comprehension or inattention [55]. The Dyspraxia scale is an instrument that provides a tool for the evaluation of simple sequences of movements without requiring a normal level of verbal comprehension or communication skills in persons with learning disability [56, 57]. It is not a test of cognitive abilities *per se*, in fact it does not attempt to assess language or comprehension skills but it is assumed that praxis could be expected to deteriorate with the onset and progress of dementia in people with mild to profound ID. The test has 62 items and directly assesses the ability of a person to perform short sequences of voluntary movements such as walking, clapping, etc. The authors reported good test re-test reliability ($r = 0.96$) item by item reliability ($\alpha = 0.97$) predictive and face validity but noticed that the validity has not been established against neuropathological diagnoses [54]. It has been used in research with community based populations of people with ID [58].

The Short Informant Questionnaire on Cognitive Decline in the Elderly (IQCODE)

The Short Informant Questionnaire on Cognitive Decline in the Elderly (IQCODE) [15] has been evaluated by Schultz and colleagues [28] for use in people with ID. In this population, they found mediocre test re-test reliability and poor correlation with

current mental status [29]. This is usually measured by informant report and a number of instruments are available. However, none have been adapted for people with ID to screen for dementia.

Test Batteries

Ten test batteries have been employed in the assessment of dementia in ID, half of which were employed in people with DS. Eight of these batteries assessed both direct and informant reports whereas two focused only on cognitive abilities. Of the eight Test Batteries that examined comprehensive abilities four also assessed behavioural as well as cognitive measures.

Neuropsychological Assessment of Dementia in Adults with ID

This test was designed by Crayton and colleagues [59] in 1997 and included a wide variety of picture identification scales testing for aphasia, agnosia and receptive language. Visual memory, object, pattern and spatial recognition was also tested in addition to conditioned associative learning tests and executive function abilities. The Cognitive Test battery was compared to the Vineland Adaptive Behavior Scale (VABS) [60] and all neuropsychological tests correlated significantly negatively with the results on the VABS. The memory tests showed significant impairment in DS subjects aged over 50 years and it has been suggested that these tests may be the most sensitive in monitoring the dementing process.

Oliver and colleagues [61], applied a similar test battery but employed the Cambridge Neuropsychological Automated Test Battery (CANTAB) and the Cambridge Assessment for Mental Disorders in the Elderly (CAMDEX) to examine learning and memory, aphasia and agnosia. Memory was tested in more detail as well as apraxia. The results showed that the deterioration in memory, learning and orientation preceded aphasia, agnosia and apraxia.

Working Groups Battery

Burt and Aylward [62] used the DMR (DLD) and the DSDS as informant tests together with three behaviour scales, the Reiss Screen for maladaptive behavior, the Scales for Independent Behavior-revised and the AAMR Adaptive Behavior Scale. The tests assessed memory and recall, spatial recognition, autobiographical memory, orientation, language and perceptual motor skills. In the first study the reliability of the assessments were not reported but since then Pyo and colleagues [63] have shown good reliability on the autobiographical memory and orientation tests.

Other test batteries have been produced by Silverman and colleagues [64], using an informant-based interview of medical history coupled with the DMR (DLD) in a large population of 273 subjects. Receptive vocabulary, verbal fluency, construction abilities visuo-spatial and episodic memory were also assessed. Behaviour was assessed by the ABS and the Reiss Screen. The test takes 2 h to administer but was able to accurately distinguish dementia sufferers from the remainder. Das and colleagues [65] showed that older subjects with DS performed poorly on a battery of tests involving memory, speech, attention and planning. Jozsvai and colleagues [66] and Johansson and colleagues [67] have also employed a number of tests in small groups of subjects, insufficient to draw firm conclusions.

Because of difficulties regarding cognitive assessment in people with ID, alternative methods of diagnosing and monitoring the progression of dementia in this population have been proposed [35]. These include assessing changes in emotional functioning [68] and adaptive behaviour [69]. Caregiver assessment of patients' overall level of functioning can also be measured by using such instruments as the DSDS [5], The Early Signs of Dementia Checklist (ESDC) [14], and the DMR (DLD) questionnaire [17]. The standardised administration of a mental status instrument is preferable to a less formal assessment of cognitive ability because it allows confident comparisons of results over time [6].

In their review of instruments for assessing memory problems, Zelinski and Gilewski [70] noted that people who are poorly educated or who have below normal IQ assessments perform poorly on test of mental status and often are likely to be described as cognitively declined when in fact they are not [71]. They have therefore proposed that the evaluation of dementia in people with ID requires use of a carer interview as well as direct assessment. Carers can report on cognitive decline independent of premorbid intelligence.

The DSDS [5] and the DMR (DLD) [12] are the best known carer assessment instruments. These two instruments together with the Modified Cambridge Cognitive Examination for Mental Disorders of the Elderly [49, 50] are recommended tools to assess severity of dementia in people with ID in the Report by National Institute for Health and Clinical Excellence-Social Care Institute for Excellence published in 2006 [72].

Psychological Tools

Commonly Used Instruments Administered to Informants

Caregivers, family members or professionals are important sources of information who can comment on an individual's past performance, abilities and observed changes in everyday functioning. Although informant-based measures should be used with caution within a retrospective assessment approach, they are useful when repeated over time. However, as baseline measurement may not be available when an individual presents with changes that might indicate dementia, this has led to a heavy reliance on informant-based measures.

As well as the above mentioned instruments based on information reported by an informant, other instruments include the ESDC [14] and the Multi-Dimensional Observation Scale for Elderly Subjects (MOSES) adapted for adults with DS [56]. These scales incorporate changes in cognitive as well as daily living skills in people with ID. Of these scales, the ESDC [14] has only been used in institutionalised adults with Down syndrome (DS), whereas the other three scales have been used in community-based adults with DS. The MOSES [56] is designed for longitudinal use only and has no cut-off score for the diagnosis of dementia.

Commonly Used Instruments Administered to ID Persons

A clinical diagnosis of dementia requires evidence of progressive deterioration in a person's cognitive abilities and daily living skills [1]. The most significant problem for the assessment of specific neuropsychological deficits associated with dementia is the variability of intellectual ability and the problems of administering neuropsychological tests to those with severe or profound ID who may not understand verbal commands [3, 73]. Poor performance on neuropsychological tests that might indicate dementia might easily be attributable to ID. This is obviously due to the fact that in the case of a person with ID, the mere presence of cognitive impairment does not equate to a diagnosis of dementia because often the impairments have been present throughout the person's life [10].

Sequential testing has been recommended in order to identify decline due to dementia in individuals with ID from a previous baseline by the administration of standardized neuropsychological tests [3]. Instruments employed for assessing dementia in the general population such as the Mini-Mental State Examination (MMSE) [47] have proved not to be suitable for diagnosing dementia in the ID population [10, 74]. Because the limitations of the MMSE and other short screening instruments in people with ID, researchers have been investigating and developing alternatives.

A recent review has assessed the effectiveness of published instruments in the assessment of dementia in this population [7]. In all, 43 schedules and tests were examined, of which 23 were direct cognitive tests, 10 were classified as informant reports and the remaining 10 were test batteries. The majority of the direct contact or subject-based tests assessed aspects of cognitive performance that were considered to be most vulnerable to a dementing process. The domains included most often involved memory, visual recognition, visuo-spatial assessment, language comprehension and executive function. The most discriminatory of these tests for memory deterioration were found to be by Devenny and colleagues [75] and Schultz and colleagues [71]. Of those tests which examined general cognitive performance the Test for Severe Impairment (TSI) [33] and the Prudhoe Cognitive Function Test [9] were able to provide a reliable quantitative measure of cognitive function in this population. The advantage of the TSI is that this schedule is able to assess the abilities of individuals with severe and profound handicaps whereas those with more

severe impairments score zero on the PCFT. However, when the PCFT is combined with a behavioural measure floor effects are largely eliminated. Furthermore, the PCFT examines all areas of cognitive ability including praxis and calculation.

Elliott-King and colleagues [7] found that tests that have been designed for use in an adult population without evidence of ID, for example the Mini-Mental Status Examination (MMSE) and the Cambridge Cognitive Examination (CAMCOG) were not able to assess those with severe impairments and the MMSE was not accurate in distinguishing cases from controls [10].

Informant reports were developed before instruments that directly assessed function. The most widely used informant test is that the Dementia Questionnaire for Mentally Retarded people (DMR), recently renamed the Dementia Questionnaire for Learning Disability (DLD) [11]. This instrument has been shown to be valuable in distinguishing those with dementia from those without [10, 71] and McCarron and colleagues [8] have shown that this test is sensitive to change in serial testing. However, as noted with many of the direct assessments an additional behavioral assessment is required to provide a comprehensive picture. The Dementia Screening Questionnaire for Individuals with Intellectual Disabilities (DSQIID) has been shown to be both reliable and valid [13, 76]. A specific test assessing behaviour, the Adaptive Behaviour Dementia Questionnaire [43] was also found to be both valid and reliable.

Elliott-King and colleagues [7] also examined what are described as test batteries. Eight of these batteries assessed both direct and informant reports whereas two focused only on cognitive abilities. Of the eight Test Batteries that examined comprehensive abilities four also assessed behavioural as well as cognitive measures [42, 59, 61, 62]. Both of the former two batteries were tested in people with Down syndrome (DS) in relatively small populations.

Although this review recommends that the most practical and efficient method of diagnosing dementia in individuals with ID is by using test batteries this opinion is not based directly on evidence. We consider that an accurate assessment of dementia in this population requires assessment of cognitive abilities assessed by both direct and informant methods as well as by a behavioural evaluation. This may be assessed by a test battery but separate assessments of these three domains may be sufficient in themselves.

Summary

When diagnosing dementia in adults with ID, the most important consideration is that the diagnosis requires a change in status from baseline functioning, not a change from ‘normal’ level. Longitudinal assessments that document baseline cognitive functioning, in addition to a change in independent functioning, is necessary before sufficient information can be obtained to make a diagnosis of dementia [3]. However, just as in the general population, it is not sufficient to identify decline in cognitive and functional skills as evidence that a person suffers from dementia as other causes

of apparent decline must be excluded before a confident diagnosis can be made. The perception of decline will also depend on the environmental demands on the individual.

As in the general population, the results of any test are meaningless if considered in isolation without its clinical context. In people with severe and profound ID, who usually fall outside the lower range of scores of most available instruments, assessment of cognitive and behavioural skills may not be possible and one may have to rely on other aspects of the history and presentation such as development of neurological symptoms such as epilepsy or dysphagia.

Perhaps we should move away from trying to develop a screening instrument that by its single use will tell us if a person is suffering from dementia and accept that we need to use an array of assessment to fully understand the nature of the process that is affecting that individual. An early diagnosis of whether a person is suffering from dementia will aid not only the individual but the carers and others to understand and adapt to the changes that inevitably accompany the illness.

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Chapter 2
Issues in Dementia Assessment Methods

Diana B. Burt

Introduction

Dementia assessment in adults with intellectual disabilities (ID) is a challenging task, but past work by clinicians and researchers has improved diagnostic accuracy. Diagnostic criteria were outlined [1] and found to be feasible and useful [2–4]. Gradations from mild to major neurocognitive disorders[1] were identified and found to affect dementia prevalence figures [5–7]. A battery of tests was proposed to identify significant declines [8, 9]. Ongoing investigations examined the sensitivity and specificity of tests from the proposed battery and additional alternative batteries [2, 3, 6, 10–25]. The purpose of this chapter is to outline and discuss general issues and factors that can affect dementia assessment either directly or indirectly [3, 11, 26]. Such issues are important to consider when evaluating tests for clinical and research purposes. As indicated in Table 2.1, a discussion of general theoretical issues will be followed by a more specific discussion of methodological issues.

[1] The term mild neurocognitive disorder will be used in this chapter to be consistent with DSM V terminology and to avoid selection of one of many terms used in research literature (e.g., mild cognitive impairment (MCI), possible dementia, probable dementia). The term dementia will be used instead of major neurocognitive disorder as it has been consistently used in the literature.

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Table 2.1 Issues in dementia assessment

Worldview/prevalence implications	Adults with Down syndrome and Alzheimer's disease, range of prevalence figures
Schedule for assessment	Single versus repeated evaluations
Purpose of assessment	Diagnosis, declines identified, dementia screening, differential diagnosis, information gathering/informant awareness
Characteristics of individuals being assessed	Intellectual level, age, etiology of ID, gender, care setting
Methods to address individual differences	Homogeneous versus stratified samples
Source of information	Informant report versus direct assessment of performance
Evaluation of assessment scales and techniques	Independent/external criterion, measures (sensitivity, specificity, predictive value), reliability, group comparisons, stages of dementia, strength/weakness profiles, evaluation across studies, clinical usefulness

Worldview/Prevalence Implications: Adults with Down Syndrome

Historically, the assessment of dementia in adults with Down syndrome (DS) was approached from two worldviews [27]. The first assumed that all adults with DS got dementia in Alzheimer's disease (DAD). Changes in functioning starting around age 30 years were assumed to be early dementia [28, 29]. The second world view, in contrast, assumed that only some adults with DS got dementia. Declines in functioning were not always indicative of dementia, particularly DAD [27, 30]. As in the general population, it was assumed that declines could be due to multiple infarcts [31–33], conditions like Parkinson's disease [34], adverse drug effects [35], or other psychiatric disorders (e.g., depression) [36–38].

Given research and prevalence figures available online, it is realistic to assume that few still adhere to the view that all adults with DS, even those in their 30s and 40s get DAD. However, differences in currently available prevalence figures could still lead to differences in the worldviews of clinicians and researchers, especially if increases in prevalence with age are not emphasized. For example, the following prevalence figures for dementia in adults with DS are listed on popular web sites (US Alzheimer's organization—75% of adults over 65, Web.MD—25% or more of adults over 35 (with unspecified increases in prevalence with age), Wikipedia—15% of those over 40 and 50–70% over 60). Research indicates figures such as 26% at age 50, up to 95% at age 68, and 70% if alive by age 70 [6, 39], with prevalence impacted by differences in categorizing adults [7].

Although clinicians/researchers do not always explicitly state their adopted worldview, it has an effect on the evaluation and use of assessment scales. According to the worldview assuming high prevalence figures, the purpose of dementia assessment is to detect declines related to dementia and to illustrate the natural history of dementia. If declines are not eventually detected on a given scale, the scale is assumed not to be sensitive enough. If dementia is presumed by a clinician lacking

expertise in ID, inappropriate scales designed for the general population could be used to verify expectations (e.g., the Folstein Mini-Mental State Examination) [40]. In contrast, advocates of the second worldview use assessment scales to differentiate clinically significant declines from those associated with typical aging. They also attempt to differentiate declines associated with irreversible dementia from those associated with other treatable conditions (e.g., depression) or environmental factors [41–44]. Standardized diagnostic criteria are designed to maximize diagnostic accuracy and to minimize the number of adults erroneously diagnosed with DAD [1, 6, 23, 28, 43]. In addition, given relatively recent emphasis on mild neurocognitive disorder in adults with ID [5, 7], consensus is needed on criteria for its diagnosis. Such consensus will allow comparisons of prevalence across sites and will hopefully further discussions on whether new methods are needed to document mild declines in cognition/memory and to determine the severity of changes in everyday functioning. As indicated in the Appendices, several scales already involve an indication of the relative severity of decline (e.g., ABDQ and CAMDEX-DS).

Other theoretical assumptions related to one's adopted worldview also influence assessment. If, for example, one assumes that all adults with DS get DAD and show the same sequence of decline (e.g., memory decline followed by motor decline [12], dyspraxia followed by other cognitive decline [45]), then tests for memory decline or dyspraxia could be adopted to screen for early signs of dementia. Any adult who showed early signs of dementia in another area (e.g., changes in emotional functioning [2, 7, 8, 46]) would not be identified by a narrow screening battery assessing only memory or dyspraxia. Similarly, if one assumes that all adults with DS or with other forms of ID get only a progressive dementia like that caused by DAD [1], then an adult who shows signs of a static dementia (e.g., related to adverse effects of medication) or mild neurocognitive disorder (without further declines) may not be identified. Whether or not all adults with DS or with other forms of ID show the same invariant sequence of declines is currently being investigated. Unfortunately, progress is slow, due to lack of collaboration across sites and the time it takes to collect longitudinal data [5]. In the meantime, effects of worldview on assessment in individual cases and in general must be considered to minimize diagnostic error in research and in practice.

Schedule for Assessment

When considering dementia assessment scales, it is necessary to examine the intended schedule and purpose of the scale. Regarding scheduling, a scale or test battery can be developed for a single administration, with performance at that one assessment presumed to be indicative of dementia status. An example of such a scale used in the general population is the Mini-Mental State Examination [40]. If an adult performs below a certain cutoff point on this scale at one assessment, they are assumed to be demented. The use of single-administration scales with adults with ID is complicated by the fact that low performance is likely to be related to

level of ID and not to dementia [3, 11, 19]. Unfortunately, anecdotal reports suggest that the Mini-Mental State is currently being used clinically by general practitioners at one assessment to evaluate adults with ID, despite consensus among experts in ID that it is not appropriate for adults with ID.

Other single-administration scales rely on retrospective reporting, like the Dementia Scale for Down Syndrome (DSDS; Chap. 4) [47]. On this scale, informants are asked to compare current behavior to remembered behavior. Although the scale is also used at repeated assessments, dementia status is based on absolute scores not on change scores indicating differences between current and previous performance. The advantage of such single-administration scales is the practicality of determining dementia status at one assessment. The disadvantage is that performance can be confounded by level of ID and by inaccuracies in retrospective reporting.

Scales can also be developed to allow direct comparisons of performance across repeated assessments. Declines in performance over time are then examined to see if they correspond to clinically significant changes indicative of dementia. Scales and tests in the battery recommended by the dementia work group [8, 9], for example, were intended for repeated assessment, with baseline performance compared to later performance. Scales have also been developed for use both at one assessment and across repeated assessments. The Dementia Questionnaire for People with Intellectual Disabilities (DLD; Chap. 3), for example, had both a scoring system for a single administration and a system for examining change scores that reflected differences in scores over repeated assessments [48]. The advantage of such a dual scoring system is that dementia status can be determined at one assessment, thus alerting the evaluator to the need for a further dementia workup [49]. The scale can also be administered repeatedly to gather further evidence about dementia status and progression. Interestingly, the two scoring systems yielded differences in sensitivity and specificity to dementia [3, 19, 50], and Evenhuis and colleagues recommended use of repeated assessments only [51, 52].

Purpose of Assessment

The advantage of single versus repeated assessments is related to the purpose of a scale or test battery. Clinicians and investigators developed scales for different purposes. A broad battery of tests may be used repeatedly, for example, to determine whether diagnostic criteria for dementia are met (i.e., memory decline, other cognitive decline, changes in emotional functioning, declines in everyday functioning). In such a battery, scales are included on the basis of each scale's ability to detect declines related to a given diagnostic criterion [3, 11]. A sentence recall task, for example, is administered to assess declines in memory, whereas a vocabulary test is included to assess declines in cognition, specifically language. An advantage of a broad battery, in addition to allowing assessment of all areas needed for dementia diagnosis, is that the clinician or researcher can examine combinations of tests and scales to see which subsets of tests lead to the greatest levels of sensitivity and

specificity [3, 11, 19, 23, 53]. In addition, when tests are administered repeatedly, performance can be examined to see if some tests detect earlier signs of dementia and thus would serve as useful screens for dementia at one assessment [3, 11, 54–59]. Performance can also be examined to see if other tests detect later signs of dementia and thus would serve best as repeated measures over time for confirmation of the presence of dementia [3, 11].

In contrast, researchers and clinicians have designed broad scales, (used alone) assessing several areas of functioning, to repeatedly assess all areas needed for dementia diagnosis [2, 48, 51, 52]. Performance on the scale is compared to other tests designed to independently determine dementia status. If one scale can diagnose dementia as accurately as a more extensive battery, then the single scale could be more efficient and cost-effective. The sensitivity and specificity of such a broad scale would need to be determined.

Test batteries have also been used with a narrower focus, for example, to detect declines on tests of memory and other cognitive functioning [54–59]. In such cases, an external criterion for dementia is needed to relate performance on the scale to dementia status to determine the test's usefulness in dementia assessment. Other batteries have been used to examine the prevention of cognitive decline, to narrow outcome measures for treatment studies [53], and to examine treatment effects [60]. Finally, performance on one scale assessing a single skill (e.g., dyspraxia) has been used to examine declines in functioning related to an intervention (e.g., with vitamin E) without necessarily relating the declines to actual dementia status [17, 45]. Such scales would also require an external criterion for dementia to determine relationships between performance and dementia status.

Regarding the purpose of scales and instruments, it is necessary to examine any rationale given for the schedule of assessment (one administration, repeated administrations, both types of administration). If the schedule of assessment is changed from that used in scale validation, then new psychometric studies are needed to examine the scale under the new conditions. Issues arising with repeated but not single assessment and vice versa would need to be considered. Single administration scales involving retrospective informant judgments, for example, do not provide baseline data for later comparisons unless specifically designed to do so (S. Deb, personal communication, January 28, 2017). That is, they often do not provide an indication or record of absolute premorbid or best typical level of functioning (e.g., dresses self independently, including all fasteners). They provide judgments of whether change has occurred relative to a baseline, which could be determined by memories of family members or an undocumented review of records.

It is advantageous to know whether a scale is useful in early detection of dementia, in the confirmation of dementia status, or perhaps both. A dementia screening instrument for early detection would ideally be less time-consuming and less expensive, so that it could be administered repeatedly without using vast amounts of scarce recourses. Dementia screening scales also need to provide information in a format easily integrated into an adult's permanent record, because screening or baseline data are only useful if they can be located easily and compared to later performance. Such dementia screening instruments could be administered when the

adult is known to be functioning optimally (e.g., young adulthood), with repeated administrations designed to determine a pattern of functioning for the individual [3, 11]. The screening instrument would then be re-administered periodically or sooner if dementia is suspected.

A test or battery of tests designed to allow confirmation that diagnostic criteria are met for either mild cognitive impairment or dementia will by definition need to be more comprehensive. To be useful, such a battery would also need to be administered at least once when an adult is known to be healthy to allow for later comparisons to baseline functioning [1, 5, 41–44]. Such a battery or broad test will also need to contain aspects that allow for differential diagnosis of dementia from other psychiatric disorders such as depression, medical conditions such as thyroid disease, adverse drug effects, or negative environmental factors [10, 13, 26, 37, 38, 41–44, 61–71]. It may not be feasible to administer such a broad battery or test on a regular schedule, because of scarce resources.

Characteristics of Individuals Being Assessed

Characteristics of individuals such as level of functioning (usually intellectual level given as intelligence quotient (IQ) or mild, moderate, severe), age, cause of ID, gender, and care setting have all been shown to influence test performance [3, 5, 7, 11, 19, 23, 72–74]. For healthy adults, such individual differences influence performance at one assessment and also affect the amount of change over time that is typical. When examining assessment instruments, therefore, one should determine what considerations were made for individual characteristics (e.g., cutoff scores for dementia calculated by level of functioning, change scores indicative of significant decline adjusted for age [33]). It is also necessary to determine the characteristics of the standardization sample to see for whom a test or scale is designed.

Intellectual Level

General reasoning as indicated by IQ was related to performance on almost all tests such that higher IQ was related to higher performance [3, 11, 19, 72, 74]. Level of functioning was also related to change in performance over time. Adults at lower levels of functioning showed improvements in performance with repeated practice, whereas higher functioning adults started at a higher level and remained at the same level [27]. Thus, amount and type of change in performance over time can be related to initial level of functioning, which would need to be considered in differentiating typical performance from that associated with mild neurocognitive disorder or dementia. Dementia cutoff scores based on informant reports, such as those on the original single administration of the DLD [48], also require adjustment for premorbid level of functioning (i.e., when healthy). The challenges in making such

adjustments include: methods for assessing level of functioning change over an adult's life span (e.g., as intelligence tests are revised), different tests are used for a given adult across time often yielding vastly different results (e.g., Weschler versus Stanford–Binet tests), and methods are not standard across countries [19].

Age

If a skill is influenced by aging, then one would expect the amount of decline over time that is typical for healthy, older adults to be different from that of healthy, younger adults. Different criteria could be needed for adults of different ages, therefore, to indicate the amount of decline that is clinically significant (i.e., greater than that typically associated with aging at that point in the life span). With age-related changes in sensory capabilities, speed of cognition and response, and perhaps motivation it is also possible that what a task measures for younger adults is different from what it measures for older adults. If test stimuli are very small or require fine hearing discrimination, the performance of older adults could be affected by sensory impairments that prevent them from seeing or hearing the stimuli [75–80]. What the scale is actually measuring at repeated assessments over a life span could change, for example, from a test of memory or reasoning to one of vision or hearing. Many older adults with ID in the current generation were not expected to wear glasses to improve vision or hearing aids to improve hearing. They often refuse to wear such aids. Thus, it is important to examine whether a test consistently measures the same thing across persons with differing ages and abilities. Factors to consider are task demands and changes in functioning that could affect the ability to meet them (e.g., fine motor skills, slowing with age, etc.). Such issues are not restricted to direct assessments for dementia. Informants asked to report on dressing skills, for example, may not mention that adults no longer dress themselves because arthritis prevents the use of their hands for buttoning, zipping, pulling, etc. An informant reporting on memory skills may not know that the adult no longer remembers events seen on television, because they can no longer see or hear well enough to do so. Thus, it is also important to include vision and hearing screening as part of any dementia assessment battery [3, 11, 13, 81].

Etiology of ID

Regarding etiology of ID [23, 74], individual differences in premorbid strengths and weaknesses profiles need to be taken into consideration in dementia assessment. Adults with DS compared to their peers without DS, for example, had a great deal of difficulty placing small, grooved pegs into a pegboard. They did not place enough pegs into the board when young and healthy to establish a high enough baseline for further detection of significant declines. Therefore, a pegboard task involving pegs that were more easily

placed was adopted for dementia assessment, which made it appropriate for adults with and without DS. Tasks that require clear speech (e.g., picture description, category fluency) are often difficult to administer to adults who have severe articulation disorders, because the Examiner cannot understand words clearly enough to know if they should be scored as correct or not. Often times, such articulation disorders are more common for adults with DS. Thus, etiology of ID has implications for task appropriateness, as well as cutoff scores for significant declines. Such differences need to be taken into account both at a single assessment and in identifying the amount of change that is typical over time. Etiology of ID can also interact with other characteristics (e.g., age, gender) so such interactive effects may also need to be considered [82].

Gender

Gender differences have been obtained on a number of tests [3, 11, 23, 73, 82, 83]. It has been suggested that lower performance in older women with ID is related to estrogen status [83]. Once again, it is important to know what is typical for adults with ID with varying characteristics so that performance indicating significant declines can be identified.

Care Setting

Although not considered highly predictive of functional decline, care setting (e.g., nursing home, community housing, home) has recently been evaluated as an individual difference worthy of further investigation [23].

Methods to Address Individual Differences

When deciding who a test is appropriate for, both on a general level and on an individual basis, characteristics such as level of functioning, age, etiology of ID, and gender should be taken into account. Researchers and clinicians have used several methods to take such characteristics into account in scale evaluation.

Homogeneous Groups

One method is to examine the use of a scale in a homogeneous group of adults, for example, all adults with DS over the age of 50 years functioning in the mild range of ID. The use of a homogeneous group eliminates some of the variability in

performance and the need to consider performance differences related to some individual differences (in this case etiology of ID, age, and level of functioning). One could conclude with greater certainty that any change over time in healthy adults is typical for this population or that relatively low performance at one assessment is less typical and thus more likely to be associated with mild neurocognitive disorder or dementia. There is still the possibility that premorbid differences in performance related to other variables are present and they need to be considered (e.g., sensory capabilities). The weakness of this homogeneous group method is that one would not know whether a scale validated on such a narrow population would be valid in other populations, such as adults without DS or adults with severe to profound ID. Any scale appropriate for adults with mild ID would also need to be feasibly administered in the later stages of dementia if the scale was to be administered repeatedly (e.g., to examine the natural history of dementia). At times, an adult performs tasks when healthy, but can no longer perform them when demented (i.e., becomes untestable on the test). In such cases, it can be difficult to differentiate "untestable" status related to dementia from that related to other conditions (e.g., depression). Untestable status can also be due to refusal to respond or to loss of the required response because of some other condition (e.g., speech, pointing response). It is best, therefore, to have a test or scale with a range of performance that can detect declines or changes related to mild neurocognitive disorder or dementia.

Stratified Sample

A second strategy for handling individual differences in performance in dementia assessment is to include a heterogeneous, stratified group of adults (e.g., adults with DS ranging from the mild to profound range of functioning). Examiners evaluate performance differences related to individual characteristics and adjustments to cutoff scores, dementia identification rules, or analyses are made [3, 7, 11, 15, 19, 23, 33]. Depending on the administration schedule for a given test, such adjustments could be needed for dementia cutoff criteria at a single assessment. They could also be needed for detection of clinically significant declines over repeated assessments. Although the stratified sample method seems advantageous, it can be quite cumbersome in practice to examine and adjust for all possible variations related to individual differences. Ideally, performance on a scale for dementia would not be affected by such individual differences [23], but as discussed previously all scales, even informant report scales, often must take such differences into account when considering dementia cutoff criteria. In evaluating dementia scales for adults with ID, therefore, it is important to determine how individual differences are handled. One needs to know whether different criteria are needed for adults with different characteristics or whether the test developer has demonstrated that the same criteria apply for all adults. One should also know whether the scale covers a wide enough range of abilities to be appropriate for most adults with ID, or if it is only appropriate for adults with certain levels of premorbid functioning.

Informant Report or Direct Assessment of Performance

An important dementia assessment issue is whether to collect information from informants, from individuals with ID themselves, or from both [2, 13, 14, 24, 41, 43, 46, 50, 53, 70, 84]. A working group on the diagnosis of dementia recommended both informant report and direct assessment for every evaluation [1, 8, 9]. They recommended informant report of emotional and everyday functioning, because most adults with ID are not able to reliably report on internal states such as emotions. Similarly, they are not able to monitor their own everyday skills to detect changes or to report on the severity of changes. Even adults who are able to report on such states may be unable to do so as dementia progresses. Changes in both emotional and everyday functioning are required for dementia diagnostic criteria to be met [1], and the severity of changes in everyday functioning could be used to distinguish mild neurocognitive disorder from dementia.

The working group recommended direct assessment of adults with ID to document memory and cognitive declines as required by dementia diagnostic criteria [1, 8, 9]. When feasible, direct assessment is usually regarded as preferable to informant report because error related to observation and reporting is not introduced into the assessment. When both informant report and direct assessment are used, consistent information obtained across the two sources is strong support for findings regarding dementia status. Inconsistent information suggests the need for further evaluation or reassessment in the near future.

Informant Report

An important issue in informant reporting is whether the report accurately reflects the functioning of the individual [2, 41, 43, 53, 85]. Bias can be introduced if informants find it emotionally difficult to report declines in functioning or depressive signs. Informants may believe that certain declines are not relevant to the person's care and thus may not take note or report them. If an Examiner asks informants to report on unobservable states (e.g., hopelessness), they are required to make an inference about internal states, which may or may not be accurate. Informant report scales on which informants' reports were compared to actual performance would be ideal. The ability of informants to report on the orientation of adults with ID (e.g., knowledge about their name, their place of residence, time) on the DLD [48], for example, was found to be fair to good [85]. For some orientation items, however, nonverbal IQ, etiology of ID, and age affected level of agreement between informant report and direct performance.

One major obstacle to the use of informant reports of functioning is the availability of consistent, knowledgeable, and reliable informants [43]. Direct care staff often have a high rate of turnover and many adults with ID have older parents who do not live long enough to report on their functioning when they become elderly themselves. Some informant report dementia scales and psychopathology scales require that informants know the individual for 6 months or longer [2, 11, 15, 86].

The informant must also work closely enough with the individual to determine and report on functioning in the last 6 months to a year [2]. Training has been successfully provided to informants to improve their ability to observe and report on behaviors and functioning relevant to dementia diagnosis [2, 26]. Although not necessary for the use of informant reporting scales, such training would be expected to improve the accuracy and sensitivity of dementia diagnosis.

In addition, reliable informant reporting depends on documentation of functioning in the adult's chart and on adequate levels of interrater reliability. A paid carer, consulting records in making judgments could be using a different decision-making process from a family member making the same judgment based on memory. Scales requiring reports on current functioning (i.e., in the 2 weeks prior to assessment) are preferable to those requiring retrospective reporting, both because of changes in care providers and because of inaccuracies in informant memory regarding past functioning. If informants make retrospective judgments, comparing current to previous premorbid or baseline functioning, the basis for such comparisons becomes blurred if the judgments are made repeatedly over time (i.e., comparing performance at first, second, and third follow-up visits to a baseline that occurred at a distant time). It is not clear what information is actually derived from comparing current to former retrospective judgments. If possible, it is ideal for the informant to indicate whether any performance consistent with a dementia diagnosis has always been typical of the individual or not (e.g., adult never knew address of living facility) [2, 3, 11].

The level of professional expertise required to complete, administer, and interpret informant report scales is another issue to consider. Lay people, such as direct care staff or family members, can complete some scales [3, 52, 70, 85, 86]. Highly trained professionals must complete or administer others (e.g., DSDS [47], most adaptive behavior scales). Some scales require two informants for clinical assessment (e.g., Reiss Screen, DSDS [47]), whereas others rely on one informant (i.e., DLD [48]). Regardless of administration procedures, most dementia diagnostic scales are interpreted by highly trained professionals. There are some scales, however, designed specifically to gather information on a regular basis that is then reported to a diagnostician [3]. Such scales are considered information gathering tools in this chapter, as opposed to dementia screens like the DLD. Dementia screens have rules for identification of dementia and psychometric data supporting such rules.

Direct Assessment

Advocates of the sole use of informant report scales often argue that direct tests of individuals with ID for dementia are not feasible or sensitive enough or are not reliable or valid [46, 87]. It has been suggested that adults whose premorbid level of functioning is at or below a mental age of 2 years are often unable to perform neuropsychological tests at a level that would allow detection of declines related to

dementia [3, 11]. In the experience of members of the working group on dementia assessment, however, most adults with ID can be assessed reliably on direct measures of memory and cognition [2, 3, 11, 17, 19, 45, 54, 84, 88, 89]. Direct assessment has been particularly useful when adults present with signs of both possible dementia and a psychiatric disorder [66, 90]. In one instance, for example, informants reported declines in daily functioning and signs of a psychiatric disorder. Direct testing over several years indicated consistent levels of memory and other cognitive functioning, with no apparent declines. Thus, in this case, test performance along with informant data indicated the presence of a potentially treatable psychiatric disorder rather than a progressive, irreversible dementia [90]. As mentioned previously, for any given test there may be individuals who cannot perform its tasks at a clinically useful level (i.e., one that would allow detection of declines), either because of low premorbid functioning or impaired sensory or motor capabilities. The fact that a test does not have universal applicability, however, does not necessarily mean that it is not useful for most adults with ID.

Some care providers report the use of videotaping to directly document changes in functioning related to dementia. Videotaping methods have been used to film assessments for purposes of supervision (i.e., checking on standardized procedures for test administration). Videotape recording and data transcription were also used to evaluate behavioral excesses (i.e., maladaptive behavior) in adults with dementia [81]. This observational method has the potential to document changes related to mild neurocognitive disorder or dementia, and could be particularly useful for lower functioning individuals or for those with sensory impairments that prevent standard assessments. The challenge would be to develop a method to efficiently provide reliable repeated assessments and clinically useful data.

When directly evaluating adults with ID for dementia, it is important to follow best assessment practices [5, 6, 23, 24, 91]. Qualified evaluators should conduct the assessment, particularly those who have experience working with individuals with ID. Untrained Examiners sometimes have biased notions about the abilities of people with ID. They may not expect them to perform tasks they are perfectly capable of completing, thus biasing results. Testing should be conducted in a room free of distractions. When selecting and interpreting the results from specific tests or scales, the characteristics of the individual should be considered (e.g., lack of speech, apparent level of motivation, etc.). Most adults with ID enjoy the one-on-one attention typical of a testing experience and benefit from reinforcement of effort. A further consideration specific for dementia assessment is time of day, given that the course of dementia varies across the day with optimal functioning often in the morning.

A question remains as to whether the sole use of either informant report or direct assessment measures is sufficient to collect information about and/or document declines in functioning, to screen for dementia, or to make diagnoses of mild neurocognitive disorder or dementia. Batteries involving both informant report and direct assessment measures led to higher levels of sensitivity and specificity than informant report alone [3, 11, 19]. Direct assessment was used to assess memory and cognitive functioning, whereas informant report was used to assess emotional and everyday functioning. Source of information was confounded, therefore, with which

diagnostic criteria were being assessed. The relative contribution of informant report versus direct assessment in the diagnosis of mild neurocognitive disorder or dementia in adults with ID, therefore, is an issue that requires further examination. It is possible that their respective values could vary with the characteristics of individuals being assessed [14, 53]. Given that each method has its strengths and weaknesses, it is strongly recommended that both sources of information be used.

Evaluation of Assessment Scales and Techniques

When examining information gathering tools, dementia screens, dementia assessment scales and diagnostic techniques, it is necessary to determine how the scale or technique was evaluated. Evaluation usually involves comparisons of dementia status obtained by using the scale to that determined by an independent source. These comparisons involve an examination of scale sensitivity, specificity, predictive validity, test–retest reliability, and clinical usefulness [2, 3, 11, 15, 20, 23, 24, 54, 56, 74]. Ideally, all of these measures would be optimized for any given scale or technique. A validation technique that is sometimes used involves group comparisons, so they are also discussed here. The role of dementia stage in the assessment process is also considered.

Independent/External Validation Criterion

To determine whether test performance or informant reported behavior are valid indicators of mild neurocognitive disorder or dementia, one must have an independent way to document whether individuals have these disorders or not. If the scale differentiates among adults, then there is support for its use. Unfortunately, there are no biological indicators for use as a gold standard for dementia [16, 43, 92]. Historically, diagnosis of dementia by an experienced clinician was used as a gold standard. There is evidence, however, that some clinicians are biased to diagnose more dementia in adults with DS than adults with other forms of ID [3, 11, 42, 61]. An alternative validation method is to combine clinician diagnosis with diagnosis based on objective test results to arrive at a consensus diagnosis of dementia [2, 16, 20]. This method has less potential for bias, particularly if the clinician is blind to the age of the adult or to the etiology of ID [2, 4]. Still others have confirmed the presence of DAD by requiring that all adults so diagnosed show declines in functioning for 2–3 consecutive years [15]. Finally, investigators have used previously developed scales with demonstrated validity to examine the validity of new methods [2, 3, 10, 18, 19, 23, 25, 73, 88]. Thus, the demonstrated validity of the new scale depends on the validity of the existing scale. Although there is currently no ideal solution for the selection of external validation criteria for dementia scales, it is important to remember that the choice of external validation criteria can have

repercussions for obtained sensitivity and specificity of tests (i.e., the extent to which a test correctly identifies those who are demented and those who are not demented, respectively). An area for future research involves the addition of mild neurocognitive disorder categories into such validation studies.

Sensitivity, Specificity, and Predictive Value

As illustrated in Table 2.2, cutoff rules are often used to indicate whether a given individual has declines in functioning consistent with dementia [2, 3, 11, 15, 20, 54, 56]. On a single test, adults with scores above a certain cutoff, for example, would be considered demented, whereas those with scores below the cutoff would be considered not demented. As seen in Table 2.2, one could set the cutoff rules liberally (i.e., a lower cutoff score) so that more adults are identified as demented. More conservative cutoff rules (i.e., higher cutoff score) would mean that fewer adults are identified as demented [2, 3, 11]. If a test battery is used, liberal cutoff rules could involve documentation of declines needed to meet any two diagnostic criteria (e.g., memory and everyday functioning). More conservative rules, in contrast, could require declines or changes such that all diagnostic criteria are met (memory, cognitive, everyday, and emotional functioning) [2, 3, 11, 19]. Adults would be classified based on the cutoff rules as demented or not demented. The dementia classifications based on the rules are then compared to those based on an external criterion (e.g., clinical judgment, existing dementia scale classifications). Scale evaluation measures are then calculated as described in the note to Table 2.2. Future work could also examine cutoff rules for adults classified as no decline, mild neurocognitive disorder, or dementia [5, 7, 46].

As indicated by the example, the use of a more liberal cutoff rule may increase the sensitivity of a scale at the cost of specificity. That is, more adults would be identified as demented. Some of them, however, would not be demented according to the external comparison criterion. Similarly, the use of a more conservative cutoff rule could increase specificity at the cost of sensitivity. In this case, fewer adults are identified as demented, but some of them are considered to be demented according to the external criterion. At times, one may want a scale or technique to be more sensitive, such as when using it as a general screen for dementia or other psychiatric disorders. At other times, one would want a scale or technique to be more specific, such as when telling family members that an adult with ID has an irreversible dementia, as opposed to some potentially treatable psychiatric disorder. When using evaluation measures such as those illustrated in Table 2.2, one must consider issues discussed previously. Did the scale and external criterion, for example, use the same source of information when evaluating dementia? If the scale was a direct assessment scale like a memory test or battery of tests and the criterion was an informant report scale like a dementia scale, lack of agreement could occur simply because of the different sources of information. Of course, if a scale is useful, one would expect it to agree diagnostically with other valid scales designed for the same population (e.g., adults with DS with mild ID) regardless of the source of information [2, 85]. When examining predictive

Table 2.2 Effect of different cutoff rules on dementia scale evaluation measures[a]

Dementia classifications based on scale cutoff rules and external criterion					
External criterion			External criterion		
Liberal cutoff rule on scale	Demented	Not demented	Conservative cutoff rule on scale	Demented	Not demented
Demented	63	20	Demented	58	2
Not demented	7	10	Not demented	12	28

Evaluation measures by cutoff rule					
Measure	Liberal rule		Conservative rule		
Sensitivity	0.90		0.83		
Specificity	0.33		0.93		
Positive predictive value	0.76		0.97		
Negative predictive value	0.59		0.70		

Note: Sensitivity refers to a scale's ability to correctly identify adults considered to be demented (i.e., 63/70 and 58/70 for liberal and conservative cutoff rules, respectively). Specificity refers to a scale's ability to correctly identify adults considered to be not demented (i.e., 10/30 and 28/30 for liberal and conservative cutoff rules, respectively). Positive and negative predictive values refer to whether demented and not demented adults identified by the scale receive matching diagnoses from an external criterion (e.g., positive predictive value for liberal data is 63/83).

[a]Data were created to demonstrate differences in evaluation measures for liberal versus conservative cutoff rules. Liberal rules applied to one scale, for example, would require a lower cutoff score as an indication of dementia compared to a more conservative higher cutoff score (with higher scores indicating more severe dementia symptoms). Liberal rules applied to a battery of tests could require that only two diagnostic criteria are met (e.g., declines in memory and everyday functioning), whereas a conservative rule could require that all diagnostic criteria are met (i.e., memory and other cognitive declines, emotional changes, declines in everyday functioning).

validity, one could determine whether dementia status on the scale agrees with that determined by the external criterion at one point in time. One could also determine whether dementia status (e.g., dementia, mild neurocognitive disorder, no declines) or declines in functioning on the scale at time 1 predict dementia status according to the external criterion at time 2 several years later [2, 3, 11]. As mentioned previously, further work is needed to determine appropriate criteria and tests/scales for examining mild neurocognitive disorder in adults with ID [5, 7].

Reliability

A dementia assessment method that is to be used repeatedly must have adequate test–retest reliability. One way to examine such reliability is simply to repeat the assessment in healthy adults to see if the scores or dementia classifications remain the same. If informant report techniques are used, the same informant would need to report on functioning at each assessment, which can sometimes be challenging because of turnover in direct care staff. When using direct assessment techniques, practice effects can affect repeated test performance even when tests are administered

after a long time interval [3, 11]. Changes in test performance that could be related to aging as opposed to mild neurocognitive disorder or dementia should also be considered when evaluating test–retest reliability. Another way to examine test–retest reliability and perhaps the validity of a more complex test is to see whether the underlying factor structure remains the same over time. For example, an informant-report dementia scale could involve assessment of depression, memory, and maladaptive behavior. Items believed to assess these three areas should have a consistent factor structure over time if they are actually measuring the same thing [3].

Finally, informant-report scales given at a single assessment or repeatedly should have adequate interrater reliability. Some informant-report scales require the use of two informants and thus allow examination of interrater agreement at each assessment (e.g., DSDS [47]; Reiss Screen [86]). If a scale requiring just one rater has demonstrated interrater reliability, then changes in informants from one assessment to the next would not be expected to result in drastic changes in reported performance, like those expected with dementia. If an adult has reported declines in performance and the informant has changed, however, it can be difficult to conclude that actual declines have taken place. Sometimes, a change in informant coincides with a change to a more restrictive or assistive living environment. If this is the case, it is difficult to separate changes in reported behavior due to informant perceptions from those due to changes in the environment.

Group Comparisons

At times dementia scales or diagnostic techniques are evaluated by comparing the performance of groups with and without dementia. A memory test is administered to adults with and without dementia, for example, and performance is compared. If the adults with dementia score lower than those without dementia, however, several issues must be addressed when interpreting such findings. First, there is the issue of confounding factors affecting performance that could differ between the groups (e.g., level of functioning, age, etiology of ID, medical health, sensory capabilities, etc.). Second, one must interpret overlapping performance between the groups (i.e., individual adults in both the demented and not demented group could remember five items). It is possible that an adult with dementia remembered 8–10 items when healthy, but declined to the current level. The adult without dementia, in contrast, could be showing optimal performance. Without an indication of performance for the individuals in the demented group when healthy, one does not necessarily know that the memory test would actually differentiate those with dementia from those without. In some cases the test being evaluated is the same test initially used to determine whether adults are demented or not (e.g., an adult with a score of 5 or lower on the memory test is demented, otherwise they are not). In such cases, group assignment is not independent of the evaluation of the scale, and reliable conclusions about the scales' usefulness cannot be made.

Evaluation and Stages of Dementia

When evaluating dementia scales and techniques, the value of the results could vary as a function of the stage of dementia (given a progressive dementia) [12, 26]. Some adults when healthy, for example, function independently in the community with a large array of academic and vocational skills. If they decline to the more advanced stages of dementia and need full time care, few individuals would argue over the presence of clinically significant declines. In such instances, high agreement would be expected among different dementia diagnostic methods. With mild neurocognitive disorder or at earlier stages of dementia, however, the individual may show some behavioral changes (e.g., uncharacteristically telling stories about what they have done or what others will do). In such instances, it is often unclear whether clinically significant declines in functioning as required by diagnostic criteria have occurred (i.e., declines in memory, other cognitive, emotional, and everyday functioning). If a psychiatric disorder is present (e.g., depression), it is also difficult to determine the extent to which losses in functioning are related to the disorder versus an underlying dementia [10, 66, 90]. One must determine whether a psychiatric disorder such as depression or a psychosis could lead to such a change in functioning [26, 42–44]. It is at this stage of dementia, specifically with psychiatric symptoms complicating diagnostic issues, when agreement among different dementia diagnostic methods would be expected to be lower. It is at this stage, however, when dementia scales could be most beneficial, because treatment could be most beneficial [5, 53, 93–99]. In addition, scales and techniques allowing classification of mild neurocognitive disorder would be advantageous at this point [3, 5, 7, 10, 11, 46, 54]. Future work and research will be needed to determine the clinical usefulness of such classifications in adults with ID.

Stage of dementia could also affect the obtained sensitivity of dementia scales. It can be very difficult for care providers and family members to detect early signs of dementia (particularly those in memory and cognition [53]). Therefore, an adult who has already shown undetected declines in functioning could be referred for information gathering or dementia screening. As such, the declines usually detected by a given scale used for information gathering, screening, or evaluation could never be detected because they occurred before the adult came to the attention of clinicians or researchers. A number of researchers and clinicians have addressed this issue by identifying and assessing only adults when they change from a healthy to a demented status. This is the ideal method for examining the sensitivity and predictive value of a scale. It is often not practical, however, because of the need to include in analyses all adults identified with dementia at a given site, because of small numbers detected with dementia. In addition, clinicians do not always have the luxury of having a baseline record of healthy functioning, and they must make diagnostic decisions based on the stage of dementia present when the adult is first evaluated. Thus, it would be beneficial when evaluating scales to determine their validity as a function of the stage of dementia. Finally, determining whether mild cognitive disorder overlaps with previously identified stages of dementia or characterizes adults who were found to meet some but not all diagnostic criteria [5, 7, 90]

will be necessary. It will not promote advances in knowledge or allow comparisons across studies if different terms/categorization schemes are used to identify the same level of decline (e.g., early dementia versus mild neurocognitive disorder) across sites and investigations.

Evaluation and Strength/Weakness Profile

Evaluation results could also be affected by premorbid level of functioning and profile of strengths and weaknesses. If, for example, adults with milder levels of ID typically scored at the ceiling of a test, the test may not be able to differentiate those who have started to show declines in functioning from others until they are in the more advanced stages. The test, however, could be an excellent indicator of mild neurocognitive disorder or dementia in lower functioning individuals. Similarly, if a test is so difficult or the instructions are so complex that healthy, lower functioning adults score at the floor of the test or at a level that would not allow detection of declines, then the test would not be a good indicator of mild neurocognitive disorder or dementia for them. It could, however, be an excellent indicator for higher functioning adults, whose functioning on such a test could be highly indicative of dementia status. Similarly, adults with a premorbid weakness in an area (e.g., due to sensory impairments or articulation disorders) could affect test evaluation results in unexpected ways. Thus, when evaluating a test it is important to consider individual differences related to level of functioning, age, gender, and etiology of ID as discussed previously [26]. At this time, it is not known whether one test or set of tests or scales is useful for adults at all levels of functioning. It is possible that tests or scales specific to level of functioning could lead to maximum levels of sensitivity and specificity, at least for adults with milder levels of ID.

Evaluation of Scales Across Studies

Recently a method to evaluate assessment scales, the Characteristics of Assessment Instruments for Psychiatric Disorders in Persons with Intellectual Developmental Disorders (CAPs-IDD), [100] was used to perform a structural evaluation of the DLD. The DLD was found to be one of the most frequently used scales across studies [101]. Use of the CAPs-IDD provided an informative summary of study methods and results. The summary also indicated future directions needed to improve evaluations and suggested a way to keep evaluation results/information up to date and accessible. Through new technologies, results of methods like the CAPs-IDD could be made accessible to researchers/clinicians and be updated when new information is available [100]. Thus, researchers/clinicians could benefit from a current state-of-the art summary of scales used internationally. In addition, the technological summary could provide a vehicle for improved communication among them.

Of course, such an evaluation across studies will not be possible without evaluation of the same scales across different sites. An additional critical review of tests/scales provides summary information and recommendations for test/scale selection [102].

Clinical Usefulness of Dementia Assessment Scales and Techniques

A final issue in scale evaluation is whether the scale would actually be useful and feasible to administer in a clinical setting. Often scales and techniques are evaluated as part of a research project, and the usefulness of the scale or technique has not been evaluated in a clinical setting. Questions shown in Appendix A address the issue of clinical usefulness.

In addition, recently it was proposed that only tests/scales specifically developed for dementia assessment in adults with ID will be clinically useful [24]. Such tests/scales are often given preference [1]. As long as tests/scales developed for other purposes (e.g., general adaptive behavior scales) are properly evaluated for dementia classification in adults with ID, the purpose of their development may not be found to affect their usefulness.

Summary

What can seem like a staggering number of issues affects the assessment of dementia in adults with ID. Such issues, however, are similar to those pertinent to the assessment of dementia in the general population for whom a considerable amount of effort and resources has been devoted [103]. Extra effort, such as that demonstrated by the authors of subsequent chapters, is required for scale development and evaluation for adults with ID. Each chapter will discuss a number of issues related to their respective tests, scales, or information gathering tools.

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Chapter 3
The Dementia Questionnaire for People with Learning Disabilities

Heleen M. Evenhuis

Introduction

To facilitate the diagnosis of dementia in persons with intellectual disabilities (ID), based on observations of caregivers, since 1980 the Dementie Vragenlijst voor Zwakzinnigen (DVZ) has been developed by Heleen Evenhuis, ID physician, and Margeen Kengen and Harry Eurlings, behavioral therapists, all working in De Bruggen center for people with ID, Zwammerdam, the Netherlands [1]. The Dementia Questionnaire for People with Learning Disabilities (DLD) is an English translation of this instrument. Formally known as the Dementia Questionnaire for Mentally Retarded Persons (DMR). After many years of distribution through De Bruggen, its publication has now been taken over by Harcourt Test Publishers [2]. In this chapter, we review the development of the DMR (DLD) along with its clinical applications.

Background

In people without a preexisting cognitive impairment, the diagnosis of dementia is primarily based upon an interview with the patient and his/her family. Collected information concerns memory, orientation, thought, mood, interest and activities, self-care, speech, and practical abilities. Completed with neuropsychological assessment, and physical and laboratory assessment to exclude physical causes of

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deterioration, a diagnosis of probable dementia can be made in an early stage in a vast majority of cases. Our practical experience at that moment, later confirmed by research, was that in principle, dementia has in people with ID the same course and similar symptoms as in other people [3, 4]. Therefore, the diagnostic procedure should be comparable. Because neuropsychological tests, at least those available in those years, were not applicable to persons with developmental ages lower than around 5 or 6 years, we considered a careful interview of observations by the family or other carers of even more importance for a diagnosis than in other people. To help us and others ask the right questions, we decided to develop a list of items, which should be normally asked in each proxy-based interview.

DMR (DLD) Designing Process

We started with the normal way in patient interviews, designing our item list accordingly: what is the situation now, and what was it before that? Before long we were confronted with the problem, that in this population, the preexisting cognitive level varies considerably between individuals. Therefore, the current functional level will always, more explicitly than in other people, have to be compared with the former level of functioning. This can only be realized in case of continuous and capable observations by persons who are familiar with the individual person and with symptoms of dementia. However, in practice, the average carer worked no longer than 2 years with the same clients, whereas, in the 1980s, nobody had any experience with dementia. Especially memory and orientation were seldom explicitly noted. As a result, observations were always incomplete and relevant data had been unsatisfactorily recorded. We concluded that looking back did not provide us with reliable, objective information, and that we had to work the other way round: structured recording of functioning before any deterioration was apparent, and again in case of deterioration. This required questions in a “here and now” format. Moreover, they had to be formulated in such a way, that they could be answered for persons with mild, moderate as well as severe ID.

These considerations resulted in a first draft with 77 items, to be completed by a family or staff member, who was familiar with the person. The questions were primarily based on first international guidelines for dementia diagnosis [5, 6] and were originally clustered in seven clinical subscales: short-term memory, long-term memory, spatial and temporal orientation, speech, practical skills, mood and inactivity, and behavioral disturbance. Further, the choice of items was based on our practical experience with interests and communicative capacities of people with mild to severe ID. Together with the methodologist Prof. L.J.Th. van der Kamp of the psychology department of Leiden University, and his graduate student Josien de Boer, the format was completed and first evaluation studies were performed. To prevent response tendencies, the items were placed in an arbitrary sequence. The questionnaire was provided with a simple linear score system, in which the items had three response categories: 0 points, no deficit; 1 point, moderate deficit; 2 points, severe deficit. Therefore, higher scores correspond to more severe deterioration. Appendix B shows the format of questions 1–5.

The subject's behavior during the past 2 months had to be judged. If an item could not be defined, e.g., in case of a lack of expressive capacities of the subject, this could be scored as "not to be determined" in the early version.

First Studies, Leading to Publication of the Final Version

In 1983, single completions of the first version of the DMR (DLD) were performed by pairs of two independent carers for 98 institutionalized older persons with mild to profound ID, to test the interrater reliability, internal consistency of the subscales, and the relationship of intellectual levels and scores. The interrater reliability appeared satisfactory (see below). Items that correlated insufficiently with the other items within the same subscale, were omitted, as well as items that in a majority were scored as "not to be determined" and items which discriminated insufficiently (i.e., mostly scored as "0"), leading to a final list of 50 questions (Table 3.1).

As expected, a negative correlation was found between intellectual levels and scores: the lower the intellectual level, the higher the scores. Based on internal consistency outcomes, the original subscale "Mood and Inactivity" was split up into the subscales "Mood" and "Activity and Interest." [7] In a second study, again with single completions, in two institutionalized populations of, respectively, 271 and 263 older persons with mild to profound ID, the relationship of the expert diagnosis "dementia" with DMR (DLD) scores was studied. Results of a discriminant analysis showed that the subscales "Short-term memory," "Orientation," "Speech," "Practical skills," and "Mood" discriminated best between groups with and without a diagnosis "dementia." If scores of all individual participants were classified according to the results of the discriminant analysis, in an average of 72% of subjects a correct diagnosis was made. A correct diagnosis based on DMR (DLD) scores appeared particularly difficult in case of a severe or profound ID, extreme apathy, or clouded consciousness [8].

Table 3.1 Dementia questionnaire for people with learning disabilities

Subscales	Min-max scores
Sum of cognitive scores (SCS)	0–44
1. Short-term memory (seven items)	0–14
2. Long-term memory (eight items)	0–16
3. Spatial and temporal orientation (seven items)	0–14
Sum of social scores (SOS)	0–60
4. Speech (four items)	0–8
5. Practical skills (eight items)	0–16
6. Mood (six items)	0–12
7. Activity and interest (six items)	0–12
8. Behavioral disturbance (six items)	0–12

Table 3.2 Interrater reliability [7]

Subscale	Pearson correlation coefficient
Short-term memory	0.84
Long-term memory	0.87
Orientation	0.86
Speech	0.68
Practical skills	0.94
Mood/activity and interest	0.74
Behavioral disturbance	0.44

Psychometric Properties

Reliability

The interrater reliability was studied by measuring the Pearson correlation coefficient for the different subscales. In this stage of the development of the DMR (DLD), the subscales "Mood" and "Activity and interest" were one subscale. The correlation coefficients for the different subscales varied between 0.44 and 0.94 (Table 3.2). Only for subscale "Behavioral disturbance," the correlation between raters was relatively low (0.44). It appeared that this low correlation resulted from differences within one of the six pairs of raters. The results for the other subscales were satisfactory [7].

"Gold Standard": Expert Diagnosis

Because no other diagnostic instruments for dementia were available, evaluated for people with ID, a specialist diagnosis by a physician and/or psychologist with expert knowledge in the field of dementia and ID was used against which to judge the sensitivity of DMR (DLD) scores. A specialist diagnosis "dementia" was made in case of a permanent and increasing deterioration of the cognitive and social functioning, according to DSM-III-R and later DSM-IV criteria [5, 9]. These criteria had to be slightly modified (Table 3.3), because of the variance of original cognitive functioning as part of the ID. Additionally, because no or hardly any neuropsychological test methods are available to reliably assess abstract thought, judgment, aphasia, apraxia or constructive insight in this population, we omitted the criterion "disturbances of abstract thought and judgment," whereas aphasia and apraxia could only be observed in daily circumstances.

Sensitivity and Specificity

In two prospective longitudinal studies, the sensitivity and specificity of different criteria for interpretation of DMR (DLD) scores have been studied in older groups with Down syndrome (DS) and with other causes of ID, both for multiple and for

Table 3.3 Modified diagnostic criteria for dementia (modified DSM-III-R) [7, 8]

A. Demonstrable evidence of decline of original level of short- and long-term memory (observed in daily circumstances)
B. At least one of the following (observed in daily circumstances)
1. Disturbance of original level of spatial or temporal orientation
2. Aphasia
3. Apraxia
4. Personality change
C. The disturbance in A and B significantly interferes with work for usual social activities or relationships with others
D. Not occurring exclusively during the course of delirium

Table 3.4 Sensitivity and specificity of the DMR (DLD) (95% confidence intervals between parentheses) [11]

	Sensitivity	Specificity
70+	7/7 (100%) (59–100)	19/26 (73%) (52–88)
DS	8/8 (100%) (63–100)	27/36 (75%) (58–88)

single completions [10, 11]. In these studies, persons with a clinical expert diagnosis of "dubious dementia" were classified as demented. The diagnosis "dubious dementia" was made in all cases of progressive functional deterioration, in which a diagnosis "dementia" could not be made according to modified DSM-III-R/IV criteria. This usually involved persons with insufficient capacities to express themselves, e.g., by severe generalized motor impairment or severe chronic depression, or persons with a beginning dementia who did not meet DSM criteria during the study, but did afterwards.

Diagnostic Criteria

The following diagnostic criterion for a diagnosis "dementia," based on scorechange as compared with original DMR (DLD) scores, led to the best sensitivities and specificities [11].

An increase of the Sum of Cognitive Scores (SCS) of 7 points or more and/or an increase of the Sum of Social Scores (SOS) of 5 points or more, independent on the original level of ID. Results of application of this criterion are presented in Table 3.4.

A sensitivity of 100% means that all cases with an expert diagnosis of dementia will be correctly identified by the DMR (DLD). A specificity of 75% indicates that 75% of persons without dementia are correctly classified as "no dementia" by the DMR (DLD). However, 25% is incorrectly classified as "dementia" (the so-called false-positives). In such cases, further diagnostic assessment usually identified a functional deterioration by other conditions. Although of course a specificity of 100% would be preferable, this is in practice realized in hardly any diagnostic instrument [12, 13]. Which specificity is acceptable, will vary per condition. For example, a false-positive diagnosis of cancer would have to be avoided as much as possible. However, in the case of dementia in persons with ID, a specificity of 75%

is acceptable. Indeed, in a majority of cases with incorrect diagnoses of dementia, further diagnostic assessment resulted in relevant and often treatable other diagnoses (severe sensory impairments, severe motor impairments, severe physical disease, and psychiatric conditions). As a conclusion, with the DMR (DLD), functional deterioration as a result of cognitive as well as noncognitive aspects is identified. Longitudinal judgment of scorechanges is more reliable than single completion and is therefore preferable.

Results of the last evaluation suggested that the DMR (DLD) is less accurate in case of specific causes of dementia, other than dementia in Alzheimer disease (DAD) (e.g., vascular dementia). However in this stage, such a conclusion can only be speculation because of the small subgroups.

Judgment by Committee on Test Affairs Netherlands

The quality of the Dutch DMR (DLD) has been recently rated by the Committee on Test Affairs Netherlands (COTAN) of the Dutch Institute of Psychologists. The purpose of these ratings is twofold. Test users are informed about the quality of available instruments, which information can help them in choosing an instrument. Besides, the ratings supply feedback to test-developers about the quality of their products. An English translation of the rating procedure has been published in the *International Journal of Testing*, 2001, pp. 155–182. Outcomes for the DMR (DLD) (2B.13 DVZ) were as follows: theoretical basis and soundness of test development procedure, satisfactory; quality of testing materials, good; comprehensiveness of the manual, good; norms, satisfactory; reliability, satisfactory; construct validity, satisfactory; criterion validity, satisfactory.

Applications of the DMR (DLD)

Dementia

The DMR (DLD) has been designed in principle for the diagnosis of dementia in adults with ID. However in practice, because DAD is the most prevalent cause of dementia, we have primarily evaluated the sensitivity for DAD. Due to small subgroups, the sensitivity for rarer types of dementia has been evaluated insufficiently.

Early Detection

Our longitudinal evaluation shows, that in all cases, a diagnosis based on DMR (DLD) scores was made prior to or at the same time as an expert diagnosis according to international criteria could be made (DSM-III-R/DSM-IV).

Screening Instrument and Effect Instrument

We stress that the DMR (DLD) is not an instrument for a definite diagnosis of dementia, because severe progressive physical and other psychiatric conditions, or a combination of less severe conditions, may influence the scores as well. Therefore, the DMR (DLD) has to be used as a screening instrument, i.e., for selection of persons for further specialist diagnostic assessment. Recently, the instrument has been proven satisfactory to evaluate effects of interventions [14, 15].

Repeated or Single Completion

The basis for a diagnosis of dementia is always a deterioration from the former individual level of cognitive functioning. Indeed, the DMR (DLD) is most sensitive in case of multiple measures.

Originally, we have also tried to develop criteria for a single completion of the DMR (DLD), which would simplify large-scale screenings, e.g., in connection with research projects. This is only possible under the condition that reliable and interindividually comparable data from former intelligence tests, performed prior to any deterioration, is available. In our own evaluation studies, the participants' level of ID had been ascertained with several tests: Stutsman Mental Measurement of Preschool Children [16], Peabody Picture Vocabulary Test [17], and Leiter International Performance Scale [18]. The results may not be completely comparable to other scales, used nowadays and in other countries to test functional levels. Therefore, a diagnosis based on a single application of the DMR (DLD) is now considered insufficiently valid, and is strongly discouraged by us.

Criteria for Persons to Be Tested

The DMR (DLD) is applicable to persons with mild, moderate, or severe ID (developmental ages around 2–10 years). It is not applicable to persons with profound ID (developmental age lower than 2 years) and to persons with severe ID (developmental age 2–3 years) combined with severe other disabilities, such as motor impairment or hearing loss. In such cases, DMR (DLD) scores may approach extreme levels before any functional deterioration ("ceiling effect").

Who Answers the Questions?

The questionnaire has to be completed by a family or staff member who is familiar with the person. Carefulness and objectivity are very important. This may be advanced by DMR (DLD) completion not by a single person, but by a family member together with a staff member, or by several carers together, and preferably guided by the investigator.

Who Interprets the Answers?

Interpretation of the results is only useful in combination with other diagnostic data, as applies for each diagnostic instrument. Therefore, this should be done by the diagnosing physician, psychologist, or behavioral therapist.

Directions for Diagnostic Use

Because longitudinal judgment of DMR (DLD) scores provides the most reliable diagnosis, it is advised to routinely perform a first scoring of the DMR (DLD) before any functional deterioration is observed. This might be done when somebody moves to a home for several persons with ID, or joins a day activity center. Any observed deterioration should prompt repeated completion of the DMR (DLD). If no scorechange is found, consistent with a diagnosis of dementia, further diagnostic assessments are to be aimed primarily at other causes of deterioration, such as a depression or sensory impairment. Dependent on the development of symptoms, a next DMR (DLD) scoring and judgment is advised after 6–12 months.

In case of a DMR (DLD) diagnosis "dementia," referral for specialized psychiatric and general physical examination is advised, according to national or international guidelines [19–22]. In any case, visual and hearing functions are to be actively tested, because of increased risks of age-related sensory impairments in this population, which are missed in many persons with ID [23, 24].

Rating

The questionnaire is provided with a simple linear score system, in which the items have three response categories: 0 points, no deficit; 1 point, moderate deficit; 2 points, severe deficit. The subject's behavior during the past 2 months has to be judged. If an item cannot be defined, e.g., in case of a lack of expressive capacities of the subject, the score has to be "2".

The items are clustered in eight subscales (Table 3.1) and placed in an arbitrary sequence, to prevent response tendencies. Combined scores on the first three subscales (short-term memory, long-term memory, and orientation) are indicated as the SCS. Combined scores on subscales four through eight (speech, practical skills, mood, activity and interest, and behavioral disturbance) as the SOS. The questionnaire is provided with a short instruction for completion. Completion takes 15–20 min.

Other Studies of the DMR (DLD)

Since the availability of an English translation of the DMR (DLD), it is clinically used in many countries around the world. Several researchers have evaluated the DMR (DLD) for their country, or used it in epidemiological or intervention studies.

The DMR (DLD) in Diagnostic Test Batteries

Since the 1990s, other diagnostic instruments, both informant-based and to be administered directly to persons with ID, have been applied or developed to assess for dementia. Most of these tests are aimed at specific symptoms, such as maladaptive behavior, memory decline, or verbal fluency, or are specifically designed for persons with DS. Combinations of such tests in diagnostic batteries have been recommended by several groups [25–28]. The DMR (DLD) in all cases was presented as the most promising informant-based screening tool in most adults with ID, including those with DS. It is the only informant-based scale available for assessing orientation [27].

Evaluations of the DMR (DLD)

Evaluations by other authors concern mostly single completions of the DMR (DLD), referencing to Intelligence Quotient (IQ) levels. It appeared that such results were less satisfactory than in our own evaluations, probably due to application of varying tests for IQ or functional levels, or other criteria for levels of ID. For this reason, Prasher proposed for persons with DS in the United Kingdom modified higher cut-off scores for single DMR (DLD) scores [29].

Burt and colleagues [30] in the United States, specifically evaluating assessment of orientation in 138 adults aged 29–82 years, found fair to good agreement between DMR (DLD) scores on the subscale “Orientation” (single ratings) and direct assessment. The level of agreement was negatively influenced by lower functioning, DS, and higher age.

Deb and Braganza [31] in the United Kingdom compared ratings on several informant-based scales with the clinician’s diagnosis among 62 adults with DS. The diagnosis according to DMR (DLD) criteria (single ratings) showed sensitivity and specificity at the 0.92 level for both categories. In this study, the observer-rated scales appeared more useful for the diagnosis of dementia than the used direct neuropsychological test.

Silverman and colleagues [32] performed a study of dementia in 273 adults with ID, applying multiple tests 18 months apart. As opposed to our own findings, single ratings of the DMR (DLD), referencing to IQ measurements with Wechsler Adult Intelligence Scale and Stanford–Binet scales earlier in adulthood, distinguished more effectively between individuals with and without dementia than scorechanges during the study period. Sensitivity of scoreschanges over the 14–18 month period was less impressive than reported in the DMR (DLD) manual. However, we suspect that in this study, the dementia process in a number of cases might have started before the first rating. As a result, no predementia baseline data were available, as is recommended in the manual. The authors recognize this: "It might be worthwhile examining change in DMR (DLD) scores for incident cases for whom a predementia baseline is available, and to rely more on single assessment scoring otherwise." In this study, effects of different IQ tests were also studied. Indeed, it appeared that the IQ testing procedure had a significant effect on classifications of nondemented participants ($p < 0.05$) and a nonsignificant effect in other dementia status groups, but the power was low.

Shultz and colleagues [33] in the United States and Canada evaluated several screening tools for dementia in a case-control study, with 38 matched participants with mild to profound ID in each group. Again, single ratings were used for the DMR (DLD), referencing to IQ measurements that were at least 5 years old, obtained with a variety of methods. Paired *t*-tests for both SCS and SOS ratings were highly significant, without correlating to gender, age, IQ level, or DS. In a logistic regression analysis of all tests used, the DMR (DLD) SOS was the variable that best predicted group membership.

Recently, Walker and colleagues [34] carried out the DMR (DLD) interview independently with 2 carers caring (at least 6 months) for 26 people with Down syndrome. Fifteen males, 11 females, mean age 50.5 years (range 40–69 years). Only 15% of the pairs of informants had good agreement. Better agreement for less abled participants. The authors recommended were not to rely only on carer interviews when assessing for dementia in persons with ID.

The DMR in Intervention Studies

Prasher and colleagues [14, 15] used DMR (DLD) scores as the primary outcome measures in a 24-week randomized controlled trial (RCT) of the cholinesterase inhibitor donepezil. The study group consisted of 27 persons with DS and mild or moderate DAD. There was a tendency that donepezil halted the rate of decline, but the sample size was too small for statistical significance. The trial was continued as an open-label study until a total of 104 weeks. Long-term use of donepezil significantly reduced the rate of decline ($p < 0.001$). A comparable 24-week effect study of rivastigmine has also been published by Prasher and colleagues [35]. Prasher concludes that the DMR (DLD) is sufficiently sensitive to measure scorechanges as a result of intervention (personal communication 2004).

An uncontrolled evaluation of treatment with different cholinesterase inhibitors in a network of specialist memory clinics for people with ID in Southwest England was recently reported [36]. Here too, the DMR (DLD) was used to monitor intervention effects, showing a significant deterioration of total scores in the last two assessments before treatment ($p < 0.01$), during a mean interval of 10.8 months. Treatment seemed to stabilize scores during a mean period of 7.4 months, whereas the SOS showed a significant improvement ($p < 0.05$).

Summary

Recently, the DMR (DLD) has been rated satisfactory to good by the COTAN. From secondary studies by other authors, we conclude that the DMR (DLD) is high-ranking in recommendations for diagnostic batteries [25, 26, 28]. Apart from cognitive items, it also scores noncognitive items. It is the only informant-based scale for assessment of orientation [27]. Authors use preferably single DMR (DLD) ratings, requiring reliable IQ levels for referencing [29, 31–33]. In that case, the choice of IQ tests or tests for functional levels may negatively influence sensitivity and specificity, because different tests lead to different dementia classifications based on the DMR (DLD) [32] Nevertheless, results in these studies are promising. Corroborated by the findings of Silverman and colleagues [32], we stress again that a sensitive DMR (DLD) diagnosis based on score-changes requires baseline ratings prior to onset of dementia and not during dementia. The DMR (DLD) is a sensitive instrument to monitor changes as a result of intervention [14, 15, 35].

Our own evaluation studies have shown that the DMR (DLD) is not sensitive in persons with profound ID, because of a "ceiling effect." To our clinical experience, there is a "bottom effect," too: in persons with very mild or borderline ID and beginning dementia, it may take years before DMR (DLD) scores reach the level of a dementia diagnosis. Apparently, the DMR (DLD) is not sensitive to more subtle functional deterioration, and the questions have been designed with capacities of people with moderate and severe ID (developmental ages 2–6 years) in mind.

During our evaluation studies, the DSM-III-R was replaced by the DSM-IV [9]. Did this influence the validity of the DMR (DLD)? In the DSM-IV, some of the former clinical criteria for a diagnosis of dementia were omitted, namely "disturbances of abstract thought in judgment" and "personality change." According to the DSM-IV, deterioration from the original level of functioning has to be more explicitly taken into account. The only change in our modified criteria would therefore be the absence of the criterion "personality change" (Table 3.3). Because this aspect in practice has hardly played a decisive role in our specialist diagnoses, it is not to be expected that outcomes of our validity studies would have shown relevant changes by applying DSM-IV instead of DSM-III-R criteria.

In 1995, we participated in an international consensus group for diagnosis of dementia in people with ID, which advocated application of ICD-10 rather than DSM-IV criteria in this population [25, 26, 37]. The reason was, that, as compared

to the DSM-IV, in the ICD-10 more emphasis is placed on noncognitive aspects of dementia (e.g., emotional lability, irritability, and apathy). In practice, these noncognitive aspects are often the first signs, reported in individuals with ID, rather than cognitive aspects. The consensus group concluded that in this way a "two-step" diagnostic procedure is introduced, in which a possible diagnosis of dementia will be reconsidered, if observed behavioral changes are not accompanied by evidence of cognitive decline. It was seen as an advantage that in this way, consideration of all possible causes of decline is required, including of those that are treatable. These recommendations are in line with the more recent recognition of the role of psychiatric and behavioral disorders in dementia syndromes in clinical research in the general population [38]. Aylward and colleagues [26] observed that ICD-10 and DSM-IV overlap completely on the part of cognitive decline. The DMR (DLD) was cited as a reliable method to detect a decline in memory and other cognitive abilities, a decline in emotional control or motivation, or a change in social behavior. Indeed, with the second part of the DMR (DLD), a range of noncognitive aspects can be assessed, among which the aspects, mentioned in the ICD-10.

We conclude that the distinction of "dubious dementia" and "dementia" in the expert diagnosis in our DMR (DLD) studies is in fact comparable to this "two-step" procedure. Our choice to classify "dubious dementia" as "dementia" for the assessment of sensitivity and specificity is in line with the considerations of the international consensus group. Therefore, it may be assumed that evaluation of the DMR (DLD) against a clinical diagnosis according to ICD-10 criteria would have resulted in comparable outcomes.

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Chapter 4
Gedye Dementia Scale for Down Syndrome

Emoke Jozsvai, Spencer Hewitt, and Angela Gedye

Introduction

The term "dementia" refers to deterioration in intellectual functioning or the development of multiple cognitive deficits affecting memory, language, comprehension, and activities of daily living. There are many types of dementia that occur in the general population and in those with intellectual disability (ID). Dementia in Alzheimer Disease (DAD) is the most common form of dementia in Down syndrome (DS). Its clinical manifestation increases with aging from 8%, in those between 35 and 40 years old, to approximately 22%, for those aged 40+. For individuals in the 60+ age group, the rate is estimated to be 69% [1–4]. Among institutionalized individuals with DS the rate of dementia has been reported to be as high as 77% [5]. However, other types of progressive dementia (e.g., vascular dementia), reversible dementias (e.g., untreated hypothyroidism), and conditions that mimic dementia also occur in adults with ID [6]. The pattern and symptoms of DAD in adults with DS are similar to those observed in the general population [7, 8], except that the decline in DS adults starts from a significantly lower level of functioning and from a younger age [9].

Unfortunately, most instruments for assessing dementia in the general population are unsuitable for use with the ID population, especially in persons with severe or profound ID. In recent decades there has been an increasing need for instruments (a) to assess for dementia in ID adults and (b) to aid differential diagnosis when cognitive decline presents.

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Background on the Development of the Scale

The development of one such instrument, the Dementia Scale for Down Syndrome (DSDS) [10] began in 1987. First, the author identified some of the psychometric concerns critical in assessing this population, especially those in the severe or profound range of ID. These concerns included:

1. The need for information that does not rely on a person's performance on tests in a person unable to follow test instructions.
2. The need to distinguish features *typical* of that person from features that indicate *loss* of functioning.
3. The need to rate severity of dementia relative to the person's premorbid intelligence.
4. The need to consider conditions that cause a *reversible* dementia or *mimic* a dementia.
5. The need for *charting over time* to detect worsening of functioning or recovery of functioning (in the case of reversible dementia).

Gedye then designed a protocol that addressed those specific psychometric concerns. She collected longitudinal data over 8 years on adults with ID (with and without DS), then did a detailed item analysis, identified item patterns reflecting differential diagnoses, and developed a scoring system that reflects stages of severity of dementia. Thereafter, reliability and validity studies were conducted in a different province on 50 adults with DS.

In 1995, the DSDS was published. Since that time, many researchers have used this scale (including those in non-English countries such as Japan, Holland, and France) and some have published results on the psychometric properties of the scale (see "Psychometric properties of the DSDS"). The DSDS was standardized and validated mostly on adults in the severe and profound range of ID, but researchers have also used it with adults in the mild and moderate range of ID. Clinicians have more testing options when assessing adults in the mild or moderate range of ID because they can be given tests that require direct performance whereas those in the severe or profound range may never have been able to do such tests. Over the years, several observer-rated instruments have been developed for people with ID [11–15]. One of the earliest and perhaps most commonly used among these is the DSDS, also known as the Gedye Dementia Scale for Down Syndrome (G-DSDS).

The Dementia Scale for Down Syndrome

The DSDS is a 60-item informant-based instrument that was designed to aid in diagnosing dementia in individuals with ID, especially those with DS. The DSDS can also be used to establish a baseline measure on individuals with ID who are at risk of developing dementia because of their age, but currently do not exhibit signs of cognitive decline. The scale is classified as a Level C test, thus clinical

psychologists with experience in the psychometric assessment of ID are qualified to administer it and interpret the results. A psychometrist with an undergraduate degree and a minimum of 2 years experience with tests of intellectual and adaptive functioning may also qualify to administer the scale. The DSDS requires that caregivers responding to the questions know the person for at least 2 years and be familiar with the person's skills of daily living. It is recommended that two people be interviewed and, if the client works, to have one informant from the person's workplace. The DSDS is available commercially in English, French and Swedish versions.

To detect the onset of dementia relative to baseline functioning, the DSDS includes many items that reflect losses in people with ID. Items on the DSDS can be rated as "typical" (if a feature is characteristic of the individual through his/her lifetime) or "not applicable" (if the feature was never part of the person's baseline cognitive or behavioral repertoire). It is important in assessing people with ID to ensure items of lifelong impairment are not misread as signs of dementia, and the DSDS was designed specifically to avoid psychometric confounding of lifelong impairments with dementia-related impairments in the list of items.

Items reflecting symptoms of dementia such as changes in interest and initiative, losses in verbal, spatial or temporal memory, decline in comprehension or language ability, may be rated "absent" or "present." The onset and progression of dementia is ascertained by tracking changes in functioning over time through follow-up assessments every 6–12 months. Questions are grouped into three categories with items that address "early stage," "middle stage," and "late stage" characteristics of dementia.

To meet criteria for the early stage, the person must have a minimum of three losses in the cognitive area and this is referred to as the Cognitive Cut-off Score (CCS). This helps eliminate people with many social/affective changes and/or physical losses—those who are perhaps depressed or showing physical declines—but who are not showing cognitive losses. It is also important to identify a time period when cognitive and other changes began. To do this, the DSDS user identifies the date of an early loss (often item #1) then typically adds 6 months to define a time period when early losses surfaced, thereby identifying the onset of dementia changes. (Occasionally the onset of dementia is very slow and the DSDS has provisions for initial changes to be spread over 12–18 months after the "first" sign of decline.) The criteria for early-stage dementia require at least three cognitive losses—a CCS of three or greater—and a total of ten changes taken from 20 possible early-stage items and 20 possible middle-stage items.

Screening for Conditions That Cause Reversible Dementia or Mimic Dementia

The DSDS aids in differential diagnosis by (a) listing clusters of test items that point to conditions that can co-occur, cause reversible dementias, or mimic dementia, and (b) providing additional questions to ask. The DSDS includes an easy-to-use section entitled Differential Diagnosis Screening Questions (DDSQ). This

section covers possible signs of hypothyroidism, pain, vision changes, hearing changes, depression, medication-induced cognitive decline, sleep apnea, and vascular dementia. Most of these conditions are fairly common in older adults with DS. The DDSQ questions can assist the DSDS user to make further inquiries and/or provide information to physicians so that other possible diagnoses can be ruled in or out. Thus, the DSDS is useful for detecting reversible types of dementia, conditions that can mimic dementia, along with DAD and other progressive dementias. One DSDS test booklet provides space for recording changes in functioning over ten assessments.

Reversible Cognitive Deterioration

1. *Hypothyroidism* is one of the most frequent causes of reversible dementia. Approximately 30–60% of those with DS over the age of 35 years have abnormal thyroid functions [16–18]. Specific symptoms of hypothyroidism include reduction in energy, motivation, and a general decline in cognitive functioning, including memory and attention.
2. *Vitamin B_{12} deficiency* can also cause forgetfulness, irritability, poor appetite, withdrawal, and a general functional decline. Symptoms of this form of dementia disappear once vitamin B_{12} therapy is administered. Vitamin B_{12} occurs in approximately 6% of people under the age of 60 and in 20% of people over 60 [19].
3. *Depression* can cause a reversible cognitive decline, but can also coexist with dementia, thus making differential diagnosis quite challenging in the DS population. It is one of the most commonly diagnosed psychiatric disorders in DS adults [20–22], and it is frequently found to be related to the elevated rate of hypothyroidism common in the syndrome. Undiagnosed depression can be misidentified as dementia and by ameliorating depressive symptoms, cognitive abilities can improve to previous levels before mislabeling of dementia [23]. Depression may be related to changes in the social milieu, such as death of parents or loss of residential caregivers. Presenting symptoms are likely to involve skill and memory declines, tearfulness, irritability, and a noticeable decrease in energy and activity level, loss of daily living skills, hallucinatory-like features [24–26]. Urinary incontinence may be associated with depression in adults with DS, and this condition also occurs in individuals with DAD [27]. Depression treatment is important in delaying the onset of dementia. Initial dementia onset in people with DS receiving antidepressants was delayed by over a year and the mean age of death by over 2 years [28].
4. *Medication-induced cognitive decline* is another concern [6, 29]. Approximately 70% of people with intellectual disabilities are prescribed regular use antipsychotics and 48% were prescribed multiple neuroleptic medications concurrently [30]. This extended use of multiple psychotropic drugs is associated with an increase in psychosis and acting out behaviours as well as decreased cognitive function.

Gedye [10, 29] described several cases of reversible dementia, and among those were cases related to seizure disorder and either short term or long-term use of neuroleptic medication. The history of cognitive decline in these cases ranged from 0.5 to 5 years, and they progressed to middle-stage features but did not progress to late-stage dementia. The majority of the individuals were under 40 years of age, but reversible dementia was also documented in adults over the age 50 years with DS and ID of other etiologies. After better seizure control or discontinuation of neuroleptic medication, all these individuals recovered their abilities.

Conditions That Can Mimic Cognitive Decline

1. *Sleep apnea* occurs in approximately 65% of DS individuals [31, 32]. It may produce behavioral changes such as irritability, depression, or paranoia. In addition, ongoing sleep disturbance can result in a significant decrease in attention and concentration, and it can produce a decline in an individual's general cognitive ability [33, 34].
2. *Hearing and visual impairment:* Adults with DS are at greater risk for both auditory and visual impairment. It has been reported that 40–70% of adults with DS likely experience sensorineuronal and/or conductive hearing loss [35], and 46% develop cataracts [36]. This compounds the already low visual acuity in the DS population [37]. These sensory impairments often produce behavioral changes such as withdrawal from regularly enjoyed activities and general apathy [38], thereby mimicking a cognitive decline.

Psychometric Properties of the DSDS

In the standardization sample 60 individuals with DS (63% male and 37% female) aged 40 years or older were selected to participate in a longitudinal study of age-related cognitive changes. The participants were selected from a provincial (British Columbia, Canada) DS population of 229 people (56% male and 44% female) who were 40 years of age or older when the study began in 1987. Ten individuals with symptoms of dementia who were under the age of 40 years were also included in the DS group and were followed for several years. A control group of 47 non-DS elderly with ID was also followed. Levels of intellectual functioning, according to the DSM-IV criteria [39], in the DS group included mild (1%), moderate (23%), severe (46%), and profound (30%) ranges. The percent distribution of levels of ID in the control group was comparable, with the least number of participants in the mild (5%) and profound (16%) categories, and the majority falling within the moderate (27%) and severe (51%) ranges. The demographics for the DS and the control group are presented in Tables 4.1 and 4.2 [10].

Table 4.1 Demographic characteristics of the DS sample

Cohort by year of birth	Number	Male	Female	Community	Institution
1919–1927	10	6	4	4	6
1928–1937	29	19	10	18	11
1938–1947	21	13	8	18	3
Subtotal	60	38 (63%)	22 (37%)	40 (67%)	20 (33%)
After 1947	10	4	6	10	0
Total	70	42 (60%)	28 (40%)	50 (71%)	20 (29%)

Table 4.2 Demographic characteristics of the ID control group

Cohort by year of birth	Number	Male	Female	Community	Institution
1909–1917	3	2	1	2	1
1918–1927	10	6	4	4	6
1928–1937	19	9	10	14	5
Subtotal	32	17 (53%)	15 (47%)	20 (63%)	12 (33%)
After 1947	5	4	1	4	1
Total	37	21 (57%)	16 (43%)	24 (65%)	13 (35%)

Reliability and Validity Studies

In the context of psychological testing clinicians are concerned with interrater reliability, the degree of agreement between results obtained by two independent raters administering the same test. An index of interrater reliability is the kappa coefficient. In the standardization study of the DSDS, [10] two clinicians independently interviewed the same caregivers. The assessments took place within a few days of one another. The obtained kappa coefficient was 0.91 for the dementia classifications, thereby indicating a high interrater reliability for the DSDS.

Validity refers to the "truthfulness" of the instrument, or the degree to which the test measures what it claims to measure. Construct and criterion-related validity are most often of interest to clinicians in applied settings. The goal of construct validation is to determine whether or not test scores provide a good measure of a specific construct. In the case of the DSDS the construct being measured is progressive loss of cognitive ability, or dementia. Gedye [10] evaluated the construct validity of the DSDS from evidence pertaining to the onset and progression of dementia. In the DSDS standardization sample, of the individuals with DS who progressed to late stages, 100% had previously met the scale's criteria for early stages and criteria for middle stages. Further evidence for construct-related validity of the DSDS can be found in the study by Temple and colleagues [40] that involved 35 adults with DS between the ages of 29 and 67 years. The participants were assessed by the DSDS and a battery of neuropsychological tests that have been shown to discriminate between individuals with DS with and without dementia [41]. The participants were followed for a minimum of 6 months, and some were followed for a total of 3 years. All of the participants had completed multiple assessments with the DSDS and/or

the neuropsychological test battery. Approximately 20% of the participants were diagnosed with early-stage dementia, and 6% with middle- to late-stage dementia. All of the participants who were assessed as having early-, middle-, and late-stage dementia showed a substantial decline on the neuropsychological tests and/or on the DSDS at follow-up.

Criterion-related evidence for validity demonstrates whether test scores are systematically related to outcome criteria, i.e., the presence or absence of dementia. In the 1993 Ontario study of the scale's psychometric properties, a psychiatrist highly experienced in working with adults with DS, rated the presence or absence of dementia in 50 older adults with DS independently from a psychologist (the author) very experienced using the DSDS, and this yielded a kappa coefficient of 0.81 [10]. Criterion-related validity can be estimated by comparing the test's results with a clinician's diagnosis, as in the above example, or with another test. In addition to using a kappa coefficient, validity can also be expressed as sensitivity and specificity of a test. The validity indexes of sensitivity and specificity can be expressed as percent agreement, and/or a kappa coefficient. Sensitivity is defined as the proportion, or the percent, of true cases (individuals with a disorder) correctly categorized by the test as having the disorder. Specificity is the proportion of true noncases (healthy individuals) correctly diagnosed as being unaffected. The probability of agreement between a clinician's diagnosis and a diagnosis derived from an instrument can also be expressed in terms of the positive and negative predicting power of the test. The positive predictive power of a test is the probability that the person with a disorder is identified by the test as having that disorder. Negative predictive power is the probability that a person without the disorder will be categorized by the test as not having the disorder (see Shultz and colleagues [42] for details of calculating these indexes).

In the standardization study of the DSDS, Gedye [10] compared the dementia ratings of two clinicians for 46 DS individuals, and found a sensitivity of 100% and a specificity of 98%. Deb and Braganza [43] compared clinicians' diagnoses of dementia using ICD-10 criteria [44] with diagnoses arrived at using the DSDS and the Dementia Questionnaire for Mentally Retarded Persons (DMR (DLD); Chap. 3) [45]. Sixty-two adults with DS, aged 35–75 years with mild (22.6%), moderate (66%), and severe (11.4%) ID participated in the study. Twenty-six of these individuals were diagnosed by a clinician as having dementia and 36 were rated as non-demented. On the DSDS, 22 of the clinician-diagnosed demented participants met the criteria for dementia, but four of the participants who met criteria on the DSDS were not diagnosed by clinicians as demented. Thus, the comparison between the DSDS criteria and the rate of diagnosis of dementia by a clinician yielded a specificity of 0.89 and a sensitivity of 0.85. Similarly, the comparison between clinician diagnosis and the DMR (DLD) criteria for dementia was 0.92 for both measures of sensitivity and specificity. A significant positive correlation ($r = 0.868$, $p < 0.001$) was found between overall scores on the DSDS and the DMR (DLD), and between DSDS scores and the main subcategories measured by the DMR (DLD) ($r = 0.82$, $p < 0.001$).

Further support for the high sensitivity and specificity of the DSDS was provided by a study of 40 DS adults, aged 26–66 years [46]. The majority of these individuals were described as functioning within the moderate (85%) and mild (12.5%) ranges of

ID. Baseline and 2-year follow-up assessments with DSDS were compared with a clinician's diagnosis of dementia using ICD-10 criteria. At baseline, values of sensitivity and specificity were 58% and 96%, respectively. Sensitivity increased to 75% at 2-year follow-up, and specificity remained at 96%. Thus, relative to a single administration, the diagnostic accuracy of the DSDS increased with repeat assessment. The disparity observed between the clinician's diagnosis and the DSDS rating occurred mostly in the cases of high-functioning individuals (mild to moderate range of ID), who showed early symptoms of dementia. But as dementia progressed from middle to late stages, there was a high agreement between the clinician's diagnosis and the DSDS.

Huxley and colleagues [46] argued that the DSDS has a lower diagnostic sensitivity for high-functioning individuals because it was originally designed to assess adults whose abilities fall within the severe and profound ranges of ID. However, with high-functioning individuals, caregivers may not notice the early signs of dementia because the initial symptoms are often indistinct. Oliver and Holland [47] conducted a review of several case reports of DS adults with Alzheimer's neuropathology and found that over 50% of the individuals had vague symptoms including depression, lethargy, and apathy. Evenhuis [48] similarly described symptoms of apathy, withdrawal, loss of self-help skills, and daytime sleepiness in a sample of adults with DS with early-stage dementia. These behavioral changes may be overlooked by caregivers, especially if contact with the rated person is infrequent. Also, most individuals with mild to moderate ID have attained a certain level of education and skill development, and therefore in the early stages of dementia they likely have the ability to compensate for skill loss, compared to their lower functioning peers. Research suggests that level of cognitive functioning may influence the expression of DAD in persons with DS and, similar to individuals without ID, higher functioning individuals with DS may experience a deferral of DAD symptoms [40].

In a study by Shultz and colleagues [42] compared the DSDS with a number of other neuropsychological assessments: the DMR (DLD), the Reiss Scale, the Shultz Mini Mental Status Exam, and the Paired-Associate Learning Task. The authors investigated the relative efficacy of each test to differentiate demented from non-demented individuals. The participants were 38 adults (45% female and 55% male), between 45 and 74 years of age, with Intelligence Quotient (IQ) scores ranging from 20 to 71. Sixty-eight percent of participants had a diagnosis of DS. The participants were assigned to one of two groups based on a clinician's diagnosis of dementia or absence of dementia using DSM-IV [39] or ICD-10 [44] criteria. The groups were matched on the following variables in order of priority: diagnosis of DS, age (within a range of 5 years), and IQ level (assessed 5 years prior to the study) within a 15-point range. The results showed that the DSDS and DMR (DLD) significantly differentiated between the two groups. For both of these tests, scores were not significantly related to age, gender, IQ or the presence or absence of DS. The DSDS showed a sensitivity of 0.65 and a specificity of 1.0, whereas for the DMR (DLD) the corresponding values were 0.65 and 0.93, respectively. The positive predictive power for the DSDS was 1.0 and its negative predictive power was 0.76. For the DMR (DLD) a lower positive (0.92) and negative (0.70) predictive power was reported. Based on these findings the investigators concluded that the DSDS and the DMR (DLD) are

both "useful in distinguishing between groups with and without dementia; and it is difficult to state simply which instrument was more effective." The slightly better ability of the DMR (DLD) to discriminate between the two groups was attributed to the relatively high proportion of high-functioning individuals in the sample.

While sensitivity and specificity are widely used measures of test validity, some investigators are skeptical about these indices. Ball and colleagues [11] argued that comparing a clinician's diagnosis with a screening test is a potentially problematic procedure. First there is no "gold standard tool to diagnose dementia in DS. Second, clinicians make clinical decisions using broadly the same assessment methods as these screening instruments" (p. 614), and thus high levels of agreement are likely, whether or not the assessments are valid. It is worth pointing out that the DSDS is not classified as a "screening tool" but a diagnostic instrument that was developed only after 8 years of longitudinal data were available on dozens of cases followed long enough to confirm progressive dementia (DAD) or not. Moreover, psychometric studies done on an independent sample from a different province were also done prior to its publication. These aspects do not make the DSDS a "gold standard," but do support its classification as a diagnostic tool, not a "screening" tool.

The DSDS in Neuropsychological Assessment

Alyward and colleagues [49] proposed that assessment of dementia in adults with DS requires the use of both caregiver interviews and direct assessment with psychometric instruments. To promote "state-of-the-art diagnostic practices and information exchange between clinicians and researchers," Burt and Aylward [50] recommended a battery of neuropsychological and adaptive behavior scales to be administered along with DSDS and/or other interview-based instruments. In response, the diagnostic sensitivity of a neuropsychological test battery in detecting dementia in adults with DS was evaluated [40]. The test battery consisted of Information and Orientation Questions, Block Design Test [51], Fuld Object Memory Evaluation [52], Grocery List, Boston Naming Test [53], Peabody Picture Vocabulary Test—Revised [54], and Test of Apraxia. The tests were administered to 35 individuals with DS to compare the group performance of older people with dementia (age 40–59 years), older people without dementia (age 40–66 years), and younger people without dementia (age 28–39 years). Dementia status of the participants was determined based on the DSDS diagnostic criteria. Participants in all three groups were within the moderate range of verbal ability. The most sensitive measures of dementia-related decline in the test battery were the Information and Orientation Questions and the Fuld Object Memory Evaluation. However, these neuropsychological tests could not be used in adults with ID in the profound range or many in the severe range. What instrument could be used with persons in the lower cognitive ranges? The DSDS is one such instrument as it was standardized on DS adults 76% of whom were in the severe or profound range plus the reliability—validity study done on a different group of DS adults 90% of whom were in the severe or profound range of ID.

Stanton and Coetzee [55] reported that the DSDS is a useful scale to include in a battery of tests to assess dementia in people with ID. Acquilano and colleagues [56] also included the DSDS in a battery of assessment tools for older adults with ID, and they mentioned that the DSDS is "sensitive for behavioral changes in the profound range of ID due to the manner of scoring" (p. 199).

Krinsky-McHale and colleagues [57] investigated age-related changes in memory functions relative to changes in memory that occur with early-stage dementia in DS. Eighty-five individuals with mild to moderate ID were administered a modified version of the Selective Reminding Test (SRT) [58]. The participants were first tested with the SRT when they entered the study (baseline) and subsequently annually. Among the participants with DS, 14 cases (ten females and four males) were diagnosed by a physician as having dementia. The DSDS was completed for 13 of these individuals. Memory decline for the dementia group exceeded the decline expected with normal aging and was steeper than the decline exhibited by members of the non-demented group. In the dementia group, for 85% of the cases memory decline occurred several years before the DSDS criteria for early-stage dementia were met, or when the physician made the diagnosis of dementia. Furthermore, in the majority of cases, memory decline preceded other symptoms of dementia by more than 1 year, and in some participants in more than 3 years. Thus, for early identification of dementia in *persons with mild to moderate ID*, test batteries should incorporate measures of memory in addition to caregiver instruments and other tests in order to evaluate multiple cognitive domains. Much earlier, Gedye [10] also advised that, with higher functioning adults with ID, psychologists using the DSDS should also "give tests to measure memory, language, visual-spatial skills and so on" in the DSDS Manual (p. 19).

Published Studies That Used the DSDS

A summary of studies that used the DSDS is presented in Table 4.3.

Advantages and Disadvantages of the DSDS

In the past two decades, a considerable amount of research has been dedicated to evaluate the efficacy of the DSDS. The psychometric properties of the DSDS are now well established. The merit of the scale is in its ability to detect and diagnose dementia in adults with DS, and to distinguish functional decline from other conditions that mimic the clinical symptoms of dementia. Another strength of the DSDS is that it allows for rating severity of dementia by identifying early, middle, or late stages. By tracking the progression of functional decline, caregivers can plan for changing support needs, and physicians have the objective means to evaluate the efficacy of pharmacological interventions designed to slow and abate the clinical

Table 4.3 Summary of studies employing the DSDS to detect dementia in adults with ID

Aylward et al. [59]	DSDS used to confirm MRI diagnosis of DAD
Burt and Aylward [50]	DSDS was found to be useful with adults with ID of etiologies other than DS; lists strengths and weaknesses of scale
Burt et al. [60]	DSDS used to aid in identification of dementia in cross-sectional design of aging in DS adults
Deb and Braganza [43]	Good positive correlation found between DSDS and DMR (DLD) scores, and between DSDS and psychiatrist ratings
Devenny et al. [61]	DSDS used to classify severity of dementia
Devenny et al. [62]	DSDS used to support diagnosis of DAD
Huxley et al. [46]	DSDS scores from baseline and 2-year follow-up were compared; accuracy of diagnosis improved with repeat assessment when dementia progresses
Huxley et al. [63]	DSDS used to assess dementia status in DS adults being evaluated for frequency and severity of challenging behaviors
Jozsvai et al. [41]	DSDS used in conjunction with neuropsychological test battery to detect presence of dementia in DS adults
Kojima et al. [64]	DSDS was translated for use in Japan; stages of dementia were evaluated and compared to prevalence rates from previous studies
Krinsky-McHale et al. [57]	DSDS used to identify early-stage dementia status
Lott and colleagues [65, 66]	DSDS used to monitor changes following the administration of donepezil in DS-DAD adults
Nelson et al. [67]	DSDS used as a criterion measure of dementia; research assistants were trained to administer and score DSDS via videotaped instruction
Shoumitro et al. [68]	DSDS used to support ICD-10 diagnosis of dementia in a study assessing the role of apolipoprotein E gene in DS-DAD
Shultz et al. [42]	DSDS and DMR (DLD) were highly correlated; DSDS was useful discriminating dementia groups
Strydom and Hassiotis [15]	Reviews the properties of the DSDS, including sensitivity and specificity in single assessments
Temple et al. [40]	DSDS scores were combined with scores from a neuropsychological test battery to assign diagnosis and to code for symptom severity
Kartakis [69]	DSDS scores were used to diagnose probable dementia; naming tests were then used to compare differences in naming errors to DSDS diagnosis

signs of dementia. The most frequent criticism of the DSDS is related to its reduced sensitivity to detect the earliest signs of dementia in individuals with mild to moderate ranges of ID. But this is not surprising for at the onset of dementia high-functioning individuals with DS, similar to individuals without ID, are often able to compensate for some loss of skills. More importantly, the DSDS was designed principally for ID adults in the severe or profound range and was standardized mostly on adults in those ranges, not in the mild and moderate range. Thus, it is no surprise if a scale is less sensitive in an area that it never claimed to cover. The strengths and weaknesses of the scale, as cited in the research literature, are summarized in Table 4.4.

Table 4.4 Advantages and disadvantages of the DSDS

Advantages
• Useful in detecting presence of dementia in DS adults
• Differentiates typical from atypical functioning and records duration of symptoms
• Does not depend on direct patient participation
• Includes analysis of item patterns and screening questions for differential diagnosis
• Evaluates early, middle, and late stages of dementia
• Good sensitivity and specificity
• High interrater reliability
• Can be used in low-functioning adults (severe or profound ID), those with little or no speech, and/or those in late-stage dementia when other instruments are unsuitable
• Allows for diagnosis on initial assessment because it focuses on losses at time of assessment, unlike other instruments currently available
• Requires administration and interpretation by a clinically trained professional which reduces the risk of false-positive and false-negative diagnostic errors
• Designed for tracking losses and recovery over time
Disadvantages
• Reduced sensitivity for mild and moderate ID ranges
• Recommended that only psychologists or psychometrists administer the scale
• Two reliable informants are recommended (not always practical)
• Scoring system yields severity rating but is not simple
• Relies on retrospective data, in that informants are required to compare current to previous levels of functioning

Recent research suggests that repeated neuropsychological testing combined with caregiver interview scales is the most promising approach to assess dementia in high-functioning individuals with DS. Improving diagnostic accuracy may lead future research to develop age-appropriate test norms for the DS population which are then used to evaluate the efficacy of currently available psychometric instruments.

Summary

The DSDS is an informant-based instrument that was designed principally for assessing dementia adults in the severe or profound range of intelligence, but has also been found useful for assessing adults in the mild or moderate range of intelligence. It has good psychometric properties as confirmed independently by other researchers. The scoring method provides a rating of severity, identifies when an individual progressed from one stage to another, and facilitates tracking recovery from reversible dementia or during treatment studies. The scale incorporates features to facilitate differential diagnosis. It is used by psychologists around the world in at least 19 countries. The restriction on it for use only by psychologists (and psychometrists) is intended to reduce misdiagnosis of intellectual decline by people

untrained in assessing intelligence. The DSDS can detect dementia in adults with and without DS and can distinguish functional decline from other conditions that mimic the clinical symptoms of dementia.

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Chapter 5
The Dyspraxia Scale for Adults with Down Syndrome

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Introduction

Dyspraxia consists of a partial loss of the ability to perform purposeful or skilled motor acts in the absence of paralysis, sensory loss, abnormal posture or tone, abnormal involuntary movements, incoordination, poor comprehension, or inattention [1]. The existence of dyspraxia is usually tested by having the patient perform some motor act on command or by imitation.

Dyspraxia is seen in Alzheimer's disease (AD) as well as in other dementias. It is related to disease severity and is particularly apparent in the mild to moderate stages of dementia, with about 10% of those with MCI also demonstrating some dyspraxia [2]. It has been recognized in persons with Down syndrome (DS) [3] who also have changes in language and communication [4] reduced speech output and gait deterioration [5], bradykinesia [6] and difficulty with walking unaided [7] and the presence of dyspraxia has been associated with AD pathology in Down syndrome [8].

The ability to measure disease severity with the Dyspraxia Scale in persons with DS may reflect the opportunity to overcome the challenges of assessment in this population [9–12] for whom there are many barriers to cognitive evaluation [13].

[†]Author was deceased at the time of publication. See Obituary p303–304.

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Rationale

The purpose of the Dyspraxia Scale for Adults with Down Syndrome is to provide a research tool for the evaluation of simple sequences of voluntary movements expected to deteriorate with the onset and progression of dementia among persons at all levels of premorbid intellectual disability (ID). The psychometric properties of the scale suggest it may be useful for longitudinal research studies. The scale also holds promise as a primary outcome measure for measuring changes in cognitive functions in clinical trials involving aging persons with DS. It taps the abilities to perform simple sequences of highly practiced voluntary movements which are involved in the skills of daily living. However, it does not depend on verbal and communication skills which would normally require a level of intellectual function outside the range of perhaps as much as one-third of all individuals with DS.

Background

The Dyspraxia Scale for Adults with Down Syndrome was an outgrowth of experience with the Western Aphasia Battery (WAB) developed by Kertesz and his associates at the University of Western Ontario in London, Ontario [14]. The WAB was designed to evaluate praxis in patients suffering from strokes [15] and later for hospitalized patients with clinical diagnoses of DAD [16]. Details are provided elsewhere of the adaptation of the WAB into a 48-item assessment tool called the Video-recorded Home Behavioral Assessment (VHB) [17, 18]. The VHB was used as the primary outcome measure in a clinical trial conducted to evaluate the effectiveness of intramuscular injections of desferrioxamine in slowing the decline in cognitive functions in patients with moderate severity DAD over a 2-year treatment period [17]. The VHB was used as the starting point for the development of the Dyspraxia Scale for Adults with Down Syndrome.

Dyspraxia Scale Construction

Several criteria were employed in the design of the Dyspraxia Scale for Adults with Down Syndrome. Each item selected for the Dyspraxia Scale had to meet the following criteria [1]. It required only a few seconds (2–5″) to perform on verbal request [4]. It was easy to administer [5]. It was easy to score [6]. It was easy to record permanently on video-tape [7]. It consisted of a sample of behavior which would normally be expected to occur in the daily life of the individual [8]. It could be easily modeled or demonstrated by the Examiner [9]. It was age appropriate [10]. It possessed adequate psychometric properties. The Scale was not designed as a speed test. Thus, no timed items were included nor were there any penalties for slow

responses. The overall strategy was to construct a scale that would reflect the best possible performance under optimal conditions from individuals being examined. Simple instructions were used. The evaluations were conducted in environments with minimal stress, such as the individual's group home, shared apartment, workshop, day treatment center, or an office which was most familiar to the individual being tested. Scoring had to be straightforward (pass or fail), response definitions had to be explicit and unambiguous. Scoring by students or direct care staff had to be easy and reliable. Training of Examiners had to be brief but effective enough to meet a high standard set by an experienced Examiner. Items were also limited to those which required minimal verbal skills, language comprehension, and which could be performed by following simple verbal commands or by imitation of the Examiner. The aim was to create a scale that would be useful throughout the course from early to advanced DAD for individuals with levels of premorbid ID ranging from mild to profound.

The structure and scoring methods for the Dyspraxia Scale for Adults with Down Syndrome are similar to those of the VHB. The Scale is divided into three parts. Scores range from 4 (maximum) to 0 (minimum) for each item. It is recommended that *Z* scores be calculated based on means and standard deviations for each part of the Scale and for a total score to permit comparisons with *Z* scores obtained on other tests by the individuals being examined. See the report by Dalton and his colleagues [19] which documents the value of using *Z* scores when other tests are used alongside with the Dyspraxia Scale for Adults with Down Syndrome.

Description and Administration of the Dyspraxia Scale for Adults with Down Syndrome

Test Materials

The test materials used for Part 1, items #1–10 require no special test materials. It is important for safety reasons to provide something that the individual can lean on for support during attempts to perform the leg lift items (#11 and 12) such as a desk, cabinet, or chair. Items #14–20 require a sheet of white paper (letter size), pencil, scissors (medium size), a paper clip (1.75″ or 4.6 cm slightly bent to facilitate handling), three dimes, a small jar with screw-cap lid and a large, yellow, baseball cap. The materials for Part 2 (items 27–40) consist of a red silk rose with a 12″ semirigid plastic stem and two plastic green leaves attached 4″ below the flower, a 4″ black plastic comb, a packaged toothbrush (adult size), a teaspoon (white plastic), a hammer (small, 10″ handle), a medium-sized padlock (about 1″ diameter) with key, a one-ounce jam jar with lid, pair of cotton garden gloves with elasticized wrist (large size), standard letter-size white typing paper. These test items should be kept in a convenient briefcase or similar container on a chair beside the Examiner. The coins used in the Coin Task of Part 3 (test items #59–62) consist of two pennies, two

nickels, two quarters, and two dimes. When used in non-United States or Canada locations, coins of the appropriate size and familiarity to persons living in these countries should be substituted for US coins. During test administration the test case containing the test materials can be placed on a chair within easy reach of the Examiner.

Detailed Scoring of the Dyspraxia Scale for Adults with Down Syndrome

Two methods were adopted for scoring. The first gives credit of 4 points for any successful response to each item, with or without "prompting," and "0" for failure on the item with or without prompting. The second method includes partial scores using prompting. Prompting consists of a graded increase in the amount of "assistance" which is provided by the Examiner to facilitate performance of the correct response by the individual being tested following a failure to perform correctly using simple verbal instructions repeated only once or twice. Prompting reduces the risk of incorrectly giving someone a "0" score for reasons unrelated to impaired praxis. This is achieved by reducing the dependence on verbal comprehension of instructions and minimizing the impact of sensory impairments, particularly hearing losses. Partial scores for incomplete responses using prompting methods are credited as follows:

4 points: A 4-point score is given for a correct response on request without any additional verbal prompts, imitation or modeling, or any form of physical assistance by the Examiner. Four points are assigned if the person correctly completes the item following the first or second request within 5–8 s.

3 points: Providing additional verbal cues and verbal hints to the person is referred to as verbal prompting. Successful performance after the use of verbal prompts decreases the score from a maximum of 4 points (unassisted) to 3 points.

2 points: Failure to obtain a correct response with verbal prompts signals the Examiner to use the next level of prompting that is, modeling. Successful performance by the individual following a modeling prompt is assigned a score of 2 points. A modeling prompt is a display by the Examiner of how the correct response should be executed. Modeling is performed when the previous verbal and gestured prompts have failed to elicit the requested behavior. The modeling is accompanied by the following verbal remarks: "Mr./Mrs…, watch me … (e.g., make a fist, salute, etc.). Now, you do it, just like I did."

1 point: If modeling fails, then the Examiner uses "physical prompting." Physical prompting is a form of "hands-on" assistance provided by the Examiner to determine whether or not the person can perform the requested item with the addition of proprioceptive and tactile cues associated with passive movement. It is used when previous prompts have failed. It represents an attempt to make the task as easy as possible by providing the maximum number of visual and auditory cues now

combined with tactile/proprioceptive cues as well. Three types of physical assistance are defined and used: (1). hand-over-hand in which the Examiner may place his/her hand over the person's hand that is holding the lid of the jar and help the person to turn the lid passively above the hand holding the open jar. (2) Moving the person in the situation requiring standing, sitting, or walking. The Examiner may place his/her hand under the person's elbow to provide support in standing up or sitting down. (3) Doing something for the person. Following the physical prompt the Examiner removes the contact and observes whether or not the person continues with the task to successful completion. A score of l point is given if the person can perform on his/her own.

0 points: Two attempts are made using physical prompting before discontinuation of the item and assignment of a score of 0 points. The individual must seem to be totally unresponsive, uncooperative, unable or unwilling to perform the required response.

Scoring Sheet

The scoring sheet (see Appendix C) is divided into columns displaying the three parts of the Dyspraxia Scale for Adults with Down Syndrome with abbreviated descriptions of each test item. There is provision for entering the name, sex, date of birth, date of the examination, location, age, and the name of the Examiner.

Detailed Administration

Part 1: Psychomotor Skills

This section consists of 20 test items. Items 1–13 are administered while the individual is standing. Items 14–20 are performed at a desk or table while the Examiner and the participant are seated. All 20 items of Part 1 are scored on the basis of decreasing independence, as defined above. It is important to use verbal approval at the end of each response such as, "That's good," or "that's fine," or "Good work," etc.

- Item 1. "Walking." The person is instructed to walk toward the Examiner (or toward a tripod-mounted video camera if one is being used). Score of 4 points for independent walking upon single command or with only 1 or 2 repetitions of the same instruction. Score of 3 points for performance with verbal prompt of encouragement. Score of 2 points for correct imitation of the model (Examiner) with: "This is what I want you to do." Score of 1 point is given if physical assistance is used such as supporting arm and elbow while providing verbal encouragement with, "If I help you a little, try to walk toward the desk (or camera)." A person who uses a cane or walker is automatically scored 1 point. Score of 0 points if the

person is unable, unwilling or refuses to complete the item. An individual who routinely uses a wheel chair automatically scores 0 points on this item.

Following item 1, the next 12 items are administered while the individual is standing. The scoring is the same as for item 1.

- Item 2. “Standing.” The person must be able to stand unassisted for 2–5 s.
- Item 3. “Look up.” The individual must use his/her eyes or head to look up. A verbal prompt such as, “Look up at the ceiling,” reduces the score to 3 points.
- Item 4. “Bend your head.” The individual must lower his/her head toward the floor upon command for a score of 4 points. Use of the verbal prompt such as, “bend your head down,” or “look at the floor,” reduces the score to 3 points.
- Item 5. “Bow from the waist.” The individual must bend slightly (2–3 in.) or completely from the waist. The use of the verbal prompt such as, “take a bow,” reduces the score to 3 points
- Item 6. “Clap your hands.” The individual must bring his/her hands together to indicate in front of the Examiner.
- Item 7. “Lift one arm over your head.” A 4-point response includes lifting the arm straight up in the air near the head, placing the hand on the head, raising the hand behind the head.
- Item 8. “Lift the other arm over your head.” A 4-point response is for behavior similar to that shown during the preceding item with the other arm.
- Item 9. “Turn your head to one side.” The individual needs to keep his/her body facing forward, toward the Examiner (camera), while he/she turns the head to the side. Idiosyncratic behavior such as rotating the torso is acceptable. If the person is unable to perform this item correctly with one or two repetitions of the command, a correct, 3-point, response can usually be emitted with the verbal prompt, “look at the wall.”
- Item 10. “Turn your head to the other side.” The individual needs to keep his/her body facing forward toward the Examiner (camera) and turn the head to the side opposite to the one in Item 9. Again idiosyncratic variation is acceptable.
- Item 11. “Lift one leg.” The individual should raise the leg off the floor and hold it in the air for at least 2 s. A score of 1 point is given if the person needs to place his hand a chair, desk, or cabinet provided for this purpose.
- Item 12. “Lift the other leg.” The individual must raise the other leg and hold in the air for about 2 s for a full score of 4 points. A score of 1 point is given if the person places his hand for support on a chair, desk or cabinet intended for this purpose.
- Item 13. “Sitting.” Upon completion of item 12, the individual is requested to sit at the table or desk (pointing by Examiner in the appropriate direction may be helpful) where the remainder of the test items are presented. This item is scored on the basis of the level of independence required to comply.
- Item 14. “Draw a circle.” The individual is provided with a standard letter-size sheet of white paper and a pencil. A circle anywhere from 4 to 15 cm in diameter is acceptable for 4-point score as long as the response is performed with verbal

command only with one or two repetitions. Three points are scored if verbal suggestions are required. If the Examiner needs to model the correct response a fresh sheet should be placed in front of the individual. The Examiner slowly draws a circle of about 10 cm (about 4 in.) in diameter in the upper half of the page and hands over the pencil to the individual with the instruction, "Now you do it, just like that, just like I did." The sheet is removed and stored as data for future analysis while the Examiner provides brief verbal approval.

- Item 15. "Draw a straight line." A new sheet of paper is placed in front of the individual. Instructions and scoring for this item are similar to item 14. At the end of the item, both the pencil and the paper are retrieved and put aside while the Examiner provides verbal approval to the individual.
- Item 16. "Clip two sheets." Two new letter-sized sheets of paper are placed side-by-side in front of the individual who is handed a large paper clip already somewhat bent outwards along one length to facilitate response. The instructions are: "Now, clip these sheets together with the clip." The individual is allowed about 30–45 s to complete the task. If necessary then, verbal prompts are provided such as: "put the sheets together. Put the clip on one sheet first, then put the other sheet under it." If modeling is required, the two sheets and clip are retrieved from the individual and the Examiner slowly demonstrates how to perform the task. Hand-over-hand assistance on this task is sometimes difficult. At the end of this item the materials are retrieved and set aside while the Examiner says, "That's good. That's fine."
- Item 17. "Cut this paper sheet." A new letter-sized sheet of white paper is placed on the table along with a medium-sized pair of scissors with the instructions, "Now, I want you to cut this sheet of paper with the scissors." A score of 4 points is given if the individual performs correctly within 30–60 s. The sheet can be cut lengthwise, sideways, in half, or in one- to two-thirds fractions as long as no further prompting other than one or two repetitions of the same instruction. At the time the materials are retrieved the Examiner says, "Good. That's fine."
- Item 18. "Three coins with one hand." A small (1 ounce) transparent jam jar is placed in front of the individual with three coins beside the jar (three dimes) with the request. "Please place each of the coins inside the jar." If the individual shifts hands during the task, the person is instructed not to do so. Picking up the coins with the thumb and the fingers or sliding the coins to the edge of the table before picking them up are acceptable responses. At the end of this item the Examiner says, "That's fine. Good," and empties the coins back onto the table with the jar next to the coins and proceeds to item 19.
- Item 19. "Three coins other hand." A repeat of item 18 but now the person must successfully perform with the other hand. Trial is terminated with verbal approval by the Examiner while the items are removed and set aside at the same time.
- Item 20. "Put on the cap/take it off." An adult size baseball cap is placed on the table in front of the individual with the instruction: "Put the cap on your head." After completion, the Examiner says, "Yes. That's good. Now, please take it off." The cap is set aside while the Examiner again provides verbal approval.

Part 2: Apraxia

These items are also administered while the person is seated at a desk in an arm-chair or in a wheel chair. For bed-ridden patients who are awake and reasonably cooperative, every effort should be made to obtain responses to as many test items as possible to help in the identification of preserved skills. About 2–4 s are allowed for each response. Each response is immediately followed by brief verbal approval from the Examiner with remarks such as those used with the previous items. Rules for defining and scoring independent performance, verbal, physical prompting, and modeling are the same as described earlier. Verbal approval after each response is also provided for each item.

- Item 21. "Make a fist." The individual must independently clench his fingers and thumb into a fist. The hand should be off the table. The shape of the fist should be roughly "rounded" with allowance for various idiosyncratic positions of the thumb.
- Item 22. "Salute." The person is required to independently raise either hand to his forehead with or without the thumb tucked in the palm. Once the hand is clearly positioned the person then should swing the hand out and away from the head for a full score. Allowance should be made for individual differences in execution.
- Item 23. "Wave good-bye." The person must hold either the right or left hand in the air and wave the hand from side to side or in an up and down motion independently for a full score.
- Item 24. "Scratch your head." The person must independently raise the hand to the head and use fingers to scratch back and forwards at least once for full score. Allowances for individual differences in response topography should be made.
- Item 25. "Snap your fingers." The person must independently snap the middle finger against the thumb with or without a sound.
- Item 26. "Close your eyes." The person must independently shut either of his eyes so that no part of the eyeball is visible. The eye closure should be visible to the Examiner (and/or camera).
- Item 27. "Sniff a flower." The person must independently hold the flower (silk rose with petals and 8″ stem) within 1–2 in. of the nose and appear to sniff the flower by holding it there momentarily.
- Item 28. "Use a comb." The person must independently hold the comb and make the appropriate stroking movements over the head. It is not necessary that the comb go through the hair. A new comb should be used with each individual being tested.
- Item 29. "Use a toothbrush." The person must independently hold the toothbrush in either hand and make either up and down or side to side movements with the brush held with the bristles toward the face near the mouth. A new brush should be presented with each new participant.
- Item 30. "Use a spoon." The person must independently pick up the spoon (soup size) from the table with either hand and bring it up toward the mouth. The person may or may not make a scooping motion with the spoon for a full score.
- Item 31. "Use a hammer." The person must pick up the hammer placed on the table in front of him/her and make one or two downward motions with the head of the hammer pointed downwards.

- Item 32. “Use a key.” A small key is placed in front of the person. The person must independently pick up the small key from the table and make turning motions in the air to pretend opening a door.
- Item 33. “Open a jar.” After a small (1 ounce) jar with a screw cap is placed in front of the person, a full score is obtained if the person independently unscrews the lid using both hands, following verbal request only.
- Item 34. “Close the jar.” The person gets a full score for replacing and screwing the lid back on the small jar without prompting. If placed incorrectly or too loosely the Examiner then proceeds with the prompting procedures outlined above and scores accordingly.
- Item 35. “Put on the (right hand) glove.” An adult-size right-hand glove is placed in front of the person with the verbal request, “please put this on.” Full credit is given if the person responds correctly without prompting. All fingers should be in their proper place. This is checked by the Examiner by reaching over and feeling through the glove for the finger positions. The person is then asked to remove the glove.
- Item 36. “Put on the (left hand) glove.” The person is required to independently put on the left hand glove after removing the glove from the right hand following performance of item 35.
- Item 37. “Unlock a padlock.” A small padlock and key are placed side-by-side and the person gets a full score if he/she independently picks up both items, inserts the key in the key hole and turns the key until the lock opens. An allowance of about 30 s is given before giving additional prompting procedures.
- Item 38. “Lock the padlock.” The Examiner presents the lock with the key still inserted in position and with the swivel open. The person is required to independently swivel the catch until it is over the hole of the padlock then to press down firmly until the catch snaps shut. About 30 s are allowed before introducing prompting procedures.
- Item 39. “Fold a sheet of paper.” The person must fold the paper (8.5″ by 11″ ordinary typing paper) neatly in half, with the edges of both sides meeting within 2–3 cm of each other. There must be a visible crease in the fold of the paper produced by the appropriate hand movement. The direction of the fold makes no difference in the scoring.
- Item 40. “Fold the paper again.” Using the same folded paper, the person gets a full score for independently making a second fold in the sheet following instruction to do so without additional prompting.

Part 3: Body Parts/Coin Task

This part of the scale consists of 18 items adapted from the WAB and 4-coin identification tasks. Items 41–58 involve pointing to various parts of the body in response to simple verbal instructions. Each response is scored as either correct (4 points) or incorrect-unable-unwilling (0 points). The instructions can be repeated up to two times for a complete score. No prompting methods are employed for these items.

Therefore, there are no partial scores for items in Part 3. However, as for all of the previous items, each response is immediately followed by brief verbal approval from the Examiner such as, "Good," or "that's fine," etc. These contingent verbal responses by the Examiner provide immediate feedback to the individual and also provide a cue indicating termination of the trial for each item, a useful feature if video recordings are involved in the test session.

The Examiner instructs the seated individual as follows: "Mr./Ms…, I am going to ask you to point to different parts of your body." The person is then requested to: "Point to your ear (item 41), nose (item 42), eye (item 43), chest (item 44), neck (item 45), chin (item 46), thumb (item 47), ring finger (item 48), index finger (item 49), little finger (item 50), middle finger (item 51), right ear (item 52), right shoulder (item 53), left knee (item 54), left ankle (item 55), right wrist (item 56), left elbow (item 57), and right cheek (item 58).

Immediately after item #58 of Part 3, the Examiner places eight coins (two pennies, nickels, dimes, and two quarters) in a random order on a letter-sized sheet of white paper within easy reach of the person. The person is then asked to, "Please give me a…" penny (item 59), nickel (item 60), quarter (item 61), and a dime (item 62). Each response is scored as either correct (4 points) or incorrect-unable-unwilling (0 points). The instructions can be repeated once or twice for a full score. No prompting methods are employed for these items. After the individual places a coin in the Examiner's hand, the coin is replaced on the sheet of paper and the coins are briefly shuffled on the page. Each response is immediately followed by brief verbal approval from the Examiner such as "Good," or "That's fine," etc.

Psychometric Properties of the Dyspraxia Scale for Adults with Down Syndrome

Validity and Reliability

Do the behaviors which are sampled in the Dyspraxia Scale for Adults with Down Syndrome constitute a representative sample? Do they include enough items or are there others which would be more intimately associated with the onset of DAD among persons with DS? It is not possible to answer these questions directly without postmortem studies of brain specimens from individuals who have died after performing the test as it is presently constructed. Thus, interpretation of the test scores obtained using this test must be cautious. It is assumed that the relatively large number of items in the Scale provides some protection against the likelihood of obtaining invalid measures. False conclusions can be minimized by careful adherence to a follow-up strategy in which baseline scores are compared with subsequent performances. Confidence in the conclusions is substantially increased by evidence of deterioration in scores. Improvements in scores over a short period of time could reflect a "practice effect" which is a characteristic of other tests of cognitive functions as suggested by Sano and her colleagues [20]. By the same token

deterioration in performance scores at follow-ups are more likely to reflect true deterioration rather than fatigue, inattention, or lapses in concentration by the person being examined because of the relatively large number of test items.

Validity

The VHB, the predecessor of the Dyspraxia Scale for Adults with Down Syndrome, revealed an average decline in overall scores of 39.4%, from an average of 91.7% correct at baseline to 52.3% correct at a 2-year follow-up among 48 patients from the general aging population with a clinical diagnosis of mild to moderate severity DAD [17]. This not only represents a clinically significant change but also demonstrates validation of the VHB against clinical diagnoses. Demonstration of similar changes in a group of persons with DS aged 54.4 years (SD = 2.62, min/max = 50/58 years) with a mean premorbid IQ = 39 points (SD = 14.1, min/max = 29/49 points) at the start, from a mean of 77.0% (SD = 6.35) correct dyspraxia items to 62.0% (SD = 28.73) correct over a 3.4-year period provides indirect evidence of the validity of the Dyspraxia Scale for Adults with Down Syndrome in persons with DS [19] because 48 of the 62 items of the Dyspraxia Scale were the same as in the VHB. However, since clinical diagnoses were not available for the participants with DS in this study the declines in dyspraxia scores could also be attributed to so-called "normal aging" rather than to early DAD. However, the "normal aging" explanation of the results with persons with DS appears unlikely because the changes observed in the aging DS group occurred over a relatively short time period (about 42 months) and a group of "elderly" persons with mental retardation without DS (mean age = 72 years, min/max = 71/84 years at the start) in a separate study [21] with the Dyspraxia Scale for Adults with Down Syndrome showed no changes in scores over the same 3.4-year test period. Moreover, Devenny and her colleagues [22] have described as "normal aging" a slow decline of less than 1% per year in the performances on a test of selective reminding and a speeded psychomotor task across test times of up to 6 years in persons with DS older than 50 years of age. Needless to say, ultimate validation of the Dyspraxia Scale for Adults with Down Syndrome as a test of DAD in persons with DS needs the support of good clinical diagnoses and neuropathological diagnoses at postmortem.

Test–Retest Reliability

In one study [23], 15 individuals (10 men, 5 women) including 9 with ID without DS and 6 with DS, with an average age of 61.9 years (SD = 14.26, min/max = 35/80 years), participated in a test–retest evaluation of the 62-item Dyspraxia Scale for Adults with Down Syndrome. Each individual was tested by the same Examiner on two occasions separated by 8 weeks. The percent agreement between first test and second test on an item-by-item basis was calculated for each participant. This analysis showed

an overall average percent agreement on 84.4% of the items (SD = 11.30, min/max = 69.4/100%). This level of agreement compared favorably with the results of a similar analysis of item-by-item agreement of 78% which was obtained using the 48-item version of the Dyspraxia Scale for Adults with Down Syndrome (the VHB) which was employed in a previously published report involving 48 patients from the general aging population with diagnoses of mild to moderate severity DAD [17].

In the second study [21], the 62-item Dyspraxia Scale for Adults with Down Syndrome was administered twice to 25 individuals within a period of 3–6 weeks by a single Examiner. Test data were collected from 10 men and 6 women with DS (mean age = 46.1 years, SD = 8.12, min/max = 35/58 years) and from 6 men and 3 women with ID without DS (mean age = 71.1 years, SD = 4.71, min/max = 64/80 years). The first test scores for each of the 62 test items were compared with the retest scores on the same items for each of the 25 persons in an overall analysis using Statistica version 5.0 Statsoft software. The results indicated that the test–retest correlation exceeded $r = 0.96$ [21], thus indicating a high degree of test–retest reliability for the Dyspraxia Scale for Adults with Down Syndrome.

Test–Retest Reliability Over 3 Years

It is possible that scores on the Dyspraxia Scale for Adults with Down Syndrome would show progressive improvements with repeated testing due to practice effects over time. Scores could also show slow deterioration indicative of "normal aging." These possibilities were examined in an unpublished study described in the *Dyspraxia Scale for Adults with Down Syndrome Manual* [2]. The study was conducted on the Dyspraxia Scale Percent Correct items scores obtained on an annual basis over a 3-year period from 14 adults with DS (eight women and six men) with an average age of 45.9 years (SD = 5.55, min/max = 40/58 years) at the start. All were healthy. None showed any signs suggestive of early DAD as determined by direct care staff familiar with each individual throughout the period of the study. The average percent correct scores from the start, year 1, year 2, and year 3, were, respectively, 82.4% (SD = 7.18, min/max = 72/93), 84.5% (SD = 9.38, min/max = 68/95), 82.8% (SD = 10.24, min/max = 64/97), and 85.5% (SD = 9.64, min/max = 70/99). The results suggested that there were no improvements in scores reflecting a "practice effect" of repeated testing over the 3-year interval. Moreover, the absence of significant deterioration in scores was consistent with the report by Devenny and her colleagues [22] where she describes as "normal aging" the small magnitude of annual (1%) changes in cognitive scores over a 6-year follow-up of adults with DS.

Split-Half Reliability

The first time scores on the 62 items of the Dyspraxia Scale for Adults with Down Syndrome were divided into two sets of 32 scores with even-numbered items in set 1 and odd-numbered items in set 2. This was done for each of 140 adults with DS

which included 109 from the standardization sample plus 31 additional individuals with DS who were referred to a clinic. The data were collected over a 2-year period by several Examiners. A split-half reliability coefficient was calculated using Statistica Version 5.0 Statsoft software. No cases were deleted from the analysis because there were no missing data. The analysis yielded a reliability coefficient of $r = 0.98$ for even versus odd items. These results raise the possibility of constructing alternate forms of the Dyspraxia Scale for Adults with Down Syndrome by using half of the test items in each form.

Internal Consistency: Cronbach's Alpha

Data from two studies were analyzed.

1. **Study 1:** The first time Dyspraxia Scale for Adults with Down Syndrome scores from a group of individuals with DS ($n = 140$) collected at various care provider agencies in rural and suburban New York State were subjected to an item-by-item analysis to determine the contribution of each item to the overall Dyspraxia score. The data from 109 cases in the standardization sample (40 women and 69 men) were combined with data from 31 adults with DS who were referred to a Staten Island clinic for evaluation of possible DAD. Most were diagnosed with DS on the basis of chromosomal studies of blood specimens while the remainder of the individuals were diagnosed on a clinical basis. None had significant mobility or sensory impairments. None had seizures or psychiatric symptoms which were not adequately controlled by medication. Nineteen (26.0%) of the individuals were classified with premorbid mild ID. Ten (13.7%) of the individuals were classified with premorbid profound ID while the remainder had scores either in the moderate or severe range of ID.
2. **Study 2:** Data on the Dyspraxia Scale for Adults with Down Syndrome from 315 persons with DS were assembled and analyzed from seven independent investigators residing in Texas, Staten Island, Boston, Birmingham England, Manhattan, and Saskatoon, Saskatchewan. The analyses included data from 162 women and 153 men ranging in age from 33 to 77 years. Premorbid ID levels were available from the records of 262 of the 316. The sample included those with borderline ($n = 2$), mild ($n = 37$), moderate ($n = 137$), severe ($n = 75$), and profound ($n = 11$) levels of ID, respectively. Clinical diagnoses of DAD were reported for 41 and clinical diagnoses of no-DAD were reported for 57. Diagnoses for the remainder were not available. Table 5.1 is a summary of the results of both studies.

Test item number is shown in the first column. The second column is a short description of each item. The third column shows the mean dyspraxia scores for each of the 62 items from Study 1. The fourth column shows the standard deviation (SD) for each item from Study 1. No similar data were available from Study 2 for this analysis. The item-to-test correlations for each item for Study 1 and for Study 2 are shown in columns 5 and 6. Cronbach's alpha values with each item deleted from the test are shown for Study 1 in column 7 and for Study 2 in column 8. Cronbach's

Table 5.1 Test of item-by-item internal reliability: Study 1 and Study 2

Item no.	Short description of item	Study[a]		Item-to-test correlation		Alpha with deleted item	
		Mean	SD	Study 1	Study 2	Study 1	Study 2
Part 1: Psychomotor skills: while standing							
1	Walking	3.94	0.73	0.50	0.54	0.94	0.98
2	Standing	3.91	0.58	0.51	0.54	0.95	0.98
3	Look up	3.51	1.34	0.73	0.67	0.95	0.98
4	Bend your head	3.52	1.48	0.77	0.72	0.94	0.98
5	Bow from the waist	3.32	1.68	0.77	0.70	0.94	0.98
6	Clap your hands	3.81	1.30	0.79	0.76	0.94	0.98
7	Lift one arm	3.36	1.35	0.77	0.76	0.94	0.98
8	Lift other arm	3.51	1.32	0.82	0.76	0.94	0.98
9	Turn head to one side	3.32	1.37	0.66	0.75	0.95	0.98
10	Turn head to other side	3.49	1.40	0.76	0.78	0.94	0.98
11	Lift one leg	3.65	1.38	0.80	0.76	0.94	0.98
12	Lift other leg	3.79	1.31	0.76	0.73	0.95	0.98
13	Sitting	4.37	1.60	0.52	0.58	0.95	0.98
Part 1: Psychomotor skills: while seated							
14	Draw a circle	3.65	1.40	0.76	0.75	0.94	0.98
15	Draw a straight line	3.53	1.54	0.52	0.69	0.95	0.98
16	Clip two sheets	–	–	–	0.61	–	0.98
17	Cut paper sheet	3.90	1.49	0.66	0.72	0.95	0.98
18	Three coins (one hand)	3.89	1.09	0.58	0.66	0.95	0.98
19	Coins (other hand)	3.78	1.30	0.65	0.71	0.95	0.98
20	Put on cap/take it off	3.95	1.04	0.71	0.67	0.95	0.98
Part 2: Apraxia							
21	Make a fist	3.33	1.52	0.74	0.73	0.95	0.98
22	Salute	3.20	1.55	0.74	0.67	0.95	0.98
23	Wave good-bye	3.68	1.28	0.82	0.76	0.95	0.98
24	Scratch your head	3.61	1.74	0.69	0.76	0.95	0.98
25	Snap your fingers	2.77	2.17	0.51	0.62	0.96	0.98
26	Close your eyes	3.69	1.62	0.42	0.68	0.96	0.98
27	Sniff a flower	3.82	1.40	0.80	0.80	0.95	0.98
28	Use a comb	3.99	0.98	0.68	0.74	0.95	0.98
29	Use a toothbrush	3.85	1.15	0.73	0.74	0.95	0.98
30	Use a spoon	3.76	1.21	0.66	0.77	0.95	0.98
31	Use a hammer	3.74	1.25	0.76	0.82	0.95	0.98
32	Use a key	3.67	1.60	0.80	0.73	0.95	0.98
33	Open a jar	3.96	1.06	0.65	0.75	0.95	0.98
34	Close a jar	3.88	1.20	0.75	0.77	0.95	0.98
35	Put on right glove	3.81	1.42	0.75	0.73	0.95	0.98
36	Put on left glove	3.81	1.43	0.73	0.73	0.95	0.98
37	Unlock padlock	3.54	1.54	0.78	0.77	0.95	0.98
38	Lock padlock	3.39	1.61	0.77	0.76	0.95	0.98

Table 5.1 (continued)

Item no.	Short description of item	Study[a]		Item-to-test correlation		Alpha with deleted item	
		Mean	SD	Study 1	Study 2	Study 1	Study 2
39	Fold a sheet of paper	3.77	1.16	0.78	0.83	0.95	0.98
40	Fold sheet again	3.73	1.43	0.79	0.80	0.95	0.98
Part 3: Body parts and orientation							
41	Point to your ear	3.56	1.76	0.71	0.72	0.92	0.98
42	Point to your nose	3.61	1.71	0.61	0.77	0.93	0.98
43	Point to your eye	3.64	1.79	0.66	0.78	0.93	0.98
44	Point to your chest	3.01	2.36	0.59	0.61	0.93	0.98
45	Point to your neck	3.33	2.11	0.72	0.72	0.92	0.98
46	Point to your chin	3.35	2.33	0.67	0.63	0.92	0.98
47	Point to your thumb	3.41	2.14	0.70	0.65	0.92	0.98
48	Point to your ring finger	1.71	2.89	0.30	0.46	0.93	0.98
49	Point to your index finger	1.54	2.85	0.12	0.35	0.93	0.98
50	Point to your little finger	2.95	2.59	0.67	0.61	0.92	0.98
51	Point to your middle finger	2.13	2.89	0.53	0.51	0.93	0.98
52	Point to your right ear	2.89	2.55	0.57	0.50	0.93	0.98
53	Point to your right shoulder	2.88	2.82	0.63	0.52	0.92	0.98
54	Point to your left knee	2.98	2.79	0.61	0.51	0.93	0.98
55	Point to your left ankle	2.83	2.90	0.69	0.46	0.92	0.98
56	Point to your right wrist	2.34	3.06	0.63	0.43	0.92	0.98
57	Point to your left elbow	3.06	2.74	0.49	0.54	0.93	0.98
58	Point to your right cheek	2.81	2.83	0.52	0.53	0.93	0.98
Part 3: Coin identification task							
59	Give me a penny	2.96	2.45	0.68	0.66	0.92	0.98
60	Give me a nickel	2.86	2.88	0.62	0.55	0.93	0.98
61	Give me a quarter	2.96	2.58	0.60	0.63	0.93	0.98
62	Give me a dime	3.14	2.65	0.75	0.60	0.92	0.98

[a]No comparable data were available for study 2 at the time of this analysis

alpha with item deleted is never less than 0.92 in Study 1 and is 0.98 in Study 2. The very high alpha values for all of the dyspraxia items in Study 2 reflect the impact of the large sample size used for the analysis. When the two studies are viewed side-by-side it is convincingly evident that the Dyspraxia Scale for Adults with Down Syndrome has a very high degree of internal consistency.

Table 5.2 Total variance explained in factor analysis

	Initial eigenvalues		
Component	Total	% of variance	Cumulative %
1	29.811	48.083	48.083
2	2.784	4.491	52.574
3	1.954	3.152	55.726
4	1.586	2.557	58.283
5	1.451	2.340	60.624
6	1.271	2.048	62.673
7	1.113	1.794	64.467
8	1.068	1.722	66.189
9	1.016	1.638	67.828

Factor Analysis

A factor analysis of Dyspraxia Scale for Adults with Down Syndrome data collected from Study 2 involving 315 adults with DS was performed in order to examine some of the structural features of the Scale. Eigenvalues were calculated and estimates of the variance associated with each component were determined. There were nine components that exceeded 1.000. These eigenvalues and the contribution of each component to the total variance are presented in Table 5.2. It can be seen that factor 1 accounted for 48.083% of the variance with factors 2 and 3 contributing 4.491% and 3.152%, respectively. These results are consistent with the idea that the Dyspraxia Scale for Adults with Down Syndrome is a one-dimensional scale.

Standardization: Normative Sample

Few, if any psychological or cognitive tests are effective in measuring functional impairment and deterioration throughout the course of DAD. Every test is likely to have either significant "ceiling effects" where the items are too easy or "floor effects" where the items are too difficult. As the dementing process progresses we must turn to direct observations of simple behaviors which are most likely to be affected by AD [21]. Assessment of simple behaviors was the aim of the Dyspraxia Scale for Adults with Down Syndrome. However, no reference point was available for the test items selected. Therefore, in order to permit cross-test comparisons of changes over time with other cognitive tests it would be useful to know the distribution of dyspraxia scores of individuals with DS whose scores are representative of the general population of persons with DS without DAD. In addition, knowing the distribution and other characteristics of a "normative" sample of individuals with DS can provide a statistical basis for defining abnormality in terms of standard deviation units from the normative mean. A carefully selected sample of individuals was studied for this purpose.

Table 5.3 Descriptive statistics for the dyspraxia standardization sample ($N = 109$)

Statistic	Part 1	Part 2	Part 3	Overall
Mean	87%	88%	60%	78%
SD	13.60	15.27	24.27	15.83
Minimum	45%	20%	0%	23%
Maximum	100%	100%	98%	98%

First time Dyspraxia Scale for Adults with Down Syndrome scores from 122 adults with DS were reviewed for inclusion in a "standardization sample." Dyspraxia scores from 13 individuals were excluded for the following reasons. Four aging individuals (50, 53, 54, and 57 years of age at the start) were excluded (1 woman and 3 men) because they showed significant impairment of learning and memory functions as determined by performances on the Dalton/McMurray Visual Recognition Test [25], a test used to detect the first memory changes associated with DAD in this population. The data from 9 of the 13 individuals (6 men and 3 women, 37–58 years of age) were excluded because 5 were uncooperative, one engaged in self-stimulatory behavior which interfered significantly with test performance, one was too young to be included (age of 12 years), and the Dyspraxia Scale for Adults with Down Syndrome was incomplete for 2 others. Each individual was involved on a regular basis in full- or part-time employment in a sheltered workshop, an industrial setting or day treatment program. There were 40 women and 69 men who met all of these criteria. The mean age of the women was 35.9 years (with SD = 10.48, min/max = 19/58 years) and for the men it was 34.9 years (with SD = 10.25 and min/max = 17/57 years). The mean intelligence quotient (IQ) scores for the women was 44 points (with SD = 15.1, min/max = 17/69 points) and for the men it was 37 points (with SD = 15.3, min/max = 16/66 points). There were no statistically significant differences between the women and men on any of the variables. Consequently, the data for women and men were pooled to permit construction of norms. Conversion to Z scores for each person's performance is done using the familiar equation (percent correct score minus the standardization sample mean) divided by the standardization sample standard deviation for the parts and overall scores. Table 5.3 provides the basic data for conversion of raw dyspraxia scores into standard (Z) scores.

Standardization Sample: Part and Total Correlations

Table 5.4 provides a numerical summary of the Pearson Product Moment correlation coefficients for comparisons between the variables of age, Dyspraxia Scale for Adults with Down Syndrome Part 1, 2, 3, and overall scores. There were no statistically significant relationships between the dyspraxia scores and age. Parts 1 and 2 of the Dyspraxia Scale for Adults with Down Syndrome were more closely correlated with each other than with Part 3, reflecting difference in difficulty or the effect of a different factor. All three parts were highly correlated with the overall score.

Table 5.4 Correlations between age and dyspraxia scores for the standardization sample

Variable	Age (years)	Part 1	Part 2	Part 3
Part 1	−0.127	–		
Part 2	−0.044	+0.778	–	
Part 3	−0.148	+0.598	+0.600	–
Overall	−0.127	+0.862	+0.866	+0.891

Review of Published Research Studies with the Dyspraxia Scale for Adults with Down Syndrome

Dalton and Fedor [24] published the first study describing the development and standardization of the Dyspraxia Scale for Adults with Down Syndrome. The study also tested the hypothesis that older individuals with DS would obtain scores consistent with signs of the onset and progression of dyspraxia when compared with younger adults with DS. Persons with DS, 40 years of age and older showed statically significant deterioration which reflected "preclinical" signs of dementia. An older group with DS with "normal" dyspraxia scores at a mean age of 54.1 years, showed deterioration which began about 3.5 years later. The scores of a group between 40 and 49 years of age were indistinguishable from a younger group between 21 and 39 years of age. The results suggested that the onset of one of the early signs of DAD could be identified at an average age of 57.9 years among persons with DS using the Dyspraxia Scale for Adults with Down Syndrome.

The aim of the same group of researchers in a further study [19] was to determine whether or not there was a specific sequence of cognitive changes over a 3-year period using three different tests. When compared with a young group of persons with DS (17–39 years at the start), an old group of persons with DS (40–58 years at the start) showed small but statistically significant changes over time suggestive of the "preclinical signs" of DAD. When the data were sorted into four subgroups on the basis of age, a more detailed analysis revealed that the subgroup that was 50 years of age and older at the start showed changes in scores which were of a magnitude more clearly indicative of early DAD. Deterioration in learning/memory scores on the Dalton/McMurray Visual Memory Test [25] began at a mean age of 54.2 years, followed later by deterioration in Dyspraxia Scale for Adults with Down Syndrome scores at a mean age of 56.9 years. Deterioration in ratings on a five-part, informant-based, maladaptive behavior rating scale, called the Multidimensional Observational Scale for Elderly Subjects (MOSES) [26] occurred at an intermediate age of 55.0 years. The results provided support for the hypothesis that persons with DS who are 50 years of age and older may develop a specific sequence of functional changes during the early stage of DAD. The possibility that those over 50 years of age showed deterioration because of between-group differences in the prevalence of one or more comorbid conditions which have a predilection for aging persons with DS was ruled out. None had a concurrent report of neurological, psychiatric, sensory or motor disabilities, other conditions and medications. These variables were routinely evaluated annually by knowledgeable nurses using a 35-item health checklist

designed for the purpose [19]. The possibility that deterioration of the old DS subgroup could be due to a lower average level of ID was ruled out. Analysis of the IQ levels of the four subgroups revealed no statistically significant differences between them. The study also illustrated the value of using norms and standard scores (Z) to enhance the usefulness of a variety of tests to evaluate DAD in persons with ID.

Brief Praxis Test

A major challenge to developing therapeutic interventions for cognitive loss and DAD in aging individuals with DS is the selection of appropriate outcome measures. Sano and colleagues [20] describe the development and application of a short version of the Dyspraxia Scale for Adults with Down Syndrome called the Brief Praxis Test (BPT) as a primary outcome measure in a double-blind clinical trial with individuals with DS. Other tests to assess cognition, behavior, and clinical global function based on previous work in DS and in DAD were also used. Measures of cognition included verbal and nonverbal memory, vocabulary, and orientation. An informant-based measure of behavior and function was adapted from the MOSES [26] and the Dementia Questionnaire for Mentally Retarded Persons (DMR/DLD) [27, 28] (Chap. 3) for use with this group. This report also describes initial experiences using these measures with 108 participants (47 women and 61 men recruited into 20 research sites in five different countries) who were enrolled in the clinical trial. A diagnosis of DAD was made independently of the outcome measures. The level of ID was the best estimate of the highest lifetime functioning. The number of individuals at each level of ID was mild ($n = 23$), moderate ($n = 46$), severe ($n = 19$), profound ($n = 9$), and missing ($n = 11$). Mean BPT scores (out of a total possible 80 points) were 65.23 (SD = 12.14) and 58.72 (SD = 15.54), respectively, for those with a clinical diagnosis of no-DAD versus those with a diagnosis of DAD. Analyses of variance revealed highly significant association between BPT scores and level of ID (with $n = 98$, $F = 10.255$, $p < 0.000$) and BPT scores with diagnosis of dementia ($n = 107$, $F = 6.166$, $p < 0.015$).

As in other populations of persons with DAD, verbal learning, memory, and delayed recall scores proved to be highly associated with the presence of dementia in the study participants. With the exception of visual memory and orientation measures (which proved too difficult to use with portions of this cohort), the tests employed proved useful in the assessment of individuals across a range of premorbid levels of ID. The authors conclude that the measures chosen for the assessment of behavior and functional ability and the use of the Clinical Global Impression appear to be appropriate for this population and comparable to instruments that have captured pharmacological benefits in other disease groups.

The BPT has been used in several studies (See Table 5.5) including the published trial of vitamin E in the treatment of older adults with DS [29]. In that study the DS cohort demonstrated the predicted deterioration over time on the BPT but no significant slowing was noted in the treatment group. Other outcome measures failed to

Table 5.5 Recent studies using the brief praxis test (BPT)

Author	Down syndrome population	Finding
Lott et al. 2012 [30]	Age: 50 years; dementia; (*N* = 53) With and without seizures Longitudinal study (2 years) using BPT and SIB	• Fewer with seizures completed either test • BPT completed by 45%; SIB completed by 4%
Powell et al. 2014 [32]	Age: 51.4 years; with and without dementia; (*N* = 20) MRI with DTI to asses frontal white matter measured fractional anisotropy (FA) Cognition assessed with BPT and SIB	• BPT & SIB lower in those with dementia • BPT but not SIB correlated with FA across all DS subjects
Walsh et al. 2015 [31]	Age: 49.8 years; with and without dementia; (*N* = 114) Mild to severe intellectual disability (ID) Evaluated RADD to assess dementia across ID BPT, SIB and other tests used for validation	• RADD correlated with BPT SIB, and other tests • RADD lower in dementia across all ID levels • 15% floor effect (RADD = 0)
Sano et al. 2016 [29]	Age: >50 years; (*N* = 337) Clinical trial: vitamin E (2000 IU/day) vs. placebo Primary outcome: BPT Secondary outcomes: CGIC, memory, orientation, vocabulary	• BPT & CGIC worsened over time • No change on other outcomes • No treatment benefit
Lin et al. 2016 [33]	Age: 48.3 years; with and without dementia; (*N* = 22) 1H-MRS to assess posterior cingulate cortex Measured neuronal (NAA, Glx) and glial (MI) biomarker Cognition assessed with BPT and SIB	• BPT & SIB lower in those with dementia • BPT and SIB correlated NAA • BPT but not SIB correlated with Glx, and NAA/MI ratio

SIB severe impairment battery, *MRI* magnetic resonance imaging, *DTI* diffusion tensor imaging, *RADD* rapid assessment of developmental disabilities, *CGIC* clinical global impression of change, *1H-MRS* proton magnetic resonance spectroscopy, *NAA* Nacetylaspartate, *Glx* glutamate-glutamine complex, *MI* myo-inositol

show systematic decline, including Orientation, the Fuld memory test, a visual memory test (The New Dot Test) and a picture vocabulary test. Of note about a third of the sample was unable to complete these tests at the end of the 3 year trial.

Other reports have used the BPT in longitudinal and imaging studies. In one report the BPT and the Severe Impairment Battery (SIB) were administered to individuals with DS and dementia to compare those with and without seizures [30]. Assessment every 6 months over a 2 year period was conducted and the primary outcome was the time to the "inability to test" on each measure. Both outcome measures identified significant difference between those with and without seizures. Though not formally compared, it appears that more individuals were able to be assessed with the BPT than the SIB across both those with and without seizures (45% vs 4%). Additionally the BPT was used to validate the Rapid Assessment of Developmental Disabilities (RADD), which is heavily weighted toward language assessment [31]. The RADD was highly correlated with the BPT but approximately

15% performed at floor level at initial assessment. While there were group differences between those with and without dementia on the RADD, unlike the BPT, the floor effect would make it difficult to use for longitudinal assessments.

In a study using MRI with DTI comparing DS individuals with and without dementia the investigators used the BPT and SIB for cognition and measured fractional anisotropy (FA) in the frontal cortex [32]. As expected lower performance was seen in those with dementia on both the BPT and SIB. Only the BPT was able to demonstrate correlations between cognitive severity and FA. Similarly in a report comparing DS individuals with and without dementia using proton magnetic resonance spectroscopy (1H–MRS) [33], the BPT and SIB were both able to discriminate between groups. Also both tests were correlated with levels of ding Nacetylaspartate (NAA), a neuronal biomarker. BPT was also correlated with the metabolite glutamate-glutamine complex (Glx), and with myo-inositol (MI), a putative glial biomarker. Results form these studies suggest the BPT is particularly useful in clinical, biological correlations in DS across a range of impairment.

Future Developments

The development of valid and reliable tests with high specificity and sensitivity for detecting dementia among persons with DS remains an important goal for the future. Almost of equal importance are tools that are highly sensitive to early changes in function. Short forms of the Dyspraxia Scale for Adults with Down Syndrome combined with a brief questionnaire should be explored in a variety of residential and clinical settings as a possible screening tool to raise the "level of suspicion" concerning the possibility of DAD. Detection of the early changes in cognition could "trigger" diagnostic referral and facilitate the planning of appropriate supports and care practices. Additional tests should be developed to tap other aspects of cognitive function. Some possibilities have been suggested elsewhere [9, 19]. Evidence-based tests of cognitive function with adequate psychometric properties can make an important contribution to the diagnostic assessment process. Such tools can also provide a source of possible outcome measures for clinical trials aimed at evaluating the efficacy and safety of research medications that could alleviate and or prevent the onset and progression of DAD in this at-risk population. Outcome measures should be developed which can be used with persons at the severe and profound levels of ID because effectiveness of treatments may vary with this factor.

Correlation of cognitive test performances with markers of pathology such as imaging and CSF biomarkers can validate the ability to capture specific dementia etiologies. Here we demonstrate the ability of the BPT to capture cross correlations in cross-sectioanal studies. One important approach for conducting longitudinal clinical and neuropathological studies has been described by Visser and his colleagues [34]. Validation of the performances also requires correlation with clinical diagnoses using uniform criteria such as those from the World Health Organization or the Diagnostic and Statistical Manual. of the American Psychiatric Association.

Additionally the approach of the National Institute on Aging-Alzheimer Association work group on diagnostic guideline encourages biomarker positivity to confirm diagnoses [35]. The development work with the Dyspraxia Scale for Adults with Down Syndrome did not frequently include candidates recruited from diagnostic clinics. This practice means that a number of interpretations of declines in Dyspraxia Scale performances are plausible. Many morbid conditions have a higher than usual prevalence among older adults with DS. These include depression [36], thyroid disorder [37], the effects of stress and delirium [37] psychiatric conditions [38], and maladaptive behaviors [39]. However, where clinic facilities are available recruitment of research participants leads to a biased selection from a population likely to have some problems. Such a practice can introduce a serious limitation on the generalizability of the research findings. The development work with the Dyspraxia Scale for Adults with Down Syndrome rarely involved the selection of clinic samples. Persons with DS were recruited from a wide variety of settings (rural, urban, suburban, day treatment programs, workshops, small residential settings), different counties in New York State and in three different countries United States, Canada, and Britain). Thus, these samples are more likely to be representative of the population of persons with DS than clinic samples. However, without clinical evaluations, attribution of dementia type must remain tentative.

The Dyspraxia Scale for Adults with Down Syndrome contains 62 items and administration can take up to an hour or so. Analyses reported above suggest that there is substantial "redundancy" in the test items. Such redundancy is important to minimize the effects of altered motivation, distraction, inattention, or momentary lapses in following instructions. A shorter version of the Dyspraxia Scale, called the BPT, consists of 20 items from the Dyspraxia Scale for Adults with Down Syndrome which can be administered in less than 10–15 min. The BPT was developed as a primary outcome measure for a randomized, double-blind, placebo-controlled trial of vitamin E [19]. Short versions of the Dyspraxia Scale, such as the BPT, could provide alternate forms useful for research applications involving repeated measures and it would shorten the length of time required for the examination of the individuals. Shorter test times would decrease the possible impact of fatigue and permit the addition of other tests as part of a less-fatiguing test session for the individual being examined.

Summary

The slow and insidious development of progressive dyspraxia is recognized as an early sign of DAD among aging persons from the general population. However, little is known about the age of onset, expression, and development of these AD-associated movement-related disorders among aging persons with DS. This report provides a brief description of the development of a Dyspraxia Scale for Adults with Down Syndrome. It includes the construction, administration, and scoring of the scale as well as the psychometric properties and the establishment of a standardization sample of 109 healthy individuals with DS. Analyses showed that

the scale is highly reliable with high internal consistency of the items. A factor analysis involving a second sample of 315 individuals with DS from seven different sites is consistent with the idea that the Dyspraxia Scale for Adults with Down Syndrome is a one-dimensional scale. Other research reveals that deterioration over a 3-year period in scores among aging persons with DS 50 years of age and older is significantly greater than that shown adults with DS younger than 50 years of age. The results further indicate that the onset of clinically significant dyspraxia can be identified at an average age of 57.9 years among persons with DS. The significance and temporal relationships between changes in dyspraxia scores, in short-term recognition memory, and in maladaptive behavior ratings are also presented. Summaries of three published reports using the Dyspraxia Scale for Adults with Down Syndrome are provided and directions for the future are suggested.

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Chapter 6
Adaptive Behavior Change, Mild Cognitive Impairment and Dementia in Down Syndrome: Case Classification Using the Adaptive Behavior Scale

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Introduction

The recognition of the facial features of Down syndrome (DS), with an incidence of approximately 1 in 691–733 live births [1, 2] has had a long history, with the first depictions of individuals with what was later determined to be the DS phenotype noted in artifacts almost 3500 years old [3, 4]. The distinctive phenotype was first noted in the *scientific literature* in the mid-nineteenth century by two physicians (Jean-Étienne-Dominique and Édouard Séguin) who were seeking to improve social and habilitative practices in institutions for intellectually disabled (ID) people, and then more comprehensively described by ***John Langdon Down*** to describe a collection of symptoms he observed in almost 10% of the children he had treated at the Royal Earlswood Asylum [5–8]. However, it was not until 1959 that Jerome Lejeune and his colleagues linked the DS phenotype to triplication of some or all of the 21st chromosome [9, 10]. (For further information on the genetics of DS see Mandava and colleagues [11–14]).

Down Syndrome

Trisomy 21 is one of the most thoroughly examined specific conditions associated with ID and has a characteristic pattern of pathophysiological sequelae [15]. Children and adults with DS have a distinctive clinical presentation that includes a small head with a flat looking face, small ears and mouth, enlarged and often protruding tongue and almond shaped eyes with epicanthal folds at the inner corners [16]. Additionally, there are consequences for cognitive development that vary in penetrance and severity, both within individuals with DS and compared with other individuals with ID [16–18]. Medical complications include an increased risk for: (a) congenital heart defects, (b) impaired hearing, (c) ophthalmic disorders, (d) endocrine deficiencies, (e) orthopedic problems, (f) skin and dental abnormalities, (g) seizure disorders, (h) certain forms of leukemia, (i) sleep apnea, (j) shortened life expectancy, and (k) Alzheimer's disease (AD) [16, 17, 19–24].

The diagnosis of DS in a newborn, even in the early-twentieth century, was synonymous with limited survival, with life expectancy of only 9 years [25]. Mortality risk was highest within the first year of life, followed by the first 10 years of life. As recently as 1975, the mean life expectancy for an individual born with DS was only 50 years, [26] in 2016 it has been estimated that the mean life expectancy may be 58 years. In fact, current projections suggest a future United States population of over 200,000 people with DS over the age of 55 years [8, 27]. This substantial increase in life expectancy is linked to a number of influences; medical factors including the swift and nearly universal application of corrective surgery for congenital cardiac problems [28] common to children with DS [29], and wide-ranging developments in medical care, nutrition, education and public health practices that have resulted in extensions in life expectancy in all Americans [27].

Dementia and Alzheimer's Disease

Dementia in old age has been recognized since the time of Hippocrates [30], however the specific neuropathology responsible for producing 60–80% of the cases of dementia in the neurotypical population and virtually 100% in people with DS was not identified until 1906, and the condition eponymously named Alzheimer's disease (AD) [31–35]. Neuritic plaques, extracellular deposits of beta-amyloid (Aβ) protein in the cerebral cortex surrounded by degenerating nerve terminals, and neurofibrillary tangles, strands of the hyperphosphorylated protein tau within individual neurons [36], are the two distinctive signature lesions in AD. Currently they are hypothesized to be the result of a cascade of processes beginning with an excess Aβ accumulation [37–39]; although alternative hypotheses have been proposed and are gaining in prominence [40–43]. Over time, these pathological processes contribute to deterioration in synaptic networks, neuronal loss and gross brain atrophy [36]. Dementia in Alzheimer's disease (DAD) is characterized by a progressive deterioration in cognitive and functional abilities caused by impaired functioning of brain networks that control thought, memory, and language abilities. With progression, serious declines become evident in an affected individual's ability to carry out daily activities, finally resulting in total inability to ambulate and communicate [44, 45]. Typically affected cognitive capabilities include deficits in memory, loss of semantic knowledge, disordered time sense, language deficits, inability to preform movements and skilled gestures, deficits in visual processing, executive dysfunction (i.e., poor planning, judgment and the ability to perform complex tasks) and disinhibition [44]. Age at onset varies; it has been estimated that 1 in 9 neurotypical adults over 65 years of age and 1 in 3 over age 85 have dementia [46, 47]. A condition called mild cognitive impairment (MCI) has been conceptually defined as a decline in functioning that is more severe than expected with typical brain aging but not severe enough to meet criteria for a diagnosis of dementia [48].

Alzheimer's Disease and Dementia in Adults with Down Syndrome

Dementia in middle—to older aged adults with DS has been noted in the scientific literature for over 100 years [49–53], and in 1948, Jervis [54] first described the clinical and pathological characteristics of AD in three adults with DS. However, as recently as 30 years ago, the study of DAD in individuals with DS did not attract much interest, given that relatively few individuals with DS lived long enough to develop DAD [55–59]. In 1987, it was found that the gene for Aβ precursor protein (APP), the protein from which Aβ is derived, resided on the proximal part of the long arm of chromosome 21 [60–62], within what many believe to be the region of chromosome 21 that must be trisomic for the full expression of the DS phenotype [14]. The extra copy of the APP gene leads to approximately a 1.5 fold increase in expression [63], and is likely to be a key contributor to the relatively early development of AD pathology in adults with DS [21, 64].

Especially strong evidence for a central role of APP overexpression in the development of AD pathology and dementia comes from two lines of research (parenthetically providing further support for the amyloid cascade hypothesis). Two case studies of older adults with DS, with microdisomy of the APP gene but with an otherwise typical DS genotype found that neither individual developed dementia throughout their lives or presented with significant AD neuropathology at autopsy [65, 66]. In addition, microtrisomy of the APP gene, as well as other APP mutations, increase risk for AD in otherwise neurotypical adults [67].

Classification of Dementia Status

Dementia, according to the *Diagnostic and Statistical Manual of Mental Disorders IV* (DSM-IV) [68] is a syndrome characterized by multiple cognitive deficits, which include memory impairment and at least one of the following: aphasia, apraxia, agnosia or disturbance in executive functioning; social or occupational function is also impaired. The Diagnostic and Statistical Manual 5th edition's (DSM-V) updated criteria for defining major neurocognitive disorder (current terminology for what was previously categorized as dementia) includes: (a) evidence of significant cognitive declines from previous levels of performance in one or more cognitive domains (complex attention, executive function, learning and memory, language, perceptual-motor, or social cognition), based upon self-appraisal or the assessment of a knowledgeable informant or clinician, with further corroboration by a second independent clinician supported by standardized neuropsychological testing, (b) that the cognitive deficits interfere with independence in everyday activities, (c) that the cognitive deficits do not occur exclusively in the context of a delirium, and (d) that the cognitive deficits are not better explained by another mental disorder [e.g., major depressive disorder, schizophrenia] (p. 603, [68]). DSM-V revisions in the criteria for dementia include increased clarity of the definition and greater emphasis on the use of objective measures to operationalize the diagnostic procedures [69, 70]. Additionally, a new diagnostic entity, minor neurocognitive disorder, was created to be analogous to Mild Cognitive Impairment (MCI), defined clinically as a decline in cognition reflecting a transitional state between characteristic brain aging and of dementia [71]. Adults with MCI have experienced noticeable declines in cognition, but not of sufficient severity to meet diagnostic criteria for dementia [72]; MCI can be distinguished from dementia by the absence of significant impairments in social or occupational functioning and activities of daily living, although mild problems in performing complex functional tasks may be present. While unambiguous operationalization of this classification remains in flux, even for older adults with neurotypical development, standard practices for diagnosing MCI in adults with substantial pre-existing cognitive impairments (i.e., ID) have rarely been reported [72]. In fact, only a few studies have focused on MCI among adults with ID and none have proposed explicit diagnostic criteria applicable to this population [48, 72–76].

Classification of Dementia Status in Adults with Down Syndrome

Distinguishing between a classification of "normal aging" and early MCI, and later-stage MCI and early dementia, is challenging even for neurotypical adults, with criteria heavily reliant on "clinical judgment" and with considerable variability in actual clinical practice. These difficulties are magnified for individuals with DS due to the presence of significant impairments in cognition and functional abilities, including basic activities of daily living (BADL; feeding, dressing, grooming, bathing, toileting, ambulating) and instrumental activities of daily living (IADL; traveling, managing finances, telephone use, meal preparation, housekeeping, laundry, shopping and medication maintenance) that vary in severity across individuals. Theoretically, MCI in neurotypical adults should not be accompanied by a substantial decline in BADLs, while occasional deficits in IADLs may be diagnostic. In adults with DS, lifelong deficits in BADLs and IADLs are the norm, so alternative definitions of MCI are necessary. While best practices for diagnosis are still in development; a comprehensive examination of the range of issues and challenges in defining MCI and dementia in adults with DS can be found in a recent review published by Krinsky-McHale and Silverman [48].

While there is increasing interest in developing classification methods that can be informative of dementia status at initial evaluation, currently classifications of dementia status are based on objective findings of a significant decline in functional and cognitive capabilities compared with previous status. Optimally, decline in functional and cognitive functioning should be established through administration of valid and reliable neuropsychological measures appropriate for this specific population [48, 77–79]. However, individuals referred for evaluations based on caregiver or personal concerns often have not undergone formal neuropsychological evaluations previously, have physical or psychiatric problems that are incompatible with cognitive testing or may be resistant to clinical evaluation; issues that may reduce the validity and reliable of dementia classification. It must be noted that classifications of dementia status presented in the following studies do not constitute differential diagnoses of DAD. With current technology, the gold standard for diagnosis of DAD entails direct evidence of characteristic neuropathology rather than just the presence of dementia. However, given the virtually universal occurrence of AD-type neuropathology in adults with DS over the age of 40 years [32] a clinical classification of dementia is tantamount to a differential diagnosis of DAD or dementia due to DAD together with another condition. These "mixed" cases are rare given that cerebrovascular diseases, one of the major alternative causes of dementia, is less prevalent in adults with DS [80].

Functional Decline and Adaptive Behavior

Neuotypically developing individuals can be expected to have baseline levels of BADLs that are relatively invariant, while individuals with ID have pre-existing impairments that may vary widely in their severity. To classify MCI and dementia

for this latter population, criteria need to consider those pre-existing impairments and document substantial declines from previous abilities. This chapter reviews the ways in which the measurement of adaptive behavior has been applied to classify MCI and dementia in adults with DS. First, descriptions of the American Association on Intellectual and Developmental Disabilities Adaptive Behavior Scale (AAIDD-ABS) [81] and AAIDD-ABS2 [82] are presented, as well as their psychometric properties, followed by a description of relevant studies that have used the ABS to examine dementia in DS. No published studies to date have used the ABS2 to classify dementia, possibly due to the longitudinal nature of many of the studies reported and the desire not to introduce experimental error by changing assessment instruments midstream. Relevant findings from our research program on aging in ID, a 30-year long endeavor, will be described in some detail; finally, we will present suggestions for future research.

Background of the Adaptive Behavior Scale

Adaptive behavior, defined as the "effectiveness of an individual in coping with the natural and social demands of his or her environment" ([83], p. 5), first became a formal component of the definition of ID in the late 1950s [84–86], and today it remains an integral element of the diagnosis. Stated simply, the purpose of adaptive behavior assessment is to obtain an inventory of an individual's strengths and weaknesses [87, 88]. The American Association on Mental Deficiency (renamed the American Association on Intellectual and Developmental Disabilities) Adaptive Behavior Scale was first published in 1967, and revised in 1974 (hereafter noted as ABS) [81], and 1993 (hereafter noted as ABS2 [82] and was "designed to provide objective descriptions of an individual's adaptive behavior" ([83], p. 5). While numerous additional adaptive behavior measures have been developed [89]; the ABS, in its various revisions has been well received, proven valid and reliable and widely used within the field of ID [82, 90–94]. Appendix illustrates page 1 of the ABS.

The ABS, in both its 1974 and 1993 versions, consists of two parts. Part 1 was designed to evaluate an individual's abilities and strengths in ten behavioral domains: (a) independent functioning, (b) physical development, (c) economic activity, (d) language development, (e) numbers and time, (f) domestic activity, (g) vocational activity, (h) self-direction, (i) responsibility, and (j) socialization. Interrater reliability coefficients for the ten domains in the 1974 ABS range from 0.71 (self-direction) to 0.93 (physical development), with a mean reliability coefficient of 0.86. Interrater reliability coefficients for the ten domains in the 1993 revision of the ABS range from 0.88 (prevocational/vocational activity) to 0.99 (independent functioning and domestic activity), with a mean reliability coefficient of 0.95. Reliability of a Part 1 total score derived by summing the ten domain scores was estimated at 0.96 [95]. Table 6.1 displays the reliability coefficients for each adaptive domain for both the 1974 and the 1993 versions of the ABS.

Table 6.1 Reliability coefficients for adaptive domains on ABS and ABS2

Domain	ABS[a] interrater	ABS2[b] test-retest	ABS2[c] internal consistency
Independent functioning	0.92	0.99	0.98
Physical development	0.93	0.96	0.94
Economic development	0.85	0.98	0.90
Language development	0.87	0.96	0.96
Numbers and time	0.86	0.97	0.94
Domestic activity	0.91	0.99	0.95
Vocational activity	0.78	0.88	0.82
Self-direction	0.71	0.92	0.94
Responsibility	0.83	0.95	0.90
Socialization	0.77	0.88	0.91

[a]Mean Pearson product moment correlations using Fisher's Z transformation
[b]Corrected reliability coefficient using Anastasi's [96] procedure for extracting error variance
[c]Mean Coefficient Alpha averaged across age groups

Part 2 of the 1974 version of the ABS was developed to evaluate maladaptive behaviors related to personality and behavior disorders, and contains 14 domains including: (a) violent and destructive behavior, (b) antisocial behavior, (c) rebellious behavior, (d) untrustworthy behavior, (e) withdrawal, (f) stereotyped behavior and odd mannerisms, (g) inappropriate interpersonal manners, (h) unacceptable vocal habits, (i) unacceptable or eccentric habits, (j) self-abusive behavior, (h) hyperactive tendencies, (i) sexually aberrant behavior, (j) psychological disturbances, and (k) use of medications (i.e., tranquilizers, sedatives, anticonvulsant drugs, and stimulants). Interrater reliability coefficients for the 14 domains range from 0.37 (unacceptable vocal habits) to 0.77 (use of medications), with a mean reliability coefficient of 0.57. In part, as a function of the less than optimal reliability of Part 2 of the 1974 version of the ABS, the 1993 ABS2 contained only eight domains related to maladaptive behavior: (a) social behavior, (b) conformity, (c) trustworthiness, (d) stereotyped and hyperactive behavior, (e) sexual behavior, (f) self-abusive behavior, (g) social engagement, and (h) disturbing interpersonal behavior. Interrater reliability was much improved, with coefficients for the eight domains ranging from 0.95 (social behavior and conformity) to 0.99 (trustworthiness, self-abusive behavior, social engagement), with a mean reliability coefficient of 0.97. Table 6.2 displays the reliability coefficients for each maladaptive domain in ABS and Table 6.3 displays the reliability coefficients for each maladaptive domain in ABS2.

Data regarding the validity of the 1974 and 1993 versions of the ABS were summarized in the 1974 and 1993 ABS manuals [82, 83, 97]. Additionally, criterion validity was investigated by Salagaras [97, 98], who demonstrated the ABS's sensitivity to quantify individual differences in adaptive and maladaptive behavior among subgroups that varied with respect to seven variables: age, sex, estimated severity of intellectual impairment, etiology of ID, place of living, the presence or absence of any mobility handicap, and the use of prescription medications (coded directly from the ABS divided into those who used no medications versus those using any medications).

Table 6.2 Reliability coefficients for maladaptive domains on ABS

Domain	ABS[a] interrater
Violent & destructive behavior	0.59
Antisocial behavior	0.68[b]
Rebellious behavior	0.55[b]
Untrustworthy behavior	0.69
Withdrawal	0.44
Stereotyped behavior & odd mannerisms	0.62[b]
Inappropriate interpersonal manners	0.47[b]
Unacceptable vocal habits	0.37[b]
Unacceptable or eccentric habits	0.57[b]
Self-abusive behavior	0.49[b]
Hyperactive tendencies	0.57
Sexually aberrant behavior	0.52[b]
Psychological disturbances	0.45[b]
Use of medications	0.77[b]

[a]At least partially computed by Phi coefficient
[b]Mean Pearson product moment correlations using Fisher's Z transformation

Table 6.3 Reliability coefficients for maladaptive domains on ABS2

Domain	ABS2[a] test-retest	ABS2[b] internal consistency
Social behavior	0.95	0.94
Conformity	0.95	0.91
Trustworthiness	0.99	0.88
Stereotyped and hyperactive behavior	0.96	0.86
Sexual behavior	0.98	0.83
Self-abusive behavior	0.99	0.81
Social engagement	0.99	0.84
Disturbing interpersonal behavior	0.97	0.90

[a]Corrected reliability coefficient using Anastasi's [96] procedure for extracting error variance
[b]Mean Coefficient Alpha averaged across age groups

Adaptive behavior items on the ABS vary in type; some require a unitary response while others require respondents to select several responses. One type of item directs the rater to circle the statement that best describes the individual's abilities in the specific adaptive behavior among several choices, while the rater can circle multiple statements (i.e., all statements that apply) in the second type of item. Item scores are summed to provide subdomain scores, and subdomain scores are summed to provide domain scores. The ABS2 norming sample included 4103 individuals with ID residing with parents, in community-based small residences, or in large congregate care residential facilities [82].

Maladaptive behavior items on the ABS are all one type; the rater is directed to circle all statements that apply to the individual being evaluated in terms of the frequency of occurrence, occasionally or frequently. Item scores are again summed to provide subdomain scores, subdomain scores are summed to provide domain scores, and percentile ranks are available for each domain.

The ABS typically is administered through an interview with a correspondent familiar with the individual being assessed (i.e., third-party assessment), however alternate methods are available (see [83] for a complete description of administration options). Briefly, first-person assessment can be used when the rater is familiar enough with the individual being evaluated to complete the form without referring to other sources for additional information. In the interview method, the rater discusses the individual's behavioral competencies and maladaptive behaviors with a correspondent familiar with the individual being assessed. After the interview, the rater independently completes the individual items on the ABS. This method of administration is not suggested for use in research when detailed information is required [83].

Research Studies

The ABS and Age-Associated Group Differences in Adaptive Behavior

In 1983, Miniszek [99] reported the first use of the ABS to distinguish between nine adults with DS over the age of 50 who were "seriously regressed" (i.e., demented) from six adults with DS over age 50 who exhibited no visible signs of dementia. Non-demented participants performed better (i.e., higher mean domain scores) than participants classified as demented. All 15 adults with DS over age 50 years exhibited equivalent (physical development) or lower ABS domain scores compared with a small group of four participants with DS under the age of 50. In spite of this successful pilot demonstration that the ABS is sensitive to dementia, it was almost 10 years until the ABS was used in other efforts to describe functional and cognitive deterioration in adults with DS [100, 101]. One such study examined ABS domain and total scores (i.e., Part 1 and Part 2), in adults with DS who ranged from 18 to over 60 years of age. Participants over 50 years of age manifested significantly poorer performance in most functional domains than did younger participants, with adults over age 60 exhibiting the lowest performance on all domains. There were no age-associated differences in ABS Part 2 domains (maladaptive behavior), with the exception of "use of medications", which was increased in the group of older adults [101]. A second study focused on language development measured by the ABS in adults with DS who had no major sensory impairments, and found that expressive language and comprehension performance was significantly reduced in older participants compared with younger participants, with the largest age-associated deficits found in comprehension skills [100]. Prasher [102], using similar procedures, also found age-associated differences in language development, however effects were equivalent in both the expressive language and the comprehension subdomains. This discrepancy may have been due to increased sensory function in the latter study, as Prasher directly assessed vision and hearing, while the earlier study was less precise.

Collacott [103] expanded these analyses to relate age of epilepsy onset (early <35 years of age versus late ≥35 years of age) to adaptive competence. Collacott suggested that late onset seizures could serve as a surrogate indicator of DAD, as this is a well-known symptom as DAD progresses into its later stages [104]. Timing of epilepsy onset was significantly related to ABS adaptive scores, with individuals with late-onset epilepsy, compared with early-onset epilepsy, exhibiting poorer performance. Again, there were no significant differences in ABS Part 2 domains (maladaptive behavior) apart from "use of medications."

Clearly, these studies showed that the ABS could document differences in cohorts defined by age and dementia presence/absence in adults with DS (i.e., inter-group differences). However, the cross-sectional nature of the types of studies reviewed thus far provided only an indirect reflection of true age-related declines (i.e., declines in the same cohort over time). Age-associated inter-group differences in adaptive competence may be due to aging and/or progression of DAD or they may be present simply because the sample members belong to different cohorts with different life experiences. These classic "cohort effects" arise because earlier life experiences can be important determinants of a person's and a population's characteristics in later life. For example, the better health care, nutrition and education provided for people with DS born 30 years ago, varied widely with the neglect and maltreatment, in general, that people born with DS experienced 70 years ago, could have caused lifelong differences between these groups that have nothing to do with either aging or DAD. In fact, the effects of aging on this population could even be underestimated due to what are termed "healthy survivor effects". These refer to a selection process such that individuals who are enrolled in a study later in life tend to represent only the healthier members of their respective birth cohorts because their frailer peers passed away at younger ages [105, 106]. To help control for these potentially confounding effects longitudinal studies of people born within the same cohort are necessary to clearly document decline over time as described next.

The ABS and Age-Associated Declines in Adaptive Behavior

Fenner [99] described one of the first studies to use the ABS to measure longitudinal declines in adaptive competence related to age in adults with DS [107]. Significant losses in functional abilities were exhibited only by one-third of the individuals over age 35 years, suggesting a more positive picture than expected given the presumed presence of AD-related brain pathology. However, the oldest participant was only 49 years old, and studies including older people seem to be necessary to address the broader extent of age-related adaptive decline characteristics of older adults with DS. Zigman and colleagues examined the age-associated incidence of significant decline in adaptive behavior and the temporal pattern of decline in specific functional skill domains in 646 adults with ID with and without DS using the ABS [108, 109]. The standard error of measurement (SEM [96]), a reflection of dispersion around an individual's true score with known statistical properties was computed, and significant

decline in adaptive behavior was defined as a reduction of two SEMs in total ABS score over a 2-year period. Cumulative incidence of significant decline in total ABS score for adults with DS increased from 4% at age 50 to 67% by age 72 years, verifying that aging throughout the 50s and 60s is not uniformly pathologic; clearly there was evidence that a significant number of adults with DS are successfully surviving into their 60s and 70s; a finding that has since been replicated (cf. [110]).

While these incidence rates for adults with DS reflect significant decline in total ABS score, and this is an imperfect reflection of true dementia, these rates are not meaningfully different from other published estimates of dementia in adults with DS [111, 112]. Cumulative incidence of significant decline in total ABS score for adults with ID without DS increased from 2% at age 50 to 52% by age 88, fairly similar to prevalence rates of DAD in the neurotypical population at that advanced age [113, 114]. There are some reports of an increased risk for DAD in adults with ID without DS [115], however variation in rates may be due to differential definitional criteria [72].

The ABS was not designed to be an assessment of dementia; therefore, various domain and subdomain scores might be differentially sensitive to progression of DAD or other causes of old age—associated dementia. An a priori descriptive content analysis of the ABS resulted in the identification of 15 separate clusters of items [108]. Changes over time were analyzed within each of the adaptive clusters only in participants who declined significantly in the overall adaptive functioning score. (An examination of non-decliners revealed that they were relatively stable over time in each of the 15 clusters). Differences in the timing and magnitude of declines were evident, with relatively large and early declines in performance in care of clothing, dressing and undressing activities, domestic activities, and vocational activities. Relatively early, but somewhat smaller declines in performance were seen in responsibility and socialization, economic activities, physical development, travel, and general independent functioning activities [108]. Proficiency in these skills also may be considered necessary to function competently in everyday activities of daily life outside the home. Clusters reflecting more basic activities of daily living skills declined slightly later. Larger declines were observed for self-direction, toilet use, numbers and time, and cleanliness. Smaller declines were seen for comprehension and social language, appearance, eating, and expression. (i.e., writing, preverbal expression, articulation, sentences and word usage).

Functional declines are first noted in skills that are more complex (i.e., IADLs) with progression to the more basic and fundamental abilities (i.e., BADLs). Not surprisingly, ability to eat, to understand spoken language, and to ambulate were among the last domains to be affected. These patterns are largely consistent with other reports describing the progression of dementia symptoms in adults with DS [116] and elderly adults experiencing progressive dementia more generally [117].

In neurotypical adults, a specific allele state (i.e., ε4) on the APOE gene, residing on chromosome 19, has been shown to be a strong predictor of risk, as well as rate of progression in AD. Schupf and Zigman and others examined the effects of APOE genotype on trajectories of change for adults with DS using available ABS data. Decline in adaptive competence was greater in those people with an APOE ε4 allele

compared to those without, regardless of whether dementia was diagnosed, suggesting that decline on total ABS scores may be a sensitive indicator of deterioration in abilities and underlying progression of AD neuropathology in adults with DS [118, 119].

Urv and colleagues examined changes in maladaptive behaviors in participants with significant changes in adaptive functioning using the ABS Part 2 items [73]. Obnoxious behavior (e.g., lying, reacting poorly to frustration or criticism, demanding excessive attention, impudent attitude towards authority), lack of boundaries (e.g., takes others' property, disrespecting others' property), and overestimating one's own abilities were significantly elevated before the occurrence of significant adaptive decline and then decreased over time. These results suggest that elevated levels of problem behaviors may anticipate significant regression in adaptive behavior and may provide caregivers with early indicators of concern. Changes that occurred concurrently with significant regression in adaptive behavior included withdrawn behavior and emotional instability (e.g., mood changes, poor emotional control). While caregivers should take notice of these types of changes in "older" adults with ID, as they may be indicative of memory problems and disinhibition, they also may be due to a host of other conditions both physiological and psychological. Conversely, the absence of these behaviors may not rule out the presence of MCI or dementia; therefore, all new behavioral symptoms should be documented and diagnosed by a physician, psychologist and/or a psychiatrist.

Overall, these findings are consistent with previous reports that suggest that selected changes in specific areas of maladaptive behavior may be early signals of dementia in individuals with DS [120–122]; however, more research on these issues is certainly necessary. Data presented to this point clearly suggest that the ABS is sensitive to age-associated differences and age-related declines over time in adaptive competence; next we will present the results of studies that measured ABS performance as a function of specific dementia classifications [123].

The ABS and Dementia Classification

Prasher and colleagues, in a series of studies based upon a longitudinal investigation of aging and dementia in adults with DS, used the ABS to measure participants' adaptive competence and maladaptive behavior [102, 116, 124, 125]. Controlling for age, results from cross-sectional analyses demonstrated that participants with dementia had significantly lower total ABS scores than unaffected adults. A subgroup of adults with DS without dementia who had no significant medical or psychiatric concerns still exhibited age-associated differences in adaptive competence, which may be demonstrating "normal" age-related changes in ability as opposed to DAD-related performance deficits; alternatively, they may just represent age-related increases in prevalence of very early stage DAD (or MCI). Longitudinal changes in ABS scores in adults with DS were described in three studies reported by Prasher [116, 124, 125]. There were several methodological differences among the studies that included the sample characteristics, duration of follow-up, and the stage of

dementia investigated. In one study, changes in adaptive competence were examined over a 2-year period ranging from 1-year before diagnosis of dementia to 1-year after diagnosis [125]. In the other two studies, changes in adaptive competence were examined as a function of dementia status (i.e., demented versus non-demented). Regardless of the differences in focus, essentially similar outcomes were obtained from all three studies. As adults with DS transition from non-demented to demented status, there are significant changes in adaptive competence and, as would be expected, the magnitude of these changes increases as dementia progresses. As noted previously, ABS Part 2 total scores were generally higher in participants with dementia compared to those without dementia.

Finally, two clinical trials examining the safety and efficiency of donepezil hydrochloride (Aricept) to slow the progression of dementia in adults with DS included the ABS as a measure of adaptive competence [126, 127]. Results of these studies demonstrated the sensitivity and utility of the ABS as a metric for change even within a relatively restricted time, unfortunately the effectiveness of the intervention was negligible.

The ABS and Dementia: The Aging Research Program

A series of multidisciplinary longitudinal studies focusing on incidence, prevalence, biological and genetic risk factors, and natural history of dementia and chronic health conditions in over 800 adults with ID with and without DS over the age of 45 have been conducted spanning a period of over 30 years [95, 110, 128–133]. This multisite program represents a collaboration of researchers from the NYS Institute for Basic Research in Developmental Disabilities, Columbia University and the Kennedy Krieger Institute/Johns Hopkins University. In this project's most recent set of studies, dementia status has been assessed at approximately 18-month intervals employing measures of adaptive and cognitive functioning, a comprehensive review of all medications and clinical records and a series of biological and genetic biomarkers. The Dementia Questionnaire for People with Intellectual Disabilities [134, 135] and Part I of the ABS has been used to measure adaptive competence and functional behavior, and the Reiss Screen for Maladaptive Behavior [136] provided an overview of possible psychopathology.

Cognitive abilities of participants have been described based upon eight direct assessment instruments. The Peabody Picture Vocabulary Test-Revised was included initially to provide a measure of receptive vocabulary [137], but this procedure was dropped due to high variability in individual performance across testing sessions. Evaluation of mental status has been evaluated using three separate instruments: (a) a modified version of the Down Syndrome Mental Status Examination developed by Haxby [138], (b) a modified Mini Mental State Exam [139] developed in our labs [140] and (c) the Test for Severe Impairment [141]. The battery also includes an adaptation of the McCarthy verbal fluency test [142], the Beery-Buktenica Developmental Test of Visual-Motor Integration [143], the Block Design subtest of

the WISC-R [144], and an adaption of the Selective Reminding Test [145]. A full description of the instrument battery and its psychometric characteristics has been published elsewhere [95, 128].

Dementia status was classified at each cycle of assessment, in consensus conferences, employing criteria broadly consistent with guidelines recommended by the Working Group for the Establishment of Criteria for the Diagnosis of Dementia in Individuals with Developmental Disability [146, 147]. Each case was classified into one of the following categories: (a) non-demented, indicating that DAD-related decline was not present, (b) MCI-DS, indicating that there was some indication of mild cognitive and/or functional decline but importantly, the observed change did not meet dementia criteria, (c) possible dementia, indicating that some symptoms of dementia were present, but declines over time were less than 100% convincing, (d) definite dementia, indicating clear and convincing evidence of substantial decline over time, (e) uncertain with complications, indicating that criteria for definite dementia had been met, but that symptoms might be caused by some other substantial concern, usually a medical condition unrelated to a dementing disorder (e.g., loss of vision, poorly resolved hip fracture, loss of social support network due to relocation), and (f) undeterminable, indicating that pre-existing impairments were so severe that detection or interpretation of declines indicative of dementia were not possible.

Classification decisions inherently included a degree of subjective judgment that is difficult to quantify. This concern could be addressed by developing objective, empirically-based criteria for case classification. Current recommendations for diagnosis of dementia recognize this by emphasizing the detection of decline from previous levels of performance [146, 147]. However, this requires either that the process of diagnosis extends over substantial time intervals (often years) or that baseline abilities have been assessed appropriately. The first of these requirements precludes rapid decision-making and intervention while the second is unlikely to occur. Therefore, findings were examined in the hope of discovering classification criteria based upon a single assessment that considered level of preexisting intellectual disability impairment in addition to those that rely on detection of decline over extended periods of time [95]. This strategy continues to show promise in ongoing investigations that are currently continuing.

The ABS and Dementia Classification

There is a broad agreement on the concept of MCI as the early stage of DAD clinical progression, during which declines in abilities exceed those associated with aging, per se, but are of insufficient severity to warrant a diagnosis of dementia. While there still is some room for debate, the key definitional characteristics of MCI as a distinct clinical entity for adults without ID have been objectively specified. For adults with DS or other conditions associated with significant lifelong cognitive impairments, the definition of MCI is just beginning to evolve [48]. Complications relating to heterogeneity within the population with presentation and clinical operationalization, and

unpredictability in its long-term outcomes continue to present challenges. Nevertheless, accurate recognition of MCI may present an opportunity to prevent irreversible damage to neural networks when and if effective treatments for DAD become available [48]. The relationship between changes in ABS scores and clinically classified transitions in status from non-demented to MCI-DS and MCI-DS to dementia have yet to be fully investigated, but a small study conducted by Zigman and colleagues [148] attempted to address this issue in a sample of 123 adults with DS who developed MCI-DS while being followed longitudinally. Over a 3-year period 38 individuals remained without MCI or dementia, 53 individuals developed MCI without progressing to dementia and 32 developed MCI and then progressed to dementia. Repeated measures analyses of variance examined group effects. Group by time interactions were consistently significant, demonstrating that the group progressing to dementia consistently showed the greatest decline over time and the group developing MCI-DS declined to a lessor, but more substantial degree than the group that maintained their non-demented status. These findings indicate that the ABS *may* provide reliable indications of MCI-DS and early DAD-related decline for adults with DS. The ability to assess dementia status based upon minimal reports or with minimal participant interaction may allow broader screening of large populations of adults with DS and perhaps ID due to other causes, enabling early interventions, once developed, to mitigate loss of function. Further replications of this result with larger samples and longer periods of follow-up are now near completion and results should be available soon.

As stated earlier, one desired outcome of our research was the development of "norms" referenced to premorbid Intelligence Quotient (IQ) that have both the sensitivity and specificity needed to successfully classify MCI and dementia in its earliest stages without depending on longitudinal follow-up or the presence of an established baseline documenting premorbid abilities. The following represents several such attempts. In one study, we developed an index that reflects the total score on ten subdomains of the ABS that were found to deteriorate relatively early in the progression of DAD (i.e., care of clothing, dressing and undressing, domestic activity, vocational activity, responsibility, socialization, economic activity, physical development, travel, and general independent functioning), hereafter called "the dementia sensitive index" [108]. Performance on the "dementia sensitive index" was plotted as a function of IQ for each dementia classification category. A function was generated that related performance on the "dementia sensitive index" to IQ and distinguished demented from non-demented individuals. This function was generated using data from the first data collection cycle and then verified with data collected in a subsequent cycle. These data are substantially less than independent, but they were useful to define a procedure to develop IQ-based dementia criteria. If the criterion scores on the dementia sensitive index (total possible score 140) was defined by this eq. (10 + (1.5 * IQ)), with a maximum score 75), the ability to correctly classify dementia in participants who were demented (i.e., sensitivity) was 0.9 and the ability to correctly classify participants who did not have dementia as non-demented (i.e., specificity) also was 0.9; for non-demented versus definite dementia cases [149]. We need to mention a few limitations of this metric: (a) these criteria were not useful with participants who had IQs less than 25, (b) the estimates of sensitivity and specificity refer to distinctions between

the two most extreme classification categories (non-demented and definite dementia) and (c) these criteria need to be validated in an independent sample to be considered anything more than preliminary.

In another study, a subgroup of 133 adults with DS were identified based upon availability of at least five cycles of data and the absence of dementia during the first and second cycle of assessment [150]. Their mean age was 50.0 at enrollment, with a mean Stanford-Binet IQ of 38.1. Dementia status was classified for everyone at their third cycle of assessment based on one-time IQ-referenced performance for the DMR-SCS or our modified MMSE. Three groups were then defined for each instrument consisting of adults who were: (a) clearly above their IQ-referenced criteria (no dementia), (b) clearly below their IQ-referenced criteria (dementia), or (c) too close to criteria for a confident classification (MCI). Longitudinal performance for these three groups was then examined over the 36 months preceding and 36 months following this Cycle 3 classification. An analysis of adaptive behavior, as measured by the ABS Part 1, generated a Group X Time interaction of $F(8, 218) = 5.54$, $p < 0.0001$. Results consistently showed that the "dementia" group changed more than the "no dementia" group. Importantly, this was true for the 36 months following assessment as well as the 36 preceding months. Findings suggest that one-time assessments of dementia status of adults with DS can be useful in diagnosis. While profiles of decline/stability in performance should remain the gold standard, the need for relatively rapid determination of diagnosis/case classification will become more and more pressing as effective treatments become available, as will impatience with having to wait 6–18 months for results of follow-up assessments.

Summary

As adults age, concerns related to diseases such as DAD increase, especially so for people with DS who are clearly at increased risk and prone to other features of atypical aging [19, 20, 27, 151]. In the over 130 years since Fraser and Mitchell [49] first discussed skill loss in middle-aged adults with DS, there has been substantial progress in research probing the complex relationship between DS and DAD. The origin of the ubiquitous AD-type pathology seen in adults with DS once they reach their late 30s is, at least in part, due to the triplication of the gene for amyloid precursor protein located on chromosome 21 [21, 61, 65]. In the early 1980s, many researchers held the belief that all adults with DS who survived into their 40s and 50s would invariably develop clinical dementia. This dire prediction has proved to be untrue, and we now know that many adults with DS are living successfully into their late 60s and, in some cases, even 70s [110].

Several factors that modify risk for AD in adults with DS have been identified [118, 129, 130, 132, 152–165], including some as cholesterol level, statin use, and bioavailability of estrogen, that may be amenable to alteration through medical intervention. Carefully controlled clinical trials are needed to test the safety and efficacy of these types of interventions, and outcome measures providing a valid indication of DAD progression will play a vital role for these studies.

Unfortunately, as this volume on neuropsychological measures of dementia in DS indicates, standard diagnostic methods used to evaluate individuals with suspected MCI or dementia in the neurotypical population are not appropriate for use with adults with DS, many of whom have never developed the specific cognitive and adaptive skills that are measured by these assessment instruments. The use of the ABS as a surrogate measure of dementia has met with considerable success [166]; similar studies using the ABS as a surrogate measure of MCI are ongoing. The other chapters in this volume demonstrate that there are multiple functional and neuropsychological measures that may be successfully used to classify dementia status in adults with DS. In fact, the emphasis of the ABS on functional behavior may result in dementia being diagnosed relatively late in the disease process. Optimally, a highly sensitive and specific assessment battery will eventually be developed that uses the most reliable and valid aspects of each instrument to classify MCI and dementia in DS at the earliest possible stage. Further research into the role of maladaptive behaviors in the identification of early signs of MCI and dementia also is warranted, and the ABS, or selected components of the ABS, is a promising candidate.

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Chapter 7
The Cambridge Examination for Mental Disorders of Older People with Down's Syndrome and Others with Intellectual Disabilities (CAMDEX-DS)

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Background

The Cambridge Examination for Mental Disorders of Older People with Down's Syndrome and Others with Intellectual Disabilities (CAMDEX-DS) is a diagnostic assessment schedule developed in response to an identified need for valid and reliable methods for the assessment and diagnosis of dementia in people with intellectual disability (ID). While the increased risk of dementia in people with Down syndrome (DS) in particular means that the schedule is especially valuable for use in this population, it can also been used when dementia is suspected in those with developmentally acquired ID for reasons other than that of DS. Accurate and consistent diagnosis is essential for both clinical practice and research and will be increasingly important as effective treatments for dementia become available.

Rationale for the Development of the CAMDEX-DS

The principal aims for the development of this assessment were to incorporate in a single schedule all the information necessary to enable an accurate clinical diagnosis of dementia in people with ID, in the context of clinical practice or research [1, 2], to provide a structured framework for collecting information on the key features of

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the dementias and of other physical and psychiatric disorders of later life, in order to aid the differential diagnosis of any observed decline, with reference to standard operational diagnostic criteria [3], and to provide a means for monitoring progress and informing social, psychological, and medical interventions.

While the diagnosis of dementia is complicated by the presence of pre-existing ID, the principle that such a diagnosis requires evidence of a progressive deterioration in memory, in a number of other cognitive domains and in daily living skills, is the same regardless of whether an individual has ID. In the general population, a diagnosis of dementia is reached on the basis of informant-based and objective evidence of progressive deterioration in a person's cognitive abilities and functional skills, the operational definitions generally accepted being those outlined in the *Diagnostic and Statistical Manual of Mental Disorders* (DSM-V) [2] and the *International Classification of Diseases* (ICD-10) [3]. Our approach in developing a diagnostic assessment for use with people with ID has been to model it very closely on an assessment schedule that is widely used as an aid to the diagnostic process in the general population; the revised version of the *Cambridge Examination for Mental Disorders of the Elderly* (CAMDEX-R) [4].

Like the original schedule, the CAMDEX-DS has been designed to be administered in community settings by mental health professionals (as part of the diagnostic process), or to be used to formalize diagnosis in the context of a research study and is designed to provide structure and support for good clinical and/or research practice. Arriving at a diagnosis of dementia requires a full evaluation and the elimination of other possible illnesses or disorders that might present in a similar manner to that of dementia. These disorders may be treatable and have a very different prognosis to that of dementia. Research studies into the relationship between DS and dementia also require a similar level of diagnostic rigor. The CAMDEX-DS is not a substitute for proper clinical assessment when dementia is suspected, but rather it is an aid to the diagnostic process, designed for use by experienced clinicians and for informed clinical researchers. The diagnosis of dementia, or of other mental or physical disorders, though aided and supported by this framework, remains a judgment based on a clinical history, direct cognitive, mental state and physical assessments, and findings from appropriate investigations. In this respect, the CAMDEX-DS differs in both its aim and format from existing observer-rated scales that have been developed specifically for diagnosing dementia of the Alzheimer's type (DAT) (cf., dementia in Alzheimer's disease (DAD)) in people with ID that work on the principle of using a cutoff score to determine whether or not an individual has dementia. Such scales can be viewed as screening tools rather than aids to the process of making a clinical diagnosis.

Development and Use of the CAMDEX in the General Elderly Population

The CAMDEX was originally developed in 1986 as a standardized instrument for the diagnosis of mental disorder in the elderly general population, with particular reference to the early detection of dementia [5]. It was subsequently published by

Cambridge University Press [6] and a revised version was published in 1998 [4]. The schedule includes an informant interview, an interview with the participant, an objective examination of cognitive function (Cambridge Cognitive Examination, CAMCOG), a standardized schedule for recording observations, and a physical examination and information on laboratory investigations.

The informant interview provides a means for collecting information in a structured manner about those areas of function that are likely to change with the onset of dementia or of any other mental disorder. It includes several questions about the informant's observations on each of the following: the person's memory, general mental and intellectual functioning, judgment, general performance, specific higher cortical functions and personality, as well as the presence or absence of specific symptoms and relevant medical and family history.

The validity and reliability of the CAMDEX informant interview for use in the general elderly population has been shown to be good. For example, reported changes in memory and mental functioning from the informant interview highly correlated with objectively measured decline [7]. On measures of interrater reliability in the general population the correlation between total scores obtained by the two raters has been found to be high ($r = 0.90$, $p < 0.001$) as has the level of agreement on individual items (median phi coefficient = 0.91, range = 1.0–0.56) [5].

The CAMCOG is a concise neuropsychological test battery for the assessment of cognitive impairment in elderly people, which forms part of the CAMDEX schedule [6]. The CAMCOG was designed to assist in the diagnosis of dementia. It covers the broad range of cognitive functions that are known to decline in dementia [8], and includes items that assess all those areas of decline specified in operational diagnostic criteria, such as DSM-V [2] and ICD-10 [3]. The CAMCOG enables the examination of profiles of cognitive performance, through the derivation of subscale scores, and permits the measurement of cognitive decline across a wide range of levels of premorbid ability, by covering a wide range of item difficulty. The CAMCOG items are divided into seven subscales, covering the following areas of cognitive function: orientation, language, memory, praxis, attention/calculation, abstract thinking, and perception. A number of these broad areas are subdivided into more specific domains. Language, for example, is divided into comprehension and expression, and memory items include those to assess remote and recent memory, intentional and incidental learning, and recall and recognition measures of retrieval. A revised version of the CAMCOG (CAMCOG-R) incorporated alternative remote memory questions for younger participants and also included two additional items to assess executive function (EF) in more detail: ideational fluency and visual reasoning. These items were not included in the CAMCOG-R total score, but enabled the calculation of a separate EFs score.

The CAMCOG has been shown to be reliable when used in the general population [9]. Total CAMCOG score has excellent internal reliability (Cochran's alpha 0.82, 0.89 in different samples) and test–retest reliability (Pearson correlation 0.86) and the reliability of individual subscales is acceptable (Pearson test–retest reliability 0.46–0.80). The validity of the CAMCOG has also been confirmed in a number of studies. The CAMCOG total score and each subscale score have been found to differ significantly between individuals with and without dementia, in an elderly population sample

[9, 10]. The CAMCOG has been used in many published studies, both clinical [11–13] and population based [14–17]. Many neuropsychological [18–20], neuropathological [21–23], and neuroimaging studies [24–26] have utilized the CAMCOG for the assessment of elderly demented and non-demented participants in the general population.

Modification of the CAMDEX Schedule for Use with Adults with Down Syndrome

The CAMDEX-DS differs from the CAMDEX-R on which it is based, by placing a much greater emphasis on the informant interview as the key to an accurate diagnosis. It acknowledges that a combination of developmentally acquired ID and the possible development of dementia may make it difficult to obtain a reliable history from the affected person him/herself. This is particularly the case for those with pre-existing severe or profound ID. In addition, in order to take into account the substantial variation in the level of cognitive and functional ability across individuals with pre-existing ID, the informant interview included in the CAMDEX-DS schedule has been modified, to place greater importance on establishing decline from the individual's best level of functioning. While a direct cognitive assessment is still included in the CAMDEX-DS, we have suggested that testing cognitive ability at a single point in time does little to aid differential diagnosis. In the non-ID population deterioration can generally be inferred from the observation of a low level of performance relative to population norms. However, for individuals with an ID, it is particularly important to establish change explicitly, since cognitive impairment may be due to the underlying ID, rather than to the development of dementia. The cognitive assessment included in the CAMDEX-DS (which retains the same structure as the original CAMCOG, but has been modified to make it more suitable for use with people with ID) is intended to form a useful adjunct to the diagnostic process, when used to detect change over time through repeated assessment.

Modification of the CAMDEX Informant Interview

The modification of the CAMDEX informant interview took into account the fact that the cognitive and functional abilities affected by dementia may be impaired prior to the onset of dementia, due to the person's pre-existing ID. Questions elucidating the presence of a particular problem (e.g., "Does he or she have difficulty in remembering recent events?") are followed up with questions to determine whether this is a deterioration in the individual's behavior or functioning, or whether it has always been a problem (i.e., "Is this a deterioration?"). There must be evidence of deterioration in that particular function (e.g., memory), as observed by the informant, if, when rating the information against operational diagnostic criteria, it is to be scored as being present as a symptom of dementia.

While the majority of the questions included in the CAMDEX-DS informant interview are based on those included in CAMDEX-R, there has been some restructuring of the schedule, in terms of section headings and the questions included within each section, to increase the ease with which the answers can be related directly to diagnostic criteria. Questions from Parts I and II of the CAMDEX-R informant interview "Items concerned with history of present difficulty" and "Questions pertaining to the subject's past history" have been redistributed into three parts "Cognitive and Functional Decline" (which includes questions relevant to the identification of features of dementia), "Current Mental Health," and "Current Physical Health" (which include questions on other potential explanations for cognitive and functional deterioration, e.g., depression, thyroid disorder, sensory impairment or serious illness). The CAMDEX-DS informant interview begins with additional questions on "Patient/Participant's Best Level of Functioning," including questions on education and employment, basic skills (such as speech, language comprehension, reading, etc.) and independent living skills (such as dressing, food preparation, housework, etc.) in order to provide an overview of the individual's level of ability prior to the onset of any decline.

The "Cognitive and Functional Decline" part of the CAMDEX-DS informant interview begins with a section on "Everyday Skills," which covers changes in usual daytime activities, e.g., employment or day-center, preparation of food and drinks, housework, shopping (incorporating questions from various sections of the CAMDEX-R, including the direct "Interview with the patient/subject," with additional questions based on items from the Activities of Daily Living Scale [27], and an explicit question on "difficulties at work, college or day-center"). The next section covers "Memory and Orientation" and includes all the questions from the "Memory" section of the CAMDEX-R (on recent memory/forgetfulness and orientation to place) with the addition of questions on remote memory and orientation in time (see Appendix E for example of CAMDEX-DS questions). The section on "Other Cognitive Skills" covers general mental functioning, language, perception, praxis and EFs and incorporates questions from the "General Mental Functioning" section of the CAMDEX-R informant interview, with additional questions on the following: slowness of thought, deterioration in reading/writing ability, language comprehension, ability to carry out familiar complex tasks, and day-to-day problem solving. These additional questions relate directly to operational diagnostic criteria regarding decline in cognitive functions other than memory. The final section covers "Personality, Behavior, and Self-Care," and incorporates questions from the "Personality," section of the CAMDEX-R informant interview, with selected questions from the "General Mental Functioning" and "Everyday Activities" sections. Additional questions on loss of personality and emotional flatness are included to relate specifically to CAMDEX criteria for dementia.

Modification of the CAMCOG

The majority of the items included in CAMCOG-DS are taken directly from the CAMCOG and the structure of the assessment remains the same. For those items that were found to be too difficult for many people with ID, the item was modified

if possible, or otherwise replaced with an easier item assessing the same area of function. The revised version of the CAMCOG (CAMCOG-R) [4] provisionally included additional tests of EF that could be used to calculate a separate EF score. These were not included in CAMCOG-DS due to their high level of difficulty. However, there are two measures included in CAMCOG-DS that can be regarded as EF measures: verbal fluency and similarities. The modifications made to the CAMCOG-DS mean that people with ID are more likely to score above the floor of the individual domains of this test (and on the test overall) than on the original CAMCOG, thereby enabling any loss of function to be determined over time.

The items that were omitted from the CAMCOG-DS because they were too difficult for the majority of individuals who took part in our ongoing study of aging and dementia in people with DS were: orientation items on date, season, county, two nearby streets, and floor of the building, comprehension items requiring a verbal yes/no response, two items on retrieval of recent information ("Who is likely to be the next King or Queen?" and "What has been in the news in the past week or two?"), the most difficult expression definition item ("What is an opinion?"), calculation items requiring the addition of two coins of different values and calculating required change and the most difficult abstract thinking item ("In what way are a plant and animal alike?"). A number of items were included in the CAMCOG-R assessment that did not contribute to the total CAMCOG-R score (but enabled the calculation of scores for alternative scales). These items were also omitted: "Write a complete sentence," "ideational praxis," "visual reasoning," "passage of time."

Minor modifications were made to a number of items, either by way of simplification or adjustments to the scoring. For the orientation questions that were retained, the scoring was changed so that 2 points were awarded if the correct answer was given without prompting and 1 point if the answer was given after a multiple choice prompt. For the comprehension questions requiring a motor response, credit was given for partially correct responses, e.g., for the item "touch your right ear with your left hand," 2 points are awarded for the correct response, and 1 point if partially correct (i.e., touches ear but with wrong hand). Some simplification was also made to the sentence construction for two of the motor response items; "before looking at the ceiling please look at the floor" was simplified to "please look at the ceiling and then look at the floor" and "tap each shoulder twice with two fingers keeping your eyes shut" was shortened to "please tap each shoulder twice with two fingers." For the tasks requiring the naming, recognition and recall of six pictures, the task structure remains unchanged but the pictures have been updated. The typewriter has been replaced with a computer and the barometer has been replaced with clock (since very few participants were able to name this item).

The retrieval of remote memories section was one that individuals with DS found particularly difficult. Little success was achieved on these questions, even using those modified for use with a younger population from the CAMCOG-R. Questions such as "Who led the Germans in the second world war?", "When did the Second World War begin?," and "Which American president was shot it Texas?" were omitted and replaced with two questions more likely to be familiar to our target population, "Who was John Lennon?" and "Which princess

died in a car crash in Paris?" The scoring system was also modified so that 2 points are awarded if the correct answer is given without a prompt and 1 point is awarded if a clue is given (i.e., he was in a famous pop group, she was married to Prince Charles). The retrieval of recent information items also caused some difficulty and only the two easiest items were retained, "What is the name of the present king or queen?" and "What is the name of the prime minister?" Again the scoring was modified, as for the remote memory questions. The remembering a name and address item was included in a modified form. The majority of participants had been unable to write down the name and address on an envelope as required in the original item, so instead the participants are shown a picture of a man, and told his name and address, asked to repeat it and told to remember it for later. At a later point, after an intervening task, the participants is shown the picture of the man again and asked "What was this man's name?" and "What was his address?" In the copying and drawing section, the 3-D house was retained, but the scoring was altered so that points were awarded for each component successfully completed—up to a maximum of 3 points.

A number of difficult items were replaced with similar but easier items included in the Severe Impairment Battery [28], a test that was developed to assess decline in people with severe dementia that has been shown to be valid for use with people with DS [29]. The expression repetition item "no ifs and/or buts" was replaced with "People Spend Money." Two attention items, "count backwards from 20" and "serial sevens" (in which the participant had to start at 100 and repeatedly subtract 7 until told to stop) were replaced with simpler items, counting to 20, counting the number of fingers held up by the Examiner and a forward digit span task (requiring the repetition of digit strings of between 1 and 5 digits in length). The most difficult reading comprehension item "If you are older than 50 put your hands behind your head" was replaced with the simpler "Give me your hand." For the copying and drawing task the linked hexagons and spiral were replaced with a simple square and circle.

Validity and Reliability of the CAMDEX-DS

Validity and Reliability of the CAMDEX-DS Informant Interview as an Aid to Dementia Diagnosis

The validity and reliability of the modified CAMDEX informant interview for use in the diagnosis of DAT in people with DS were examined using data from a population-based study [30]. The concurrent validity of the instrument was found to be good. Diagnoses based on the CAMDEX informant interview were validated against objective evidence of decline in cognitive functioning. Decline was measured over a period of approximately 6 years prior to diagnosis, using the CAMCOG neuropsychological test battery. Diagnostic category was found to discriminate well between those who had previously shown decline of greater than the mean change

+1 SD in CAMCOG score and those who had not. Those with a diagnosis of DAT were at least eight times more likely to have shown decline in neuropsychological test performance over the preceding 6 years, than those without a diagnosis of DAT. Point estimates of sensitivity and specificity for the CAMDEX informant interview were shown to be high (0.88 and 0.94, respectively) and comparable with the levels found for the Dementia Questionnaire for Mentally Retarded Persons (DMR) and Dementia Scale for Down Syndrome (DSDS) [31]. However, the small number of participants with DAT in the study, and resulting width of the 95% confidence interval for the sensitivity score, mean that these should be interpreted with caution.

The predictive validity of informant interview-based diagnoses was also shown to be good. None of the DAT diagnoses made at the baseline assessment were reversed at follow up approximately 6 years later. Those with a diagnosis of DAT at baseline were at least six times more likely to be diagnosed with DAT (or have died following DAT) at follow up than those without a baseline diagnosis of DAT. The follow-up diagnoses were all made blind to knowledge of previous diagnoses, thus ruling out potential bias. Although numbers were too small for the authors to draw any firm conclusions, the study also provided some support for the accuracy of the CAMDEX in predicting cognitive decline.

Only three participants with DAT at baseline were able to participate in the neuropsychological assessment at follow up. However, all three showed a decline of more than the mean +1 SD on the CAMCOG, and this degree of decline was found to be significantly more likely to occur in those with DAT at baseline than in those without ($p < 0.005$, Fisher's exact). A number of participants were shown to have developed DAT in the 6-year period between baseline and follow-up assessment. These also showed decline in neuropsychological test performance.

Interrater reliability was also examined and shown to be very good. Data were reported for a subset of 20 people with DS, four of whom had DAT. The responses of the informants were rated simultaneously and independently by a psychiatrist, who conducted the informant interview, and a psychologist (one of the authors, SB) who observed. For each participant the ratings were compared for all items in the interview. Agreement between raters was shown to be excellent, with 91% of items falling within the "near perfect" range (Kappa >0.8) and all items showing an agreement of Kappa >0.6 (substantial), as defined by Landis and Koch [32].

Although the results of the study are highly supportive of the validity of the informant interview, the relatively small number of participants with DAT in the study limits both the strength of the conclusions that can be drawn and the degree to which validity measures can be compared with those of other methods of diagnosis. However, the CAMDEX informant interview is currently the only tool for the assessment of DAT in DS to have been evaluated with regard to predictive validity and to use internationally agreed criteria to make DAT diagnoses. This study is also the first in this field to have demonstrated validity as measured against objective evidence of neuropsychological decline (a much stronger comparison than clinician's diagnosis).

Psychometric Properties of the CAMCOG-DS

As discussed above, the CAMCOG-DS is included in the CAMDEX-DS schedule to provide additional information that is useful in the diagnosis of dementia in people with DS. Findings have been published regarding the ability of the CAMCOG to differentiate cross-sectionally between older and younger participants with DS [33]. Scores on the CAMCOG have been found to be well distributed, with only eight participants (11%) scoring zero on the test. This contrasted favorably with performance on the Mini-Mental State Examination [34] where there was a narrower range of scores and a higher percentage scoring zero. There was a significant difference in cognitive performance between younger (30–44 years) and older (45+ years) participants on the total CAMCOG score and on six out of the seven CAMCOG subscales. The study found that the CAMCOG, with minor modifications was a useful test to assess those areas of cognitive function known to decline with dementia. Apart from those with preexisting severe ID, severe sensory impairments and/or already advanced dementia, people with DS were able to score above the floor of the test.

In addition, participants with a diagnosis of dementia have been shown to decline to a greater degree on the CAMCOG than those without. The CAMCOG-DS included in the published CAMDEX-DS schedule has been further modified, to ensure that the majority of people with DS are able to score above the floor of the tests, thus better enabling the detection of cognitive decline. Validity not established for further modified CAMCOG-DS. Items with floor effects removed and replaced with easier items—should have the effect of making the measure more sensitive to the presence of dementia.

It should be noted however, that the CAMCOG-DS has limited diagnostic value at a single assessment, as without a baseline measure, it is not possible to determine the extent to which poor cognitive function is a consequence of a person's ID, any developing dementia, or any other disorder that might affect cognitive ability. However, the charting of decline in cognitive test scores over time provides a useful adjunct to the diagnostic process and may be constructive in informing support strategies.

Using the CAMDEX-DS

Administration of the Assessment Schedule

As outlined above, the CAMDEX-DS assessment schedule comprises an informant interview and a direct assessment of the patient/participant.

The CAMDEX-DS informant interview is a structured interview, comprising the following four parts: (1) best level of functioning, (2) cognitive and functional decline, (3) current mental health, and (4) current physical health. It has been designed to be carried out in the absence of the patient/participant, with a relative or

Q No.	Does he/she have difficulties with ...?			Is this a deterioration?		Slight deterioration	1
		Yes	1 →		Yes →	Great deterioration	2
		No	0		No 0		
		DK	8		DK 8		
		N/A	9		N/A 9		

Fig. 7.1 Example question

carer who knows him/her well. The interview should be face-to-face whenever possible, but satisfactory information can be obtained from telephone interviews. The interview consists of approximately 150 questions in total and takes around 40 min to complete. Its aim is to facilitate the systematic collection of information about the presenting symptoms and clinical history. As illustrated in Fig. 7.1, in the section of the interview focused on cognitive and functional decline, each question is in two parts, the first establishing whether there is a problem in a particular area (e.g., recent memory) and the second establishing whether this is a deterioration.

Due to the particular emphasis placed on the importance of observed change from the individual's baseline level of functioning, the interview should be carried out with an informant who has known the person with DS well, for at least 6 months. For each question, answers are coded as follows: "no" = 0, "don't know" = 8, and "not applicable" = 9. Positive responses are coded either as 1 for "yes" or are graded in terms of severity, e.g., for questions regarding whether there is a deterioration, "slight deterioration" is coded as 1 and "great deterioration" is coded as 2. These codes are intended to aid the recording and storage of data. It should be noted however, that they are not intended to contribute toward a total score. Diagnosis should be based on the rating of responses against diagnostic criteria as described below.

The section of the CAMDEX-DS schedule, that is, completed directly with the patient/participant him/herself, includes both subjective report and objective measurement of decline in function associated with dementia or other mental or physical disorders and comprises the following three parts: (1) clinical interview, (2) cognitive assessment, and (3) interviewer observations. The clinical interview is a brief structured interview with the patient/participant, consisting of 13 questions, covering basic background information, current mental state, and additional information regarding presenting symptoms of dementia. Interviewer observations regarding present mental state, appearance, and demeanor are recorded using a standardized schedule. For both, answers are coded as for the informant interview and information is intended to provide additional support to carer observations when rating against diagnostic criteria for dementia.

The cognitive assessment (CAMCOG-DS) has been modified from the original CAMCOG, as described above, with the aim of assessing all the cognitive deficits specified in operational diagnostic criteria, i.e., memory impairment, aphasia, apraxia, agnosia, and disturbance in thinking (EF), using tasks that are suitable for use with people with a preexisting ID. Items within each cognitive domain are graded in difficulty to permit assessment within the full range of cognitive ability. The assessment covers the following domains: orientation, language (comprehension and expression), memory (new learning, remote, and recent), attention, praxis

(drawing of complex figures and ability to carry out complex tasks), abstract thinking, and perception, all of which are known to decline with dementia. CAMCOG-DS provides subscale scores for hypothetically dissociable functions, as well as a total score with a maximum of 109. Each item contributes between 1 and 6 points to the relevant subscale and to the total score. Comparison of scores over assessments repeated at intervals of 6 months or more is intended to supplement subjective information regarding cognitive deterioration when making a diagnosis.

Diagnostic Process

In addition to the assessment schedule described above, the CAMDEX-DS pack also includes guidance regarding how to use the information gained through this assessment to inform the clinical diagnosis of dementia. The process of diagnosis essentially has three stages, as listed below, each of which is covered by one or more of the sections in the CAMDEX-DS:

1. A systematic history from the person him/herself and from an informant who has known that person over time, to establish the onset and course of the presenting problem (CAMDEX-DS patient and informant interviews).
2. A physical and mental state examination and cognitive assessments (CAMCOG-DS) and other investigations to enable the evaluation of present functioning and the identification of other possible causes of decline. The medical investigations should be guided by the clinical picture but invariably include investigations of a person's basic physical state (e.g., kidney and liver function and the presence or not of anemia) and specific tests, such as measures of thyroid function, or specialist assessment of hearing and/or vision. Where the clinical picture is unusual or the diagnosis is in doubt a computerized tomography (CT) or magnetic resonance imaging (MRI) brain scan may be indicated.
3. A detailed formulation and the evaluation of findings against known criteria for dementia and for other physical and mental disorders in order to arrive at a definitive diagnosis. For people with DS, three particular disorders are common and their presentation may mimic that of dementia as well as coexist with dementia and thereby make the disabilities associated with the development of dementia significantly more pronounced. These are depression, underactive thyroid gland (hypothyroidism), and visual and/or hearing impairments.

The diagnostic process leads to a formulation that brings together information from the various assessments and investigations and finally determines the likely cause of the observed clinical changes and sets them in the context of the individual. This is then the basis for developing an individualized care plan given the diagnosis and knowledge of the individual and his surroundings.

Incorporated in the CAMDEX-DS pack, are CAMDEX, DSM-IV, and ICD-10 criteria for dementia. Each set of criteria takes the form of a systematic checklist (as

A	**EVERYDAY SKILLS** **Progressive failure in performance of the common activities of every day life, not due to impairment in health or physical handicap.** **General deterioration of mental processes manifest as impairment or loss of:** • Skills necessary for usual activities at work, college, or day center • Ability to use household utensils and equipment

Area of Decline	Question		Slight Decline	Great Decline
Usual Daytime Activities	33	Usual daytime activities at work, college, or day center	↓ ☐	↓ ☐
	34	Special skill or hobby	↓ ☐	↓ ☐
Use of household Utensils / Equipment	36	Making a cup of tea	↓ ☐	↓ ☐
	37	Housework, e.g., dusting, dishwashing	↓ ☐	↓ ☐
	38	Preparing simple meals/snacks	↓ ☐	↓ ☐

40	Duration of changes greater than 6 months	**Yes** ☐
137–156	Changes due to ill health or physical handicap	**No** ☐

Criterion A Met	**Yes** ☐	**No** ☐

Fig. 7.2 Example diagnostic criterion

illustrated in Fig. 7.2). This is included as an aid to summarizing the information gained using the assessment schedule, as it relates directly to each criterion. The numbers of the relevant questions from the informant interview associated with each criterion are presented, and the amount of decline (slight or great) can be recorded. A judgment can then be made as to whether each criterion is met, and a diagnosis made on the basis of this information, in conjunction with information from physical and mental state examinations, cognitive assessments and other investigations. As stressed above, the aim of the CAMDEX-DS is not to provide a substitute for good clinical practice by rather to provide a framework to support the diagnostic process.

Guidance for Postdiagnosis Intervention

In recognition of the fact that the diagnosis of dementia marks the beginning rather than the end of a program of ongoing health and social care support, the CAMDEX-DS pack also includes a section providing guidance on postdiagnosis

Table 7.1 Summary of guiding principles for postdiagnosis intervention

1. Keep the person with dementia at the center of care planning
– Look at the person not the diagnosis and individualize care based on specific needs
2. Ensure all relevant people and agencies are working in partnership
– Family, advocates, GP, care manager, staff, professionals from community team
3. Forward thinking: prepare by being informed and anticipating change
– Consider where the person lives, daytime activities, training of care staff
4. Effective interventions, tailored to the individual
– Consider peer, family, and staff support, effective communication, memory books, interpreting challenging behaviors, environmental alterations, medication
5. Review and revise the person's needs and support strategies

intervention. Dementia diagnosis is the starting point for the development of a detailed and integrated plan to meet the continually changing needs of the person with dementia and his or her family. The first part of this section provides a summary of the key points that should be considered when planning support (outlined briefly in Table 7.1), while the second part consists of an example of an environmental checklist for residential homes, to help rate the suitability of the home for people with ID that develop dementia.

Review of Research Studies Using the CAMDEX-DS

The main application of CAMDEX-DS so far, has been its use in population-based research into dementia in people with DS. Diagnoses based on the informant interview that forms a major part of the CAMDEX-DS, have provided the basis for published estimates of the prevalence and incidence of dementia within this population. The informant interview and cognitive assessment (CAMCOG) have also been used longitudinally to chart the course of dementia in individuals who have been affected, providing valuable information regarding the sequence and timescale of decline in distinct areas of cognition, behavior, and functional ability.

Prevalence rates have been reported in the range of a few percent in those aged 30–39 years, between 10 and 25% in the 40–49 age group, between 20 and 50% in the 50–59 age group and between 30 and 75% in those over 60 [35–38]. Variations in these rates can be explained in terms of differences in diagnostic criteria and selection bias in the subject groups studied. In an attempt to overcome these problems, Holland and colleagues [36] carried out an unbiased population-based study of individuals with DS using a slightly modified version of the CAMDEX informant interview to diagnose dementia using standard criteria (including ICD-10, DSM-IV, and CAMDEX criteria for DAT) and provisional criteria for frontal-type dementia (FTD) [39]. Adults with DS over the age of 30 on July 1, 1994, within the catchment area (population 280,000), were identified through examination of health authority records, contact with community learning disability teams, contact with local private and voluntary services and direct contact with residential services for people with ID. Seventy-seven individuals met the inclusion criteria for the study and, of these, 75 agreed to take part.

Age-specific prevalence rates of dementia were found to vary according to the diagnostic criteria used, with more cases meeting CAMDEX compared to ICD-10 [3] and DSM-IV [2] criteria. Using CAMDEX criteria for DAT, prevalence rates were found to increase from 3.4% in the 30–39 age group to 10.3% in 40–49 age group and to 40% in the 50–59 age group. These rates are similar to those observed in the general elderly population but shifted forward by 30–40 years. However, in addition to those participants who met criteria for DAT, a number of participants met provisional criteria for FTD, showing changes in personality or behavior in the absence of decline in memory. While age-specific prevalence rates for DAT were found to be higher in participants over 45 years of age, prevalence rates for FTD were higher in the younger age group (<45 years), a finding that was taken to suggest that the presentation of AD in people with DS may differ from that in the general population.

In a follow-up study, Holland and colleagues [40] used the modified CAMDEX informant interview to determine the extent and nature of changes in memory, personality, general mental functioning, and daily living skills over an 18-month period. At the first assessment, carers of 35 (71%) of the 49 participants for whom changes had been reported, stated that the first change they had noticed was in personality or behavior rather than in memory or other areas of functioning. At the second assessment, estimated incidence rates for a clinical presentation resembling FTD (characterized by personality/behavior changes) were shown to be high and greatest in the youngest age group, while incidence of DAT occurred predominately in the older group. On the basis of these findings, the authors hypothesized that functions served by the frontal lobes are the first to be compromised with the progressive development of Alzheimer-like neuropathology in people with DS, perhaps as a result of the known underdevelopment of this brain region in people with DS [41]. It was suggested that the lower reserve capacity of the frontal lobes in this population, may increase the vulnerability of frontal lobe functions to the effects of AD neuropathology resulting in a clinical presentation resembling that of FTD occurring prior to the development of the full features of AD. It is important to note that it is not suggested that individuals with DS develop FTD (which, in the general population, is associated with neuropathology that differs from that associated with AD) but that AD-like neuropathology results in a presentation similar to FTD in the early stages of AD in this population.

A further follow-up of the same population sample approximately 5 years later [42] has provided further support for this hypothesis. Dementia status was reassessed using the CAMDEX informant interview and documentation of progression in clinical presentation suggested that the clinical course of dementia begins with early changes in personality or behavior and is followed by an increase in characteristics associated with frontal lobe dysfunction, prior to the development of the full features of DAT. Participants who met criteria for FTD (with five or more reported changes in personality/behavior) were found to be at a significantly increased risk (1.5 times) of progressing to a diagnosis of DAT over the following 5 years compared to those who did not meet FTD criteria. What is more, participants whose personality and behavior changes were insufficient for a diagnosis of FTD (i.e., for whom 1–4 changes were reported) were found to be at a significantly increased risk (1.5 times) of progressing to a more severe diagnosis (e.g., FTD or DAT) over this period than

those without such changes. This suggests that even limited evidence of change in personality or behavior is sufficient to increase the risk of dementia 5 years later.

In addition to examining clinical progression by way of informant reports, the CAMCOG cognitive assessment was completed at baseline and follow-up assessments, to provide a measure of decline in global cognitive function. Two additional measures were derived from the CAMCOG to examine specifically the sequence of decline in frontal lobe-associated executive function (EF) and memory. The EF measure combined scores for the abstract thinking and attention/calculation subscales and scores for the verbal fluency item and the clock drawing item, which has been found to have a strong EF component [43]. The memory measure combined scores for the memory and orientation subscales. Degree of decline on these measures over the 5 years prior to diagnosis was compared across groups based on diagnosis and age. The sample was divided into five groups as follows: those who met CAMDEX criteria for DAT, those who met criteria for FTD, those who showed personality/behavior changes insufficient to meet FTD criteria, those with no reported changes who were younger than 50 years, and those with no personality/behavior changes who were older than 50 years.

Participants who met FTD criteria and those with 1–4 personality changes had shown a degree of decline on the CAMCOG that was intermediate between that of those with no reported changes and those with DAT, and had shown a specific decline in EF with no significant decline on the memory measure. The DAT group, however, had shown a significant decline in both EF and memory over the preceding 5 years, but had show a significantly greater degree of decline in memory than in EF. These findings provide further support for the hypothesis that features similar to those associated with FTD are a precursor to the more marked cognitive deterioration associated with clinically diagnosed DAT. Interestingly, the group of older participants with no informant-reported changes decline to a greater degree than younger participants, but had shown a more generalized pattern of deterioration than individuals with informant-reported changes, with no significant difference in the degree of decline in EF and memory. This suggests that while age is likely to have an effect on cognitive function, such age-related changes appear to be distinguishable from preclinical AD.

The use of the modified CAMDEX in this longitudinal study has enabled the direct comparison of the clinical course of dementia in DS with that of dementia in the general population. This exploration of the differences and similarities that exist between the presentation of DAT in these two populations may serve to inform strategies for supporting individuals with DS who develop dementia and help to identify individuals at an early stage in the development of AD. This second benefit is likely to become increasingly important as new treatments become available that may halt or slow the progression of AD pathology.

In addition to the work carried out by Holland and colleagues, a number of other studies have also used the CAMDEX informant interview as a means of diagnosing dementia in this group. In a paper on the development of the Adaptive Behavior Dementia Questionnaire (ABDQ) [44], a brief 15-item questionnaire tool for screening for dementia in DS, Prasher and colleagues report that the CAMDEX informant interview was used to aid the diagnosis of DAT according to ICD-10 criteria. This diagnosis

served as the standard against which the validity of the ABDQ was established. Similarly, the usefulness of MRI as an aid to diagnosing DAT in people with DS was assessed by comparing MRI findings between individuals with and without clinically diagnosis of DAT (made using the CAMDEX informant interview) leading to the conclusion that the role of MRI was limited [45]. This method of diagnosis has also been used in a study investigating biological risk factors for dementia in DS. Rubenstein and colleagues [46] reported that apo-E genotypes are associated with similar risk effects in DS as they are in the general population, with apo-E4 allele carriers at increased risk of developing dementia (diagnosed on the basis of the CAMDEX informant interview) and apolipoprotein-E2 allele carriers at decreased risk. In a study examining behavioural disinhibition, apathy and executive function in pre-clinical Alzheimer's disease, Ball and colleagues [47] used CAMDEX-DS both for dementia diagnosis and for collecting information regarding the number and nature of informant report of behavioural/ personality and memory changes. The authors found out that disinhibition and apathy were correlated with impaired performance on tasks involving executive function. Landt and colleagues [48] investigated the relationship between plasma dehydroepiandrosterone concentrations, age and the risk of dementia in adults with DS (using CAMDEX-DS for dementia diagnosis) and observed a direct association. People with DS had lower concentrations of plasma dehydroepiandrosterone when compared with age-matched controls while those with DS and dementia had lower concentrations than those with DS without dementia. In addition, both CAMDEX-DS and CAMCOG-DS were administrated for dementia diagnosis and cognitive status in a study using positron emission tomography (PET) with Pittsburgh compound-B (PIB) in 49 participants with DS aged 25–65 to better understand amyloid deposition in their brain [49]. The authors concluded that abnormal PIB binding became evident from 39 years onwards and first appeared in striatum. No participants exhibited abnormal PIB binding in the hippocampus. Amyloid deposition was strongly associated with dementia (diagnosed on the basis of the CAMDEX-DS) and cognitive function (assessed by CAMCOG-DS).

In other studies, the CAMCOG assessment has been used as a measure of cognitive functioning in adults with ID. Beacher and colleagues [50] measured the association between concentration of myoinositol in the hippocampus and performance on the CAMCOG in older adults with DS and controls. The serum sodium/myoinositol cotransporter gene is located on chromosome 21, and myoinositol affects neuronal survival and function. In this study adults with DS were found to have significantly increased concentration of myoinositol compared to controls, and concentration of myoinositol was negatively correlated with cognitive performance. The authors suggest that further studies are required to relate myoinositol concentration to risk for AD in people with DS. Hassiotis and colleagues [51] describe the setting up of a memory clinic for older people with IDs, in which the CAMCOG is one of the instruments used to monitor cognitive function and decline over time. Oliver and colleagues [52] in their 4-year prospective study of age-related cognitive change used CAMCOG to analyze changes in orientation over time confirming the association between general cognitive deterioration and age. Another study using the longitudinal comparison of CAMCOG in all domains concluded that the occurrence of behavioural changes attributed to bereavement following the loss of the primary caregiver significantly increases the probability of cognitive decline in

individuals with DS [53]. Nevertheless, due to their ID, some participants performed poorly on some of the CAMCOG subscales that are suitable for the general population, leading the authors to confirm the appropriateness of adaptation of the instrument to the specifics of the population of individuals with ID, as it is the case for the modified CAMDEX-DS. In an exploratory study with individuals with ID subtests of the CAMCOG-DS were also used as a means of comparison with a visual association test [54].

The CAMDEX-DS has been translated and validated for the Spanish population obtaining good validity and reliability [55]. The instrument has also been translated into Portuguese and is currently in the final phase of validation for the Brazilian population.

Pros and Cons of the CAMDEX-DS

The major benefits of using the CAMDEX-DS to assess and diagnose dementia in people with DS are that (1) it enables the collection of information that maps directly onto standard diagnostic criteria for dementia; (2) it provides a structure for the collection of information regarding other potentially reversible disorders (e.g., depression), enabling a differential diagnosis to be made; (3) in relying on a formalized process for clinical diagnosis rather than a cutoff score, it enables the identification of individuals who may be suspected to be in the early or preclinical stages of dementia and who require close monitoring for further changes; and (4) it goes beyond diagnosis to provide guidance on intervention and support strategies for people with DS who are diagnosed with dementia.

However, the CAMDEX-DS is not a substitute for good clinical practice and does not eliminate the need for a full clinical assessment, with particular focus on those areas in which concern is highlighted through the use of the schedule. Clinical judgment remains the most important part of the diagnostic procedure. Furthermore, the schedule has been designed to provide a framework for a comprehensive dementia assessment and is therefore necessarily more time-consuming to administer than brief screening questionnaires.

While the CAMCOG direct cognitive assessment provides a quantitative score, which can be tracked longitudinally as an objective measure of decline, the diagnosis of dementia, based on the schedule as a whole, is a qualitative judgment so "degree of dementia" cannot be tracked in a quantitative manner as is the case for "scores" on dementia screening questionnaires. However progression in clinical presentation can be observed and reported on the basis of qualitative shifts over time.

Clinical Experience

Our experience in using this interview has been that carers and relatives of people with DS, who have known them for sometime are generally very perceptive to the subtle changes that occur in the person they care for abilities and behavior and,

Table 7.2 Summary of findings for cases mapped on CAMDEX-DS criteria for DAT

CAMDEX-DS criteria for dementia of Alzheimer's type (DAT)	Case 1	Case 2	Case 3
Progressive failure of the common activities of everyday life		?	?
Decline in memory sufficient to impair functioning in daily life			?
Progressive impairment in cognitive functions other than memory			
OR			
Deterioration of personality or general behavior			
Clouding of consciousness/delirium not present most of the time			
Gradual onset			
Deterioration not accounted for by other disorders		? Hearing, depression	? Eyesight, depression, antiepileptic medication
Diagnosis	DAT	Does not meet DAT criteria—possible preclinical features	Possible DAT

when prompted are able to provide specific examples of the kinds of changes that have occurred. In Appendix F, three case studies are presented that show examples of changes in behavior reported by carers, obtained using the CAMDEX-DS informant interview.

Also presented, in Table 7.2, is a summary of how these reported changes map onto CAMDEX-DS criteria for DAT, illustrating the degree to which moving from specific examples of change to a diagnosis requires the use of clinical judgment, supported by the framework provided.

What these case studies illustrate is that the information gained from the informant interview, may not in itself be sufficient to reach a diagnosis but may highlight areas of concern that need to be investigated before a diagnosis of dementia can be made or ruled out. In the case of Michael (Case 2) for example, the informant interview indicated that he suffered from a hearing impairment that was corrected by the use of a hearing aid. Such a finding should prompt an investigation as to whether the hearing aid is functioning properly or whether any reported changes could be due to hearing difficulties. The observation that he is now much more prone to crying than he used to be, in conjunction with the fact that he is currently on antidepressant medication should prompt a review of this medication and a full investigation into the presence of other features suggestive of depression.

In the case of Mary (Case 3), who has a severe ID, the range of her abilities was so limited prior to any signs of dementia that it is difficult to establish whether deterioration has occurred. However, when prompted the carer was able to come up with specific examples of change, such as the fact that she has stopped singing songs (previously her favorite activity). Clinical judgment is required in order to conclude whether such changes are sufficient for diagnostic criteria to be met. Again, potential

explanatory factors such as antiepileptic medication, features of depression and poor eyesight as a result of cataracts are highlighted as requiring further investigation.

Summary

With the changing age structure of populations, dementia and other illnesses related to old age are now the focus of very considerable research and policy attention in general. Given that people with DS have this high risk of DAT at a relatively young age their needs and the effectiveness of any new treatments for DAT must be considered in this population. Reliable diagnosis and the ability to track decline is central to both treatment development and treatment trials and is also important in informing social care policy and support strategies. We believe that the following research and clinical issues require particular attention: (1) the identification of other individual or environmental risk or protective factors that modify the age of onset and course of dementia in people with DAT; (2) ethically and clinically sound trials in people with DS of treatments, as they are developed, aimed at the prevention or the amelioration of DAT; (3) the education and training of paid and family carers about the relationship between DS and DAT, how it presents, and what support strategies are known to help maintain the quality of life of people with DS and dementia; and (4) the ultimate goal is establishing the underlying mechanism that accounts for the high risk of DAT in people with DS, and specifically whether overexpression of the amyloid precursor protein gene (located on chromosome 21) is the main etiological factor. Only then will new treatments be developed that are tailored specifically to people with DS. Each of these objectives, to varying degrees, requires the involvement of people with DS and their carers and the ability to detect with a high degree of certainty whether dementia is developing or has developed. Research therefore requires instruments such as the CAMDEX-DS and strong partnerships between people with DS, their families and paid carers, clinicians, and basic scientists.

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Chapter 8
The Test for Severe Impairment

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Introduction

It is expected that the number of people with dementia will reach 66 million by 2030 and 115 million by 2050 [1]. It has also been noted that the number of cases of early onset dementia and young onset dementia (under 65 and under 45 respectively), while still infrequent are rising [2]. Overall prevalence rates of 5–7% have been reported in those over 60 [3] with prevalence rates of 2–10% for those under 65 years reported [1]. As has been widely reported, these figures are starkly different to those seen in a population of people with intellectual disabilities (ID), and in particular for those with Down syndrome (DS). Individuals with DS have a third copy of chromosome 21, trisomy 21, and this leads to 4–5 time the expression of amyloid precursor protein (APP) [4]. This overexpression of APP leads to increased amyloid deposition in the brain. This in turn leads to an increase in amyloid B deposition [5], where Amyloid B is known as a key contributor to Alzheimer's disease (AD). A number of positron emission tomography (PET) studies have shown abnormal

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amyloid binding in individuals with DS [6] with abnormal binding found in those as young as 39 [7]. Thus, while these studies have shown neuropathological hallmarks prior to the fourth decade, the average age of clinical diagnosis has been found to be between 51 and 56, with an average duration of 3.5–6 years [8–10].

Strydom [11] in a review of published studies (1997–2008) reported prevalence rates accelerating from 9% in those under 49 years [12], 5.7–10.3% in ages 40–49 years [13, 14], 30.4–40% in ages 50–59 years [12–14] and 42–50% in those aged 60–70 years [14]. An earlier report of a 100% prevalence rate in adults aged 65 years and over [15] appears largely confirmed in a recent 20 year follow-up of 77 women with DS. Findings were that 97.4% developed dementia with a mean age at diagnosis of 55 years (SD = 7.1) and an 80% risk of dementia was estimated at age 65 [16]. There are therefore pressing needs for assessment and diagnosis of dementia in persons with DS, usually at ages where such assessment has not been traditionally considered for the general population.

As noted earlier, there is growing recognition that there is a small and growing number of persons in the general population who will have early and young onset dementia with reports that diagnosing such dementia is often fraught with difficulties and timely diagnosis poses numerous challenges for clinicians, confounded by patient heterogeneity, different clinical features and often confusion with mental health problems [2]. Such challenges are further confounded in people with ID and complicated by pre-existing intellectual impairment, difficulty in using standardised tests, communication difficulties, care environment concerns, lack of base line performance data and the consequences of high staff turnover [17].

Despite the growing emphasis for the general population on seeking short screening instruments [18], the growing consensus for diagnosing dementia in people with DS and non-DS ID is predicated on having an understanding of decline/change from the individual's previous level of functioning [19] supported by a reliable baseline measure of functioning against which to compare and a key informant who has known the individual over an extended period of time. The needed tools are rarely available; baseline measurement of functioning is more often an exception rather than the norm, and both frequent staff changes in out-of-home placements and lack of regular assessment in family situations often means there is poor knowledge, understanding, or measurement of decline/change.

Standard neuroimaging such as computerised tomography or magnetic resonance imaging scanning generally used to support diagnosis in the general population is another option but it too has proven less helpful in people with ID. The most consistent structural change of early AD in the general population is atrophy of the medial temporal lobe, but among people with DS, for example, medial temporal lobe atrophy occurs at an earlier age and is totally unrelated to dementia. Also, the lack of standardization within our understanding of dementia in different syndromes associated with ID has led to conclusions that neuroimaging is of limited value to the diagnosis of dementia in people with ID [20].

There is even greater diagnostic uncertainty in older age as many adults with ID, especially those with DS, are also at increased risk of other health conditions which often mimic dementia and/or confound diagnosis such as hypothyroidism, sensory impairments, B12 and folate deficiency, and depression [21]. There are also chal-

lenges for persons with ID who also have been diagnosed with head trauma or brain injury [17]. Achieving accurate diagnosis will only happen if there is a consistent approach to diagnosis, and accurate information available to measure and evaluate change in the context of the individual's premorbid level of functioning. The availability of useful instruments with demonstrated properties is critical to meeting this need as is the availability of protocols and mechanisms for routine expert assessment. Prompt and accurate diagnosis of dementia is also frequently associated with memory clinics for the general population [22] but has been less so for people with ID with reports that communication barriers, lack of experience in interviewing people with ID, poor understanding of decline in the context of pre–existing impairment, and age-based criteria to access services [23]. Now, however, there are some memory clinics within ID services [16, 19, 24–26] and the present analysis draws data from such a memory clinic.

Burt and Aylward [27] reported the findings of a working group, established under the auspices of International Association for the Scientific Study of Intellectual Disability (IASSID) and the American Association on Mental Retardation (AAMR), which proposed a battery of tests to aid the diagnosis of dementia in individuals with ID. They identified that differentiating between changes associated with normal aging from dementia-related changes posed a significant challenge. Both informant-based scales that report on an individual's functioning and tests for direct assessment were included in the proposed battery. The importance of informant information was stressed, as was the need for longitudinal assessment. In order to establish a healthy baseline, the authors recommended that all individuals with DS should be assessed for the presence of dementia before the age of 40 years and before the age of 50 years for those with other causes of ID. Periodical reassessment should then occur depending on the age and symptoms of the individual. These recommendations continue to be endorsed [28]. Nevertheless despite continued efforts in the development and validation of both informant-based (carer-rated) and objective test instruments (client-rated), there is as yet no agreed consensus on the optimal battery of test instruments to be used in detecting and diagnosing dementia in persons with varying degrees of ID. This chapter reports on work to better understand the value of the client-based assessment, the Test for Severe Impairment.

Development of Test for Severe Impairment

The Test for Severe Impairment (TSI) (see Appendix G) was developed to provide a test of cognitive function for people with severe cognitive impairment [18]. Work with nursing home patients has further supported its sensitivity in measuring moderate to severe impairment [29]. In addition to being a valid and reliable tool, the test was designed to be nonthreatening, appealing, easily administered, time efficient and uses small readily available objects. The TSI is a 24-item cognitive test that takes 10 min to administer. It tests a broad range of cognitive functions and was designed for use in people from the general population whose MMSE score is less than 10 out of 30. The level of difficulty of the TSI is such that most people with

moderate and severe ID should be able to score on it unless they are at an advanced stage of dementia. Also, a wide range of skills is tested including: language, memory, conceptual ability, and spatial skills. In total, the test contains six subsections each containing four items: well-learnt motor performance (fine and gross), language comprehension, language production, immediate and delayed memory, general knowledge, and conceptualization. Each item is either scored correct or incorrect. Each subsection has four questions, giving a total maximum score of 24, but the test was not designed to generate discrete subscale scores. Only eight out of the TSI's 24 items require the subject to answer a question verbally. This may be of benefit when testing those persons with DS who have limited verbal abilities.

Albert and Cohen's original study involved 40 residents of a chronic care facility with a variety of types of dementia [30]. The MMSE was administered and only subjects scoring in the severe range (<11) were included in the study. Construct validity, external reliability, internal consistency, and factor structure were all studied establishing the TSI as a reliable and valid instrument. The internal consistency (alpha coefficient = 0.91) was considerably higher than that for the MMSE in the severe range ($a = 0.56$). It was suggested therefore that the TSI could complement the MMSE and give reliable scores for persons where the MMSE is exhibiting floor effects.

Foldi and colleagues [31] reassessed the TSI and compared it to the Dementia Rating Scale (DRS) [32]. The DRS was developed to measure more severe impairment than the MMSE, however it requires training and time to administer making it less practical for use in a long-stay facility. They investigated the TSI's validity, reliability, and range. When criterion validity was calculated using the TSI and DRS total scores, the resulting correlation supported it being considered a valid screening tool ($r = 0.88$). A strong correlation was particularly noted in the memory and conceptualization domains, but weaker, nonsignificant correlations were found for language comprehension and production items. Indeed, when compared to the DRS scores the only item not to reach a significant level of correlation was the TSI item on language comprehension. Test–retest reliability and internal consistency reliability calculations both yielded high reliability scores. Further analysis suggested the TSI to be a tool applicable across a wide range of ability, not just for those with severe impairment. An additional aspect of this study correlated the TSI total score with the Boston Naming Test (BNT) [33] in an attempt to determine how well the TSI captures changes in naming skills. The total TSI and BNT scores correlated and in particular the language production score of the TSI correlated highly with the BNT. The authors suggested that the TSI may be of particular use when there are time constraints, more severe language impairment or the Examiner is not formally trained in psychological testing.

Jacobs and colleagues utilized the TSI in a longitudinal study of those with dementia but without ID [34]. Scores on the TSI were compared to results from the MMSE and the modified MMSE (mMMSE) [35]. The mMMSE was constructed to strengthen perceived weaknesses in the MMSE, namely in the language, attention, and construction subsections. The TSI and the MMSE were found to be highly correlated ($r = 0.83$). Of particular note was the greater range of scores on the TSI for those obtaining very low scores on the MMSE. The mMMSE also correlated well with the TSI ($r = 0.82$). Those scoring in the severely impaired range of the mMMSE

also produced a wide range of scores on the TSI, further supporting the relative robustness of the TSI in avoiding floor effects.

A modified version of the TSI (mTSI) was administered with the MMSE to 130 elderly females with moderate- to end-stage dementia but without ID [29]. In the modified version a facilitating cue was offered if the first response was incorrect; however, the number and content of the items were unchanged from the original TSI. Two points were scored for an outright correct answer and one point was offered for a correct answer following the facilitating cue. No points were given if the answer was incorrect or not given. Therefore the maximum score for the mTSI was 48 points. The mTSI score was different from zero significantly more often than the MMSE. In addition, only 9.2% of TSI items required a facilitating cue to give a correct answer. A limitation noted for both the MMSE and the mTSI was that approximately one-third of subjects were not tested due to behavioral concerns or the severity of their dementia. Appollonio surmised that this was likely to be a general limitation of performance-based instruments. A further study compared the performance-based mTSI with the observer-based Bedford Alzheimer Nursing Severity Scale (BANS-S) [36, 37]. Neither test was optimal, the mTSI appearing more useful in moderate to severe dementia, whereas the BANS-S mean scores only worsened in the later stages of the disease.

Validity and Reliability

Reports on the ease of administration and the likelihood of finding a range of scores among people with severe impairment suggested that the TSI might be a useful tool in the investigation of dementia in those with ID [38]. In an initial use of the TSI in the DS population, Cosgrave and colleagues assessed its validity and reliability in 60 older persons with DS [39]. The Down Syndrome Mental Status Examination (DSMSE) was administered in conjunction with the TSI. The DSMSE tests recall of personal information, orientation to season and day of the week, short-term memory, language, visuospatial construction, and praxis [40]. Comparing the results of both tests administered by the same rater indicated the convergent validity of the TSI for all subjects as 0.94. Interrater reliability for the TSI was satisfactory at 0.97 and test–retest reliability over 2 days yielded a concurrence of 0.98. Internal consistency of the TSI measured using Cronbach's alpha was 0.89. It was further reported that the TSI was brief and easy to administer and yielded a range of scores across all groups tested with the exception of those with severe ID and dementia. However, when compared with the DSMSE, the TSI provided a greater range of scores in the severe ID group. This finding in particular suggested the TSI would have greater utility as a tool in longitudinal testing, as it appeared less susceptible to the floor effects found in other instruments. Of concern, however, was that well-learned motor performances appeared to be retained until the later stages of the disease leading to recommendations that it should be used in conjunction with an observer-based rating instrument such as the Early Signs of Dementia Checklist [41, 42].

Rates of change on TSI scores in those with dementia in the general population were previously noted to be greatest in the middle stages of the disease with an aver-

age annual rate of change of 3–4 points [34]. Cosgrave and colleagues found a similar rate of score change of 3.2 points per year on the TSI in a 5-year study following 80 individuals [41]. However, the change was not linear with more modest reductions in early and late stages of dementia. The earliest items of the TSI to be affected in those with moderate ID and dementia included stating the number of weeks in the year, delayed memory, name writing, and counting to ten. In those with severe ID many items were answered incorrectly at the time of diagnosis except for learned motor responses. With the progression of dementia the last TSI item to be lost was shaking hands with the Examiner.

Tyrrell and colleagues also reported on the use of the TSI in a cross-sectional study of 285 persons with DS; of these, 185 lived in an institutional setting and 100 in the community [14]. At baseline testing, 38 cases of dementia were diagnosed according to DSM IV criteria giving a prevalence of 13.3%. The data gathered were subjected to logistic regression analysis yielding a model where scores on the Daily Living Scale Questionnaire (DLSQ), age and presence of epilepsy yielded the best fitting model for predicting dementia [43]. Neither the TSI nor DSMSE scores appeared predictive of dementia when analyzed in this manner. At year 2 there were 266 persons in the study including 46 persons with dementia, of whom 14 were newly diagnosed. Delayed and short-term memory, comprehension, and expressive language all appeared significantly impaired between year 0 and year 2. The annual rate of change of scores on the TSI was 1.4 (SD = ±2.6) for the dementia group and −0.35 (SD = ±1.2) for the non-dementia group representing a significant difference ($p = 0.0001$). These changes were modest when compared with reports from the general population. Cosgrave and colleagues' findings with persons with DS also included a higher annual rate of change than calculated by Tyrrell and colleagues [14, 41]. Tyrrell and colleagues' findings may reflect that the baseline scores may have included those who already had severe dementia and that the 24-month follow-up period was too short; detection of changes may require a longer time frame to become manifest.

The cohort studied by Cosgrave and colleagues is also important because it initiated a larger, longitudinal, and cross-sectional study. The 20 years of data collected to date in this larger study is presented here focusing on the findings in relationship to the TSI.

Methods

At time of entry into the study there were 80 women with DS living within the programs of one large service providers. Three individuals were lost to follow up and thus were not included in analysis; results are presented on the remaining 77 individuals. The mean age of subjects at commencement was 47.7 years (SD 8.4, range 35–71 years). Thirty-six of the subjects were living in long-stay residential type units, 24 subjects were living in a community setting, and 17 were living in campus group homes.

The same experienced clinician periodically assessed each subject over a 20-year period for the presence of dementia. Upon identification of symptoms, a dementia-

specific team including a psychiatrist, psychologist, and physician reached consensus on the diagnosis of dementia using ICD criteria. Comorbid conditions likely to mimic dementia and known to be more common in aging persons with DS were ruled out as recommended by Pary [44].

Measures

Following the recommendations by Burt and Alyward [47] a comprehensive assessment was conducted including a clinical examination along with a review of medical records. This review included the establishment of level of ID, an information on history of cardiovascular health, lunch disease, diabetes, epilepsy, depression, vision and hearing impairment and gastric disease. Then a full physical examination included urinalysis, geriatric blood tests and mental health assessments. Finally, standardized measures of cognitive functioning, repeated annually, track any decline.

The study protocol was maintained over the 20 year period, allowing for purposeful and consistent tracking of any changes that may have occurred. A diagnosis of dementia was given only after all other options had been exhausted and following a consensus meeting with members of the person's multi-disciplinary team including the clinical nurse specialist in dementia, caregivers/family, consultant psychiatrist, and psychologist using the modified ICD criteria. All data was analysed using SPSS 21.

The cognitive measures used in the study were:

The Daily living Skills Questionnaire (DLSQ) [43] which measured activities of daily living (ADL).

The Dementia Questionnaire for People with Learning Disabilities (DLD) [45, 46] was first administered in 2005 and then repeated annually.

The Down syndrome Mental Status Examination (DSMSE) [30] is an objective measure and was repeated annually.

Finally, and of interest for this chapter, the Test for Severe Impairment (TSI) [19] was administered as a second objective measure.

Calculation of Rate of Change

The method utilized to calculate the annual rate of change on the TSI, DLSQ and DLD has been reported in previous longitudinal studies of this population [8] and was applied consistently here. Annual changes in scores for the entire follow-up period for each person regardless of dementia status was calculated by dividing the change in score over this time by the numbers of years of follow-up. Not all the data collected for each subject were used in this approach; however, the "restricted two-point estimate" [48] was deemed statistically more preferable. The results could potentially have been skewed by uneven contributions as the number of assessments and time points differed by person.

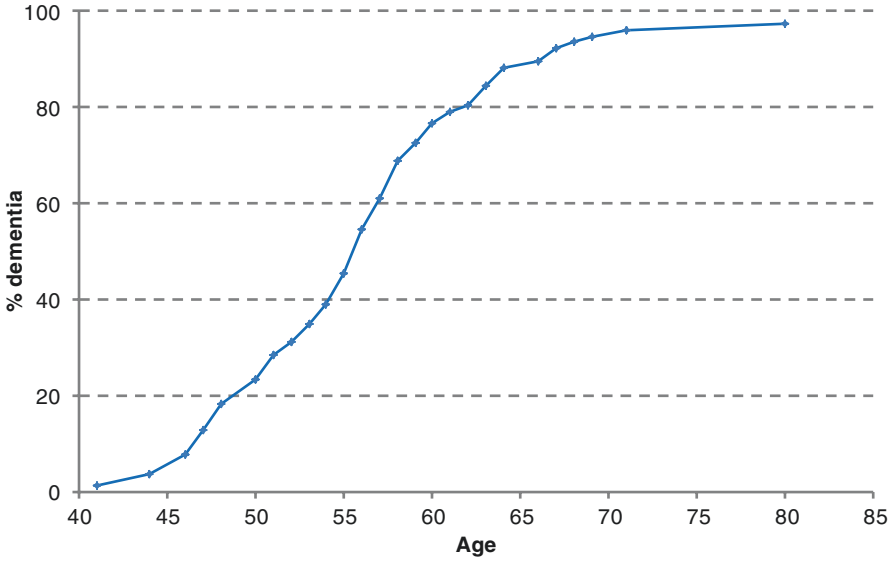
For people with a previous diagnosis of dementia, a baseline score was utilized and was defined as the score in the year prior to the diagnosis of dementia or their year 1 score if they entered the cohort with dementia. The annual rate of change was calculated for those without dementia over their entire follow-up period. The scores of subjects whose scores had already "floored" were excluded from the analysis beyond that point as no further change was possible and further inclusion of scores would depress the change score of interest.

For the TSI, changes in scores 2 years prior to diagnosis and 2 years after were examined as were the TSI scores for individuals at least 4 years prior to their diagnosis to further assess what items in the TSI appeared to change in the period immediately prior to diagnosis.

Results

Of the 77 individuals in the study, 97.4% developed dementia over the course of the 20 years study period. For those with moderate ID, 96.7% (59/61) developed dementia and 100% (15/15) of those with severe ID developed dementia. The average age of diagnosis was 55 years. At the time of follow up, one individual had died without dementia and another was still alive without a diagnosis of dementia [16].

Age specific incidence rates were also calculated where is was found that the risk of dementia was 23.4% by age 50, increasing to 45% by age 55 and at 88% by the age of 65.



Due to the longitudinal nature of the study, it was possible to track scores on the DLSQ, DLD, TSI and DSMSE from 5 years prior to diagnosis and 5 years post

diagnosis. Pre diagnosis data was unavailable for 18 individuals who already had a dementia diagnosis prior to the beginning of the study.

An increase in scores on the DLD indicates lower level of functioning with DLD scores plotted on the secondary axis on the graph below. Scores on the DLD declined over the period of 5 years pre diagnosis, while scores of the DLSQ declined 3–4 years prior to diagnosis. The differences in these scores were not statistically significant. Decline was more pronounced for those with a moderate ID than for those with severe ID. Given that original scores were higher, decline may have been more observable for these individuals.

For 15 participants there was TSI data available for between four and 11 years prior to diagnosis. There were minimal or no changes in scores during this premorbid period other than a small decline for some in the conceptualization subscale. On the other hand for 20 participants for whom there was TSI data available 2 years prior and 2 years after diagnosis, examination of the scores revealed both increases and declines in all subscale scores in the 2 years prior to diagnosis.

Discussion

Over the 20 years the approach has proven effective in tracking what was ultimately significant with declines 97.4% of the 77 adults with DS developing dementia, which corresponds to a risk of 23% at age 50; 45% at age 55; and 88% at age 65. Age was the principle risk factor for developing dementia was age but with epilepsy present in 77.9% of the participants and in 75% of cases it was notable that both diagnoses were established within 3 years. Epilepsy was a predictor of mortality and by the end of the study 74% of the participants had died, all but one with a diagnosis of dementia of mean duration 6 years. The 20 year follow-up offered an opportunity to observe the value of various assessment scales, particularly their ability to predict later diagnosis.

A consistent approach through a memory clinic model to assessment and diagnostic work up for dementia in persons with ID also appears confirmed as critical in these findings. It appears that with such follow-up the chances for accurate and timely diagnosis, and, in turn, opportunities for discussion with the person and their family/staff carers it means that plotting of trajectory of dementia and likely prognosis as well as more timely application of symptomatic therapies, both pharmacological and non-pharmacological, is more likely and may be tailored to improve quality of life for the individual.

The success of such a process is heavily dependent upon the assessment instruments used. The TSI has been previously recommended as a sensitive tool to measure change in persons with ID [30]. Use in both cross-sectional [14] and in 5-year and 14 year follow-up studies [8] have already suggested that the TSI is a useful instrument for persons with DS. The findings here further support use of the TSI as a reliable and valid dementia test in this population although comparison of findings, particularly with the DLSQ and the DLD suggest there are some limitations.

The administration of the TSI takes 10 min yet it assesses six different cognitive domains and the equipment is easy to carry and readily available. The test is short, easy to use and the findings here support that it is applicable across the range of dementia and levels of learning disability. The need for intact speech is minimized in comparison to other performance-based assessment tools and its range of use enhances its utility in longitudinal studies. However, ease of use does not reduce the need for training in using the TSI to ensure consistency in application and scoring. A continuing limitation, but less prominent than for other instruments, is that the TSI was found here and in other studies to demonstrate a ceiling effect in persons with upper moderate and mild ID, and a floor effect in those with the most severe level of ID. Indeed, the TSI was found to produce a range of results in situations where subjects scored at or near zero in other tests. These are important findings; such sensitivity suggests for example that it may be feasible to use the TSI to monitor the effects of pharmacological interventions in dementia. As has been previously recommended [14], the TSI should be augmented with an additional test of memory such as the Modified Fuld Object Memory Evaluation [49] and measures of functional ability. The findings here also support use in particular of the DLSQ and the DLD.

Despite the reported strengths, the findings here are also consistent with other reports that a minority of subjects are unable to participate in testing due to severity of dementia or the presence of behavioral difficulties. Similar to floor effect concerns, with the TSI, this proportion however, appears to be less than with other tools and may be a general limitation of performance-based tools for persons with ID. The authors continue to be concerned that sensory deficits such as color blindness or deafness may confound the administration of the TSI in a small proportion of subjects and they encourage attention to sensory deficits as potential sources of both the difficulties being investigated as possible signs of dementia and as barriers to effective use of assessments. A further limitation is that while the TSI provides gross evidence of decline, it gives no indication as to the potential cause. The content of the TSI may also benefit from slight alteration, as certain items such as stating the number of weeks in a year, appear universally difficult at baseline. In addition a measure of orientation would enhance the usefulness of the test without compromising its benefits.

Frequently with screening instruments clinical cutoff scores are established, sometimes to facilitate diagnosis and more often to support the need for additional assessment. Given the range of cognitive disability already present in persons with ID, creating such cutoff scores for the TSI would not be possible or useful. An alternative strategy has been to give greater attention to developing annual rate of change scores. In the general population the rate of change on TSI scores in those with mid-stage DAD have been reported as 3–4 points annually [34]. As was noted earlier, Cosgrave and colleagues reported a similar annual rate of change over 5 years for persons with DS [41]. However, Tyrrell and colleagues in a larger study found over 2 years a rate of change of approximately 1.4 per annum.

The results here are more mixed. On the one hand examination of data 4–11 years prior to diagnosis in an admittedly small [15] group of individuals

prior to dementia diagnosis found very stable scores year to year. For a group [20] with dementia where we could establish scores 2 years prior to and 2 years after diagnosis, while there was an overall decline in scores similar to previous reports, there were also fluctuations in scores with increases as well as declines year on year both before and after diagnosis although decline was more prominent after diagnosis. Perhaps it is change in scores rather than a decline per say that would be a signal to be concerned that dementia symptoms may be present and deserve closer investigation. Equally the more consistent decline in TSI scores after diagnosis may suggest that the TSI be considered as a tool for confirming transitions through the stages of dementia in people with DS. Daily functioning scores for the group with dementia in this study more consistently declined over time and for the group without dementia there was some decline but at a slower rate and DLD scores most consistently declined in the last 2 years before diagnosis suggesting that these instruments may be more useful for tracking the decline to diagnosis. One factor that has not been accounted for in this study is the effect on the rate of decline of antidementia medications that have been introduced in recent years.

Summary

The longitudinal use of the TSI and the monitoring of rates of change appear to confirm its usefulness with an early baseline for each individual serving as a marker against which to compare later scores. However other scales may better identify initial decline so it should not be used alone. The TSI may instead be more useful for tracking further decline post diagnosis, particularly when other scales more quickly hit floor and ceiling effects. Given these findings, the continued regular application of the TSI in clinical practice is recommended. However, measures of memory, adaptive behavior, and informant-based measures should be included to expand the clinical picture. When used in the advised manner the TSI appears to be a reliable and valid tool likely to aid in the diagnosis of dementia in those with ID.

Case Vignette

From 1996 and up to 2009, Ms. A was described as being a very independent woman who required minimal support in day-to-day activities of living. She could be relied upon to look after all personal care needs, and would actively participate with the various chores around the home, all with reasonable frequency and without reminding.

She would always ensure that her peers attended to their daily chores to her exact standards, but she was also very maternal and caring.

She was orientated to time, place and person. She was familiar with the routine of the day/week/month, could structure her day independently, and had no difficulty in remembering appointments, birthdays and scheduled activities. Cognitive skills were good and she enjoyed participating in a varied activity programme in her workplace and in the home setting. She had a very successful career in Special Olympics, and was described as being a 'social butterfly', who thoroughly enjoyed every social opportunity that presented itself.

Both long and short term memory were good. She could describe in detail what had happened over previous days and what her plans were for the coming days and weeks.

In 2009 (55 years old) staff noted hard to describe changes in personality; any changes in daily routine caused irritability and there were occasional instances of apathy and loss of self-direction.

In 2010 (56 years old), staff reported subtle decline in day-to-day activities of daily living and occasional episodes of confusion and forgetfulness. She became mostly apathetic about previously enjoyed activities. There was increased evidence of reduced emotional control, with accompanying irritability, and she had great difficulty in describing her feelings. She became quite 'faded' and this previously 'bubbly' lady was beginning to withdraw from her social networks.

Ms. A required increased levels of prompting to carry out routine tasks. Although her ability to carry out household chores remained meticulous she had significantly slowed down. Instructions had to be repeated several times over before she was fully aware of what was being asked of her. Routine monitoring of cognitive and functional status using established scales measured change from previous level of functioning and a full diagnostic work up carried out in June 2010 included a full physical examination, psychiatric review, neuroimaging and a full geriatric blood screen. Other potential causes of decline were subsequently out ruled. A consensus meeting of the team which included consultant psychiatrist, clinical nurse specialist in dementia, key worker and family member input and review concluded that presenting changes were consistent with early stages of Alzheimer's dementia.

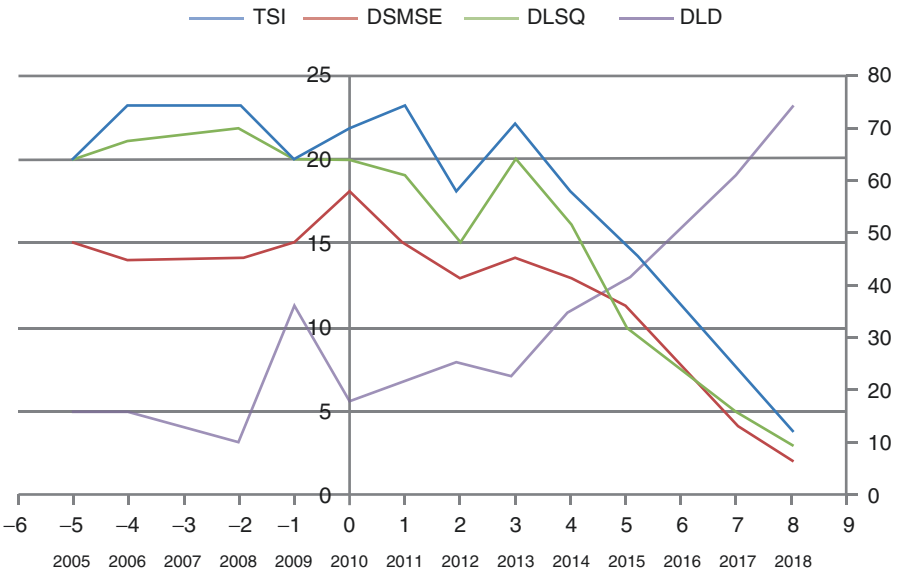
Ms. A continued to present with on-going decline in attending to personal hygiene, dressing, and self-care. There was notable decline in motivation and she was not as interested or meticulous about her overall personal appearance. She would wear the same clothing over and over—which was very out of character.

Continued decline from 2010 to 2014 was gradual and decline was supported in scores on neuropsychological objective test instruments. See table below.

	2017	2016 Aug	2016 Jan	2015	2014	2013	2012	2011	2010	2009	2008	2006	2005
TSI	4	8	14	15	18	22	18	23	22	20	23	23	20
DSMSE	2	4	4	11	13	14	13	15	18	15	14	14	15
DLSQ	3	5	4	10	16	20	15	19	20	20	22	21	20
DLD	74	61	58	42	35	23	25	22	18	36	11	16	16

From 2014 Ms. A's decline was at an increased pace. Concentration was poor and it was increasingly more difficult to engage and focus her attention. She moved home settings to support her growing level of need.

As may be seen in Graph below, prior to diagnosis, Ms. A's scores on the TSI, DSMSE and DLSQ began to fluctuate, with notable declines at 2 years post diagnosis (2013). Between 2014 and 2016 decline was accelerated and compressed. There was on-going evidence of confusion, impaired memory and forgetfulness. She was disorientated to time and no longer knew the days of the week, or the routine of the day. There was evidence of reduced emotional control, emotional liability and apathy. She required increased supports for all activities of daily living, as reflected in the DLSQ score (2015).



In 2016, agnosia were becoming more and more apparent, and she had obvious difficulty in immediately recognising family members when they came to visit. Simple everyday tasks became all but impossible. While she remained independent in toileting, she could not dress/shower herself without on-going prompting and support from staff.

With declining emotional control, episodes of tearfulness and unexplained crying became more and more apparent, and when staff attempted to alleviate her distress, Ms. A had great difficulty in describing what exactly was upsetting her. In extreme situations she would hit out or threaten her peers and became notably less tolerant of noise in the home. Accusations about peers were becoming an everyday presentation, all of which was very out of character for this lady. With the progress of dementia, Ms. A was finding it increasingly difficult to make sense of her world. Her progressive cognitive impairment overwhelmed any logic.

Episodes of confusion and disorientation to time were impacting her sleep/wake cycle, and she would only sleep for 2–5 h per night; however, she was also sleeping during the day and getting sufficient hours of sleep over a 24-h period. Disorientation to place and way finding in the familiar home environment was becoming more obvious. She had difficulty in locating her bedroom or the bathroom area.

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Chapter 9
The Cued Recall Test: Detection of Memory Impairment

Darlynne A. Devenny, Sharon J. Krinsky-McHale, and Adeniyi Adetoki

Introduction

Memory decline is a characteristic of normal aging as well as an early symptom of dementia in Alzheimer's disease (DAD) in both individuals from the general population and in individuals with intellectual disabilities (ID). The determination of decline in individuals with ID is difficult because they have a compromised memory system even when young and healthy and because there are substantial individual differences in level of functioning. The Cued Recall Test, a list-learning task that presents test items in a controlled learning paradigm, has both concurrent and predictive validity and is promising as a research and as a clinical diagnostic measure for the identification of memory impairment in adults with ID.

The first issue we address is the identification of memory impairment associated with DAD in individuals with Down syndrome (DS). The diagnosis of DAD was made by community physicians independent of the findings of the Cued Recall Test. Both cross-sectional and longitudinal findings show that the Cued Recall Test can discriminate individuals with DAD from those without the diagnosis. The second

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issue is the identification of memory impairment in individuals with DS prior to a diagnosis of DAD. Longitudinal data indicated progressive declines in performance on the Cued Recall Test in some individuals, suggesting that they may be in a preclinical phase of the disease. Finally, we examined the effectiveness of the Cued Recall Test in detecting changes associated with normal aging in adults with DS. Older adults were poorer than younger adults with DS on the free recall of test items, one of the component measures of this test.

Background

Memory impairment is a behavioral signature of DAD and is frequently the first sign of change in individuals from the general population [1, 2]. Establishing an "impairment" in individuals with ID is difficult because performance on memory tasks is related to level-of-cognitive functioning which varies considerably among these individuals. Setting a level for "impairment," then, is problematic when baseline memory ability is compromised. In spite of these problems in measurement, recent longitudinal studies have determined that, as in the general population, memory impairment is also one of the first signs of change associated with DAD in adults with DS [3–5].

Performance on memory tasks not only depends on memory ability but is influenced by other cognitive functions, such as attention, processing capacity and efficient use of strategies, [6] abilities which show attenuation with normal aging and which may be selectively or globally impaired in individuals with ID at any age. In the general population, procedures that induce semantic processing (e.g., providing category cues for test items) have been shown to reduce the influence of these other cognitive abilities on memory in healthy older adults and thus reduce the overall effects of aging on memory tasks. These category cues are most efficient when they are provided both as a support for encoding and for retrieval [7]. In a typical paradigm of controlled learning, items on a memory task are introduced with a category cue and the same cue is provided when initial spontaneous retrieval of an item fails [6, 8, 9]. That is, the encoding of each task item is enhanced by focusing attention on a semantic association, and retrieval is enhanced by its close alignment with the context of encoding. While there have been several variations of this paradigm, the procedure, in general, has been found to be effective in identifying specific memory impairment associated with Alzheimer's disease (AD) in older adults from the general population [8–12].

In our longitudinal study, of aging in adults with ID we administered a memory test that employs a controlled learning paradigm [13] that was modeled on a procedure developed by Grober and Buschke [14]. In this study we were particularly interested in the course of aging among adults with DS as both premature aging and a high risk for AD are associated with this syndrome. The goals of the study were to examine changes in cognitive functioning associated with normal aging and to distinguish these changes from those associated with early-stage dementia. Since declines in memory are one of the primary and earliest signs of change in dementia,

we focused our efforts on evaluating and developing tasks that could detect the earliest changes in memory and in identifying areas of cognitive ability that influence memory performance.

Identification of Memory Impairment in Adults with Dementia in the General Population

Memory measures found to be most sensitive to age-associated declines focus primarily on episodic memory. Episodic memory is related to the acquisition of information obtained in a specific time and place [15] and is dependent on the integrity of the hippocampus and its connections with the frontal lobe [16]. List-learning tasks are used to assess this type of memory; the items on the list, while within the vocabulary of the individual, are uniquely associated with the event of the specific testing situation. In the controlled learning paradigm the items to be recalled are presented with a category cue that is related to the test item. In the learning phase there is an opportunity to learn the test items over repeated trials. In the testing phase, for each trial, free recall is followed by cued recall in which the category cue is provided for each item that was not recalled spontaneously.

Initial studies of the cued recall procedure in the general population found that it discriminated between individuals with and without a diagnosis of DAD [2, 8, 14]. In a version with a maximum score of 48, a cutoff score of ≤44 identified all participants who had a diagnosis of dementia [6]. Because the presence of the category cue was so effective in facilitating retrieval in individuals who did not have dementia, Grober and Kawas [9] found a ceiling effect in using the Total Score (Free Recall + Cued Recall) in individuals who were in a preclinical phase of DAD. Follow-up testing conducted 3 years later, however, showed a decline for those participants with DAD relative to healthy elderly control participants.

In order to make the test more difficult and to eliminate the ceiling effect, Buschke and colleagues [8] modified the procedure in the cued recall task by increasing the number of items to 64 and providing four exemplars for each of the 16 categories. This modified version of the task provided good sensitivity and specificity in distinguishing individuals with mild DAD from healthy participants. The controlled learning procedure, then, facilitates encoding specificity in older, healthy adults but not in those with DAD.

Age-Associated Memory Impairment and Down Syndrome

In adults with DS, the effects of aging on the memory system are imposed on those that are pre-existing due to an atypical developmental history [17]. Although the investigation of the memory system in relation to aging in adults with ID is relatively recent, initial findings show a pattern of performance that mirrors that seen in

the general population. In longitudinal studies, older adults with DS showed small age-related declines in episodic memory [5, 13, 18–20] and recent cross-sectional studies have shown that older adults with DS are poorer than their younger peers on measures of visual short-term memory [21–23]. In contrast, auditory short-term memory span shows little or no decline either with normal aging [3, 4, 19] or with early-stage dementia in adults with DS [3].

Diagnosis of Early-Stage Dementia in Adults with Down Syndrome

Declines in episodic memory are frequently the earliest symptom of change associated with DAD and are distinguished from declines associated with normal aging by the degree of impairment [13, 20]. Identifying the early stages of dementia with neuropsychological tests is difficult because dementia has an insidious onset, there is heterogeneity of initial cognitive deficits, and many areas of early deficits are shared with normal aging and with dementia from other causes [24]. In adults with DS there is the additional difficulty of distinguishing changes in cognitive function associated with dementia from those related to precocious but normal aging, and from those attributable to lifelong cognitive impairments. Typically, baseline measures from which to assess change are unlikely to be available for most patients seen in diagnostic clinics. Because we employed a longitudinal study design, we were able to use individuals as their own controls and to look for the sequence and magnitude of decline on multiple measures of cognition and memory. One aim of our study, then, was to develop tests that will be clinically useful when administered as a one-time measure to identify memory impairment.

Early Identification of Significant Memory Impairment in Adults with Down Syndrome

A second aim of our study was to develop measures that would identify individuals with DS who have Mild Cognitive Impairment (MCI). In the general population, MCI refers to a concern regarding a change in cognition with impairments in one or more cognitive domains while the individual shows intact or slightly impaired functional abilities [25, 26]. In addition, studies have shown that deficits in higher cognitive abilities associated with language, judgment and problem solving may coexist with memory deficits [27–29]. MCI attempts to account for the gray area when cognition is not intact but not sufficiently impaired to be considered dementia. Identifying MCI is of interest because for some individuals it represents the early preclinical period for DAD. Individuals from the general population who are identified as having MCI appear to be at higher risk for developing dementia than older

adults without significant memory declines. When MCI was based on memory impairment, an estimated rate of annual conversion from MCI to DAD ranged from 6 to 25% [26, 29, 30]; however, in those individuals whose memory impairment was accompanied by declines in additional cognitive abilities, the conversion rate increased to a range of 40–60% [28, 29, 31].

The distinction between MCI and a "preclinical" stage of dementia is currently far from clear. "Preclinical" refers to the period of cognitive decline prior to when an individual meets the criteria for a diagnosis of dementia and is estimated to have a duration from 6 to 10 years [27, 32]. Although a cognitive profile of deficits may not easily distinguish prospectively between MCI and preclinical DAD, retrospectively it is possible to evaluate decline from the time of diagnosis of dementia and determine those individuals who were "preclinical." The sequence of cognitive decline associated with both MCI and preclinical DAD appears to have some regularity, with deficits in memory, and particularly delayed recall, occurring several years before diagnosis [27, 32]. Intervention at this stage to prolong the period before the onset of dementia is a goal of many recent clinical trials [25].

Identifying MCI in adults from the general population is based on a premise of being able to define a narrow expected range of "normal" memory functioning. A specified deviation from what is defined as "normal," then, constitutes memory impairment. In adults with DS this premise is untenable because of the variability among individuals in their initial baseline level-of-functioning. However, despite this variability, recent longitudinal studies have shown that our measures of memory that have been adapted for use with adults with DS are able to identify individuals with significant memory declines [13, 20].

Measures of Episodic Memory in Adults with Down Syndrome

The first test of episodic memory we administered, the Selective Reminding Test (SRT), consisted of eight items from a single semantic category (food or animals). On the first trial of this test, the items were presented auditorily and the participant was asked to recall them. On each of the subsequent five trials, the participant was reminded of only those items not recalled on the previous trial. We have administered this test at each 12- to 18-month assessment cycle for the past 19 years of our longitudinal study. We have found this to be a test with good reliability [18] and with the ability to detect age-associated decline in memory when administered longitudinally [18, 20]. We were also able to specify significant decline in memory associated with early-stage dementia. We have established a criterion of a 20% decline from an individual's previous highest score for two consecutive years as indicating the amount of decline associated with early-stage dementia [20]. This criterion, however, relies on having at least one baseline score that is representative of the individual's memory ability on this test, administered at a time when the individual was healthy and free from dementia, and on having two subsequent evaluations conducted over a 2-year period.

The Cued Recall Test is also a list-learning task, but differs from the SRT in some critical elements. The Cued Recall Test has an increased number of items (12 test items), and each item is from a different semantic category. In addition, the Cued Recall Test facilitates the encoding of items into storage because there is an initial learning phase in which items are systematically presented in small units (four at a time), the test items are presented both auditorily and visually (picture format), and participants are re-presented with the category cue that was provided in the learning phase if they are unable to spontaneously retrieve an item. This category cue, if efficiently utilized by the participant, constrains the internal search for a test item and may prompt recognition, a component of memory that is less vulnerable to declines associated with normal aging.

Procedure

We modeled the Cued Recall Test on a measure developed by Grober and Buschke that identifies memory problems in adults from the general population [14, 33]. The stimuli are 12 black and white line drawings [34] with each item representing a distinct semantic category. There are two versions of the test that are alternated across test cycles in our longitudinal study.

Version 1		Version 2	
Test item	Category cue	Test item	Category cue
Grapes	Fruit	Cake	Eat
Helicopter	Flies	Iron	Hot
Candle	Gives light	Star	In the sky
Screwdriver	Tool	Tree	In the garden
Hat	Wear	Tie	Wear
Lips	Part of face	Hand	Body
Scissors	Cut	Top	Toy
Rabbit	Animal	Squirrel	Animal
TV	Furniture	Dresser	Bedroom
Pot	Kitchen	Pitcher	Table
Guitar	Musical instrument	Barn	Farm
Bike	Has wheels	Swing	Ride

In comparison with the original test for the general population, we chose items and categories appropriate for the vocabulary of individuals with mild and moderate ID, we reduced the number of items from 16 to 12, and we modified the training procedures. In our procedure, the participant is presented with the category cue only once during the initial presentation of the items. If any subsequent learning trials are required only the specific name of the item is repeated.

The testing procedure for the Cued Recall Test involves a learning phase and a testing phase. In the learning phase the goal is to achieve encoding specificity by

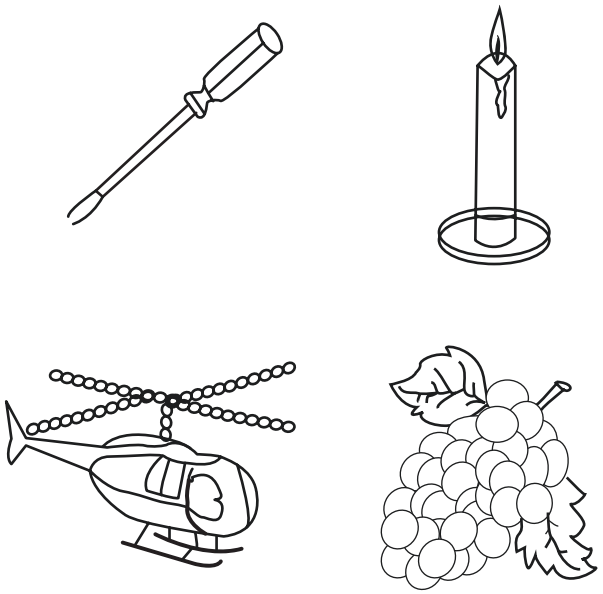

Fig. 9.1 An example of a card with stimuli presented during the learning phase

providing the participant with the same category cues that will be used to prompt retrieval. Four pictures are presented at a time, one in each quadrant of an 8″ × 11″ card and the participant is asked to inspect the card and name the picture corresponding to the verbal category cue (e.g., "Which one is fruit?") (Fig. 9.1). After the participant has pointed to and named each of the four items, the card is removed and he is asked to immediately recall the four test items from memory. Presentation of the cards for the learning of the items ceases when all four items are correctly recalled, or after three trials of the presentation of each card. The number of items that are recalled on each trial are noted. Typically, the four items are learned by the third trial. If, however, by the third trial all the items are not recalled, the participant is shown the card one more time with the items not recalled pointed out, but no further recall trials are given for that card. After the learning phase the stimuli are removed from the view of the participant.

The testing phase immediately follows completion of the learning phase and consists of three trials of free and cued recall. Each trial begins by asking for free recall of all 12 test items in any order. The free recall portion of a trial ends when the individual either indicates that he/she does not remember any more items or the individual begins to repeat items already named. For each item not recalled during the free recall trial, the category cue is provided (e.g., "What was the animal?") and the individual is given the opportunity to respond. This acts as a focused reminder and in individuals without significant memory impairment the category cue is usually sufficient to prompt the recall of the specific item. If the individual does not retrieve the item with the cue, the participant is reminded of the missed item (e.g., "The animal was a rabbit."). Two scores are generated for each trial, a Free Recall Score and a Total Score (Free Recall Score + Cued Recall Score).

Table 9.1 Participant characteristics of etiology and mean age and IQ with standard deviations in parentheses

Etiology	Status	N	Age	IQ
Unspecified ID	Healthy	61	59.2 (11.7)	58.4 (11.5)
	DAD	1	75.7	86
	MCI	2	58.2 (10.7)	58.5 (5.0)
Down syndrome	Healthy	61	48.2 (8.3)	54.2 (11.2)
	DAD	32	55.4 (5.4)	53.6 (11.2)
	MCI	14	52.6 (6.8)	48.5 (10.4)

Participant Characteristics

Participants in our longitudinal study consisted of adults with DS and those with ID with unspecified etiologies. Inclusion criteria for the study are: (1) no suspicion by caregivers of declines in functioning; (2) no uncorrected serious sensory impairments; (3) absence of uncontrolled seizure disorder; (4) age ≥30 years; (5) IQ ≥30; (6) attendance at a community program, such as independent employment, workshop or day treatment program.

During the course of the study, three individuals developed chronic medical conditions that could contribute to a profile of cognitive decline (clinical depression, stroke, and transient ischemic attacks) and these individuals were eliminated from our analyses. Over the course of the study 32 individuals with DS have developed dementia. Among participants with ID from unspecified etiologies, one individual has received a diagnosis of DAD. In addition, we have some individuals who have substantial declines in memory ($N = 14$ with DS; $N = 2$ with unspecified etiologies) that suggest they may be in the preclinical period and we have identified them as having MCI (Table 9.1).

Participants were divided into those with DS and those with an ID that does not have a known etiology. A status of "healthy" indicates that no declines have been identified in cognitive/memory or adaptive functioning. A diagnosis of DAD was provided by a physician once declines in memory, other cognitive abilities and activities of daily living were established and other causes of decline were ruled out. A diagnosis of MCI indicates significant memory impairment without declines in adaptive functioning.

Psychometric Properties

Reliability of Different Versions of the Test

For the past 6 years we have alternated between two versions of the Cued Recall Test [13]. Our first assessment was to determine if the two versions were comparable. We examined the scores from 95 individuals (with DS and with unspecified ID)

who have remained healthy over this period and for whom we had at least three sets of scores within the first three test cycles. A comparison of the two different test versions administered approximately 1.5 years apart showed a Pearson correlation coefficient of 0.564 for the Total Score ($p < 0.001$) and a coefficient of 0.469 for the Free Recall Score ($p < 0.001$). These somewhat reduced coefficients may reflect variability in performance associated with aging, with individuals with DS expected to show declines at relatively earlier chronological ages. We then selected only those individuals with ID with unspecified etiologies who were younger than 60 years of age ($N = 33$) and, therefore, were not expected to show declines in performance associated with normal aging, and found a Pearson correlation coefficient of 0.683 for the Total Score ($p < 0.001$) and a coefficient of 0.641 for the Free Recall Score ($p = 0.001$). These correlation coefficients indicate an acceptable level of comparability between the two versions of the Cued Recall Test.

Reliability of Retesting with the Same Test Version

Test–retest reliability was examined by comparing test scores of the same version separated by an interval of 3 years. When all healthy participants were included, the Pearson correlation coefficient was 0.390 for the Total Score ($p = 0.001$) and 0.388 for the Free Recall Score ($p = 0.002$). These scores are influenced, in part, by the longer interval between the administrations of the tests when the amount of decline may be amplified and when decline may be occurring at a faster rate in some individuals. Among individuals with ID from unspecified etiologies the coefficient for the Total Score increased to 0.428 but was not significant due to the reduced number of participants; the coefficient for the Free Recall Score of 0.623, however, was significant ($p = 0.002$). Overall, the Cued Recall Test appears to have modest test–retest reliability. A better measure of reliability would be to conduct repeated testing across different versions just days apart with a large sample, but this has yet to be done.

Diagnostic Efficacy

In the initial evaluation of our version of the Cued Recall Test there were 19 individuals with DS who had a diagnosis of DAD [13]. Based on their performance on this test, in comparison to that of their healthy peers with DS, our data suggested that a cutoff Total Score of ≤23 distinguished between the two groups. This cutoff score, however, reflected the performance characteristics of this specific group of participants. Since the publication of these findings, an additional 13 individuals have developed and received a diagnosis of dementia. For this new group of individuals we employed the same criteria for establishing the date of onset of DAD as previously; that is, the date of a physician's diagnosis. We then examined performance on

the Cued Recall Test and found that the cutoff score of Total Score ≤23 identified all individuals with a diagnosis of DAD in this new group. In fact, the age at which 11 of these individuals, who had received a diagnosis of DAD, met the cutoff score preceded the age of diagnosis from 9 to 84 months ($X = 29.2$ months); in the remaining two individuals it was concurrent with the diagnosis. This prospective group contributes to the validity of our choice of the particular level of the cutoff score.

We then examined the relationship between age at performing at the level of the cutoff score and age at diagnosis of DAD among all our participants with DS ($N = 32$). Twenty-nine individuals with DAD met the cutoff score at the time of their diagnosis and their scores on the Cued Recall Test continued to show subsequent declines. Indeed, many of these individuals met the cutoff score even prior to receiving their diagnosis and on subsequent testing their Total Score typically remained less than 23, indicating that the cutoff score represents a threshold. Three out of the 32 individuals with dementia did not quite reach the cutoff score at the time they received a diagnosis (Total Scores ranged from 24 to 26) although their scores at the time of diagnosis represented a decline from a previous level and their Total Score just prior to their diagnosis was ≤23 (Fig. 9.2). In general, a decline in Total Score to ≤23 should be considered a significant memory impairment in adults with DS and mild or moderate ID.

Sensitivity and Specificity

Sensitivity refers to the ability of a diagnostic test to identify individuals who have the disease and is directly related to the level of the cutoff score. In the case of a memory test that has the potential to be used as a screening test, it is important to set the cutoff score at a level high enough that it will identify all individuals who are in need of a diagnostic evaluation. To determine sensitivity, we examined only adults with DS because in our sample we have only one individual with ID of unspecified etiology who had a diagnosis of dementia. To test the sensitivity of the Cued Recall Test employing the cutoff score of ≤23, we examined performance on this test at the time of the clinical diagnosis, that is, when individuals first met the criteria for DAD. Since DAD is characterized by progressive decline, anyone with a diagnosis will eventually show global cognitive impairment and poor performance on any test of memory or cognition. It was, therefore, important to evaluate the Cued Recall Test at a time when individuals first met diagnostic criteria. For all other participants in our longitudinal study we used their score at their most recent testing.

The first estimate of sensitivity compared the participants with the diagnosis of DAD to all our other participants with DS. Among the group of participants without a diagnosis, there were very likely some individuals in a preclinical phase of DAD [35]. While recognizing this, we were interested in determining how effective the Cued Recall Test was in identifying individuals with DAD. In this analysis, the Total Score on the Cued Recall Test had a sensitivity of 91%, indicating that it detected most of the individuals who had a diagnosis of DAD.

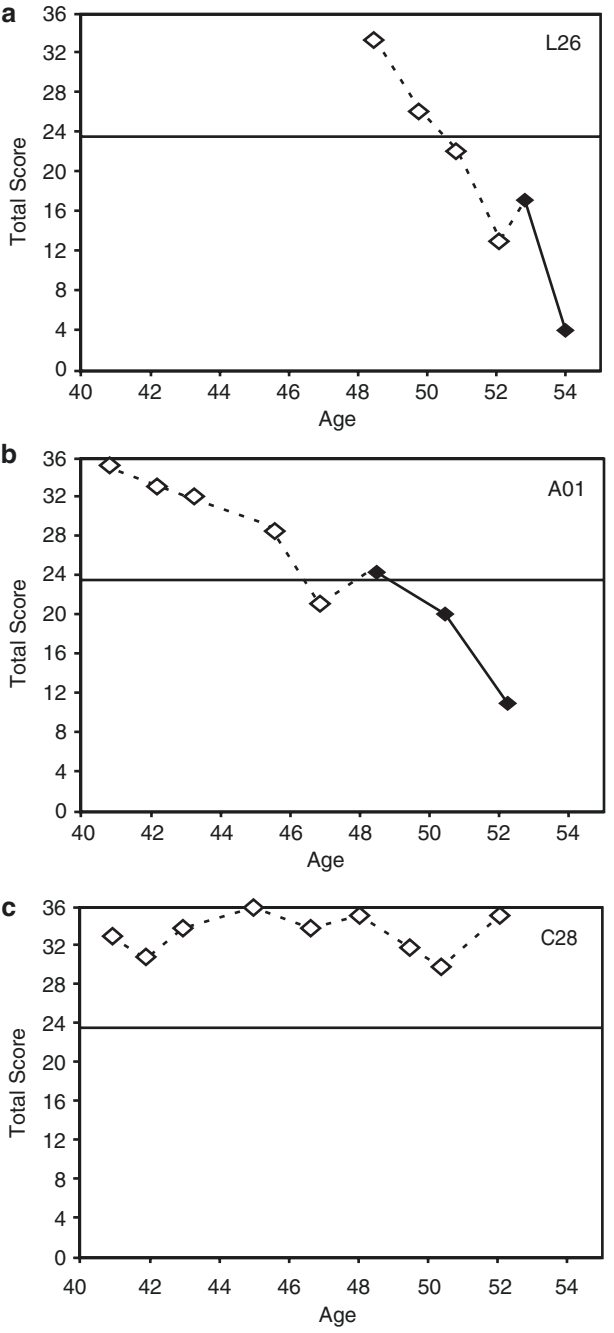

Fig. 9.2 (**a**–**c**) Individual performance profiles across test times of Total Score on the Cued Recall Test prior to (*open diamonds*) and after (*closed diamonds*) diagnosis of dementia. L26 met the cutoff score 3.2 years prior to a diagnosis and represents 83% of cases with dementia; A01 had a score close-to but not below the cutoff score at the time of diagnosis and represents 8.5% of cases. C28 has remained healthy

Specificity refers to the ability of a test to correctly identify individuals who are without the disease. High specificity for a test can contribute diagnostic information by assisting in ruling out the presence of a particular disease. The specificity of the Cued Recall Test among the participants with DS was 72%. This lower score reflects the inclusion of some individuals who have MCI.

Positive predictive value (PPV) refers to the likelihood that a positive test result will be correct and is a measure of the efficiency of the screening test. Predictive values are related to the prevalence of a condition or disease, with higher prevalence rates related to higher PPVs and lower negative predictive values (NPVs) [36]. A comparison of all participants with and without a diagnosis gave a PPV of 58%. This score also reflects the inclusion of individuals with MCI who have memory impairment but who are for this analysis included in the nondemented group. A NPV refers to the likelihood that a person obtaining a score higher than the cutoff value is correctly identified as an individual without the disease. The NPV was 94%.

Mild Cognitive Impairment

Among our participants with DS there were 24 individuals who obtained a Total Score below the cutoff at the time of testing but did not have a diagnosis of DAD. For 22 of these individuals, we had longitudinal data that showed that their Total Score on the Cued Recall Test, at the most recent testing, represented a mean decline of 13.5 (SD = 8.5) points from the highest score they had received on previous testing. Their current performance, therefore, represented a change in their memory ability and can be correctly interpreted as memory impairment.

Fourteen individuals from the group with low scores on the Cued Recall Test also showed significant memory declines on a separate memory test, the SRT [18, 20]. In a second analysis of the sensitivity of the Cued Recall Test we identified these 14 individuals as a group with MCI because there was independent verification of memory impairment but, since their skills of daily living were sufficiently preserved, they did not meet the criteria for a diagnosis of DAD. We compared their performance to individuals identified as "healthy," that is, participants without declines on the SRT (Table 9.2). In this analysis, sensitivity of the

Table 9.2 Comparison of sensitivity, specificity, positive predictive value (PPV) and negative predictive value (NPV) for two and three trials on the cued recall test

Comparison	N	Sensitivity	Specificity	PPV	NPV
Three trials					
Demented vs. nondemented	107	91	72	58	94
MCI vs. healthy	75	79	84	52	94
Two trials					
Demented vs. nondemented	107	91	69	56	95
MCI vs. healthy	75	86	82	36	96

The cutoff score for two trials was a Total Score of ≤15 and for three trials was ≤23.

Cued Recall Test to identify individuals with MCI was 79% and the specificity was 84%; the PPV was 52% and the NPV was 94%. The lower PPV in this analysis may be due to the stringent criterion for significant memory decline on the SRT which has the effect of reducing the prevalence of individuals identified as having a "memory impairment."

Longitudinal Data

We examined the longitudinal performance of ten individuals with a diagnosis of DAD for whom we had data for three complete test cycles across a time period from when they were thought to be healthy up to and after receiving their diagnosis. The interval we examined spanned a mean of 2.85 ± 7.3 years. These individuals met the cutoff score on an average of 20 months (range = 0–39 months) prior to receiving a diagnosis of DAD. Two individuals (8.5%) met the cutoff score at the time of their diagnosis. The remaining two individuals (8.5%) did not meet the cutoff score at the time of their diagnosis although their scores were close (24 and 25) and their performance represented a decline from a previous level (see Fig. 9.2 for atypical profiles of performance).

The longitudinal analysis included scores from 48 individuals who were healthy (mean age = 44.9 ± 7.1 years; mean IQ = 53.4 ± 12.1), 9 with MCI (mean age = 51.6 ± 4.1 years; mean IQ = 50.7 ± 9.9), and 10 with a diagnosis of dementia (mean age = 53.4 ± 3.6 years; mean IQ = 54.1 ± 12.5). Analysis of covariance (ANCOVA) with Total Score across three test cycles as a repeated measure, with dementia status at the third cycle (healthy, MCI, dementia) as a between-subjects factor, and age and IQ as covariates showed a significant overall effect of dementia status ($F(1,62) = 57.528$, $p < 0.001$) that was modified by a dementia status × test cycle interaction ($F(4,122) = 8.509$, $p < 0.001$). While differences across test cycles in performance were not significantly related to age, there was an effect of IQ ($F(1,62) = 9.595$, $p < 0.01$) in which higher intelligence quotient (IQ) scores were, in general, associated with better performance on the test. Post hoc comparisons indicated that participants who were healthy had significantly better performance across test cycles than those with MCI ($F(1,53) = 53.808$, $p < 0.001$), but the difference between participants with MCI and those with DAD was not significant (Fig. 9.3).

In this longitudinal analysis, participants were categorized based on their dementia status at cycle 3. Within each of the groups, where individuals were classified as either MCI or demented, there is a pattern of decline that reflects their changing dementia status. In the group with MCI five of the nine individuals did not have memory impairment (defined as meeting the cutoff score) at cycle 1 and two did not have memory impairment at cycle 2. In the group with dementia, four of the ten individuals did not have memory impairment at cycle 1 but all had either MCI or dementia at cycle 2. The findings demonstrate a continuum of progressive memory impairment prior to and including the onset of dementia which was reflected in the Total Score of the Cued Recall Test. Further, these findings indicate that this test has good predictive validity.

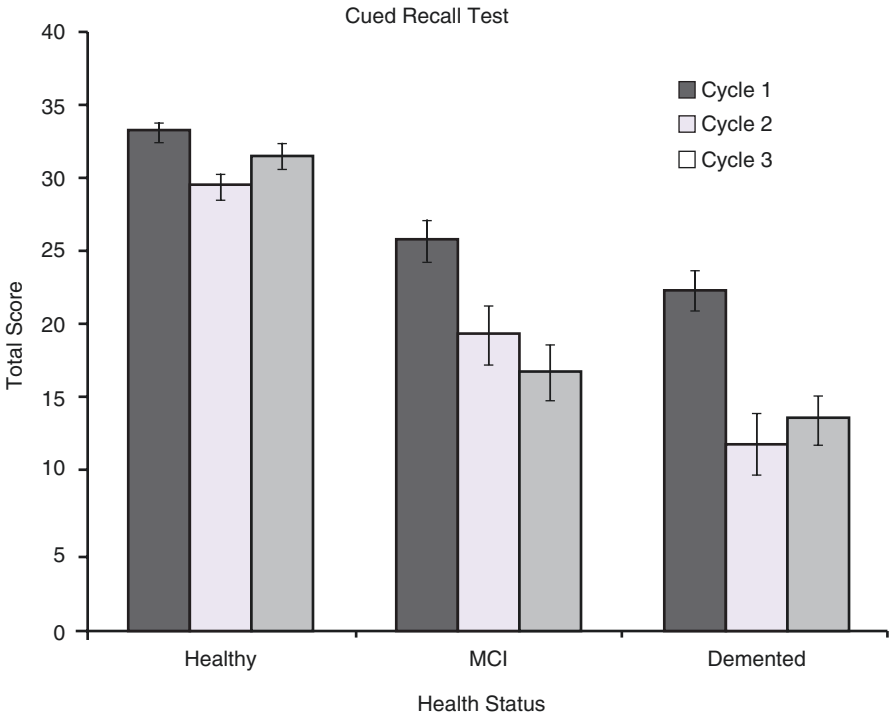

Fig. 9.3 Means (with standard error bars) of longitudinal Total Scores on the Cued Recall Test for three groups of individuals with Down syndrome (DS). The healthy group is not suspected of decline, the group with Mild Cognitive Impairment (MCI) has significant memory declines on the Selective Reminding Test, and the group with dementia has a diagnosis of Alzheimer's disease

Normal Aging and Free Recall

Thus far we have discussed the role of the Total Score in identifying memory impairment associated with the preclinical and early-stage dementia. The Cued Recall Test also measures free recall and we first examined performance on this component in relation to age-associated changes in memory among those participants who have remained healthy and are not suspected of declines in functioning. An ANCOVA examined the Free Recall Score with etiology (DS, ID of unspecified etiology) as a between-subjects factor and age and IQ as covariates. Although adults with DS were poorer on Free Recall (X = 12.73, SD = 8.35) than adults with ID from unspecified etiologies (X = 18.67, SD = 9.16), this difference did not reach significance. There was a main effect of age ($F(1,117) = 19.43$, $p < 0.001$) that was modified by an etiology x age interaction ($F(1,117) = 6.36$, $p = 0.01$). Post hoc analysis of this interaction revealed age-associated declines on this memory measure for adults with DS ($F(2,58) = 12.61$, $p < 0.001$), but not for those with ID from unspecified etiologies.

Next, we examined whether the Free Recall component can distinguish between the groups with and without memory impairment among the adults with DS.

We found that, once again, there were significant main effects of age ($F(1,102) = 20.01$, $p < 0.001$), IQ ($F(1,102) = 5.42$, $p = 0.02$), and dementia status ($F(1,102) = 20.35$, $p < 0.001$). Post hoc analysis indicated that the healthy group was significantly different from both the MCI and dementia groups, but the latter two groups did not differ from one another.

These findings of performance on the Free Recall component essentially correspond to those of the Total Score. However, the Free Recall Scores have sufficient overlap across health status groups that it was difficult to determine an effective cutoff score with this measure.

Evaluation of Two Trials

In an effort to reduce the testing time for the Cued Recall Test, we examined our data to determine if we could achieve the same discrimination among the groups using scores from only the first two trials. We repeated the ANCOVA among the participants with DS with dementia status (non-demented, MCI, demented) as a between-subjects factor and age and IQ as covariates employing a Total Score cutoff of ≤15 (out of a possible 24) and found a significant main effect of dementia status ($F(2,102) = 51.860$, $p < 0.001$). Once again there were significant effects for age ($F(1,102) = 7.793$, $p = 0.006$) with older participants having poorer scores, and IQ ($F(1,102) = 14.921$, $p < 0.001$) with lower IQ scores associated with poorer scores. Post hoc analysis showed that non-demented participants performed significantly better than either the group with MCI or dementia.

We repeated the calculations for sensitivity and specificity (Table 9.2) and found that a cutoff Total Score of ≤15 for only two trials was adequate to discriminate between individuals who were demented from the group that was not demented (non-demented and MCI) and also between individuals who had MCI from those who were non-demented. The PPV, however, reflected the relatively high number of false positives with the cutoff score of ≤15.

Findings from Recent Studies

There are now findings on the Cued Recall Test from three new studies of adults with DS. Benejam and colleagues [37] translated the Cued Recall Test into Spanish and tested 75 healthy adults (mean age = 36.1 ± 9.8 years) and 15 adults with DS and DAD (mean age = 51.1 ± 5.1 years). They found that, similar to the study just described, among non-demented individuals Free Recall Scores were associated with aging, with a decline beginning in the 5th decade of life; Total Scores were uniformly high ($x = 34.9 \pm 1.3$) and were not associated with individual level-of-functioning. Individuals with DAD had significantly lower Total Scores ($x = 17.5 \pm 5.2$) with all individuals, except one, in this group scoring below the suggested cut-off score of 23.

In addition, individuals with DAD had more intrusions on Cued Recall. While it is still anecdotal, an incorrect response after a cue has been provided (designated as an 'intrusion') may be an early sign of memory impairment. It should be noted that some non-demented individuals who had low intellectual functioning and/or severe cognitive declines associated with DAD were not able to perform the test and, therefore, were not included in this sample.

In a second new sample, the Cued Recall Test is one of the measures in an ongoing, multi-site, longitudinal study that is examining the relationship between neuropsychological functioning and the deposition of amyloid-β in adults with DS. The first report by Hartley and colleagues [38] of this study included adults with DS who were not suspected of having dementia. In addition to the administration of the neuropsychological battery, these individuals participated in neuroimaging (MRI/PET scans) to assess brain amyloid-β deposition using the Pittsburgh compound B (PiB). Participants were divided into two groups: PiB+ individuals had deposition in one or more regions (frontal cortex, anterior cingulate gyrus, parietal cortex, lateral temporal cortex, precuneus cortex) that exceeded a cut-off value established in the general population as being associated with AD (n = 22; age = 44.3 ± 3.8 years); PiB− individuals were below regional thresholds (n = 41; age = 34.5 ± 5.2 years). On the Cued Recall Test, the Total Score for the adults with DS who were in PiB+ group was 30.7 ± 5.5, while for the PiB− group it was 33.2 ± 5.8. These findings were consistent with the previous studies of non-demented adults with DS on this this test.

A 3-year follow-up report of this same group of individuals showed that a significant increase in the PiB level of the precuneus region (t = 3.54, p = 0.002) was correlated with a decrease in the Total Score of the Cued Recall Test ($r = 0.41$, $p = 0.04$) and an increase in the Reiss Maladaptive Behavior Scale ($r = 0.46$, $p = 0.02$) [39].

In one other pilot study conducted by Cooper and colleagues [40] the purpose of which was to demonstrate the feasibility of a drug trial of simvastatin for the prevention of DAD in adults with DS, the Cued Recall Test was included in the evaluation battery. The preliminary findings showed that the placebo group ($n = 11$; mean age = 53.7 ± 3.2 years) had initial Total Scores comparable to other studies (x = 34.1 ± 4.5) with a decrease when re-tested 12 months later (x = 28.7 ± 13.0). The intervention group ($n = 10$; mean age = 54.9 ± 3.1 years) had lower scores initially with a large standard deviation (x = 21.2 ± 12.7); 12 months later their scores were substantially poorer (x = 12.7 ± 12.7) suggesting that some individuals may have had memory impairment when they entered the study. This is not surprising given the advanced age of the participants in this study.

Pros and Cons of Cued Recall Test

The Cued Recall Test is appropriate for the evaluation of adults with developmental disabilities in the mild to moderate range of ID. Verbal ability is required but, in our experience, some individuals with a receptive vocabulary as low as an age

equivalent of 2.5 years (as measured by the Peabody Picture Vocabulary Test—Revised [41]) have successfully performed on the Cued Recall Test. The principal advantage of this test is its ability to distinguish between individuals who have significant memory impairment from those who do not. Further, the test appears to be sensitive to the memory impairment that precedes the onset of the symptoms that are the basis for a diagnosis of dementia (DSM-IV [42]; ICD-10 [43]). In fact, the cutoff score of ≤23 on the Total Score identifies most individuals with MCI. Early diagnosis is useful in planning for individuals and, when treatment becomes available, it will be essential.

More importantly, this test has the potential to be a useful screening tool because of its ability to identify memory impairment at a single evaluation. If the overall level-of-functioning of an individual can be established to be within the mild to moderate range of ID then the Total Score on the Cued Recall Test can be interpreted without reference to a baseline score. The ability of this test to detect memory impairment may facilitate an earlier diagnosis of dementia because the score indicating "memory impairment" is not based on an interval of documented decline.

Ideally, however, adults would be administered this test while they are healthy to establish a baseline of their performance, and then would be periodically retested. Systematic, longitudinal assessments would firmly establish a decline in memory ability. For adults with DS, the suggested age for a baseline test administration is 40 years, with retests every 3 years.

The three-trial version of the Cued Recall Test requires about 20 min to complete and should be administered by an Examiner familiar with testing procedures, in general, and with testing individuals with intellectual impairment, in particular. Our findings indicate that a cutoff score based on the three trials is efficacious. However, a preliminary analysis showed that two trials may be sufficient, but this needs to be confirmed with prospective data.

With respect to the specific value of the cutoff score, we set it at a Total Score of ≤23 for three trials based on our longitudinal study. In at least two of the recent studies with independent samples, non-demented adults perform well above this cut-off score. The study from Barcelona largely confirmed the efficacy of this value for determining memory impairment associated with DAT.

On the other hand, if the Cued Recall Test is employed as a screening tool for identifying individuals who need a clinical evaluation, then a somewhat higher cut-off score might be needed. In our sample, a cutoff score of ≤26 would have identified all individuals with a diagnosis of DAD (but would have increased the false-positive rate, also). An optimum cutoff score will be determined, in the future, when the Cued Recall Test is employed by investigators in diverse settings. It may be that a conservative cutoff score would be of more use for researchers, while a more liberal value would be more useful in clinical screening programs.

While the strength of our longitudinal study has been an ability to follow, carefully, adults with DS over an extended period of time, the study has a relatively small number of individuals. In the current analysis, we applied a criterion we previously established to a new group of individuals who were showing declines and found it to be applicable. Future studies will broaden the data base to include

individuals from a variety of contexts, including clinical settings, and individuals with various etiologies.

Studies comparing scores from different testers and test versions, and test–retest on the same version, will also be needed in order to rigorously determine reliability. Our longitudinal studies have shown that even healthy individuals have small variations in performance on this test (see performance of participant C28 in Fig. 9.1). Reliability studies will be useful in interpreting individual variability. However, even without these reliability studies in hand, the Cued Recall Test appears to be able to assign adults with DS and mild or moderate ID to the categories of either memory impaired or memory unimpaired.

The Cued Recall Test is not useful for individuals with few or no verbal abilities, nor is it applicable to individuals with IQs below 30. In addition, we have evaluated only individuals with DS and, therefore, do not know if the criterion we have established is appropriate for individuals with ID from other etiologies.

Summary

Findings from our study employing the Cued Recall Test indicate that individuals with even very early-stage DAD are unlikely to achieve a Total Score greater than 23. Having a test with a cutoff score will be very useful to assessment protocols whereby memory impairment can be established at a single evaluation. Although there was variability in performance across test cycles that reduced reliability assessments, healthy participants, in general, maintained scores above the cutoff score. We were also able to demonstrate that individuals who received a diagnosis of DAD had a history of decline in memory performance on this test.

Individuals with MCI and individuals with early-stage DAD had significant memory impairment and were distinguished from the healthy participants by their performance on Total Scores on the Cued Recall Test. To distinguish between an individual with MCI and an individual with early-stage DAD, evidence is needed of cognitive and functional decline on additional measures.

The Cued Recall Test will be a useful component of a screening test battery for older adults with DS. It is relatively easy and quick to administer, is noninvasive, and it identifies most individuals who are in need of further evaluation for DAD. Further, it identifies individuals in an early stage of memory decline at a time when intervention would be most beneficial.

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Chapter 10
The Adaptive Behavior Dementia Questionnaire (ABDQ)

Vee P. Prasher

Introduction

The concept of adaptive behavior has been defined by Heber in 1961 [1] as "the effectiveness with which the individual copes with the nature and social demands of this environment" and by Gunzberg in 1977 [2] as "the extent to which an individual is able and willing to conform to the customs, habits and standards of behavior prevailing in the society in which he lives; by the degree to which he is able to do so independently of direction and guidance and by the extent to which he participates constructively in the affairs and conduct of his community." Adaptive behavior scales (ABSs) assess an individual's current abilities as they are manifested in a given situation. Several measures and patterns of behavior are assessed in different situations to give an overall assessment. Individual items are grouped together into domains. Such domains include, for example, communication, dressing, feeding, and toileting.

Background

Zigman and colleagues in Chap. 6 give a full and detailed review of the role of adaptive behavior in the assessment of dementia in persons with intellectual disability (ID). This chapter will focus specifically on the *AAMR Adaptive Behavior Scale* (ABS) [3] and how it was used to develop the Adaptive Behavior Dementia Questionnaire (ABDQ).

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Several studies have recently been published investigating the assessment of adaptive behavior in persons with ID (Table 10.1). The majority of studies have used the ABS as the measure of choice. It is designed to provide objective descriptions and evaluations of an individual's behavior in coping with the natural and

Table 10.1 Recent reports investigating adaptive behavior in the intellectually disabled population

Authors	Sample (population)	Age-range (years)	Residence	Main findings
Schupf and colleagues [4]	99 DS individuals 99 non-ID individuals DS and ID controls)	20–69	Institution community	DS adults over 50 had significant greater regression than controls and younger DS individuals during the last 3 years of life
Brown and colleagues [5]	130 (DS)	1–59	Institution community	Age-related decline present. Least decline for individuals resident in institutional settings
Rasmussen and Sobsey [6]	56 DS individuals 64 ID individuals (DS and ID)	–	Institution	Decline in skills for individuals over 40 years of age. Particularly in self-help and communication skills. Adaptive skills more stable for ID groups
Burt and colleagues [7]	34 (DS)	22–56	Community	No age-related decline in nondemented middle-aged DS individuals. Level of ID significant factor in analysis
Roeden and Zitman [8]	115 (DS and ID)	31–62	Community	Loss of skills in adults with DS >50 years. Dementia factor in loss. Nonsignificant loss due to visual decline
Prasher et al. [9]	128 (DS)	16–72	Institution community	Decline in skills for middle-aged DS population over 3-year period of assessment. Only one significant factor for decline dementia
Oliver et al. [10]	36 (DS)	30–64	Community	More frequent deficits and excesses and greater management difficulty and effects on the individual in a dementia group than age comparable and younger groups
Zigman et al. [11]	248 DS individuals 398 ID individuals (DS and ID)	30–84	Institution community	Cumulative incidence of significant decline for adults with DS increased from less than 0.04 at age 50 years to 0.67 by 72 years, whereas cumulative incidence of significant decline for adults with ID without DS increased from less than 0.02 at age 50 to 0.52 at age 88
Kirk et al. [12]	12 DS individuals 76 ID individuals (DS and ID)	41–86	Community	Significant relationships between dementia questionnaire for mentally retarded people and the adaptive behavior scale

Table 10.1 (continued)

Authors	Sample (population)	Age-range (years)	Residence	Main findings
Ghezzo et al. [13]	67 (DS)	11–66	Community	Adaptive skills lower in non-demented DS adults over age 40 years as compared to adults younger than 40 years. Particular in language and short memory skills, frontal lobe functions, visuo-spatial abilities
Makary et al. [14]	33 (DS)	16–56	Community	Age-related adaptive declines. Practical skills maintained with age. Social and conceptual skills associated with declines

DS Down syndrome, *ID* intellectually disabled

social demands of his/her environment. The ABS consists of two parts. Part I (independent functioning) is designed to evaluate an individual's skills and habits in ten behavior domains considered important to the development of personal independence in daily living. The 10 behavior domains and 21 subdomains are given in Table 10.2. Part II (maladaptive behaviors) of the scale is designed to provide measures of maladaptive behavior related to personality and behavior disorders. Part II consists of 14 domains (Table 10.3).

The scale is completed by a person familiar with the person with ID or by a semi-interview assessment with the interviewer filling out the scale item-by-item while obtaining information from the person familiar with the subject. In the latter case it is possible to clarify and extend the questioning about individual items. The ABS is one of the most widely used and best standardized instruments and has been shown to have good reliability and validity [15–17].

A number of researchers have previously used the ABS to assess age-related changes in adults with DS (Table 10.4).

Miniszek [18] was able to show that elderly persons with DS (N = 15, age >50 years) scored lower on the ABS than did younger DS (N = 4 age <50 years) subjects in every area of adaptive functioning except in domestic functioning. The elderly DS group was divided into nine residents, judged to be severely regressed, and six who were still functioning relatively well. The regressed group scored much lower in all areas. An individual subject could, on comparison of their ABS profile with the above profiles, be reasonably diagnosed as having regression and dementia if other causes of regression were excluded.

Collacott [20] examined age-related changes of adaptive behavior in 308 adults with DS who were identified through the Leicestershire Mental Handicap Register. Scores for each domain of ABS Part I were analyzed for each age-related cohort. Mean scores for older subjects (>30 years) were compared to those below this age. Collacott found a significant reduction within the domain of physical development (which included sensory impairment and locomotor disability) for the cohort aged 40–49 years. For those in the age cohort 50–59, deterioration occurred in all

Table 10.2 Adaptive behavior scale part I domains

I. Independent functioning
A. Eating
B. Toilet use
C. Cleanliness
D. Appearance
E. Care of clothing
F. Dressing and undressing
G. Travel
H. Independent functioning
II. Physical development
A. Sensory development
B. Motor development
III. Economic activity
A. Money handling
B. Shopping skills
IV. Language development
A. Expression
B. Comprehension
C. Social language
V. Numbers and time
VI. Domestic activity
A. Cleaning
B. Kitchen duties
C. Domestic activities
VII. Vocational activity
VIII. Self-direction
A. Initiative
B. Perseverance
C. Leisure time
IX. Responsibility
X. Socialization

Table 10.3 Adaptive behavior scale part II domains

I. Violent and destructive behavior
II. Antisocial behavior
III. Rebellious behavior
IV. Untrustworthy behavior
V. Withdrawal
VI. Stereotyped behavior and odd mannerisms
VII. Inappropriate interpersonal manners
VIII. Unacceptable vocal habits
IX. Unacceptable or eccentric habits
X. Self-abusive behavior
XI. Hyperactive tendencies
XII. Sexually aberrant behavior
XIII. Psychological disturbances
XIV. Use of medications

Table 10.4 Principal studies using the ABS to assess aging in persons with DS

Authors	Sample (population)	Age (years)	Residence	Main findings
Miniszek [18]	19	34+	–	Older (>50 years) DS persons scored lower on the ABS than younger (<50 years) persons. Regressed persons scored significantly lower than nonregressed controls
Cosgrave et al. [19]	128	35–75	Institution community	No association between aggressive behaviour and dementia or severity of dementia
Collacott [20]	308	18+	Institution community	Age-related exponential decline
Prasher and Chung [21]	201	16+	Institution community	Age-related decline found. Significant causative factors were aging, severity of ID, and presence of DAD. Absence of a medical illness was a predictor of higher scores
Collacott [22]	351	18+	Institution community	DS persons with late-onset seizures had lower adaptive scores than older control group and the early-onset seizure DS group
Prasher and colleagues [9]	128	16–72	Institution community	Decline in skills for middle-aged DS population over 3-year period of assessment. Only one significant factor for decline dementia
Prasher [23]	57	17–71	Institution community	Significant decline in ABS scores over 5-year period for persons with dementia as compared with controls

domains. Statistical significance was found for the domains of physical development, economic activity, numeracy and time sense, domestic activities, and vocational activities. After the age of 60 years, significant deterioration occurred in all domains. No consistent age-related changes were found for maladaptive behavior. For the total population decline with age followed an algebraic curve in which the overall ABS score was a function of the square of the individual's age.

Prasher and colleagues [9, 21, 23] in a number of articles investigating changes in adaptive behavior in 201 adults with DS over a 5-year period confirmed the association between decline in adaptive skills and aging and dementia in older adults with DS. The researchers were able to specifically correlate decline in ABS scores with onset and deterioration in dementia in Alzheimer's disease (DAD). Particular domains of ABS Part I, which showed significant change, were independent functioning, numbers and time, self-direction, and responsibility.

Zigman and colleagues [11] analysed information from the Office of Mental Retardation and Developmental Disabilities. The ABS was used to describe the adaptive behavior of subjects. Two hundred and forty-eight adults with DS (mean age 51.5 years) and 398 non-DS adults (mean age 60.6 years) participated. The ABS was administrated four times of the course of the study. For the adults with DS cumulative incidence of significant decline in adaptive behavior to age 72 years

was 0.67. A rapid increase in cumulative incidence was seen during the late 50s. Earlier indicators of decline included dressing/undressing, domestic activity, vocational activity, responsibility, economic activity, physical development and travel and general independent functioning. Late indicators included self-direction, toilet use, numbers and time, cleanliness, comprehension, appearance, eating and expression.

Following research over a period of 30 years, using the ABS to assess change in adaptive behavior in older adults with DS it was apparent that the ABS could be used as a neuropsychological measure to detect and monitor DAD in adults with DS. Particular domains of ABS Part I were, therefore, used to device a questionnaire to screen for DAD in adults with DS. This is described below.

Development of the ABDQ

Sample Group

One-hundred and fifty adults with DS, living in the same geographical region, were recruited. Baseline demographic data of age, gender, karyotyping for DS, residence, and severity of ID were available. Severity of premorbid ID was assessed by (1) review of previously reported intelligence tests, (2) previous level of functioning as determined by review of medical notes, from carer interview and from the mental state examination of the individual. Severity of ID was classified using ICD-10 criteria [24].

Of the 150 adults with DS who participated, 83 (55%) were male and 67 (45%) were female. The mean age of the sample at the start of the assessments was 44.0 years (SD 11.46; range 16–76 years). All individuals had physical stigmata of DS with 92% trisomy 21 (of 135 tested) and 6% of those tested had translocated form of DS. Sixty (40%) were resident in their family home, 57 (38%) in community group homes, and 33 (22%) resided in the hospital. Twenty-seven (18%) individuals had mild ID, 104 (69%) moderate, and 19 (13%) severe ID.

Assessments

All persons were being followed up on an annual basis as part of ongoing clinical care with detailed reassessments of their physical and mental health, adaptive behavior, and social needs. Carers and individuals were interviewed to elicit any evidence of any significant medical condition. As part of the care provision individuals underwent annual (where compliant) venepuncture for routine hematological (including B12 and folate levels), biochemical (including plasma glucose), and thyroid screening. Brain magnetic resonance imaging was also undertaken in a number of cases. Psychiatric assessments were undertaken by

using Part 1 Section H of the Cambridge Mental Disorders of the Elderly Examination (CAMDEX) schedule [24], standard mental state examination of individuals, and completion of the ICD-10 Symptom Checklist for Mental Disorders [25]. As recommended by the international research community [21] all available information was reviewed annually to determine the presence of mental disorder according to ICD-10 criteria, and in particular DAD [26]. Adaptive functioning of the individuals was assessed annually for five consecutive years using the ABS [3].

Findings for the absence or presence of DAD were compared to change in the ABS measurements over the 5-year collection period to determine which items of the ABS best correlated with deterioration in intellectual functioning and could be subsequently used to develop a screening questionnaire.

Development of Questionnaire

In order to diagnose DAD there must be evidence of decline in any given criteria. For this reason the differences in the ABS scores across the 5-year period were analyzed to see if any pattern emerged. The differences that were examined were those of the scores obtained in year 1 subtracted from those of years 2, 3, 4, and 5. Hence a decline in any area was reflected by a negative difference. To compare the ABS findings with the diagnosis of DAD, a new variable called "DCHANGE*i*" (where *i* = year 2, 3, 4, or 5) was introduced. This variable took 1 or 2 possible values and these were assigned as in Table 10.5.

Only patients who were nondemented at the beginning of the 5-year period and were still alive after the data collection period participated in the subsequent data analysis. Four adults died by year 3, 11 died by year 4, and 19 persons died during the 5-year period. This part of the analysis looked at the change in the DAD state, i.e., comparing those that remained nondemented over the 5-year data collection period ($N = 103$) to those who were initially nondemented but were diagnosed with DAD ($N = 16$) at some point in the time period. This diagnosis of DAD was independent to information obtained from the ABS data.

In order to get a spread of items from all Part I, ten domains of the ABS, each domain was analyzed individually to see which of the items in each domain were the best at predicting the onset of DAD. The analysis to find the best predictors was performed by using two primary methods of analysis, logistic regression analysis and stepwise discriminant analysis.

Table 10.5 Scoring criteria for DCHANGE*i*

Is the patient demented in year 1?	Is the patient demented in year *i*?	DCHANGE*i*
No	No	0
No	Yes	1

Identification of Significant ABS Items

Using logistic regression analysis, individual items of the 66 items of Part I of the ABS which produced significant results for a particular time period were identified. For example for domain I, items 3 and 42 (drinking and time, respectively) were significant when the difference in the scores obtained over the time period covering years 1–2 was considered. Thirty-one items appeared to be predictors for the onset of DAD. It was attempted to remove the least possible items from the questionnaire to make it less parsimonious but at the same time obtaining some useful results with the logistic regression analysis. After examining correlations between the 31 items, it was possible to reduce further the number to 16 items. Therefore, the 16 Part I ABS items whose change was shown to differentiate individuals with DS who develop DAD from those who do not were:

1. Tooth brushing
2. Dressing
3. Control of hands
4. Purchasing
5. Conversation
6. Time
7. Food preparation
8. Table clearing
9. Job complexity
10. Job performance
11. Initiative
12. Persistence
13. Personal belongings
14. Cooperation
15. Participation in group activities
16. Social maturity

To confirm that changes in the final 16 ABS items were of clinical significance, logistic regression analysis on these items with DCHANGE5 as the response and the items above as the model was performed. The model also included the patient's age, sex, place of residence, and severity of ID, as there was reason to believe these factors would have an effect on the outcome.

If the differences over the 5-year time period are considered, a list of each individual's probability of getting DAD over the 5 years can be obtained. The descriptive statistics for this is given in Table 10.6, split according to DCHANGE5.

Table 10.6 Descriptive statistics for each individual getting DAD over the 5-year period

Variable DCHANGE5	Number	Mean	Median	Standard deviation	SE mean
Probability 0	103	0.03044	0.00002	0.08127	0.00801
1	16	0.8040	0.8657	0.2548	0.0637

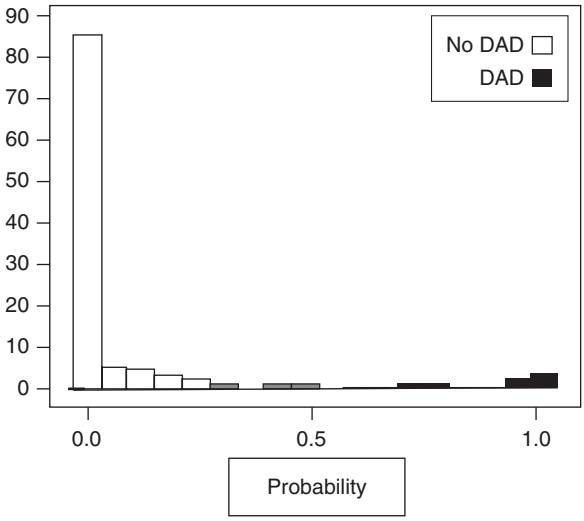

Fig. 10.1 Histogram showing probabilities of dementia computed using the 16 ABS items

Figure 10.1 shows the probabilities, split according to DCHANGE5. A clear distinction between the probabilities for the nondemented (white bars) and those that are diagnosed with DAD (black bars) over the 5-year time period are evident. This finding is also emphasized when considering the sensitivity (15/16 = 94%) and specificity (88/103 = 85%) of the questionnaire. These are both very high when a cutoff probability of 0.5 is used indicating an accurate test.

Composing the Questionnaire

Identification of change in 16 items of the Part I of the ABS had been shown above to be good predictors for the development of DAD in adults with DS. The 16 items were now compiled into a questionnaire format that focussed on decline in these 16 items over time. Each item consisted of a question asking whether the respondent had recently experienced a change in a particular behavior on a scale ranging from "better than normal" to "much worse than normal" (where "normal" referred to when the respondent was well and before the onset of any recent ill-health). Errors due to "tendency to agree" were reduced by avoiding the use of a bimodal response scale and "error of central tendency" was eliminated by having an even number of response categories. The four-point response scale was treated as a multiple-response scale (Likert scale) with scores of 0, 1, 2, and 3 assigned to the four positions.

Calibration of the questionnaire was undertaken using two calibration groups—DS adults who were well, and those who had a clinical diagnosis of DAD according to ICD-10 criteria [26]. The latter group consisted of mildly demented, moderately demented, and severely demented persons. This approach was necessary to save items which could discriminate people who were well from those with mild DAD but at the same time be sensitive to various degrees of severity of DAD.

The questionnaire was sent out to a further sample of 100 DS individuals, with targeting to those DS persons with DAD, selected from those known to the clinical service. This sample included individuals were had not participated in the initial part of the study. Seventy-four completed questionnaires were returned (48 from non-DAD persons and 26 from patients with DAD). Each questionnaire was completed by the principal carer (family or paid carer). For those individuals who were also cared for by a second carer who also knew the patient in question well, the second carer was asked to complete and return the questionnaire independently of the first carer. Forty-two questionnaires were sent to a second carer, of which 36 were returned.

Questionnaire Analysis

It was apparent after the tests were returned that a significant proportion of carers had difficulty in answering the first question "Are they able to brush their teeth?" If the person had no teeth then "decline" in this behavior was not possible. Since approximately 15% of the returns had difficulty in completing this question, it was decided that this question would be excluded from any subsequent analysis, leaving 15 questions. The final 15 questions of the ABDQ are given in Appendix H.

The responses were coded as 0 (better than normal), 1 (same as normal), 2 (worse than normal), and 3 (much worse than normal). Analysis using the total of the 15 items was initially undertaken but improvement was found if a weighted total of the 15 items was used. A suitable weighting was derived as follows.

A series of l6 logistic regressions were performed each using two independent variables; the total of the 15 items and an individual item score. The most significant individual item was then identified and its weighting varied in the total score in line with its coefficient in the corresponding logistic regression. With this new "total" the above process was repeated until the weighting of each item had been reviewed. The weightings finally derived are shown in Table 10.7.

Figure 10.2 illustrates dot plots that show the total weighted score (TWS) obtained by the patients split according to DAD status using the ABDQ questionnaire. They show a clear distinction between the scores obtained by the nondemented and demented patient. Using a cutoff score on the TWS of greater than 78, a sensitivity for the ABDQ questionnaire to detect DAD was 89% and a specificity of 94%. The positive predictive value was 89% and the negative predictive value was 94%. The overall percentage correct identification (accuracy) of DAD and non-DAD cases was 92%.

Using the weighted items the questionnaire was developed further to categorize individuals into non-DAD mild DAD, moderate DAD, and severe DAD. The cutoff scores for the ABDQ for an ordinal logistic regression with severity of DAD according to ICD-10 criteria [26] and the weighted totals of the 15 items are given in Table 10.8.

Table 10.7 Weightings for questions in questionnaire

Question	Item	Weighting
1	Are they able to dress themselves better/same/worse than normal?	1
2	Can they use their hands better/same/worse than normal?	4
3	Is their ability to buy/shop better/same/worse than normal?	1
4	Are they able to have a conversation better/same/worse than normal?	1
5	Is their awareness of time better/same/worse than normal?	4
6	Do they help to prepare food better/same/worse than normal?	1
7	Do they help to clear the table better/same/worse than normal?	6
8	Are they able to perform simple jobs better/same/worse than normal?	4
9	Do they carry out simple jobs better/same/worse than normal?	5
10	Is their initiative in doing activities better/same/worse than normal?	1
11	Is their persistence in doing activities better/same/worse than normal?	1
12	Do they take care of their personal belongings better/same/worse than normal?	3
13	Is their cooperation better/same/worse than normal?	3
14	Do they participate in group activities better/same/worse than normal?	1
15	Is their ability to do things independently better/same/worse than normal?	1

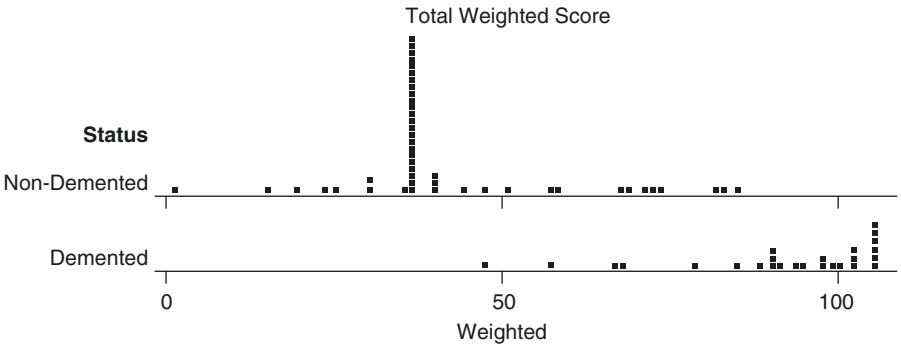

Fig. 10.2 Dot plot for demented versus nondemented DS individuals

Table 10.8 Questionnaire cutoff scores for severity of DAD

Severity of DAD[a]	Cutoff scores of ABDQ
No dementia in Alzheimer's disease	<78
Mild dementia in Alzheimer's disease	78–89
Moderate dementia in Alzheimer's disease	90–99
Severe dementia in Alzheimer's disease	≥100

[a]Dementia in Alzheimer's disease

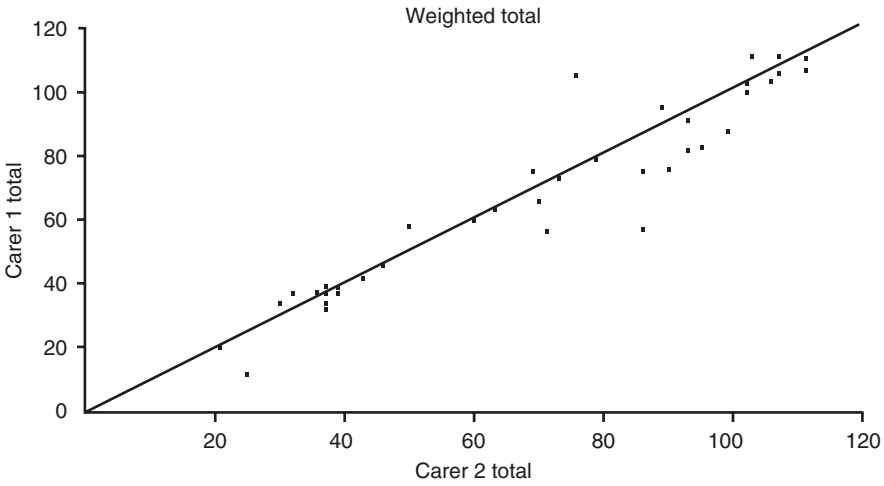

Fig. 10.3 ABDQ interrater reliability

Psychometric Properties of ABDQ

Interrater Reliability

The TWS from one carer was correlated with that reported by the second carer ($N = 36$) (see Fig. 10.3). Pearson correlation was 0.954 ($P < 0.01$). The findings demonstrate that the ABDQ questionnaire has good interrater reliability.

Validity

Face Validity

It has now been well established in the literature [27, 28] that onset and deterioration of clinical AD in adults with DS is associated with a significant decline in adaptive behavior. Further the principle instrument that has been used to measure adaptive behavior has been the ABS [3]. The items of the ABDQ questionnaire were derived from the ABS [3] and therefore do have good face validity. The 15 selected items which make up the ABDQ involve the detection of change in many of the different areas of abilities which are known to be affected in DAD, e.g., in orientation to time, attention, speech, self-care skills, social and occupational skills. Deterioration in adaptive behavior can, therefore, reflect decline in intellectual, social behavior, personal activities and emotional aspects of DAD.

Split-Half Validity

In order to verify the face validity, split-half validity was undertaken. To do this 74 patients were randomly split into two halves with 24 nondemented and 13 demented patients in each half. One half was then used to derive a weighting as above and the other half used to determine the validity of the weighted totals. All the items that were found to have weightings >1 were those that had greater weighting previously. There was good agreement between the two sets of results. The results of the binary logistic regression using the new weighted items gave an overall accuracy of 94%, and was comparable to 92% found previously.

Research Studies

Although the ABDQ has been widely used clinically, few research studies using the ABDQ have been published. Makary and colleagues [14] recently published findings using the ABDQ in their of adaptive behavior in adults with DS without dementia. The ABDQ was used to screen and to assess for dementia. Adaptive functioning being significantly and negatively associated with ABDQ scores.

Future Issues

Further field trials investigating the psychometric properties and clinical accuracy of the ABDQ to detect DAD in adults with DS are recommended. However, readers should be aware that the ABDQ has not been tested on non-DS adults with dementia, in persons with deterioration in physical health or onset of non-DAD psychiatric disorders, or investigated for the effects of demographic variables (e.g., age, race). Researchers are encouraged to investigate the use of the ABDQ in these areas.

Summary

Previous research by the authors [9, 27] over a 10-year period using the ABS [3] has demonstrated that this instrument can significantly measure deterioration of DAD in adults with DS. This work has led to the development of the ABDQ questionnaire which can be completed on all older adults with DS, irrespective of the underlying ID or degree of test compliance, which is an informant-based questionnaire, which has now been shown to have good reliability, validity, and accuracy. It is user-friendly and takes approximately 10–15 min to complete.

There continues to be ongoing methodological issues on flaws relating to research in the field of ID. The recruited sample size was 150 adults but was reduced to 119 persons for appropriate data analysis. However, this size remains a large sample compared to other previous studies in the field. This sample was found to be representative of adults with DS as it included a wide age range, males and females, different degrees of severity of ID and individuals resident in different settings. A significant number of individuals were diagnosed as having DAD (44 individuals) during the period, and again although this number may appear small compared to studies in the non-ID population, it is relatively large for studies of people with ID.

The problem of a gold standard in the diagnosis of DAD is an ongoing issue. The diagnostic process used was that recommended by the international research community [29]. Further, in this study individuals diagnosed with DAD were followed up (up to 6 years) after the diagnosis was made which allowed the reliability of the diagnosis according to ICD-10 criteria [26] to be further validated. The diagnosis of DAD was independent of the adaptive behavior assessment, and the analysis of the ABS data only took place after the 5-year period, ensuring no cross-contamination of data. Individuals with other causes of dementia other than DAD were excluded as part of the diagnostic process for DAD, and therefore, these findings reflect the specific screening for DAD in adults with DS, and not just for a general dementia disorder.

The diagnosis of DAD in the ID population requires further research. At present no definitive antimortem measure is available, and although a number of other neuropsychological measures to screen for dementia have been developed, virtually none have been accepted internationally and are not designed to specifically detect DAD. The Dementia Questionnaire for Mentally Retarded Persons [30], or now known as the Dementia Questionnaire for People with Learning Disabilities (Chap. 3), is widely used for screening for dementia. Conflicting results have been found regarding its validity [31, 32] to detect DAD although it may prove to be of value as a tool to assess treatment response in drug trials [33–36]. Other measures such as the Dementia Scale for Down Syndrome [37] (Chap. 4) have been produced, but again their reliability and validity needs to be independently researched [38]. The ABDQ has been developed from over 10 years of research investigating changes in adaptive behavior in adults with Down syndrome. It can be used for all adults with ID, irrespective of the severity of ID or DAD. It is "user-friendly" and specifically screens for DAD not just dementia per se.

Acknowledgment This chapter revised from original article. Prasher, V.P., Farooq, A, Holder, R (2004). The Adaptive Behavior Dementia Questionnaire (ABDQ): Screening Questionnaire for dementia of Alzheimer's disease in adults with Down syndrome. *Research in Developmental Disabilities*, 25, 385–397. Printed with permission from Elsevier.

Copy of the ABDQ available from author.

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Chapter 11
National Task Group Early Detection Screen for Dementia (NTG-EDSD)

Lucille Esralew, Matthew P. Janicki, and Seth M. Keller

Introduction

Alzheimer's disease, one of the major causes of dementia, is a progressive degenerative disease which causes loss of neurons in the brain and leads to neurocognitive dysfunction. As noted by Alzheimer's Europe [1], the symptoms may eventually manifest as dementia of the Alzheimer's type which impacts cognition, function and behavior, becomes progressively worse over time and cannot be reversed. The World Health Organization (WHO) [2] has noted that the prevalence and incidence projections indicate that the number of people with dementia will continue to grow, particularly among the oldest old and that the total number of people with dementia worldwide in 2010 is estimated at 35.6 million and is projected to nearly double every 20 years, to 65.7 million in 2030 and 115.4 million in 2050. Further, dementia has a devastating impact on adults with an intellectual disability (ID) as well as on their families, friends, housemates, and service provider staff who often provide key long-term support and care; and that community services' providers are facing a 'graying' of their service population, many of whom are affected by cognitive decline and dementia, and are challenged to provide the most effective and financially viable daily supports and long-term care [3]. Further, specialized assessment and diagnostic resources are needed to help more effectively identify adults with an ID and dementia and a common screening instrument would be useful for the early detection and follow-through to assessment and diagnosis [3].

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Alzheimer's disease is but one cause of dementia (or neurocognitive disorder), albeit a primary one. One recent study [4] of some 3.1 million health services beneficiaries in the United States found that 43.5% were diagnosed with Alzheimer's dementia, 14.5% with vascular dementia, 5.4% with Lewy body dementia, and 1.0% with frontotemporal dementia. Other types of dementia made up the balance. Studies show that persons with ID are as susceptible to the causes of dementia as are other adults, with the relative distribution of etiologies mirroring those of other adults [5]. One exception is among adults with Down syndrome (DS) who tend to express primarily dementia of the Alzheimer's type. Although these various dementias may have some variations in expression and early phenotypic markers, they all generally result in changes in progressive neurocognitive dysfunction and eventual death. The challenge, as noted by the WHO is to identity early those adults susceptible and affected so that assessment and diagnostic work-ups can be undertaken [2]. When behavioral changes are evident, the next step is to identify whether they are the result of a neurodegenerative disease or condition or whether some other factor (endocrine, psychiatric, pharmacological, neurotoxicity, etc.) is contributing to the decline.

To get to the stage where assessment or diagnostics are warranted, clinical and other services need to implement some form of screening to identify at-risk individuals. These same challenges present among adults with ID. While assessment processes and scales are in place for the general population [6], screening tools generally rely on brief patient assessments and surveys, such as the Mini Mental State Examination [7]. However, most, if not all, are inefficient for persons with ID [8, 9] due to intellectual variations, communication and performance challenges, and discordant context. Thus, the challenge: First, what type of dementia screening instrument might be useful to use with the range of functions expressed by older adults with ID? Second, what benefits may accrue from using a dementia screening instrument?

Screening

Early identification of signs and symptoms of cognitive and functional decline associated with dementia is an important first step in managing the course of the diseases causing dementia and providing quality care. Family and professional carers should work with the adult's health care provider to share information about observed changes. A screening tool can be used to substantiate changes in adaptive skills, behavior and cognition. In the United States, early detection is one of the aspects stressed by the *National Plan to Address Alzheimer's Disease* [10]. With early detection, assessment and diagnosis can be carried out to determine whether cognitive changes are the result of a neuropathological process related to disease or trauma to the brain, or attributable to other causes, often treatable and reversible. However, early detection among persons with lifelong cognitive impairments can often be difficult and problematic. With respect to screening, specialized measures

are needed that help take into account lifelong impairment and assist in picking up on subtleties in dysfunction.

Screening and the accompanying activities are important to effective early detection of cognitive decline and general dysfunction. To effectively address dementia-related decline, there are a number of steps that should be undertaken [5, 11, 12]. First, establish a baseline of 'personal-best' functioning and have staff who are familiar with the individual or family complete a screening tool in order to capture information about change. Second, share information from the tool with all members of the support or care team and with the adult's health care provider, and if the individual has had a rapid change in mental status consider that there may be a medical condition that warrants an immediate medical assessment. If the individual appears to be depressed or disoriented, have the person evaluated for medication interactions and ascertainment of whether depression is present. If there are sensory deficits these maybe contributing to decline in adaptive functioning, and these too require further assessment. Such factors may lead to decline and most likely are not neuropathologies. Thus, screening can help with starting the triage approach process in determining whether seemingly classic symptoms of dementia are potentially something else, or may actually represent the expression of dementia. Re-current screening then may be the first of many steps in determining the cause of functional and behavioral change in adults who are suspected of dementia.

The National Task Group

The National Task Group on Intellectual Disabilities and Dementia Practices (NTG) is a collective composed of over 300 agency personnel, academics, government officials, family members, and persons affiliated with various associations and organizations—most of whom are resident in the United States. The members are from medical and non-medical disciplines. The NTG is associated with several organizations in the United States (the American Academy of Developmental Medicine and Dentistry, the American Association on Intellectual and Developmental Disabilities, and the University of Illinois at Chicago's RRTC on Developmental Disabilities and Health) as well as numerous other university centers and national organizations (see www.aadmd.org/ntg). When the NTG was formed in 2010, its members recognized that no systemic and cross-cutting national-level plan existed in the United States that addressed the needs of adults with ID affected by dementia, and that these needs warranted systemic advocacy and attention. Its initial role was to address this issue and the growing requests for information and policy direction. Serendipitously, a new federal law that called for a 'national action plan' was enacted in 2011 and the NTG's role was expanded to advocate for the inclusion of ID in this national plan [3, 10].

The National Alzheimer's Project Act required the creation of a national strategic plan to address the rapidly escalating Alzheimer's disease crisis and called for coordination of Alzheimer's disease research and carer support efforts by the federal government. One of the considerations in this national plan was the promotion of an

assessment tool for detection of cognitive impairment as part of the annual wellness visit under the U.S. Patient's Bill of Rights and Affordable Care Act (PL. 111–148). As a result, the NTG undertook to develop an administrative screening and early detection instrument for dementia among adults with ID that could be easily used by family carers and service provision staff. This chapter describes the development and use of the National Task Group Early Detection Screen for Dementia (NTG-EDSD) screening instrument, and explores factors associated with and benefitting from screening.

The benefits of differentiating types of dementia as part of the assessment and diagnostic process, include diagnostic precision; pharmacological treatment applications; projections of residual life years; assemblage of care management plans to address expected behavioral presentations and progression, communication and interaction variations and projecting expectations for change in care needs; and referrals for diagnosis and introducing post-diagnostic measures.

The National Task Group Early Detection Screen for Dementia (NTG-EDSD)

The NTG-EDSD is an informant-based rating tool for use with adults with intellectual and developmental disabilities who are suspected of experiencing changes in thinking, behavior, and adaptive skills suggestive of mild cognitive impairment or dementia [11, 13]. The form is a compilation of general information about the adult, health status information, pharmacological usage, and a variant of the DSQIID [14, 15]. It is considered an administrative, and not a clinical assessment, tool. The NTG-EDSD was developed to collate behavior and health information, capture early changes in function, and specialize in accounting for subtleties in these changes [11, 13].

The Historical Basis for the NTG-EDSD The NTG-EDSD has its roots in a meeting held in the mid-1990s, which was the first time a collective of international researchers interested in dementia and ID came together. In 1994, a conference grant from the National Institute for Health helped support a meeting held in Minneapolis, Minnesota, held in association with the Fourth International Conference on Alzheimer's Disease and Related Disorders, which was one of the early iterations of the international conference on Alzheimer's disease now known as the AAIC (Alzheimer's Association International Conference—see https://www.alz.org/aaic/) [16]. The outcomes and products of this meeting included a number of reports and publications as well as the formation of an informal network of the researchers in the field of ID and dementia. One of the papers that resulted from the meeting was co-authored by a team led by Drs. Elizabeth Aylward and Diana Burt [17]. While the work of this group was useful to researchers and professionals conducting dementia assessments, it left open what might be applicable for use by lay

workers and family carers. Over the years, there evolved a growing interest in the early recognition of cognitive, behavior, and adaptive changes that could be substantiated by family and staff carers. Provider agency staff indicated that they needed an instrument for early detection and initial screening that could be used by direct support workers and families. The original instruments cited in the 1996 effort were direct and informant-report assessments requiring professional level administration or interpretation. Many agency staff and families did not have access to psychologists and other practitioners who had the expertise to conduct such extensive assessments and noted the need for something that could serve as a less complex early detection measure. Furthermore, there was increasing demand within the field in general for a rating instrument that could help capture information about changes that could then be shared with clinical teams and health care practitioners to advance services. Subsequently, a number of short form symptom identification measures of varying complexities were developed and introduced into the field [4, 12, 18].

When the NTG was organized in late 2010, among its first tasks was to identify a basic user-friendly screening tool that could be widely used as a first pass screen for early detection of changes and which would identify individuals who needed an additional and more comprehensive assessment. Consequently, the NTG charged a working group to undertake a process to recommend such a screening tool. During this process the working group sought input and involvement from some of the original members of the 1994 workgroup on diagnosis and assessment and others regarding tools that were in current use and which had proved helpful in identification of individuals who might have dementia. The outcome, following a review of various extant instruments, was the endorsement of the use of a tertiary modification of the Dementia Screening Questionnaire for Individuals with Intellectual Disabilities (DSQIID) [14], as well as inclusion of other information pertinent to screening (such as demographic information, health and function status, co-incident conditions, living arrangement, and medication usage). The original work in adapting the DSQIID in the United States was done by a group in Philadelphia, Pennsylvania, with the help of Carl Tyler of the Cleveland Clinic [3]. The instrument was also field tested at various sites in the USA and Canada [16], and was the subject of an efficacy study in Austria and Germany [19].

The NTG-EDSD was designed as a way of collecting key information and enabling the recording of indicators and signal behavioral markers of significant change [13]. The purpose was to give family and professional carers a tool that would enable them to capture objective data on changes in function when suspicions arose and prior to making a referral for a comprehensive assessment. As such, the NTG-EDSD is regarded as an administrative rating tool and not an assessment instrument. In the United States, the NTG-EDSD could also present helpful data which can be shared during the annual wellness visit under the U.S. Patients' Bill of Rights and Affordable Care Act, as many service agencies were looking forward to that process to help them with identifying any significant potentially neuropathologic functional and cognitive changes among the individuals whom they support.

The Uses of the NTG-EDSD When applied, the NTG-EDSD provides an opportunity to review relevant information that can be used by the team and healthcare practitioner to aid in shared decision-making and planning training, services, and supports. The NTG-EDSD was not designed to diagnose dementia, but to be a help in the early identification and screening process, as well as to provide information to begin the dialogue with clinical teams and health care professionals. Persons who complete this instrument are asked to indicate whether they have observed the occurrence of new problems or a worsening of problems that have previously been observed. The items are associated with changes in cognition, behavior, mood, and activities of daily living.

The NTG-EDSD is considered an 'administrative' tool as it is not a clinical determination instrument—it doesn't result in a validation of a clinical impression or a diagnosis. It, however, permits the user to note dysfunction in key areas generally associated with the expression of dementia, helps confirm suspicions of change in behavior by concordance with key marker items, and helps with statistical reporting of functional areas of concern. It also enables carers to have a means of creating a summary of behavioral changes and events and health shifts that can help form a basis of discussion among carers and between carers and clinicians. The completed form can also be entered into the adult's medical record or program plans and successive iterations compared for variations from the baseline. Specific information on the interpretation of items and processes to use to collect the data for the NTG-EDSD are found in the instrument's manual [13].

A tool, such as the NTG-EDSD, is meant as a first-pass screening to identify individuals who might need more comprehensive assessment. Each service setting can develop its own protocol regarding how information from this assessment can best be utilized on behalf of the consumer. However, it is conceivable that care paths might include sharing the information with the adult's physician, deciding if there needs to be a change in programmatic or personal care supports, a reallocation of resources, or recognizing the implication for the residential setting. The adult's team may want to adopt a "watchful waiting" approach in which certain areas of identified change are further monitored through additional data collection via the use of the NTG-ESDS or other means. As many service agencies indicated that they did not have access to professionals who could provide a cognitive screening, the NTG wanted to the tool to be accessible to carers who were not necessarily trained to do assessment, but had valuable information regarding day-to-day changes in functioning—or experience or observed telling behavior (see Table 11.1). The tool needed to be easy to administer, could not be time consuming, and should be sufficiently robust to yield information that could be used as an aid in shared decision-making. The items that make up the NTG-EDSD are associated with the changes typically observed in dementia [14] (see Table 11.2). Via the use of this screening tool caregivers or staff can substantiate if a person with ID manifests these changes and can then share the information with health care providers.

Components of the NTG-EDSD The NTG-EDSD is composed of four primary sections containing some 40 questions or question groupings about relevant subject

Table 11.1 Noted symptoms and notable incidents that lead to suspicions of dementia

Common symptoms	Notable incidents reported by group home staff	Notable incidents reported by family carers
Memory loss	Wandering	Falling
Difficulty performing familiar tasks	Falling	Difficulty eating
Problems with language	Decline in general abilities	No longer talking
Disorientation to time and place	Short term memory loss	Increased aggression
Poor or decreased judgment	Increased aggression	Short-term memory loss
Problems with keeping track of things	Increased conflicts with peers	Throwing self on the floor
Misplacing things	Called fire department instead of taxi for outing	Decline in general abilities
Changes in mood or behavior	Safety issues	Undressing inappropriately
Changes in personality	Stealing others possessions	Difficulty getting out of bed
Loss of initiative	Medical problems	Increased conflict with peers
	Uncommon behaviors (stuck head in toilet to wash hair)	Becoming disinterested in activities
		Medical problems (e.g., seizures, incontinence)
		Other problems (such as 'trying to make guests leave house')

characteristics, ratings of health, mental health and life stressors, a review of multiple domains associated with adult functioning, and a review of chronic medical conditions (see Appendix I for the original American English language version[1]). It also provides for a notation on the number and nature of medications being taken, and permits comments on observations to be entered. Specifically, the NTG-EDSD contains ten basic demographic items (such as identification data, personal characteristics, diagnostic, and residential setting information), eight health and function items, and the adaptation of the DSQIID (including queries as to activities of daily living, language and communication, sleep-wake change patterns, ambulation, memory, behavior and affect, the adult's self-reported problems, and notable significant changes observed by others).

The NTG-EDSD includes a listing of chronic illnesses from the University of Illinois at Chicago Longitudinal Survey [20], which can be used to note co-incident conditions (these include the following categories: bone, joint and muscle; heart and circulation; hormonal; mental health; pain-discomfort; sensory; and other). The co-occurrence of chronic illness and neurocognitive disorder, for instance, cardiovascular issues and diabetes are among highly co-incident conditions for dementia of the Alzheimer's type in the general population. These data are

[1] Other language versions of the NTG-EDSD can be found at www.aadmd.org/ntg/screening.

Table 11.2 Correspondence with prevalent markers/indicator of dementia and the NTG-EDSD

Feature	Descriptors	Applicable NTG-EDSD area
Behavioral and psychological symptoms of dementia (BPSD)	Behavioral symptoms include physical, social inappropriateness, hitting, pushing, scratching, kicking and biting, throwing things, wandering/pacing, hoarding, verbal screaming, cursing, temper outburst, complaining or whining, repetitive sentences, verbal sexual advances, constant request for attention, rummaging, and nighttime wandering. Psychological/psychiatric symptoms can include anxiety, depression, hallucinations or delusions.	[24] Behavior and affect (p. 4)
Memory	Decline in memory. Memory changes and increased confusion resulting in problems with accepting personal assistance;	[23] Memory (p. 4)
Sensory impairment	Changes in vision and hearing and loss of sensory acuity can be related to increased confusion and agitation. The inability to process information about the environment through our senses can either increase agitation or present as increased lethargy and disengagement.	Conditions present (p. 2) Items 30–32 sensory (p. 5)
Psychosocial stressors	May be evident via significant losses and significant changes. Occur when situations exceed the person's ability to adaptive or effectively respond and may be expressed by agitation, confusion, or being overwhelmed to the point of being less functional.	[14] Significant recent (in past year) life event (p. 2)
Seizures	May be evident among adults with a history of seizures, but after a long period of seizure inactivity, may show breakthrough seizures. Also, may occur in new late-onset seizures in someone who never previously had seizures (particularly in adults with Down syndrome).	Seizures (p. 2)
Gait and balance	Neurological changes may be evident in an increase in gait and balance problems.	[26] Notable significant changes observed by others (p. 4)
Changes in Activities in Daily Living (ADLs)	Individuals with neurocognitive disorder may show difficulties in sequencing (affects dressing, eating, and toileting independently), visual spatial (safe ambulation and finding way within environment); Language and communication; verbal memory problems lead to loss of words; impoverished speech; lack of spontaneous speech; receptive language issues. Sleep-wake pattern changes due to changes in circadian rhythm.	[19] Activities of daily living (p. 3)

important as effective treatment of chronic medical conditions can increase quality of life for the person who does have a neurocognitive disorder. Also, the data create an opportunity to see the patterns of co-occurring medical problems with dementia. The last section of the NTG-EDSD also contains an item on current medications; a place to note comments related to other notable changes or concerns, as well as

next steps and recommendations. Lastly, there is an area for notations on form completion.

Completion and Use of the NTG-EDSD The NTG-EDSD can be completed at any point in time on an adult with ID. The NTG recommends that the instrument be used with adults with DS beginning with age 40 (or earlier—age 35—if functional decline is suspected), and with other at-risk persons with intellectual or other developmental disabilities when they are suspected of experiencing cognitive change. The form can be completed by anyone who is familiar with the adult (that is, has known him or her for over 6 months), such as a family member, agency support worker, or a behavioral or health specialist, using information derived by observation or from informants, as well as the adult's personal and medical record. Carers and staff with close knowledge and familiarity are more likely to be aware of subtle changes in behavior and functioning that may signal important information for health care providers. What should carers or staff observe? First, they should look for changes from characteristic baseline behaviors in cognition (memory, attention, problem solving), behavior (social and control of impulses), and emotion (mood, emotional regulation). Secondly, they should look for changes in expected function in activities of daily living. The estimated time necessary to complete this form is between 15 and 60 min. Minimally, it can be used on an annual or on an "as-indicated" basis when there is a suspicion of cognitive change.

Use with Medical Visits The NTG-EDSD can also be used in preparation for medical visits. Having concise information available for the examining physician can help instigate queries and any follow-up assessments. Moran and colleagues [21] and Prasher [22] covered the manner of examining for potential dementia; the NTG-EDSD can be instrumental for conveying key health and dysfunction information by informants as a preliminary during such an assessment visit [13].

In situations where the healthcare provider may not know the individual, especially if a new practitioner or specialist is being consulted, data aggregated from the NTG-EDSD can be most useful to complement any interview data. This will be particularly helpful with practitioners who do not have experience in the care of individuals with ID and may not be able to discern functional decline associated with normative aging from that of an underlying pathology, such as that seen in early onset Alzheimer's disease. Also, when a practitioner may have unintended biases and stereotypes and apply "diagnostic overshadowing," the data can be used to offset this when the adult with ID is brought into the office or clinic for evaluation. Carers also may also be susceptible to "diagnostic overshadowing"—that is, they may have observed change but interpret such to be in line with aging. Such diagnostic overshadowing can occur when there is an automatic tendency to attribute all changes to the adult's primary ID and thereby overlook other medical and psychiatric issues that may need attention.

As part of this interview and assessment visit, to complement the data shown on the NTG-EDSD, it is helpful for the carer or family member also to provide a general life synopsis or life story that includes some details about the adult and his or her history, which should include information on dietary patterns, language skills,

family history, social history, psychiatric history, medications, substance abuse, and past medical history [21]. The NTG-EDSD data can then be presented as current information about issues or problems noted, with any changes noted. This collection of information can then be put into a fuller context with respect to whether the change(s) noted is a natural sequalae of the ID, a function of aging, an effect of a comorbidity, or the result of possibly emerging neurocognitive disorder. Thus, the findings from the NTG-EDSD can be contextualized within the person's life story and information regarding changes from his/her previous highest baseline. The importance of this process is key since it will now be up to the practitioner to decide what the next steps should be [23].

Consequently, the practitioner may decide to track the individual's behavior and function to ensure that decline is indeed occurring, but not rush to any immediate conclusion and wait for a subsequent assessment visit. However, if it has been made clear via the NTG-EDSD and other data that decline has absolutely taken place, then it is up to the practitioner to institute a work-up and evaluation appropriate for adults with ID looking for possible explanations for these changes. In this context, the carer and family can add their voice as advocates and supporters and speak up on behalf of the adult as they helped to provide baseline information and other facts, including the NTG-EDSD.

How to Use Observations Captured by NTG-EDSD? Suspicions of cognitive and functional changes are often aroused by everyday events (see Table 11.1). Families and staff may perceive select behaviors that are out of norm or context and question them. There might be a need to reflect on antecedent event(s) which may have triggered the behavior. Observing to see if the behavior repeats or whether it is triggered by something unrelated is important and should lead to a discussion with persons providing supports to the adult. If warranted, this may call for further tracking of changes on the NTG-EDSD in key areas of functioning and using the information gleaned for advanced planning regarding staffing, residential, and programmatic decisions. It can be helpful if the initial review using the NTG-EDSD can be accompanied by notes indicating onset of conditions.

Following the initial review which would serve as a baseline, the carer or staff person completing the form can indicate whether there has been a change since the last review. If concerns are raised and the individual is determined to need a specialized assessment, a referral should be made for more comprehensive work-up that would include medical and psychological testing. The team can share ratings of "new symptoms" or "always but worse" with the examiner and discuss among members of the team implications for programming, personal assistance, residential placement, services, and supports. With the issuance of the APA's Diagnostic and Statistical Manual of Mental Disorders -5th edition (DSM-5) [24] and the Diagnostic Manual for Intellectual Disability (DM-ID-2) [25], the health care practitioner can link documentation of change with updated criteria for the diagnosis of dementia. The DM-ID-2 is the preferred manual to consult regarding neurocognitive disorders in adults with ID.

Case Studies

The following case scenarios are offered as illustrations of how the findings from the NTG-EDSD can be used to guide the decision-making activities by members of the adults' support teams.

Example	Back story related to ratings on the NTG-EDSD	Screening/Impressions
Case presentation #1: Laura	Laura is a 55-year-old woman with a history of mild ID and DS who had been living with her mother in their family home until that parent needed placement in a nursing home… and then Laura went to live with her older sister Mary and Mary's family. Mary has noticed that Laura is not attentive to her hygiene and appears to take showers irregularly. Mary reports that Laura often seems "…out of it" and spends most of her time while at home in her room. Laura works at Tesco. Laura's work supervisor called with concerns that Laura has gotten into a few arguments with customers, recently, one of whom filed a complaint. Although Laura has several friends with whom she had been socializing or with whom she had maintained phone contact, she has not kept up with social dates for several months.	Further investigate the impact of psychosocial stressors. Track changes with more frequent use of NTG-EDSD and look for signal changes that may warrant assessment Ask to what extent might she have displayed earlier decline in adaptive skills that were either compensated for by the mother or unobserved and unaddressed? Investigate extent to which this is an adjustment reaction to changes in her living situation with accompanying depression
Case presentation #2: James	James is a 67-year-old male with a history of moderate ID who lives in a group home. He has worked at a production center for 18 years and has always had the reputation of being a quick, productive and efficient worker. Over the past 6 months he has been slowing down and earning less on his paycheck; he has been displaying difficulty learning the names of new staff at his program and residence; he has been forgetful with regards to doing routine chores in the group home. He wears hearing aids and glasses; he has lost both these devices and both are in the process of being replaced. He has a history of diabetes and arthritis.	Further investigate the role of sensory deficits in his performance. Develop a routine for storage of hearing aids. Use NTG-EDSD to coalesce data from the work and home sites – To review for disparities Work supervisor might assess to see if there are any modifications in work task demands or opportunities for a change in day activities that may be beneficial. Consideration given to possible shift to 'retiree' status

(continued)

Example	Back story related to ratings on the NTG-EDSD	Screening/Impressions
Case presentation #3: Stephanie	Stephanie is a 70-year-old woman with a history of bipolar disorder and mild intellectual impairment and spectrum disorder. Although maintained on medication, she is displaying more rapid cycling including periods when she appears to experience psychotic features while manic. She has become verbally aggressive, property destructive and combative with staff–all of which had been noticed during manic episodes but are now occurring on a regular basis despite apparent mood stability. She has displayed episodes of disruptive and impulsive behaviors leading members of her team to believe that she can no longer safely remain in a supervised living situation.	Further investigate medication management of bipolar disorder. Look for collateral signal data on NTG-EDSD beyond psychiatric symptoms Has she become a "rapid cycler" which may render current regimen insufficient. Investigate how can the support team provide positive routine and realistic limits to behavior that promote safety.

A number of questions and queries for follow-up can surface following use of the NTG-EDSD (see case studies). For example, the user and the team can ask whether the adult has displayed new symptoms in at least two domains on the NTG-EDSD. Alternatively, it may be noted whether the adult has gotten worse for symptoms already noted in the two areas. Other factors to consider is whether delirium and/or depression have been ruled out? Delirium would be notable if the confusion and other disruptions in thinking have had an abrupt onset. Depression typically would involve lack of affect ('down mood') and a lack of interest in what is going on around the person. At times, hallucinations may be a factor. Hallucinations would be evident if the adult sees or hears someone speaking who is not there. Often hallucinations may occur in more severe forms of dementia and are generally indicative of Lewy body or Parkinson's disease dementia. Individuals with dementia of the Alzheimer's type can also experience hallucinations and paranoia that appear psychiatric in presentation; however, these symptoms are related to the brain changes associated with dementia. Another rule-out is the adverse effects of medications. Some medications may interact and create adverse interactions or have side effects that may mute or alter behavior. Also, there may be circumstance in the adult's life or environment or in relationships that may adversely affect behavior in some of the domains. All of these considerations should be factored in when discussing the information obtained by the NTG-EDSD.

Usage to Date The NTG-EDSD has been adopted for screening by various services agencies and organizations across the world [26, 27]. The form is available in multiple language translations (all forms are available at www.aadmd.org/ntg/screening) and organizations are welcome to translate or adapt the form into their language

as long as the core items are not changed and due credit—is given to the NTG. As an example of use for broad screening, NHS Health Scotland [26] has recommended that local screening efforts using the Scottish adaptation of the NTG-EDSD be undertaken as follows (a) establish a baseline assessment of functioning against which to compare future suspected changes and that this should begin at age 30 with adults who have DS; (b) screen adults with DS over the age of 40 every 2 years and annually after the age of 50 (as recommended by the British Psychological Society because of the increased risk of dementia and the prevalence of undetected but treatable illnesses) and that this screening be linked to the person's overall health action plan; and (c) for individuals with ID other than DS, a baseline assessment should be conducted at age 50 with no further action or screening until concerns are raised.

Applications have also included inclusion in research where the NTG-EDSD is used as one of the instruments in both small and large population studies. Small, such as the Wichita Project study of small group homes for community-based dementia care being undertaken by the University of Illinois at Chicago [28, 29] and large, as the US National Institute on Health's US$37 million dollar multisite 'Biomarkers of Alzheimer's Disease in Adults with Down Syndrome' study [30]. Further, as a screening tool, it has been recognized as having utility among other tools generally used to identify early signs of dementia [9, 31, 32].

Applications

Staging and Etiology Data drawn for one or more successive administrations of the NTG-EDSD can help with framing suppositions about possible type of dementia and about staging. Why is it important to know about etiology (type of dementia) and staging of dementia with respect to planning care? Both can provide insights into the nature of the behavior and losses of function that the adult may experience and consequently impact staffing, programming, interventions, and adaptation in the living environment. Although the NTG-EDSD should not be used for diagnostics, preliminary assumptions about the type of dementia might be drawn from the nature of the domains that show particular changes. For example, data showing progressively declining sharpness in memory, sleep impairments, and other adverse functional changes might suggest the presence of Alzheimer's disease, whereas, memory consistency, but marked changes in performance, may suggest frontotemporal dementia.

As the NTG-EDSD is not a diagnostic tool, but a screen, any suppositions should only be used to help to stimulate discussion within the team and for framing the referral to a clinician for an assessment or diagnostic work-up. Particularly for individuals with no known psychiatric history, the onset of paranoia, hallucinations or behavioral aberrations from known characteristic baseline should alert the team to the need for further assessment and diagnostic clarification. The same comment applies to staging. At times, the adult may present for screening whilst already showing clinical features of progressive or mid-stage dementia (as opposed to mild cognitive impairment [MCI] or early stage dementia). In such cases, experienced

clinicians and team members may be able to make some assumptions about staging, particularly if there is a strong presence of hits on markers on the NTG-EDSD and other clinical indicators (and there is evidence of changes from pre-morbid functioning). Thus, getting validation of the potential etiology and confirmation of staging can be beneficial to planning the interventions to be developed, for estimating duration of being affected by dementia, and planning staffing and environmental modifications. Research is needed that would substantiate that general ratings on the NTG-EDSD align with different stages of neurocognitive disorder.

To offer some context, these features warrant consideration. Dementia is a description of a clinical phenomenon of significant decline from pre-existing baseline functioning in cognitive, behavioral or social skills that interferes with daily functioning. By capturing information on changes in behavior, emotional and social functioning and everyday behavior, the NTG-EDSD can be used to identify items that lead to suspicions of dementia for a particular person.

Dementia is not a clinical diagnosis; it is a clinical description of observed change in functioning. Dementia does not indicate etiology of decline, it denotes a state of significant changes in cognition, function, and/or behavior that interfere with the individual's independence and pursuit of daily routine and relationships. Dementia describes the effects of neurocognitive disorder such as probable Alzheimer's disease, cerebrovascular dementia, frontotemporal dementia, and other dementias [33].

As the NTG-EDSD includes a listing of chronic conditions and illnesses from the University of Illinois at Chicago's Longitudinal Health in Intellectual Disability Survey (LHIDS) [20], the notation of the co-occurrence of chronic conditions and illnesses may signal some aspects associated with neurocognitive disorder. For instance, cardiovascular problems and diabetes are among highly co-prevalent conditions for dementia of the Alzheimer's type. The presence of such conditions may be telling, as studies have shown that adults with ID and dementia tend to have about twice as many comorbidities as those age- and function-matched adults absent dementia [34, 35]. Tracking such chronic medical conditions and providing effective treatment can increase quality of life for adults with a neurocognitive disorder.

Neurocognitive disorders are progressive and deteriorative. As the adult moves through stages of dementia regardless of the etiology, the adult will need increased personal assistance and supervision. Thus, offering training toward competency in provision of dementia-capable services is warranted. Staff working with adults with dementia should have a grounding in facets of normal vs. pathological aging, neurocognitive disorders, variations of dementia and staging, health and social care practices, as well as day-to-day care management of people with dementia.

Commentary

A few closing comments. Screening and the accompanying activities are important to effective early detection of cognitive decline and general dysfunction. To effectively address dementia-related decline, there are a number of steps that should be

undertaken. First, establish a baseline of 'personal-best' functioning and have staff who are familiar with the individual or family complete the NTG-EDSD in order to capture information about change. Second, share information from the rating scale with all members of the care team and with the adult's health care provider, and if the individual has had a rapid change in mental status consider that there is a medical condition or cerebrovascular accident present and not necessarily dementia. If psychiatric symptoms are evident, such as depression, have the person evaluated for medication and psychosocial approaches to depression management and eliminate any factors that may be leading to decline that are not neuropathologies.

Findings from the NTG-EDSD should be reviewed by a health care practitioner who knows something about the profile of change for dementias and about appropriate assessment/evaluation methods in adults with ID. He or she can therefore steer further inquiry and assessment in thoughtful ways. The Diagnostic Manual for Intellectual Disability-second edition (DM-ID-2) [25] has an extensive chapter on neurocognitive disorders. Findings from the NTG-EDSD can be linked with the criteria within the DM-ID-2. Guidelines can be provided to medical and non-medical healthcare practitioners linking findings from the EDSD with treatment and support planning.

The NTG-EDSD is an evolving instrument. Since it is a "work in progress," studies and reports of usage outcomes are welcome, as are questions which can help guide further development of the tool. Although Deb and colleagues [14] did propose a threshold 'score' when using the DSQIID, there is no comparable "score" for the overall NTG-EDSD. Whereas the DSQIID is intended as a diagnostic screening, the NTG-EDSD is intended to collect broader information and to be an administrative tool to support healthcare decision-making. The ratings on the tool correspond to observed changes in functioning. Family and professional carers can share these ratings with a healthcare provider. Currently, DSM-5 and DM-ID-2 criteria for dementia can be used to determine if there has been "significant change" to warrant recommendation for further evaluation or if other recommendations are indicated to address issues that affect cognitive and adaptive functioning that may not be related to dementia.

Lastly, the authors have received correspondence from worldwide sources providing anecdotal support for the NTG-EDSD as a useful means for capturing information about change. Future directions include surveying groups that have identified themselves as "superusers" of the instrument in order to determine how findings are being used to advance the healthcare and support needs of persons with disabilities with suspected neurocognitive disorder.

The ratings from the NTG-EDSD can be used to train carers on case-based characteristics of change over time for individuals with suspected neurocognitive disorder [13]. Moreover, the rating schema within the NTG-EDSD highlights which behaviors to observe for change; this can serve to advance carers' ability to advocate for individuals with dementia with their healthcare providers [23]. Research undertaken could examine the utility of serial re-assessments and the influence of variations in staff/family completers, impact on clinical determinations, and relationships among marker items, as well as compare the validity of the threshold score cited by Deb and colleagues [14] with that potentially derived via the adapted DSQIID items

within the NTG-EDSD. We encourage the open access to and use of NTG-EDSD and trust that both practitioners and researchers will find ways to enhance the instrument and undertake studies illustrating its utility and reliability.

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Chapter 12
The Rapid Assessment for Developmental Disabilities

Christy L. Hom, David M. Walsh, Eric Doran, and Ira T. Lott

Introduction

The *Rapid Assessment for Developmental Disabilities* (RADD) was developed to address the challenges of measuring cognition in individuals with intellectual disabilities (ID). As opposed to individuals with ID, commonly used intelligence tests such as the *Wechsler Intelligence Scale for Children* (WISC) [1], the *Wechsler Adult Intelligence Scales* (WAIS) [2], and the *Stanford-Binet* (SB) [3] were designed for use with the general population [4]. By design on these measures approximately 95% of the population have IQ scores that fall within two standard deviations (SD) from the mean (scores between 70 and 130; Fig. 12.1). Individuals with ID typically have IQ scores more than two SDs from the mean and they represent less than 2.5% of the total population, forming a statistically rare group. Commonly used intelligence tests were not designed to differentiate between the varying levels of ability among individuals at this lowest end of the intelligence spectrum. The RADD was developed to fill the need for measuring the cognitive abilities of individuals with IQ scores more than two standard deviations below the mean (scores below 70).

Traditional intelligence tests often require over an hour to administer and can be problematic for individuals with ID who may have brief attention spans, maladaptive behaviors, or have limited abilities to comply with instructions or requests [5, 6]. Often these individuals have more serve cognitive impairments and because of floor effects, the WAIS, WISC, and SB do not have the ability to differentiate between severe and profound levels of ID [7]. Additionally, the high incidence of sensory, motor and speech impairments among individuals with ID [8] complicates

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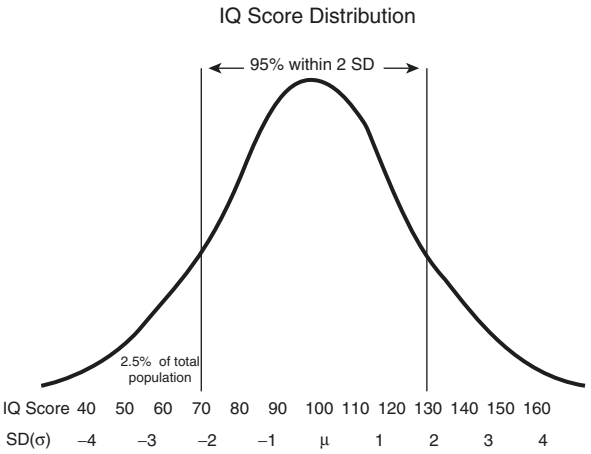

Fig. 12.1 RADD designed to measure statistically rare individuals

the interpretation of test results because these batteries rely heavily upon language skills [9].

When using mainstream IQ tests for individuals with ID, one method of addressing these challenges has been to depart from the standardized protocols, and instead, use only specific components of the WISC, WAIS, or the SB. In such instances, examiners may select tests geared toward the individual's strengths and omit tests that require physical, sensory or speech abilities that are reflective of the developmental deficit [6]. Another alternative is to utilize a fixed battery such as the *Slosson Intelligence Test* (SIT) [10], which was designed for use with individuals with more severe cognitive deficits. While the SIT can be more rapidly administered than other intelligence tests, its heavy verbal loading and poor psychometric qualities may be problematic [11]. Departing from the standardized protocols may result in considerable challenges in comparing individuals across the spectrum of ID and between studies.

Rationale

The RADD was developed to provide a brief, valid and reliable instrument for measuring cognitive functioning in individuals with ID. It provides information about a wide range of functional abilities including receptive and expressive language, orientation, registration, recall, attention, self-identification, motor skills, imitation, abstract reasoning, number skills, comprehension and short-term memory. The entire battery takes less than 25 min to administer, and can be administered to individuals who are non-verbal. It can distinguish between all levels of ID, including severe and profound. This capacity of the test is particularly useful in the evaluation of declining cognitive functions in individuals with Down syndrome (DS) and dementia.

Development of the RADD

The RADD consists of 76 items selected from the following neuropsychological tests: the *Standardized Mini-Mental Status Examination* (MMSE) [12], the *Severe Mini-Mental State Examination* [13], the *Expressive One Word Picture Vocabulary Test—Revised* (EOWPVT-R) [14], the *Peabody Picture Vocabulary Test—Revised Form M* (PPVT-R) [15], the *Merrill-Palmer Scale of Mental Tests* (MPSMT) [16], the *Hawaii Early Learning Profile* [17], and the *Wechsler Intelligence Scale for Children—Third Edition* (WISC-III) [1]. Scoring on the RADD is consistent with protocols from the original source tests (0 = incorrect, 1 = correct), with one exception. The WISC-III protocol allows scores for Similarities and Comprehension items to range from 0 to 2; however, the RADD deviates from the WISC-III protocol and assigns a score of 0 or 1.

Two-hundred and seventy one individuals with ID participated in the RADD's development. As part of the formative phase of test development, 68 individuals with ID were evaluated with items selected from the source tests listed above. The items that differentiated between levels of ID were retained in the final version (Table 12.1). The RADD standardization population consisted of 203 individuals with ID, including DS [18]. The RADD's validity specifically for individuals with DS, with and without dementia, was subsequently explored through a second study [19].

Table 12.1 RADD items and original test sources

RADD subtest	Items	Source
Orientation	Week, month, year, location, state of residence	Standardized mini-mental status examination
Registration	Ball, flag, tree	
Recall	Ball, flag, tree	
Attention—forward	C-A-T	The severe mini-mental state examination
Attention—backward	T-A-C	
Self-identification	Language item 4 All or none item 10	Merrill-Palmer scale of mental tests
Movement	All or none items 13 and 14	
Imitation	Gestural imitation section	Hawaii early learning profile
Expressive language	1, 6, 11, 12, 25, 26, 31, 33, 42, 46, 54, 56, 60, 62, 70, and 73	Expressive one-word picture vocabulary test—revised
Receptive language	1, 3, 16, 17, 45, 48, 65, 67, 81, 82, 91, and 92	Peabody picture vocabulary test—revised, form M
Similarities	1, 6, 7 and 8	Wechsler intelligence scale for children-third edition
Arithmetic	6, 7, 9, 12 and 14	
Comprehension	1, 4, 6, 7 and 9	
Digit span	1, 2, 3 and 4 (both trials)	

Materials Needed for Administration of the RADD

A hand-held mirror and the picture cards from the EOWPVT-R and PPVT-R used for the expressive and receptive language subtests are the only materials needed.

Psychometric Properties

The RADD's validity and reliability were first evaluated among individuals within the full spectrum of developmental ID. The psychometric properties were then examined among individuals with DS, with and without dementia.

Study 1 [18] The RADD standardization was focused upon 194 participants with ID, after nine subjects were excluded due to missing data. The standardization sample was 63% male, with an average age of 37.63 years (standard deviation; SD = 11.93). Approximately half of the sample (47%) lived in the community and were referred to a developmental disabilities clinic to address behavioral disturbances, psychiatric comorbidities, or polypharmacy. The remaining sample of individuals (53%) lived in a state residential facility for individuals with ID. Data such as gender, age, scores from most recent standardized intelligence tests, and level of ID were gathered from participants' medical records. Level of ID was placed into a four-point ordinal scale, with "1″ = "Profound," "2″ = "Severe," "3″ = "Moderate," and "4″ = "Mild".

Study 2 [19] The RADD was compared to informant-based and direct measures of cognition and dementia among 114 individuals with DS with and without Alzheimer's type dementia. Approximately 55% of the sample was male, with an average age of 49.8 years (SD = 8.9). Trisomy 21 was independently verified. Premorbid ID levels were gathered from participants' medical records. The sample consisted of participants with mild (35%), moderate (39%), severe (23%), and profound ID (3%). Approximately 62% of participants were diagnosed with dementia. The time interval between dementia onset and date of testing ranged from 3.7 to 79.8 months, with a mean interval of 29.1 months (SD = 17). Individuals who were diagnosed as demented met the criteria from the *International Classification of Diseases, Tenth Edition* (*ICD-10)*; [20] and *Diagnostic and Statistical Manual of Mental Disorders, Fourth Edition, Text Revision* (*DSM-IV-TR*) [8]. Participants with medical conditions that might cause symptoms mimicking dementia during the study's medical and neurological examinations were excluded. The final diagnosis of dementia was made by a board-certified neurologist, and was determined independent of RADD testing.

Validity

To demonstrate how well the RADD subtests differentiated between levels of ID, a Multivariate Analyses of Variance (MANOVA) with four levels of ID as an independent variable and all 14 subtests as dependent variables revealed a significant

between groups effect (Wilks $\Lambda = 0.135$, F (42, 525.832) = 12.075, $p < 0.001$). Subsequent one way analysis of variance (ANOVA) tests revealed all differences to be significant at the $p < 0.001$ level. The adjusted R^2 for the subtests ranged from 0.411 to 0.668 with a median of 0.566. An inspection of the mean plots for the 14 subtests found, in all cases, large increases in mean scores from profound to mild ID. Scheffe post-hoc analyses compared the four diagnostic levels across all 14 subtests. In a total of 84 post-hoc analyses (6 diagnostic pairs × 14 dependent variables), 71 of 84 pairwise comparisons were significant at $p < 0.001$. Scores were not significantly different between mild and moderate ID on the recall, imitation and self-identification subtests, between moderate and severe ID on the imitation subtestand between severe and profound ID on the recall, similarities and arithmetic subtests. In Fig. 12.2, subtest scores were converted to z-scores in order to depict the relative performance of the four ID groups on all 14 subtests. Inspection of Fig. 12.2 revealed no overlap between the four diagnostic groups on any subtest.

In order to ascertain how the test was influenced by participants' characteristics, a three-way between subjects ANOVA was completed with the RADD total score as the dependent variable and residential setting, gender, and levels of ID as independent variables. A main effect was detected for levels of ID (F (3, 178) = 79.60, $p < 0.0001$) with no significant interactions. There were no significant main effects

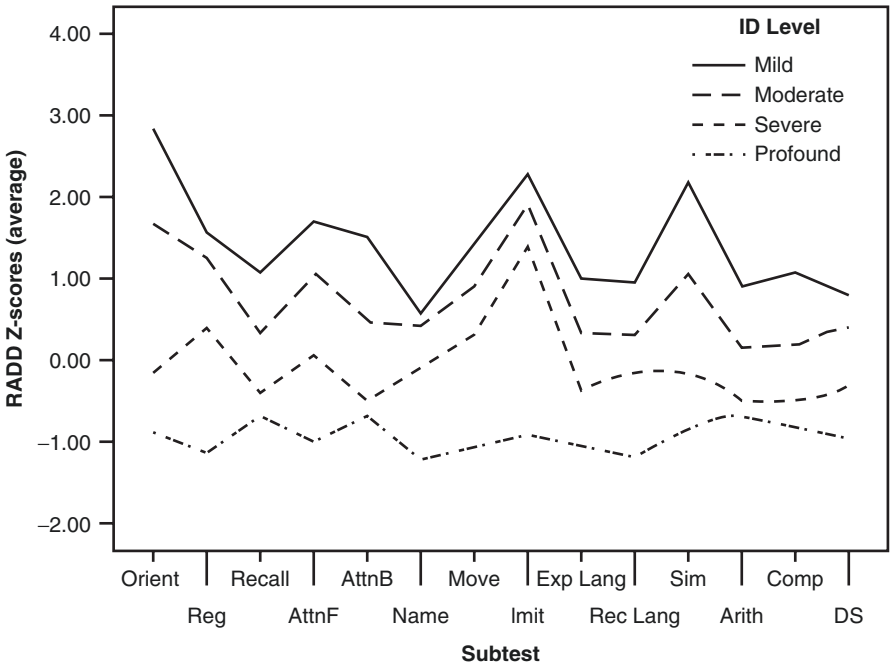

Fig. 12.2 Profile of cognitive abilities by level of intellectual disability (n = 194)

Table 12.2 Correlations between the RADD and other direct and informant-based measures among participants with Down syndrome based on dementia status

Measure	RADD scores total sample (n = 114)	RADD scores non-demented (n = 43)	RADD scores demented (n = 71)
Brief praxis test	0.842[a]	0.789[a]	0.852[a]
Severe impairment battery	0.921[a]	0.862[a]	0.930[a]
Dementia questionnaire for persons with mental retardation—sum of cognitive subscale	−0.889[a]	−0.855[a]	−0.827[a]
Dementia questionnaire for persons with mental retardation—sum of social subscale	−0.683[a]	−0.337[b]	−0.661[a]
Bristol activities of daily living scale	−0.812[a]	−0.675[a]	−0.769[a]

[a]Pearson correlations significant $p < 0.01$
[b]Significant $p < 0.05$

for gender (F (1, 178) = 0.271, $p < 0.603$) or residential setting (F (1, 178) = 2.03, $p < 0.156$).

In order to specifically illustrate the test's validity for individuals with DS, RADD scores were correlated with scores from other direct and informant-based measures. As shown in Table 12.2, the RADD exhibited high correlations with other measures of cognitive functions used in DS (with and without dementia), such as the *Brief Praxis Test* (BPT) [21], *Severe Impairment Battery* (SIB) [22], and *Dementia Questionnaire for Persons with Mental Retardation* [23, 24]—Sum of Cognitive subscale (DMR-SCS) and Sum of Social subscale (DMR-SOS); ranging from 0.84 to 0.92. Furthermore, the patterns of correlations between RADD and other measures remained consistent regardless of the presence or absence of dementia.

Reliability

The RADD has robust internal consistency. Items within each of the 14 subtests are highly correlated, Cronbach alpha coefficients ranged from a low of $r = 0.82$ to a high of $r = 0.97$, with a median alpha of $r = 0.93$. For the RADD Total Score, Cronbach alpha was $r = 0.99$ (Table 12.3). Correlations between the RADD subtest scores and Total Score are presented in Table 12.4. They ranged from $r = 0.72$, to $r = 0.94$, with a median correlation of $r = 0.865$.

The 6 month test re-test reliability was examined among a subset of individuals with DS, and found to be robust (r (41) = 0.95, p < 0.001). The mean scores from the first and second administrations were 19.4 (SD = 18.7) and 17.6 (SD = 20.1), respectively; they were not statistically different.

Table 12.3 Psychometric characteristics of subtests (n = 194)

	Number of items	Low	High	*M*	*SD*	Alpha
Orientation items	5	0	5	1.84	2.01	0.90
Registration items	3	0	3	1.78	1.43	0.97
Recall items	3	0	3	0.79	1.13	0.82
Attention items forward	3	0	3	1.58	1.46	0.97
Attention items backward	3	0	3	0.92	1.33	0.96
Name items	3	0	3	1.28	0.94	0.82
Movement items	3	0	3	1.72	1.31	0.88
Imitation items	4	0	4	2.82	1.72	0.96
Expressive language items	16	0	16	6.67	6.05	0.96
Receptive language items	12	0	12	6.09	4.55	0.94
Similarities items	4	0	4	1.45	1.71	0.92
Arithmetic items	5	0	5	1.13	1.66	0.87
Comprehension items	5	0	5	1.51	1.75	0.86
Digit span items	8	0	8	2.93	2.85	0.92
Total score	76	0	75	32.51	26.14	0.99

Table 12.4 Subtest score correlations with RADD total score and level of intellectual disability (n = 194)

	RADD total score	ID level (rho)
Orientation items	0.89	0.77
Registration items	0.88	0.73
Recall items	0.72	0.63
Attention items forward	0.90	0.76
Attention items backward	0.77	0.68
Name items	0.85	0.74
Movement items	0.84	0.72
Imitation items	0.81	0.71
Expressive language items	0.94	0.81
Receptive language items	0.93	0.80
Similarities items	0.84	0.72
Arithmetic items	0.76	0.64
Comprehension items	0.88	0.76
Digit span items	0.89	0.72
RADD total score	1.00	0.86

Factor Analysis

A principal components factor analysis was performed on the 76 items of the RADD in Study 1, which involved adults with ID. The initial factor selection criterion was to select Eigenvalues greater than one followed by a Varimax rotation. The

Table 12.5 Total variance explained in factor analysis (n = 194)

	Initial eigenvalues		
Component	Total	% of variance	Cumulative %
1	41.55	54.68	54.68
2	5.63	7.41	62.09
3	2.48	3.26	65.35
4	2.10	2.78	68.13
5	1.40	1.84	69.97
6	1.33	1.75	71.72
7	1.24	1.63	73.35
8	1.04	1.44	74.79
9	1.01	1.37	76.16

nine-factor solution accounted for 76.16% of the variance (Table 12.5). Inspection of the data revealed a pronounced general factor (eigenvalue = 41.55, 54.68% of variance). Factor 1 consisted of items that measured registration, brief auditory attention span, receptive vocabulary, imitation, and motor coordination. Factor 2 (eigenvalue = 5.63, 7.41% of variance) was verbal reasoning and concept formation. Factor 3 (eigenvalue = 2.48, 3.26% of variance) was numerical reasoning ability. Factor 4 (eigenvalue = 2.10, 2.78% of variance) was expressive vocabulary. Factor 5 (eigenvalue = 1.40, 1.84% of variance) was working memory. Factor 6 (eigenvalue = 1.33, 1.75% of variance) was short-term memory. Factor 7 (eigenvalue = 1.24, 1.63% of variance) was longer auditory attention span (the more difficult Digit Span items that required the participant to repeat four or more digits). The remaining factors accounted for an additional 2.83% of variance. Interestingly, items that measured orientation (Orientation items from the MMSE and participants' understanding of social norms (Comprehension items from the WISC-III) did not load on any single factor.

Sensitivity and Specificity

In Study 1, the RADD's ability to differentiate between severity levels of ID was ascertained by computing receiver operating characteristic (ROC) curves. In a ROC curve, the true positive rate (i.e., sensitivity) is plotted as a function of the false positive rate (100 minus specificity) for different cut-off points. Each point on the ROC curve represents a sensitivity/specificity pair corresponding to a particular decision threshold. A test with perfect discrimination (no overlap in the two distributions) has a ROC curve that passes through the upper left corner (i.e., 100% sensitivity, 100% specificity). Therefore, the closer the ROC curve is to the upper left corner, the higher the overall accuracy of the test [25]. In Fig. 12.3, the false-positive and false-negative performance of the RADD are depicted as the area under the curve (AUC). The RADD effectively differentiated participants with severe and profound ID (AUC = 0.922; $p < 0.001$).

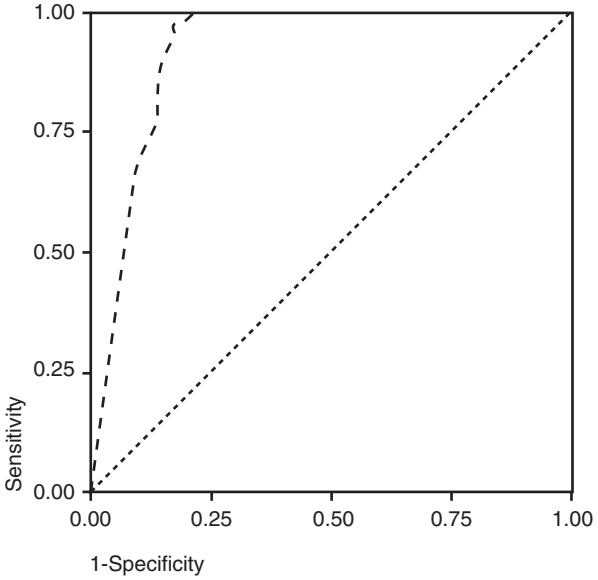

Fig. 12.3 Receiver operating characteristic curve depicting the differentiation between participants with profound or severe intellectual disability (n = 98). A RADD total score of 5 has 82.7% sensitivity for participants with severe to profound ID. Approximately 98% of participants with profound ID were correctly classified; 17% of participants with severe ID were incorrectly classified as profound ID with this cut-off

Sensitivity reached 82.7% when the horizontal cut-off line was set at a score of 5 points correct on the RADD. Approximately 98% of participants with profound ID were correctly classified and about 17% of participants with severe ID were incorrectly classified as profound with this cut-off. Furthermore, the RADD effectively differentiated participants with mild and moderate ID (AUC = 0.825; $p < 0.001$). Sensitivity was 73.5% when the cut-off was set a score of 57. Approximately 81% of participants with moderate ID were correctly classified and 29% of participants with mild ID were incorrectly classified as moderate using this cut-off (Fig. 12.4).

The RADD also differentiated severe and moderate ID (AUC = 0.78; $p < 0.001$). Sensitively of the RADD was 55.3% with a cut-off score of 44. Approximately 92% of participants with severe ID were correctly classified and 45% of participants with moderate ID were incorrectly classified as severe using this cut-off (Fig. 12.5). The AUC results for all comparisons are summarized in Table 12.6.

Use of the RADD in Neuropsychological Assessment

The RADD total score was strongly correlated with the level of ID level (*rho* = 0.86) indicating the test was a valid measure of participants' cognitive abilities. Factor analyses indicated the battery measured a diverse set of cognitive abilities including

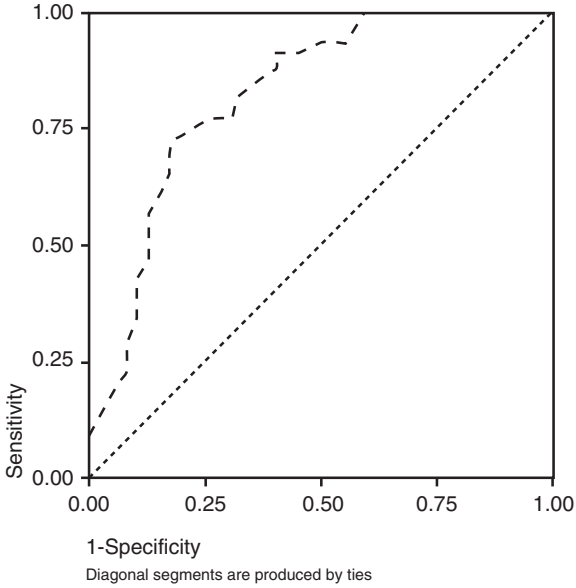

Fig. 12.4 Receiver operating characteristic curve depicting the differentiation between participants with moderate or mild intellectual disability (n = 96). A RADD total score of 57 has 73.5% sensitivity for participants with mild to moderate ID. Approximately 81% of participants with moderate ID were correctly classified; 29% of participants with mild ID were incorrectly classified as moderate ID using this cut-off

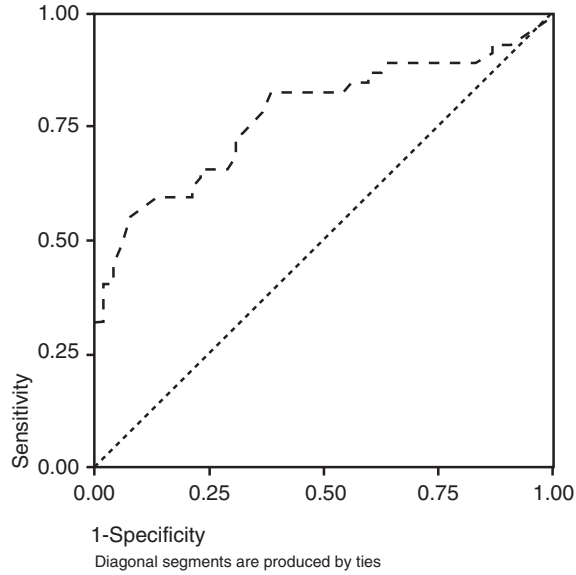

Fig. 12.5 Receiver operating characteristic curve depicting the differentiation between participants with severe or moderate intellectual disability (n = 99). A RADD total score of 44 has 55.3% sensitivity for participants with moderate to severe ID

Table 12.6 Area under the curve comparisons across intellectual disability levels (n = 194)

Differentiations	AUC	p-value
Moderate versus mild ID	0.825	0.001
Severe versus moderate ID	0.780	0.001
Profound versus severe ID	0.922	0.001

ID Intellectual disability, *AUC* Area under the curve

general intelligence, verbal reasoning, numerical reasoning, expressive language, working memory, short-term memory, and auditory attention span. Internal reliability estimates from the RADD total score and from individual subtests were highly significant. The standardization sample included participants with all levels of ID, from both community and residential settings, with psychiatric and medical conditions, as well as speech, motor, and sensory impairments typically experienced by individuals with ID. Our results indicate that the battery is appropriate for most individuals with ID, with the noteworthy exception of those with significant hearing impairments.

Use of the RADD in Dementia Assessment

Study 2 compared RADD assessments to informant-based and direct measures of cognition and dementia, commonly utilized in research with individuals with DS. The informant-based measures consisted of the DMR (DLD) and the *Bristol Activities of Daily Living Scale* (BADLS) [26]. The direct cognitive measures used for comparisons were the SIB and the BPT.

The mean RADD Total Score was 30.3 (SD = 21.6), with individual performances ranging from 0 to 73 (of a maximum possible score of 76). MANOVA were utilized to evaluate the possible effect of gender on the individual scores from each of the direct and informant-based measures. There were no significant gender differences [$F^{(6, 100)} = 1.05$, $p = 0.40$]. MANOVA using dementia status as the independent variable and the direct and informant-based measures as dependent variables was significant [$F^{(6, 100)} = 10.89$, $p < 0.001$]; individuals with dementia exhibited more severe impairment on all measures. In order to set the RADD apart from other tests, a two-way ANOVA was completed with level of ID and dementia status as independent variables and RADD Total Score as the dependent variable. The model was significant [$F^{(5, 109)} = 45.45$, $p < 0.001$] and accounted for 67.8% of the overall variance. There were significant main effects for both ID level ($p < 0.001$) and dementia status ($p < 0.001$), with a non-significant interaction effect ($p < 0.10$). Post-hoc analyses for ID level found progressive gains from severe-profound to mild ID (Fig. 12.6).

In order to demonstrate the RADD's ability to differentiate between participants with DS based upon their dementia status, three ROC curves were calculated based on ID level. These curves plot sensitivity, which is the proportion of true dementia cases correctly identified, against 1-specificity, which is the proportion of false positives. ROC curves for individuals with mild, moderate and more severe levels of ID are plotted in Figs. 12.7, 12.8, and 12.9, with the accuracy of the RADD quantified as the AUC. As shown in Fig. 12.7, results indicted the RADD effectively differentiated mild ID participants based on their dementia status (AUC = 0.944; $p < 0.001$). Sensitivity was 0.95%, specificity was 0.79, and 87.5% were correctly classified with the RADD cut-off score of under 60 indicating presence of dementia. Figure 12.8 provides comparable results for participants with

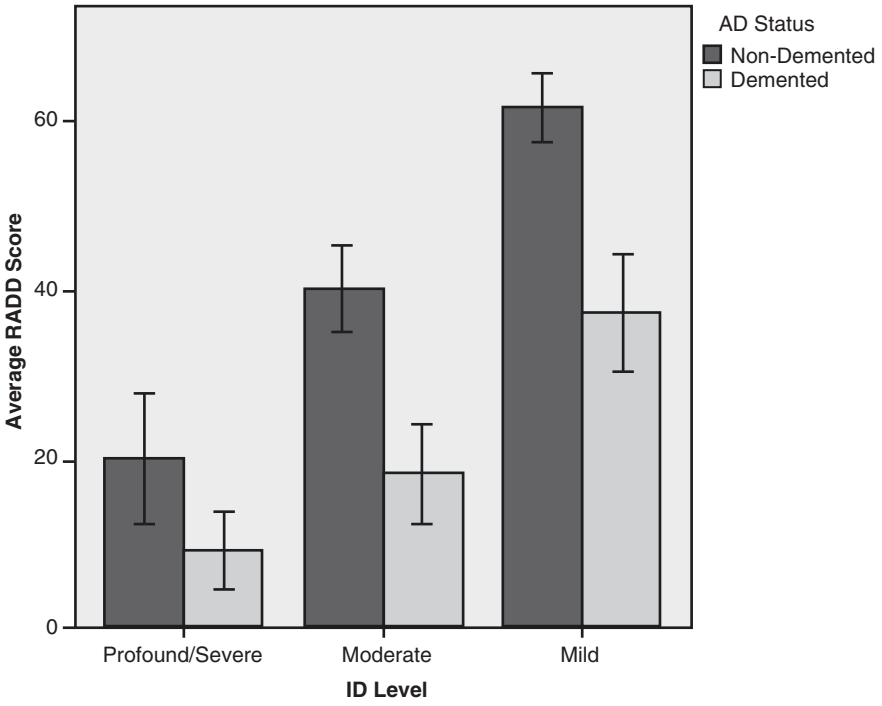

Fig. 12.6 RADD scores across intellectual disability levels among participants with Down syndrome based on dementia status (n = 114)

moderate ID (AUC = 0.87; $p < 0.001$). Sensitivity was 0.79%, specificity was 0.87, and 81.8% were correctly classified with the RADD cut-off score of under 30 indicating presence of dementia. As shown in Fig. 12.9, the RADD differentiated dementia status among participants with severe ID (AUC = 0.83; $p < 0.009$). Sensitivity was 0.89, specificity was 0.75, and 84.6% of participants with severe ID were correctly classified with the RADD cut-off score of under 20 indicating presence of dementia. Participants with profound ID (n = 4) were excluded from the ROC analyses due to the limited sample size and performance at floor independent of dementia status. The AUC results for all comparisons are summarized in Table 12.7.

To ascertain if these RADD criteria for classifying dementia status were sensitive to relatively early stages of dementia, sensitivity was recalculated for only cases diagnosed within the 2 years immediately preceding the RADD assessment (n = 30). Cases included 11 adults with mild ID, 10 with moderate ID, and 9 with severe ID. Overall sensitivity was 0.73, with the criteria for the mild ID group remaining quite high at 0.91 but with estimated sensitivity considerably lower for the moderate ID subgroup (0.50). However, these ID-level differences could reflect imprecision in estimates associated with small sample sizes rather than true effects.

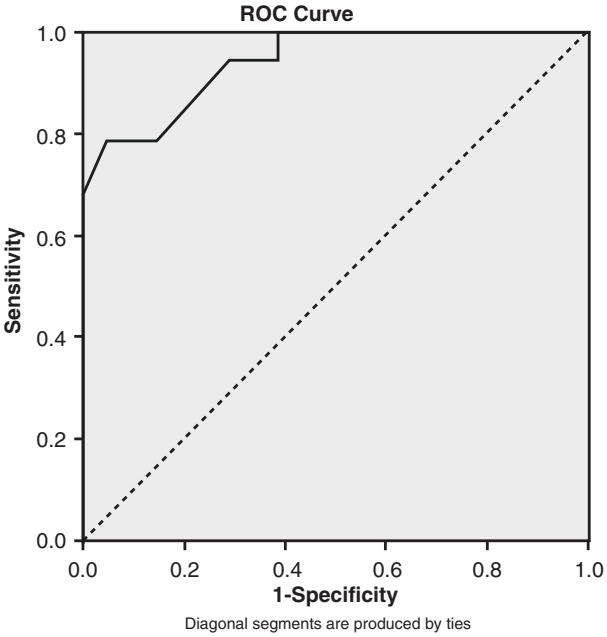

Fig. 12.7 Receiver operating characteristic curve depicting differentiation of dementia status among participants with down syndrome and mild intellectual disability (n = 40). A RADD total score of <60 has 95% sensitivity and 79% specificity; 87.5% of participants with mild ID were correctly classified as having dementia with this cut-off

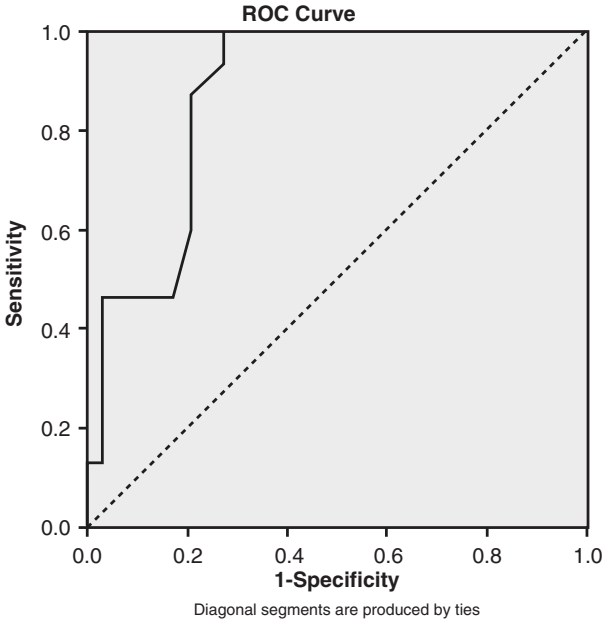

Fig. 12.8 Receiver operating characteristic curve depicting differentiation of dementia status among participants with Down syndrome and moderate intellectual disability (n = 44). A RADD total score of <30 has 79% sensitivity and 87% specificity; 81.8% of participants with moderate ID were correctly classified as having dementia with this cut-off

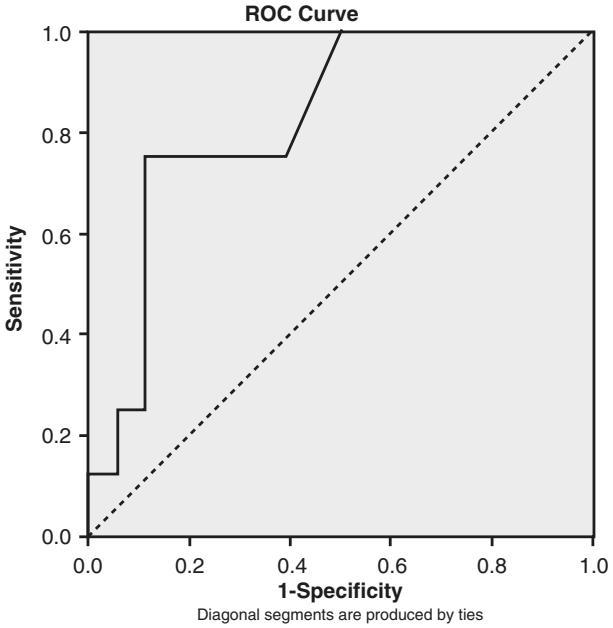

Fig. 12.9 Receiver operating characteristic curve depicting differentiation of dementia status among participants with Down syndrome and severe intellectual disability (n = 26). A RADD total score of <20 has 89% sensitivity and 75% specificity; 84.6% of participants with severe ID were correctly classified as having dementia using this cut-off score

Table 12.7 Area under the curve comparisons across intellectual disability levels and dementia status (n = 114)

Differentiations	AUC	p-value
Mild ID, dementia versus non-dementia	0.944	0.001
Moderate ID, dementia versus non-dementia	0.870	0.001
Severe ID, dementia versus non-dementia	0.830	0.009

ID Intellectual disability, *AUC* Area under the curve

Interpretation of Results for Down Syndrome and Dementia

Standard dementia diagnostic assessment methods designed for the general population are typically not informative for individuals with DS because their premorbid scores are often too low for clear interpretation of subsequent decline [27]. Also, traditional scales and tests do not have normed data for this population, which prevents meaningful interpretation of such assessment results [28]. Consequently, some adults with DS without a decline in cognitive function may be incorrectly diagnosed as having dementia when they do not have an underlying disease (i.e., misdiagnosis). In others, there may be an under diagnosis of dementia because cognitive decline is confused with "normal aging" in an individual with ID [29, 30].

There seems to be general agreement that early detection of dementia is a priority (see Krinsky-McHale and Silverman [27] for a review). Dementia assessment in individuals with DS needs to include direct assessment of neuropsychological function as well as informant-based measures [30–33].

Direct assessment is usually regarded as preferable over informant report due to errors related to observation and/or recollection, and biases in reporting [34]. Older adults with DS may not have parents who are alive or can report on their everyday function and abilities because their parents are no longer the primary caregiver. Family members may also find it emotionally difficult to report declines in functioning or incorrectly believe that certain declines are not relevant to the person's care, and therefore, do not take note or report them. Paid care staff may have a high rate of turnover, making reporting of changes in multiple domains difficult when several informant-based dementia and psychopathology scales require that the caretaker know the patient for at least 6 months. In addition, there are a growing number of individuals with DS who live independently for whom there may be no available informant with adequate knowledge of their current functional abilities.

However, most direct cognitive measures that have been used with individuals with DS only assess one specific domain. For example, a list learning task only assesses memory, and a naming test only assesses expressive language. These tests may also lack normative data for individuals with ID, precluding the possibility of their use as single-administration tests; as opposed to their use as a repeated measure of change over time. Therefore, most dementia assessments for persons with ID rely upon informant-based measures and/or an extensive battery of tests to assess function across a broad range of areas [34].

The RADD exhibits criterion-referenced validity by way of strong correlations between RADD scores and premorbid IQ levels determined during prior standardized IQ testing, as well as by differentiation between participants with and without dementia [13]. This is a significant advantage over other direct cognitive tests that have been used with the ID population as many of them had problems with floor effects when participants were classified as having premorbid severe ID [35–37]. The RADD is also positively correlated with informant-based measures that have been demonstrated to be useful in determining dementia diagnosis among individuals with ID [13, 27], namely, the DMR (DLD) and the Adaptive Behavior Scale [38]. The RADD also provides critical information beyond what can be obtained with most standardized IQ tests as it also measures an individual's receptive and expressive language, orientation, registration, motor coordination, and imitation skills.

In summary, using the RADD in dementia assessments for individuals with DS has several advantages over traditional neuropsychological tests and informant report measures. The RADD is: (1) a direct measure, standardized with the ID population and specifically with individuals with DS, that is not subject to informant report errors or biases, (2) its total score strongly correlates with level of ID, (3) it can be sequentially administered to identify deterioration in subtest and total scores from a previous baseline, and (4) it can differentiate between individuals with DS with and without dementia.

Future Application of the RADD for Cognition and Dementia in Down Syndrome

The future application of the RADD will depend on the utility and information which the test can provide to practitioners, particularly those who have neither the training nor the resources to carry out full scale neuropsychological testing. Practical requirements for rapid cognitive assessments include the ease of administration by a technician, the lack of a requirement for clinical judgement by the examiner and the ability of the test to distinguish levels of disability [39]. Neuropsychological measures in DS should be able to measure a wide range of skills. The RADD has been shown to profile a wide range of functional abilities including receptive and expressive language, orientation, registration, recall, attention, self-identification, motor skills, imitation, abstract reasoning, number skills, comprehension and short term memory. Edgin and colleagues [40] have indicated that cognitive tests in DS should afford adequate test-retest reliability and should be resistant to confounding factors such as poor motivation and language impairment. The RADD meets these test-retest criteria and also demonstrates construct and criterion-referenced validity via convergent correlations with other established measures of cognitive functioning for individuals with DS.

There are many causes of cognitive decline irrespective of dementia in DS. For example, poor cognitive performance may be seen as an outcome of obstructive sleep apnea in DS [41]. Individuals with DS who have congenital heart defects show lower cognitive scores [42]. There are other known comorbidities in adults with DS that can affect quality of life and cognition including hypothyroidism, vision and hearing impairments, and depression [43, 44]. As opposed to dementia, many of these entities are readily treatable and even curable. The RADD could be used to follow pre-and post-treatment effects of many of these medical comorbidities in DS.

Repeated cognitive assessments that can detect changes in core domains are required for clinical trials in dementia [45]. In the general population, the US Food and Drug Administration has indicated that reliable cognitive measures may be used to test AD drugs before functional impairment becomes evident [46]. However, challenges may exist when repeated measures are derived using cognitive tests not designed for the specific population. This is particularly true for the early diagnosis of dementia in DS. Neuropsychological testing in DS cannot rely on tests that track cognitive decline in the typical population [30]. The RADD has shown predicted effects across intellectual levels and appears suitable to be employed as longitudinal cognitive measures for clinical trials in DS.

There are several requirements published by *The International Working Group on Harmonization of Dementia Drug* Guidelines [47] that have applicability to therapeutic trials for dementia in DS. These include validity (the instrument must measure the disease-relevant cognitive functions), test-retest reliability, appropriate sensitivity range and the availability of longitudinal data. On these accounts, the RADD meets the required criteria.

A paradigm shift towards Personalized Medicine will use a variety of biomarkers to predict when to begin therapeutic intervention. Increasingly the application of Personalized Medicine will be extended towards people with DS [48]. It is likely that the RADD will be employed as an independent cognitive measure taking its place in a battery to measure therapeutic response across the entire range of ID.

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Chapter 13
The Severe Impairment Battery

Nick Hutchinson

Introduction

The term dementia is an umbrella term used to describe a clinical syndrome that consists, primarily, of significant, progressive and irreversible deterioration of cognitive functioning (learning and memory, language, perception, executive functioning, attention) from a higher level of premorbid functioning, which is of significant severity to interfere with independent living skills across a range of domains (instrumental, domestic, self-care, social). Decline in cognitive functioning can also be accompanied by behavioural and personality changes [1, 2]. The National Institute for Health and Care Excellence [3] defines dementia as "a progressive and largely irreversible clinical syndrome that is characterised by a widespread impairment of mental function … as [dementia] progresses [people] can experience some or all of the following: memory loss, language impairment, disorientation, changes in personality, difficulties with activities of daily living, self-neglect, psychiatric symptoms and out-of-character behaviour" (p. 5). There are many different types of dementia caused by a number of diseases of the brain (for example Alzheimer's disease, Frontotemporal degeneration, lewy body disease, vascular disease), the most common cause being Alzheimer's disease [3]. In the most recently published diagnostic manuals [2] the 'dementias' are subsumed within the category of Major and Mild neurocognitive disorders.

People with Intellectual Disabilities (ID) are at a higher risk of developing dementia compared to people in the general population [4]. Dementia, more specifically Alzheimer's disease type, is particularly prevalent in people with Down syndrome (DS) who are at significant risk of developing early onset dementia. The British Psychological Society [5] has summarised the main studies into the

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prevalence of dementia in people with DS and report prevalence rates of a few percent for people aged 30–39 years, increasing to between 30 and 75% in adults aged 60 years or older. Studies consistently show that the average age of onset of dementia in people with DS is between 50 and 56 years [5–7]. The pattern and progress of Alzheimer's disease in people with and without ID/DS is similar [8, 9], although research now supports clinical observations that changes in personality and behaviour associated with executive dysfunction can precede deterioration in memory in people with DS [10, 11].

The assessment and diagnosis of dementia in people with a pre-existing ID is fraught with many challenges [12, 13] and, as such, the recommended approach to assessment is one of baseline assessment (reactive or premorbid) and prospective monitoring which involves repeated assessment at different time points in order to identify change in functioning from baseline and increase the likelihood of early diagnosis [5]. Where possible, direct neuropsychological assessment of cognitive functioning should be included as part of an assessment battery alongside information gathered through questionnaire/interview based informant measures of dementia onset [5].

There are a range of direct neuropsychological tests that have been developed or adapted for use in dementia assessment with people with ID, the most widely used of which are covered in this book. This chapter will review the Severe Impairment Battery (SIB) [14, 15].

Background to the Severe Impairment Battery (SIB)

The SIB [14, 15] is a measure designed for the assessment of cognitive functioning of people with dementia in the general population. At the time of its development in the late 1980s/early 1990s there were very few neuropsychological tests suitable for assessing people with dementia and moderate to severe cognitive impairment. Saxton and colleagues [14] developed the SIB to enable the assessment of cognitive deficits seen in people with severe impairment due to dementia—a patient group who at the time were viewed by most clinicians as 'untestable.' In their seminal paper published in 1990 [14], Saxton and colleagues report that the SIB was developed "to assess a range of cognitive functioning in patients who are unable to complete existing standard neuropsychological assessment scales. It was designed for the severely demented patient and takes into account the specific behavioural and cognitive deficits associated with severe dementia" (p. 299).

The SIB consists of 39 test items, some of which are two or three part questions, measuring nine domains of functioning. There are seven domains relating to areas of cognitive functioning—memory, orientation, language, attention, praxis, visuospatial ability and construction—and two brief measures of social interaction and orienting to name. The Severe Impairment Battery also gives an overall total score out of 100. Table 13.1 provides a summary of SIB subtests and scoring.

Table 13.1 SIB domains and test items

SIB domain	Test content/administration	Scoring
Social interaction	The patient's interaction when they enter the testing room or when the test begins is assessed through the patients ability to shake hands and follow initial directions to be seated and get comfortable ready to begin the testing.	Brief subtest consisting of 1 three-part item scored 2 (correct), 1 (prompt), 0 (incorrect).
		Maximum score 6.
Memory	This includes items relating to immediate retrieval of verbal (examiners name, recall of short sentence) and visual information (recognition of two presented objects (cup and spoon), recognition of a presented shape and coloured block and also longer term recognition of the two earlier presented objects and delayed recall of the examiners name.	7 items scored 2 (correct), 1 (prompt), 0 (incorrect).
		Maximum score 14.
Orientation	Items relate to person, time and place. The patient is asked to state their name, the present month, and name the City in which the testing is taking place.	Brief subtest consisting of 3 items scored 2 (correct), 1 (prompt), 0 (incorrect).
		Maximum score 6.
Language	This subtest includes items relating to language expression and comprehension: Ability to write name; state the months of the year; verbal responsive naming (for example: Item 8—"what do you call the thing you drink coffee from/eat soup with"); object naming from pictures and presented objects (cup and spoon); simple verbal repetition (e.g. item 11a: "Now say this: People spend money". Item 11 b: "Now say this: Baby"); Ability to read and comprehend a brief line of text ("Give me your hand", item 9) written on a card presented to the patient; verbal fluency (item 13: "Tell me all the things you like to eat"); Colour naming when presented with three different coloured blocks; shape identification when presented with three different shaped blocks and free discourse (at the very end of the test the examiner engages the examinee in conversation and asks "How have you been").	This is the longest subtest and contains 16 items, a number of which are in two-part or three-part question format.
		Items scored 2 (correct), 1 (prompt), 0 (incorrect). 2 items scored 1 (correct) or 0 (incorrect)
		Maximum score 46.
Attention	This includes a measure of digit span (verbal repetition of one digit to five digit series), auditory span (Examinee counts along to the examiner tapping on a table) and visual span (the examinee is asked to count the number of fingers being held up by the examiner)	3 items scored 2 (correct), 1 (prompt), 0 (incorrect).
		Maximum score 6.

(continued)

Table 13.1 (continued)

SIB domain	Test content/administration	Scoring
Praxis	The examinee is asked to demonstrate through gestures and actions how to use a cup and a spoon when presented with these two objects (pictorial and object form)	4 items scored 2 (correct), 1 (prompt), 0 (incorrect).
		Maximum score 8.
Visuo-spatial ability	The examinee is asked to match and discriminate between different coloured blocks and also between different shapes.	4 items scored 2 (correct), 1 (prompt), 0 (incorrect).
		Maximum score 8.
Construction	The examinee is asked to draw a circle and a square.	Brief subtest consisting of 1 two-part item scored 2 (correct), 1 (prompt), 0 (incorrect).
		Maximum score 4.
Orienting to name	At the end of the test, while preparing to leave, or when walking back to the waiting area, the examiner stands directly behind the examinee and calls his/her name.	Brief subtest consisting of 1 item scored 2 (correct), 1 (prompt), 0 (incorrect).
		Maximum score 2.
Total score		Sum score of the 39 items.
		Total score 100.

The SIB is very brief with an administration time of between 20 and 30 min for people with ID. As the SIB was developed for the assessment of cognitive functioning in people with severe dementia related impairment, the test items involve 'low level' tasks to ensure that these patients are able to perform successfully across the range of domains. The test has simple, one-step command instructions accompanied by gestural cues and prompts if necessary.

There have been three versions of the full SIB. Early data on the psychometric properties of the first version of the SIB for use with people with severe dementia in the general population were published in 1990 by Saxton and colleagues [14]. In a preliminary study sample of 41 participants with dementia (40 had a diagnosis of Alzheimer's disease, one participant had a diagnosis of multi-infarct (vascular) dementia) they report that the SIB has very high interrater reliability, high test-retest reliability (mean test-retest interval of 14 days) with a correlation coefficient of 0.85 ($p < 0.001$) and adequate construct validity when SIB performance was compared to scores on the Mini-Mental State Examination questionnaire (MMSE) [16]. Saxton and colleagues [14] reported that even those patients classified as being very severely impaired on the basis on MMSE performance were able to

score on the majority of the SIB subscales and conclude that the SIB was a useful clinical assessment tool for measuring a range of cognitive functions, and monitor cognitive decline longitudinally, in people with severe dementia who would perform at the floor level on other tests. Saxton and colleagues [15] present psychometric evaluation data from the second version of the SIB in the SIB manual. These data show the SIB to have very high inter-rater reliability (Spearman's rho correlation coefficients for all subtests above 0–89; SIB total score r = 0.99), test-retest reliability (r = 0.90), and construct validity when performance on the SIB was compared with the Mini-Mental State Examination (MMSE) [16] (correlation between SIB and MMSE score 0.76) and the Mattis Dementia Rating Scale [17] (correlation between SIB and DRS score 0.88).

The SIB is widely used by clinicians within the UK and internationally. It has been translated into different languages and evaluated for use in dementia assessment in a number of countries across the world including Italy [18, 19], Spain [20], France [21], Greece [22], Germany [23], Norway [24], Korea [25], Brazil [26], and China [27]. There are also short-forms of the SIB, which again have been translated for use across the world [18, 26–29].

Following studies specifically investigating the reliability and validity of the SIB for use within clinical trials [30, 31], the SIB has been extensively used in randomised control trial intervention studies investigating the effectiveness and efficacy of a number of 'anti-dementia' medications such as Donepezil [32, 33] Rivastigmine [34, 35], Memantine [36, 37] and Galantamine [38] for the treatment of Alzheimer's disease. It has also been used as an outcome measure in a few studies investigating psychosocial interventions, such as quality of life within nursing care homes [39] and music interventions [40] for people with dementia in the general population.

Evaluation of the SIB for Use with People with Down Syndrome

As already mentioned, the SIB was developed for use as part of the neuropsychological assessment of adults with dementia and severe levels of cognitive impairment in the general population. The measure was not developed specifically for use with people with ID. The test is administered with people with ID using the standard format and test instructions, and the SIB has not been adapted or revised in any way for use with people with ID, as some neuropsychological tests developed for use in the general population have been (for example the Cambridge Cognitive Examination [41] (see Chap. 7). Because of the low level nature of the test items, and the fact that

the test is structured in a way so as to ensure its administration incorporates simple one-step command instructions, gestural cues, and simple prompts, it has been proposed that the SIB is a suitable test to enable people with ID, with pre-existing additional communication difficulties, sensory and physical impairments—in other words people who would struggle to complete standard neuropsychological assessments which would produce 'floor effects'—to engage in direct neuropsychological assessment of dementia [42].

From clinical experience, and review of the existing literature, it would appear that clinicians started to use the SIB with people with ID in the mid-late 1990s. Witts and Elders [42] carried out the first evaluation of the utility and psychometric properties of the SIB when used with people with DS. Using a within-subject repeated measures design with 33 adults with DS, Witts and Elders [42] showed that the SIB has good test-retest reliability (Spearman's rho = 0.89). They used the Vineland Adaptive Behaviour Scales (VABS) [43], which has well-established psychometric properties and is widely used by clinicians and researchers, concurrently with the SIB, and demonstrated that the SIB has good criterion validity (Spearman's rho = 0.68) when used with people with ID. Witts and Elders [42] conclude from their findings that the SIB "can successfully be used with adults with DS to assess cognitive functioning over a wide range of ability and may be useful, if used longitudinally, in assessing for deterioration in cognitive functioning associated with dementia" (p. 213).

McKenzie and colleagues [44] undertook the next study investigating the usefulness and validity of the SIB specifically as a measure of cognitive decline in people with DS. They carried out a group comparison study whereby SIB scores for two groups were compared—a 'deteriorating' group (n = 10) who had shown a decline in their adaptive behaviour, according to their VABS scores, over at least a 2 year period, and a 'non-deteriorating' group (n = 14) whose adaptive behaviour had remained stable over this time period. The results of this study showed that the 'deteriorating' group showed a significant decline in SIB scores from baseline to 12 and 24 months; the 'non-deteriorating group' SIB scores remained stable. Interestingly, McKenzie and colleagues [44] suggest that the SIB orientation domain, in particular the item relating to the patient's ability to name the city they live in, may have discriminant validity as an early indicator of dementia related cognitive decline in adults with DS.

In sum, the studies by Witts and Elders [42] and McKenzie and colleagues [44] suggest the SIB has utility, validity and reliability as a neuropsychological measure for use in dementia assessment with people with DS. However, it is important to note (as the study authors do) the limitations of these studies. Firstly, although the VABS [43] is a well-established measure, it is a measure of *adaptive* functioning, rather than *cognitive* functioning and, as such, the appropriateness of the use of the VABS as a measure of deterioration [44] and as a concurrent measure for

establishing criterion validity of the SIB [42] can be questioned. There are also methodological issues relating to small sample sizes, potential bias towards more able participants coming forward or being put forward by caregivers to participate in the study and there are a number of issues that need to be taken into account when interpreting the results of group comparison studies [45] as undertaken by McKenzie and colleagues [44].

The third study evaluating the usefulness and validity of the SIB as a measure of cognitive functioning with people with DS was carried out by Hutchinson and Oakes [46]. Following up on the suggestions for further research made by previous researchers [42], Hutchinson and Oakes [46] used an informant questionnaire that specifically assesses areas of cognitive functioning—the Dementia Questionnaire for Mentally Retarded Person (DMR—as was; this questionnaire has since been revised and is now called the Dementia screening questionnaire for people with Learning Disabilities, DLD; see Chap. 3) [47–49]—as the concurrent measure to establish the criterion validity of the SIB. Using a cross-sectional correlation design with 37 adults with DS, Hutchinson and Oakes showed the SIB to have good criterion validity, with statistically significant correlations (Spearman's rho correlations ranged from −0.54 to −0.67) found between the DMR (DLD) Sum of Cognitive Scores (SCS—represents the sum of the short-term memory, long term memory and spatial and temporal orientation subscales) and five of the major SIB domains (memory, orientation, language, attention, visuo-spatial ability). The total SIB score also showed a statistically significant correlation with the DMR (DLD) SCS (Spearman's rho = −0.73).

One particular strength of the study carried out by Hutchinson and Oakes [46] is the use of a cognitive measure concurrently with the SIB as a measure of its criterion validity. However, methodological limitations such as small sample size, issues relating to the reliability of informant measures and the concurrent use of informant measures to evaluate the psychometric properties of a direct assessment measure [45, 50], remain an issue.

To summarise, the literature evaluating the reliability and validity of the SIB for use in the direct neuropsychological assessment of dementia in people with DS is sparse and researchers have acknowledged that further evaluation research is required. To date the sensitivity and specificity of the SIB has not been investigated. However, from the three studies summarised here (see also Table 13.2 for more details), it is fair to conclude, based on the current evidence, that although the SIB was not developed for use with people with ID, the available research suggest it has good test-retest and criterion validity when used as a measure of cognitive functioning in people with DS. Based on the literature—to be covered in the next section—and clinical experience it can be said that the SIB does have utility and feasibility for use in clinical settings and in research.

Table 13.2 Studies evaluating the SIB

Author(s) Date Country	Aim(s)	Design	Participants	Evaluation	Measures	Key findings
Witts and Elders [42] United Kingdom	1. To examine the utility of the SIB with people with DS. 2. Examine the test-retest reliability for the SIB with people with DS.	Within-subject repeated measures design	33 adults (18 male, 15 female) with Down syndrome (non-dementia) completed the SIB Mean age 36 years (range 22–53 years) Care staff members completed the Vineland Adaptive Behaviour Scales (VABS) [43]	Psychometric properties Test-retest reliability (T1 and T2 separated by 30 days) Criterion validity	SIB Vineland Adaptive Behaviour Scales (VABS) [43] were used to establish criterion validity.	High SIB test-retest reliability (Spearman's rho = 0.89) Significant correlation between VABS age equivalent and SIB total score (Spearman's rho = 0.68) indicating high criterion validity. No floor effects: Mean SIB total score 80.94/100 (range 34–100)

McKenzie, Harte, Sinclair, Matheson, Patrick and Murray [44] United Kingdom	1. To examine if significant differences exist in SIB scores at baseline and follow-up, for participants with DS showing decline in their adaptive skills. 2. To compare these results to those of a group of participants with DS who do not show decline in adaptive skills.	Repeated measures and cross sectional design.	24 adults (7 male, 17 female) with DS completed measures every 12 months for at least 2 years. Split into 2 groups: (a) 'deteriorating group' (n = 10, mean age 51.9 years) who showed decline in adaptive behaviour over at least a 2 year period and met criteria for Alzheimer's disease. (b) 'non-deteriorating' group (n = 14, mean age 44.2 years) showed no decline in adaptive behaviour over this period of time	Group comparison—Deteriorating versus non-deteriorating group Psychometric properties—Discriminant validity	SIB Vineland Adaptive Behaviour Scales (VABS) [43]	Deterioration group significantly older than the non-deterioration group (unrelated t-test, t- 2.838, d.f. = 19.97, $p < 0.01$) Significant decline in SIB orientation subtest scores between baseline and 12 months (Wilcoxon signed rank test $Z = -2.428$, $p < 0.01$) and baseline and 24 months (Wilcoxon signed rank test $Z = -2.414$, $p < 0.01$) in the deterioration group. In particular, there was a significant decline in one of the SIB orientation subtest items—Ability of participant to name the city they lived in—Between baseline and 24 months (Wilcoxon signed rank test $Z = -1.667$, $p < 0.05$) SIB orientation subtest might have discriminant validity as an early indicator of cognitive decline in adults with DS.

(continued)

Table 13.2 (continued)

Author(s) Date Country	Aim(s)	Design	Participants	Evaluation	Measures	Key findings
Hutchinson and Oakes [46] United Kingdom	1. To examine the concurrent criterion validity of the SIB	Cross-sectional correlation design	37 adults (21 male, 16 female) with Down syndrome (non-dementia) completed the SIB. Mean age 38.97 years (range 20–58 years) Caregivers completed the Dementia Questionnaire for Mentally Retarded Persons (DMR) (now republished as the Dementia Questionnaire for people with Learning Disabilities—DLD) [48–50].	Psychometric properties -concurrent criterion validity Descriptive analysis of SIB scores	SIB Dementia Questionnaire for Mentally Retarded Persons (DMR)/ Dementia Questionnaire for people with Learning Disabilities (DLD) completed to establish criterion validity.	Significant correlations between 5/6 SIB subtests measuring cognitive functioning and the DMR (DLD) Sum of Cognitive Scores (SCS) (Spearman's rho ranged from −0.54 to −0.67 $p = 0.001$) indicating high concurrent criterion validity Significant correlation between DMR (DLD) SCS and SIB total score (Spearman's rho = −0.73) indicating high concurrent criterion validity. No significant correlations between SIB scores and DMR (DLD) Sum of Social Scores (SOS) No floor effects: Mean SIB total score of 78.97/100 (range 41–100)

Studies Using the SIB with People with DS and ID

There are now a number of studies that have been carried out looking at the use of the SIB in research and clinical practice with people with ID.

Research has shown that the neuropathological changes associated with Alzheimer's disease—senile plaques and neurofibrillary tangles—will be evident in the brains of people with DS by the age of around 40 years, and often well before the clinical signs and symptoms of dementia are observed [51]. The SIB has been used as an outcome measure in a number of studies further investigating the neuropathology and neurobiology of people with DS with and without Alzheimer's disease. For example, the SIB has been used as a cognitive baseline measure or outcome measure in brain imaging research into the levels of beta-amyloid protein in the brains of people with DS (with and without Alzheimer's disease) compared to other people with ID or in the general population (see, for example references 51–54).

The SIB has also been used as a primary outcome measure in other neurological/neuropsychological research, such as studies investigating the association between epilepsy and cognitive decline in people with DS and dementia [55]. Individual subtests of the SIB have been used in research as part of broader assessment batteries. For example, Ball and colleagues [10] used the SIB memory sub-test, to measure short and long-term memory, alongside memory items from other neuropsychological tests, in their study investigating executive dysfunction in adults with DS and Alzheimer's disease.

The SIB has been used as a measure of cognitive functioning in studies relating to best practice in clinical assessment and intervention for dementia in people with ID and DS. Clinical guidance [5] does recommend the SIB as one of the direct neuropsychological tests useful in the assessment of dementia in people with DS and intellectual disabilities. Over the past decade or so, the SIB has been increasingly used or considered for use within clinical service settings as part of dementia assessment care pathways for people with DS (see for example references 56–58). The SIB has been utilised as a measure of cognitive functioning in intervention studies of the efficacy of 'anti-dementia' medications in people with DS and dementia [59, 60]. For example, Prasher and colleagues [59, 60] used the SIB as a measure of secondary efficacy during their 24-week, double blind, placebo controlled trial of donepezil for people with Down syndrome and Alzheimer's disease which showed donepezil to be possibly efficacious and should be considered in treatment for this patient group. Safety and efficacy of donepezil was further demonstrated in the extended 80-week open label study [60]. The SIB was again used in this open trial as a secondary outcome measure [60].

Test Selection: Strengths and Limitations of the SIB When Used with People with ID

Before using any direct neuropsychological test, it is important for the clinician/researcher to be aware of the range of factors that must be taken into consideration when choosing which test to use with an individual with ID. The British Psychological Society (BPS) [61] have written guidance on the neuropsychological assessment of

people with ID and make clear that test selection will depend upon a range of factors including the characteristics of the individual being assessed (including age, apparent level of learning disability, method of communication, language comprehension) and the characteristics of the tests (standardised vs non-standardised, availability of normative data, ceiling and floor effects, single tests vs test batteries).

As is evident from the chapters in this current text, there are a number of direct neuropsychological tests available for use in dementia assessments with people with ID. In order to guide the clinician/researcher in their test selection and to aid clinical decision making on whether or not to use the SIB, this section gives an appraisal of the strengths and limitations of the SIB when used with people with ID. This appraisal is based on the literature that has evaluated the use of the SIB with people with ID [42, 44, 46, 57, 58] and clinical experience of the author using the SIB in clinical practice in a dementia assessment clinic for people with DS. Table 13.3, below, outlines the main strengths and limitations of the SIB for use in neuropsychological assessment of people with ID and DS.

Table 13.3 Strengths and limitations of the SIB when used with people with ID

Strengths	Limitations
Brief: 20–30 min administration time—Limited risk of test fatigue.	Originally designed for the assessment of adults without ID but with severe dementia. Test difficulty might not extend high enough for it to be useful in longitudinal assessment of more able examinees (ceiling effect)
Brief: 20–30 min administration time—Can be completed alongside other measures in a single testing session.	Due to the low-level tasks and simple one-step commands, some test items might appear patronising to more able examinees. For example: Baby
Well-structured with an interview style format that is engaging for the examinee	Scoring—reading item (language subtest)—a fail on 1 part of the question means the examinee automatically lose fails the next parts—resulting in 6 lost points
Simple one-step commands accompanied by gestural cues allows for people with range of levels of intellectual ability and communication skills to score on the test (limited floor effects)	Some items impacted by deficits in language comprehension e.g. immediate and delayed memory objects test.
Simple low-level tasks allows for people with range of levels of intellectual ability and communication skills to score on the test (limited floor effects)	No normative data, instead individual baseline used for repeat assessments
Allows for non-verbal and partially correct responses thus enabling people with a range of levels of intellectual ability and communication skills to score on the test (limited floor effects)	SIB attention subtests incorporated into the CAMDEX-DS (CAMCOG)
Allows for longitudinal assessment of a range of cognitive functions and a total SIB score and appraisal of scores over repeat assessments	
Easy to score—All but two items are scored on a 3-point scale (2 = correct, 1 = partially correct, 0 = incorrect)	

It is important during a neuropsychological assessment that the examiner pays close attention to ensuring that the testing environment enables the examinee to perform to the best of their ability. Consideration needs to be given to engagement and rapport building, the length of the testing session, and tests used taking into account the person's attention span, concentration levels, motivation and fatigue levels [61].

The time it takes to complete a neuropsychological test with a person with an ID will vary depending on the person's level of ID, but the SIB should only take between 20 and 30 min to complete with a person with ID, thus reducing the possible impact of testing fatigue. Another benefit here is that it is possible to complete the SIB alongside other direct neuropsychological assessments (for example the CAMDEX-DS/CAMCOG—see Chap. 7) or informant interviews within the same assessment session, with regular breaks if the assessment sessions are going to go on beyond an hour [61].

The structure and administration of the SIB makes it a particularly useful test to use with people with ID. Saxton and colleagues [15] in the SIB manual, state that the "presentation of the items is intended to be performed in a smooth flowing manner, drawing out a response naturally or automatically" (p. 4). It could be said that this style of eliciting test responses serves to put the examinee at ease and helps to maintain engagement in testing. There is a view that many people with ID may have had difficult past experiences within assessment or testing situations, which may 'shape their approach to new challenges' (64, p. 18), including neuropsychological testing situations, during which people might expect failure or look for reassurance from other people with regard to test performance. It could be argued that the SIB structure and style of administration—its flowing, conversational presentation of test items—is conducive to helping the examinee relax and feel more confident in a testing situation.

Another major strength of the SIB is that people with a range of levels of learning disability and additional needs, such as communication problems and sensory/physical impairments are able to perform and score on the SIB (see for example vignette 3, below) and research shows that the SIB is not subject to floor effects [42, 46].

Finally, the SIB provides a total score and also scores for a number of domains of cognitive functioning that are expected to deteriorate with onset of dementia, and thus the test can be used for baseline and then repeat assessments carried out prospectively to track neuropsychological change over time (see vignette 2, below).

In terms of limitations, the difficulty level of the SIB may not extend high enough for people with more mild level of ID, reducing the sensitivity of the tests to identify longitudinal cognitive decline indicative of dementia. For the most able people with ID, the SIB could be susceptible to ceiling effects [50] and as such it might be difficult to detect the early signs of cognitive deterioration in these individuals (see vignette 1 below). Another challenge for more able people with ID is that the structure, delivery of instructions or content of the SIB could be experienced as patronising or even infantilising due to the simplicity and low-level nature of the test [42]. To illustrate this point, Witts and Elders [42] refer to the word repetition items, which form part of the language subtest. Here the examinee is asked to repeat a simple phrase—'people spend money,' and word 'baby.'

Test limitations are also present for people with more severe ID. Firstly, with regard to scoring, a number of authors have noted that on one of the language subtest items, that involves the examinee having to read aloud a message on a printed card, failure to read the card leads to the person subsequently failing the next two items. This results in six lost points on the language subtest [42, 50]. Secondly, language comprehension difficulties can affect an individual's ability to perform on 'non-language' related test items. For example, as part of the memory subtest, examinees are presented with two objects (cup and spoon) which they are asked to remember. Immediately after the initial presentation of these two objects, and then after a time delay, the examinee is asked to identify these two objects when they are presented alongside additional objects. Clinical experience has shown that a common error is for examinees to proceed to name all of the items presented to them, rather than the two original objects they were asked to remember. This results in failure on these test items and, as such, a reduced memory score influenced by language comprehension difficulties, rather than poor memory ability per se.

The SIB can be used in clinical practice and in research studies in combination with other neuropsychological tests. If the SIB is being used in conjunction with the CAMCOG assessment (see Chap. 7), the examiner must be aware that the SIB attention subtest items are incorporated into the CAMCOG meaning both these tests include the same test items relating to the attention domain. This raises the potential for practice effects and/or could impact on an examinees engagement and motivation level due to them being asked the same questions on more than one occasion, possibly within the same testing session!

Case Vignettes

In this final section, three case vignettes are presented to highlight how the SIB can be used for a range of people with ID, in conjunction with informant measures, as part of longitudinal, prospective neuropsychological assessment of dementia. The case studies also illustrate some of the strengths and limitations outlined in the previous section.

Vignette 1: James: Mild Cognitive Impairment (MCI) [62]

James is 57 years of age and lives in supported accommodation with two other people. He has a mild ID, and good verbal communication skills (comprehension and expression). He attends a day service 3 days a week and enjoys a range of activities and interests in his spare time. In terms of everyday skills, James is able to prepare simple meals and make hot drinks, and he can take care of all of his self-care needs without assistance.

James was first seen for neuropsychological assessment 10 years ago when he was 47 years of age. The assessment was prompted by concerns expressed by care staff about James having problems concentrating during activities, and mixing up days of the week resulting in him missing appointments and activities at the day service. James completed the SIB and care staff completed a range of informant measures.

Information gathered during the first assessment indicated that the presenting concerns were related to psychological difficulties that James was experiencing after going through significant and stressful events in his life—day service closure resulting in him moving to a new service, and his mother's ill-health that resulted in him moving into a short-term respite placement and then into supported accommodation. James scored 94/100 on the SIB on the first assessment.

James was referred again for neuropsychological assessment when he was 56 years old. Care staff reported that James takes more time to remember where items are kept in the kitchen, and sometimes needs prompting from staff; James takes longer to remember recent events, for example what he did yesterday, and he sometimes misplaces his belongings. Care staff did not report any changes in James' everyday skills, self-care ability, behaviour or other areas of cognitive functioning. Once again, James scored 94/100 on the SIB. Concerns were apparent on informant measures, but these did not meet criteria for onset of dementia.

The team have given a provisional diagnosis of Mild Cognitive Impairment (MCI) [62] and James continues to be seen for review and repeat assessments.

Vignette 2: Patrick: Alzheimer's Disease

Patrick is 62 years old and he has a diagnosis of mid-stage Alzheimer's disease. He received this diagnosis approximately 2 years ago. Patrick lives in a large residential home that has 20 residents. He has lived there for about 15 years and prior to this he lived with his mother.

Patrick was previously very independent with his everyday skills and he was able to attend to his self-care needs without any assistance. He enjoyed a range of activities and could go out independently in the local community and travel independently to his day service.

Patrick was first referred for neuropsychological assessment around 10 years ago when he was in is early 50s. Care staff reported that Patrick was in need of slightly more support with his self-care, in particular prompts and minor assistance with shaving; he had also hit out at other people on a few occasions and was less interested in his activities, hobbies. Patrick completed the SIB during the first assessment and care staff completed a range of informant measures, the results of which did not indicate the onset of dementia. Following this initial assessment, Patrick continued to be monitored and the assessment was repeated every 6–12 months.

Table 13.4 Patrick's SIB scores

SIB score	Time 1	Time 2	Time 3	Time 4	Time 5	Time 6	Time 7
Social interaction	6/6	6/6	6/6	6/6	6/6	6/6	5/6
Memory	13/14	14/14	12/14	10/14	10/14	12/14	4/14
Orientation	6/6	5/6	5/6	6/6	6/6	6/6	2/6
Language	39/46	39/46	39/46	39/46	38/46	36/46	30/46
Attention	6/6	6/6	6/6	4/6	4/6	6/6	5/6
Praxis	4/8	6/8	8/8	7/8	6/8	5/8	0/8
Visuo-spatial	8/8	6/8	8/8	8/8	4/8	8/8	7/8
Construction	4/4	4/4	4/4	4/4	4/4	4/4	4/4
Orienting to name	2/2	2/2	2/2	2/2	1/2	1/2	2/2
Total	88/100	88/100	90/100	86/100	79/100	84/100	59/100

Patrick's SIB scores for repeat assessments are shown in Table 13.4 above.

It is evident from looking at the scores in Table 13.4 that Patrick's SIB scores remained stable across assessment time points 1–3. Between time points 3 and 4 there was a 4 point decline in Patrick's total SIB score, with decline particularly evident in his memory score. At this time, the informant assessment results suggested possible onset of dementia; however, Patrick was experiencing a range of physical health problems, including frequent urinary tract infections and, to add to this, his mother was experiencing health problems, which meant she was no longer able to visit Patrick as often as usual. Patrick was very tearful and withdrawn and it was felt that depression, coupled with the impact of physical health problems, possibly accounted for the decline in scores on informant measures and on the SIB.

However, once Patrick's physical and psychological health needs were addressed, he continued to present with decline in his everyday skills, cognitive functioning, self-care and behaviour. The diagnosis of Alzheimer's disease was given around time point 6, based on the results of the SIB and informant assessment. Further progression of dementia was evidenced in Patrick's SIB scores at time point 7.

Patrick continues to live well, and receives excellent support from care staff in his home.

Vignette 3: Alice: Non-Dementia

Alice is 49 years old and lives at home with her mother. She is described as having a moderate ID. She has good language comprehension; however, her speech is limited to single words and short phrases. She also has visual impairment caused by a corneal disorder—keratoconus. Alice enjoys doing craft activities and copy writing at home and attends a day service during the week.

Alice was referred for neuropsychological assessment about 5 years ago due to concerns that she was having problems associating activities with days of the week; she appeared slowed down, less interested in activities, required more prompts with everyday skill, such as making drinks, and was experiencing episodes of low mood and tearfulness.

Alice completed the SIB and informant measures were also completed as part of the assessment. Alice scored 72/100 on the SIB on the first assessment. The initial assessment did not indicate onset of dementia. Instead, hypothyroidism and deteriorating vision were thought to be causing the presenting concerns.

Alice was seen for a repeat assessment 2 years after the initial assessment. Results of the repeated informant and direct assessment did not indicate onset of dementia and her SIB total score remained stable 73/100.

Due to Alice being at significant risk of developing early onset dementia she continues to be monitored and will be seen again soon for repeat neuropsychological assessment.

Summary

Best practice guidelines on the neuropsychological assessment of dementia in people with ID recommend the use of both informant and, where possible, direct measures to track changes in neuropsychological functioning and daily living skills over repeated assessments across time, with the aim to increase early identification and diagnosis of dementia [5]. There are a number of direct neuropsychological assessments now available to the clinician or researcher for use in the assessment of dementia in people with ID. This chapter has described and reviewed one such test—the Severe Impairment Battery (SIB) [15].

The SIB was designed as a measure of neuropsychological functioning in people with severe dementia in the general population, so it was not originally designed for use with people with ID. Although there are only three studies that have directly addressed the psychometric properties of the SIB, and there still has not been any research into the sensitivity or specificity of the test, the evidence that is available shows the SIB to have good reliability and validity when used as a neuropsychological assessment of dementia in people with DS. This is further supported by the fact that researchers have started to use the SIB as a criterion validity measure in the development of new tests [63].

The usefulness and utility of the SIB for use in research and clinical practice has been demonstrated, but of course, there are many factors that the clinician/researchers must consider when selecting a neuropsychological measure. To help here, this chapter has reviewed the strengths and limitations of the SIB when used with people with ID and presented some short case vignettes to demonstrate the utility of the SIB for neuropsychological assessment of dementia with a range of people with DS and ID.

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Chapter 14
Strengths of Previous Work and Future Challenges

Diana B. Burt

Introduction

The dementia tests and scales described in previous chapters are an impressive representation of work conducted to improve dementia diagnosis in adults with intellectual disability (ID). In this chapter, the strengths of work represented in this book and new issues that have arisen since the first edition of this book are discussed. Anticipated future challenges are also delineated. The discussion of strengths is not intended to be exhaustive. Instead, the following highlights and related future challenges will be considered: identification of onset of decline, differential diagnosis, monitoring progression, breadth of functional areas assessed within and across instruments, modified administration and scoring techniques, and scale evaluation methods. New issues include: evaluation criteria for tools used at different stages of practice, changes in validated scales, baseline definition/documentation, mild neurocognitive disorder in adults with ID, and collaboration among clinicians/researchers. Additional general challenges faced by clinicians and researchers involved in dementia assessment include longitudinal research methods, multidisciplinary expertise, and funding.

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Strengths of Previous Work

Identification of Onset of Decline

The Gedye DSDS was designed to allow the clinician/researcher to identify the date of early skill loss, thus identifying the onset of mild neurocognitive disorder or dementia (Chap. 4). The ability of informants to retrospectively report on the onset of declines, which can be gradual and difficult to detect, however, is a matter for future investigation. Evaluation of efficient informant-report scales to regularly monitor functioning in healthy adults and thus detect early signs would be valuable (e.g., ABDQ (Chap. 10); NTG-EDSD (Chap. 11)). In addition, it would be beneficial to evaluate scales found to be sensitive to early declines [1–3] by adding them to existing batteries across sites. Comprehensive procedures with repeated assessments, such as those from the CAMDEX-DS (Chap. 7) and scales such as the SIB (Chap. 11) are designed to maximize the chances of detecting mild neurocognitive disorder, which could be the preclinical stage of an impending dementia (see SIB vignettes). Extensive batteries and independent clinical assessments, such as those used by Devenny and colleagues, also allow an examination of early signs of dementia, such as preclinical changes in memory functioning [3–5].

Identifying the onset of declines is important for clinical and research purposes. Detection of early change indicates that the adult would benefit from differential diagnostic procedures such as those built into the CAMDEX-DS, DSDS, and NTG-EDSD. Early change also indicates that the adult needs more extensive follow-up and monitoring than individuals without such change. Clinically, treatments could be most effective in the earliest stages of dementia if detection of such early change is possible. In addition, if declines are related to some treatable condition (e.g., hearing loss due to ear wax or allergy-related congestion, sleep apnea), then earlier detection leads to earlier treatment minimizing disruptions in functioning. For research purposes, if investigators are examining biological substrates of dementia, it is often necessary to detect the onset of clinical signs. If a certain biological process is associated with dementia, changes in a biological marker should coincide with or precede the onset of dementia. Finally, accurate identification of onset is important in research on the prevention of declines [4].

Monitoring Progression

The ability to monitor the progression of declines is a strength of several scales. The DSDS, for example, allows one to record ten assessments on the same form. The interval between repeat assessments is determined by behavior reported at each assessment. If signs of decline are reported, the interval between assessments is reduced to 6 months. Diagnostic accuracy improved on the DSDS with repeated assessment, so it is a strength of the scale to be able to record repeat observations on

one form. The DSDS also provides guidelines to identify stages of dementia based on the scores obtained at each assessment.

As mentioned previously, comprehensive schedules like the CAMDEX-DS and multiscale batteries (Devenny, Zigman and colleagues) allow one to detect declines in functioning and to determine when some but perhaps not all areas show decline [6–8]. The RADD and SIB (Chaps. 12 and 13) provide sub-scale scores that also allow one to detect such declines. These tools allow one to describe the natural history of mild neurocognitive disorder and dementia across individuals to determine whether there is any universal pattern or invariant sequence of decline. The DLD (Chap. 3) also provides separate scores for cognitive versus social functioning, and subscale scores in each area can be recorded. Thus, the Examiner can determine whether changes occur in both areas simultaneously or whether declines in one area precede the other. Holland and colleagues suggest that changes in personality or behavior precede cognitive changes. Zigman and colleagues suggest that maladaptive changes precede changes in adaptive behavior and that within the adaptive behavior domain not all skills decline at the same rate. Devenny and colleagues indicate that declines in cued recall occur early. McCarron and colleagues conclude that declines in delayed memory, writing ones name, and counting to 10 precede well-practiced motor tasks like shaking hands. Additional research is needed, using overlapping scales at multiple sites, to determine whether there is an invariant progression of decline in adults with and without DS. In addition, it will be useful to determine whether stages will be identified such as those reported on the DSDS. As demonstrated by the work here, declines can only be detected in areas being assessed repeatedly. If an investigation in cognitive/memory change does not assess early personality or behavioral changes, for example, it is difficult to determine the actual sequence of decline. If scales are not used at multiple sites, it will not be possible to examine reported discrepancies in areas of early decline.

Differential Diagnosis

The inclusion of techniques in a scale to aid in differential diagnosis is valuable. The techniques highlight the fact that conditions other than Alzheimer disease cause changes in functioning. It is the clinician/researcher's responsibility to consider all possible causes of any behavioral changes, and to provide treatment or to refer adults for treatment as appropriate. As with many of the procedures involved in a comprehensive dementia evaluation, attention to differential diagnosis may be standard practice for most clinicians/researchers. The inclusion of standardized procedures for their consideration, however, ensures that they will not be overlooked and allows others to replicate and evaluate them. Two assessment instruments and an information gathering tool contain techniques to aid in differential diagnosis. The DSDS includes questions to help in the identification of conditions such as hypothyroidism and depression. The CAMDEX-DS provides a structure to collect information on physical conditions, psychiatric disorders, and sensory impairments that can affect

functioning in later life. Holland and colleagues also provide case studies to illustrate that conditions such as hearing difficulties or depression affect functioning. The CAMDEX-DS also provides guidance for post diagnosis intervention, highlighting the fact that the diagnosis of dementia is just the beginning of the clinical process. The NTG-EDSD gathers information on recent stressors/changes, sensory impairments, sleep history, and psychiatric/physical conditions. Illustrative vignettes demonstrated the use of the SIB in making diagnostic judgments (e.g., declines related to dementia versus treatable medical conditions). Finally, Lott and colleagues (Chap. 12) indicated that the RADD could aid in differential diagnosis.

Breadth of Functional Areas Assessed

Other than tests designed specifically to assess one area of functioning (e.g., Chap. 9), most of the instruments were designed to assess and document declines in several areas of functioning as required by dementia diagnostic criteria [9]. One of the most comprehensive assessment schedules, the CAMDEX-DS (Chap. 7), involves informant-report of functioning in a number of areas (e.g., memory, mental functioning, everyday skills), interview of the adult with ID, direct assessment of seven areas of cognitive functioning (e.g., praxis, language, memory), standardized observations of the adult with ID, and physical examination (including laboratory investigations). The schedule was designed to collect all information needed for a clinician to make a diagnosis of dementia over repeated assessments. Similarly, informant-report dementia scales (Chaps. 3 and 4) as well as an information gathering tool (Chap. 11) involve requests for information about multiple areas of functioning (e.g., orientation, social skills, memory performance). Informant-report scales for adaptive behavior also assess functioning in several areas (e.g., independent functioning, language development, economic activity; Prasher; Zigman and colleagues). Regarding direct performance tests, the TSI, RADD, and SIB (Chaps. 8, 12, and 13, respectively) assess several functional areas (e.g., language, memory, conceptual ability).

The inclusion of multiple functional areas makes a scale more useful for several reasons. First, the scale is more likely to include areas needed to document declines for mild neurocognitive disorder or dementia diagnosis. Second, items/tests usually differ in terms of task demands (e.g., verbal versus nonverbal responses). Thus, some of them are likely to be more useful than others for assessing adults with differing sensory abilities, premorbid levels of functioning, and strengths/weaknesses profiles. If an individual does not have or loses understandable speech, for example, items/tests that require nonverbal responses can be administered and scored separately. Multifaceted batteries or tests also allow the Examiner to create profiles of functioning to determine premorbid strengths and weaknesses. Finally, decline may occur at different rates for different skills across individuals. Such differences in the area and rate of decline were observed on the TSI and the Adaptive Behavior Scale (Prasher, Zigman, and colleagues).

To benefit from breadth in an assessment instrument, however, the instrument must allow documentation of performance on subscales or tests that assess different

abilities (e.g., CAMDEX-DS, RADD, SIB). Scores for different functional areas must also be recorded and available for future comparisons. One summary or composite score for all abilities is less useful than a set of scores, because declines in one but not all areas can be masked by a summary score. If so, then the summary score would be less sensitive to the onset of mild neurocognitive disorder or dementia than individual subtest scores. It is most useful if the repeated scores are raw scores as well as standardized scores and that they are accompanied by actual descriptions of performance (e.g., dresses self-completely including tying and buttoning, remembers five words in correct sequence in a sentence). In longitudinal research, instruments often undergo revisions which change the items included and the way summary scores are computed (e.g., the Maladaptive Subscale of the Adaptive Behavior Scale). On retest one needs to be able to compare current to previous performance and the more detailed the information, the more useful it will be. In addition, if an adult moves, the assessment procedures could change and with behavioral descriptions one can better judge whether changes in functioning have occurred. Finally, diagnostic criteria (e.g., DSM-V, ICD-10) change over time, perhaps requiring knowledge of performance on subscales.

In addition to strengths in the breadth of functional areas in individual scales, their breadth as a group is a strength. Examples of dementia scales, a dementia screen, adaptive behavior scales (standard versus shortened version), cognitive scales, and an information gathering tool were presented. Challenges for future work are whether the scales would provide improved diagnostic accuracy if combined into a battery or schedule such as the CAMDEX-DS [6, 7]. It is unknown at this time what combinations of scales allow optimal diagnostic accuracy for which adults with ID. Although progress has been made in identifying useful tests or areas of assessment [1, 10], there is rarely collaboration across sites or countries [4]. Finally, it is highly possible that different tests or groups of tests will be needed for adults with different characteristics (e.g., sensory abilities, levels of intellectual functioning, speech skills) [11].

Administration Techniques

Another strength of the tests and scales was the innovative way in which administration techniques were developed and used. Multiple sources of information were described. In addition, flexible administration rules were standardized so they would be appropriate for adults with differing capabilities.

Source of Information

As has often been the case historically [12], informant-report scales of adaptive behavior, everyday functioning, and emotional functioning were described. Direct assessments were described for tests of memory and other cognitive functioning.

Such a dichotomous splitting of source of information allows a group of tests to be administered to adults functioning at many levels and with different abilities. As long as informant-report and direct testing of adults are used to assess mutually exclusive areas of functioning (i.e., adaptive, everyday, emotional versus cognitive, memory) all of which are involved in dementia diagnostic criteria, both sources of information will be needed to accurately assess dementia. Advantages and disadvantages of the use of the two sources of information were discussed in Chap. 2. Challenges for the future involve assessment of the veracity of informant reports and exploration of ways to improve such reports [13, 14].

With few exceptions (e.g., CAMDEX-DS, NTG-EDSD), the adults with ID were not asked to report on their own memory, cognitive, everyday, or emotional functioning, nor were direct observations of functioning made a part of the assessment. Clinicians and researchers may routinely make observations or interview adults with ID as part of their clinical or research procedures. Such observations, however, were not integrated into the standardized scales or procedures themselves. Direct observation of the adult with ID is a critical part of any dementia assessment, and the standardization of observation and interview procedures is a strength because it allows procedures to be replicated and evaluated across sites.

An additional strength of informant-report work was the use of multiple informants to allow a determination of inter-rater reliability and the examination of differences in perspective (Prasher; Gedye and colleagues). The DSDS, for example, recommends informants from two different settings. Interestingly, there is a difference across scales in terms of the specified amount of time for reliable informants to have known the adult with ID. The DSDS requires that informants know the adult for at least 2 years, whereas the CAMDEX-DS interview and the NTG-EDSD require only 6 months of association with the adult being described. In practice, unless adults with ID have close contact with parents or other family members, it is difficult to find an informant knowledgeable about all aspects of the adult's functioning. The length of time required for an informant to observe and adequately report on functioning for a dementia assessment is an area for future research and discussion. Promising work has been done to train informants to improve the accuracy and reliability of their reports [13].

An additional strength of several of the informant-report scales is that informants indicated whether current functioning represents a change from typical functioning (ABDQ, DSDS, CAMDEX-DS, NTG-EDSD). It is very important to determine whether current behavior represents a decline or change from typical behavior as required by dementia diagnostic criteria [4, 9, 15–17]. Such requests for judgments about typical functioning, however, often require a certain amount of retrospective reporting on the part of informants. Retrospective reporting is subject to memory errors or bias. As behavioral changes become more remote, it is possible that errors in memory will become more pronounced (e.g., the parent or other informant could forget what their child could previously do, adjusting their expectations to changing performance). At an initial evaluation for dementia, previous assessment information is not always available or in a format to allow for evaluation of changes in performance. Thus, retrospective reporting is the option of choice. It is beneficial as

in the CAMDEX-DS, therefore, to allow informants to report that they don't know whether behavior is typical or not. They should also be allowed to respond that a question is not applicable to the adult in question. Such choices in responses are important so that an informant is not forced to make a response that does not accurately describe an individual.

Flexibility of Instructions

Investigators developed multiple forms of their scales or built in modifications to their administration procedures to allow for administration across individuals with different capabilities (e.g., sensory impairments, language spoken). Administration flexibility increases the applicability of the scale across individuals. It also makes it more likely that a scale will remain useful to chart the progression of dementia once skills are lost (e.g., loss of speech or the loss of a pointing response). Dalton, for example, built flexibility into the administration procedures for his Dyspraxia Scale for Adults with Down Syndrome [18] (Chap. 5). Instructions allow different levels of prompts for adults with differing sensory abilities. In this way, the Examiner is able to differentiate declines due to dementia from those due to sensory loss. Similarly, Holland and colleagues made modifications to a number of the direct assessment items on the CAMCOG portion of their schedule. Points were awarded on the basis of answers given with and without the Examiner's prompting. The SIB uses gestural cues and prompts as necessary. The RADD was found to be useful for nonverbal adults and those with sensory impairments (except significant hearing impairments). Finally, the TSI allows the Examiner to repeat the instructions three times to engage attention. Regarding considerations for differences in language or cultural background, Dalton built flexibility into his Dyspraxia Scale for Adults with Down Syndrome that allows for international administration (i.e., changing the coins used). The DLD and NTG-EDSD allow flexibility in administration, because they are available in many languages [19]. Similarly, the DSDS is commercially available in English, French, and Swedish and has been administered in several non-English speaking countries such as Japan, Holland, and France.

Scoring

Approaches to scoring were innovative. Dalton [18], (Chap. 5) for example, computed *z*-scores for his Dyspraxia Scale, so that performance on it could be compared to performance on other tests in his battery. McCarron and colleagues computed and examined annual rate of change scores on the TSI. Such scores provide a benchmark against which to compare repeated performance on the test. An examination of profiles of cognitive performance is possible on the CAMDEX-DS because of the derived subscale scores. The RADD and SIB also provide subscale scores and the

SIB has different scoring for use of prompts. Profiles of adaptive and maladaptive performance can be examined on the Adaptive Behavior Scale as demonstrated by Zigman and colleagues. Such comparisons allow an examination of the natural history of dementia. Relative decline is examined to see whether specifiable functional areas show decline first.

Innovative scoring procedures also allow one to determine which tests are best for screening early signs of dementia versus later confirmation of dementia (i.e., documentation that diagnostic criteria are met). On the ABDQ, for example, Prasher used weighted item scores when he discovered that some items were more or less predictive of dementia than others. Such a weighting technique makes scales more useful and can make them adaptable for adults with different capabilities (i.e., if different weights are assigned based on level or cause of ID, age, gender).

It is important to remember that standardized scores, such as *z*-scores, must be interpreted with caution when they are obtained at one administration. Differences in scores could represent premorbid intra-individual differences in strengths/weaknesses profiles. If an adult with DS at one time of testing, for example, has strong dyspraxia performance compared to memory or fine motor performance it does not necessarily mean that the adult has had declines in memory or fine motor performance. It could be that dyspraxia has always been a relative strength for the individual. It is important to remember that there can be large inter-individual differences in strength/weakness profiles as a function of sensory abilities, speech, and cause of ID (DS or other conditions). Such premorbid differences must not be confused with documented declines related to dementia.

Several investigators (Devenny, Zigman, Lott and their respective colleagues) evaluated the use of cutoff scores or formulas at one time of assessment to differentiate adults with dementia from those without dementia. This procedure eliminates the need to rely on retrospective reporting (cf., ABDQ, DSDS, NTG-EDSD). As mentioned by the investigators, however, there is usually overlap in the scores of adults with and without dementia. Consequently, a certain number of false-positive and false-negative diagnoses occur (as with any technique). Zigman, Lott and their colleagues took intellectual level into account when interpreting performance on the ABS and RADD, respectively. Cutoff scores used by Devenny and colleagues were said to apply for adults with IQ scores of at least 30. On the DLD a single assessment approach, using different cutoff criteria as a function of IQ, was originally adopted and abandoned (Evenhuis and colleagues). Challenges for future work, therefore, will be to determine the advantages and disadvantages of using scoring systems designed for diagnosis based on one assessment. As mentioned by Devenny and colleagues, cutoff scores and formulas based on a research sample could be different from those needed for adults who have never seen the instruments or scales before.

Interpretation of scores from a dementia assessment is often a challenge. The inclusion of DSM and ICD diagnostic criteria for mild neurocognitive disorder and dementia in a checklist, such as in the CAMDEX-DS, aids in the diagnosis and differential diagnosis of dementia. Such a checklist reminds practitioners that it is feasible to apply diagnostic criteria. It also provides a framework for reporting

results from one assessment to another (i.e., which diagnostic criteria were met at a given time) and for comparing results across research studies. The CAMDEX-DS also provides guidance on how to use the information gained through the assessment in making a judgment about dementia status. Such guidance is valuable, because it is sometimes unclear how to use test scores either singly or in combination with others when making a diagnosis of dementia, or in deciding that a further dementia assessment is indicated.

Evaluation Methods

Impressive techniques were used to evaluate dementia tests and scales. Investigators, for example, used external criteria for dementia status that were independent of the scale being evaluated (Devenny and colleagues, Holland and colleagues, Prasher, Lott and colleagues, Hutchinson). Holland and colleagues also used their direct assessment data to evaluate their interview schedule. Thus, they were able to avoid possible errors involved in using clinical judgment as the sole external criterion. Investigators also used extensive diagnostic procedures to determine that adults identified with dementia did not have other conditions that could account for declines (e.g., depression). When examining DAD specifically, investigators also ruled out other types of dementia. In addition, scales and dementia identification criteria were developed using one sample of adults, and then evaluated in completely independent samples (Prasher) or in samples of adults with new onset dementia (Devenny, Zigman and their respective colleagues).

A number of techniques were used to make scales more efficient and accurate. Prasher, for example, discerned that some of the items on his scale were less useful than others, so he eliminated them (i.e., reports of toothbrushing skill). Devenny investigated ways to shorten the assessment process by evaluating the use of two versus three trials of data. Holland and colleagues eliminated items from the original CAMCOG that were less useful for the assessment of adults with ID (e.g., serial sevens). Dalton and colleagues (Chap. 5) combined data on his Dyspraxia Scale from several sites, so that he could determine which items were most useful. He then shortened the scale to improve efficiency and ease of administration. Evenhuis and colleagues compared the value of single versus repeated administration of the DLD, and changed their recommendations regarding optimal testing schedule based on their findings. They also determined that their scale was most useful in the mid-range of ID, because of floor and ceiling effects in adults in the profound and mild levels of functioning, respectively.

Scales have been used and evaluated at numerous sites (ABS, DLD, DSDS, and TSI). Thus, evidence is accumulating as to their validity and reliability. The DSDS, for example, has been used in a number of research studies, with evidence suggesting that it is most useful for adults similar to the normative sample who are functioning in the severe or profound range of ID. A future challenge is whether the scale needs modification to increase its sensitivity across a wider range of capabili-

ties. The DLD has also been evaluated in a number of studies, with some inconsistency regarding the usefulness of single versus repeated administration. Such multisite evaluation is the type of research that is needed to determine the usefulness of existing and newly developed dementia scales [20].

Instruments were identified as being most useful as either dementia screening or diagnostic tools. The CAMDEX-DS and the DSDS were designed and evaluated for the diagnosis of dementia. In contrast, the Dyspraxia Scale for Adults with Down Syndrome, the DLD, and the ADBQ were described as dementia screening instruments that could be used to detect early signs of dementia. The NTG-EDSD was described as an information gathering tool. The Cued Recall Test detects preclinical or early declines in memory (Devenny and colleagues) suggesting that it would also be useful in a battery to screen for mild neurocognitive disorder or dementia. Assessment of maladaptive behavior (Zigman and colleagues) could also be useful in a dementia screening battery. Future research is needed to determine the combination of screening and diagnostic tools that will lead to early, efficient, and accurate diagnosis of dementia in adults with ID. Data mining or hierarchical linear modeling (HLM) could be useful techniques for identifying such tools [7, 10, 21].

New Issues Identified

Evaluation Criteria for Tools Used at Different Stages of Practice

Evaluation criteria have been proposed for individual tests/scales (e.g., Chap. 2) [20, 22] and for tests/scales identified for inclusion in consensus batteries [9]. Criteria have not been outlined for new information gathering tools, which were not intended to be dementia diagnostic screens (e.g., Chap. 11). If information from such tools is shared with health professionals and stored in permanent medical records, however, it would be best evidence-based practice if the tools also met proposed validation criteria [23]. Once information from such tools is conveyed, there could be little control over how it is used. In the absence of assessment expertise and additional assessment, informal data would probably be used in current or future diagnostic judgments, especially by health providers who lack experience with dementia assessment in adults with ID.

Changes in Validated Instruments

At times, scales/tests have been changed and/or used in ways that are different from their validation form (e.g., items added, cut-off score not used, schedule of assessment changed). The question then becomes whether new validation studies are

needed. If information gathering tools or tests/scales are developed from previously validated scales; then inclusion, administration, and scoring of the scale in its original form would provide needed reliability and validity until a new validation study can be conducted (S. Deb, personal communication, January 31 and February 1, 2017). New items or areas of assessment could be added, and the scale could be scored and evaluated with and without the new items. Diagnostic cut-off scores, for example, could be evaluated to see how well they function as referral cut-off scores for information gathering tools. Scales used for a new purpose (e.g., information gathering) could be compared to existing validated scales [23]. The risks of using modified scales without new validation include: unsubstantiated decision making, lack of sensitivity in referral/diagnostic judgments, and lack of data for evaluation/sharing purposes.

Baseline Functioning Issues

Informant-report scales designed and validated for use at one assessment often involve retrospective judgments about whether changes have occurred in functioning (e.g., ABDQ), Dementia Screening Questionnaire for Individuals with Intellectual Disabilities (DSQIID) [24]). Recently, such scales or their modifications were proposed for use as baseline and repeat follow-up tools. Thus, questions arise about what type of baseline information a retrospective scale provides, what baseline information informants use when making repeated judgments, and what type of administration instructions/details are needed to ensure reliable research/practice. Initial retrospective judgments (i.e., how is adult with ID doing now compared to time in past) are not an absolute indication of baseline functioning. For repeated administration, clinicians/researchers using such scales emphasize the importance of comparisons of current to premorbid baseline functioning, not current to functioning at previous retrospective assessments (S. Deb, personal communication, February 1, 2017). Differences in how such judgments are made would be expected to affect most psychometric properties of a tool.

A tool involving retrospective informant judgments could provide an indication of baseline functioning, if some record of absolute past functioning is included. The CAMDEX-DS designed for repeated assessments, for example, provides absolute judgments on current behaviors which provide the basis for future comparative decisions. As indicated in Appendix E, current behavior is rated as yes-no on an item such as “has difficulty remembering location of items.” The same item then assesses comparisons of current to past performance, based on whether there has been a deterioration and how severe the deterioration is. Future research is needed to explore issues related to maximizing the reliability of retrospective informant ratings, which are often the only type of informant rating available.

Mild Neurocognitive Disorder, Healthy Aging, and Diagnostic Consensus

DSM-V diagnostic criteria for mild and major neurocognitive disorders [25] raise issues about the use of dementia diagnostic categories in adults with ID. When healthy adults, who later show declines in functioning, are followed longitudinally, symptoms of dementia do not all arise at one period of time [4, 5, 26, 27]. Rather, declines can occur over several years, with early declines failing to meet diagnostic criteria for dementia. Adults with such early changes have been classified as having early dementia, mild cognitive impairment, possible dementia, etc. The inclusion of such diagnostic categories has been shown to affect prevalence rates and the ability to compare rates across sites [5]. Discussions about the use of such important classification systems, and about diagnostic criteria and methods needed for their use (e.g., Cued Recall Test) are best left to an international consensus group. Such classifications are important for treatment and prevention research, because adults with mild neurocognitive disorder could provide different results from those who meet criteria for dementia. Thus, the time is right for an international consensus group to convene about these classifications, about the continued need for differentiation of significant declines from typical aging [5], and about other fruitful directions for future work in this area.

Collaboration

Recommendations have been made in the past for multi-site collaboration on assessment issues among clinicians/researchers. Some progress has been made in this area [5]. The Cued Recall Test, for example, is being used in a multi-site longitudinal study to examine relationships between neuropsychological functioning and the deposition of amyloid-β in adults with DS (Chap. 9). Such investigations of the biological basis for dementia and treatment or prevention studies provide funded opportunities for scale evaluation, which will hopefully be pursued further. Another example of multi-site use is the evaluation of NTG-EDSD usability across international sites [19]. Further validation studies will hopefully follow in the near future. In the absence of multi-site collaboration, the most one can hope for is the consistent use of scales across studies so that new evaluation techniques and critical evaluations of scales can be conducted [20, 22, 28]. In addition, new methods for communication and information sharing are needed [22].

Future Challenges and Directions

Several challenges in future work for clinicians/researchers were mentioned in the discussion of new issues and in the strengths of work presented in previous chapters. Three additional challenges related to longitudinal methods needed to examine assessment procedures are now discussed.

Longitudinal Research

There are several challenges inherent in the longitudinal assessment required to evaluate dementia scales (i.e., repeated assessment over time). The first challenge is to integrate collection of useful baseline data into assessments that are already being conducted for adults with ID (e.g., transition to workplace assessment or in medication follow-up clinics in the United States). Logistic issues also arise, such as storing information/data for long periods of time in a format that is useful. Another logistic issue is the loss of investigators, who lose funding or become older themselves and retire from clinical or research work. One issue illustrated by several of the investigators (e.g., Evenhuis and colleagues, Dalton and colleagues, Prasher, Zigman and colleagues) is change in scoring techniques or scale modification over time. A clinician/researcher could collect and store information about adults in one format. They could, for example, store data indicating whether adults meet cutoff scores for dementia on the DLD on the basis of one assessment, total scores on the full Dyspraxia Scale for Adults with Down Syndrome, and total scores on the adaptive behavior scale. If the clinician/researcher then wanted to use the newly modified scoring criteria, they would have to change their scoring procedures (i.e., use only change scores on the DLD, use the shortened dyspraxia or adaptive behavior scale scores; use the maladaptive behavior scale with improved reliability). Such modifications are only possible if the clinician or researcher has access to previously collected raw data. Data or clinical information must be stored in a form as close to that which was collected as possible to allow for maximum usefulness across time.

Interdisciplinary Expertise

Longitudinal, cutting-edge research on dementia assessment requires expertise across a number of disciplines. Given that the diagnosis of mild neurogenerative disorder or dementia is a clinical judgment (Holland and colleagues; Prasher; Gedye and colleagues), it is imperative that someone on the clinical/research team has actual experience with the challenges of diagnosing dementia in adults with ID. Using clinical judgment improves with time and experience. Developing and evaluating dementia assessment scales requires further expertise. Unfortunately, clinicians and researchers with an interest and expertise in the diagnosis of psychiatric disorders in adults with ID are rare. Training programs for their education are also rare. The type of research needed, involving repeated assessment of the same procedures, is not the type of innovative, creative process that easily attracts and holds the attention of researchers. Communicating cooperatively and extensively with other researchers and integrating methods across sites is not a commonly taught skill. From a research standpoint, a relatively small number of participants with dementia of the Alzheimer type are identified after years of costly research. Thus, researchers who are sometimes required to compete with each other for funds and publication space are going to need to collaborate across sites to make true progress. It is

difficult to attract clinicians and researchers to the field of dementia assessment in adults with ID in the first place, and retaining them is another challenge.

When conducting evaluations of instruments or diagnostic methods, members of the evaluation team need to be up to date on research and statistical methods needed to examine longitudinal data. There are a number of issues to be considered and research needs to be ongoing to better illustrate what are typical or atypical patterns of performance over time. Test–retest effects, for example, must be considered [12, 29] as well as the effects of attrition due to death or survival due to above average health [30]. In conducting analyses to examine the sensitivity, specificity, and predictive validity of instruments or techniques, it is important to remember that how one defines mild neurocognitive disorder or dementia has considerable effects on these measures. For example, if adults with diagnoses of mild neurocognitive disorder are included in a group with dementia, the results could be very different than if such adults were excluded all together or considered to be in the not demented group. It is sometimes helpful to be flexible in terms of analyses, by conducting them several different ways to see if diagnostic grouping has significant impact on conclusions drawn. A diagnosis of mild neurcognitive disorder or dementia involves judgment, and it is best to remember that one is evaluating groupings that are almost certain to contain some erroneous assignment of adults (i.e., false-positive or false-negatives). A team member who is cognizant of changes related to aging versus those related to dementia is a valuable asset to help in making decisions about group assignment. Finally, an area for new and future research is the examination of associations between test performance and any biomarkers that aid in the diagnosis of dementia (Dalton, Devenny, Zigman and colleagues). Once again, members of the research team or consultants to the team would need the expertise required to perform such research.

Funding

Interested researchers and clinicians find it difficult to acquire funding for their assessment work. As mentioned, longitudinal research designed to examine instruments to assess dementia is costly, and consistency is currently more important than creativity. A number of scales and instruments have been developed/evaluated and a logical next step is a large collaborative evaluation of them. Clinicians who are also researchers are in an ideal position to collect and examine data if funding is available (e.g., Prasher, Holland and colleagues). To obtain such funding, in the United States, however, it could require a grassroots call for funding like that launched regarding autism diagnostic issues in the past.

Dalton demonstrated the usefulness of incorporating scale evaluation in clinical trials. Such work could have greater funding potential than assessment evaluation research, but the researcher must have the interest and tenacity required to make scale assessment a subsidiary goal of the research. Researchers have the responsibility to make the greatest use of data collected. The need to share data further

illustrates the need to store data in formats that others can use, both for scale development and evaluation purposes [18]. Such sharing requires clinicians/researchers to take a leadership role in fostering scale development and evaluation. The ability to provide such leadership requires energy, tenacity, experience, and expertise such as that demonstrated by the authors in this book.

Conclusions

In conclusion, the clinicians and researchers whose work is represented in this volume have made outstanding progress toward improving the diagnostic accuracy of mild neurocognitive disorder and dementia in adults with ID. There are considerable future challenges for additional strides to be made. At present, competitive work is being done with researchers making progress at various sites. Cross-site collaboration occurs occasionally. Perhaps it is not realistic to expect collaboration across national boundaries, when such collaboration does not exist for dementia assessment in the general population. The relatively small size of the population of adults with ID and dementia, existing disparities in the quality of health care, and the need for active advocacy for needed research indicate that collaboration in the population with ID will be required. Leaders in the area need to convene international workgroups to generate goals for mild neurocognitive disorder and dementia research in the next decade and beyond, and to identify mechanisms to reach such goals.

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Appendix A: Questions on Clinical Usefulness of Scales, Tests, and Techniques

(A) Practical issues

- How much time is required for completion of the scale or technique (e.g., assessment battery) at each assessment?
- Is the scale completed in one assessment or does it require repeated assessments over time? If the scale requires repeated assessments, is clinically useful information obtained at each individual assessment? For example, follow-up assessments could indicate if any noted declines are consistently observed, if new areas of decline occur, or if performance plateaus.
- What cost is involved? Is any technology involved (e.g., computers with special programs)?
- What level of expertise is required to administer and interpret results from the test? How much clinical judgment is required to determine whether an adult has dementia (declines in functioning) or not? If more judgment is required, more expertise and experience is needed and one could expect greater differences across raters.
- What source of information is being used, the adult with ID or an informant?

(B) Purpose

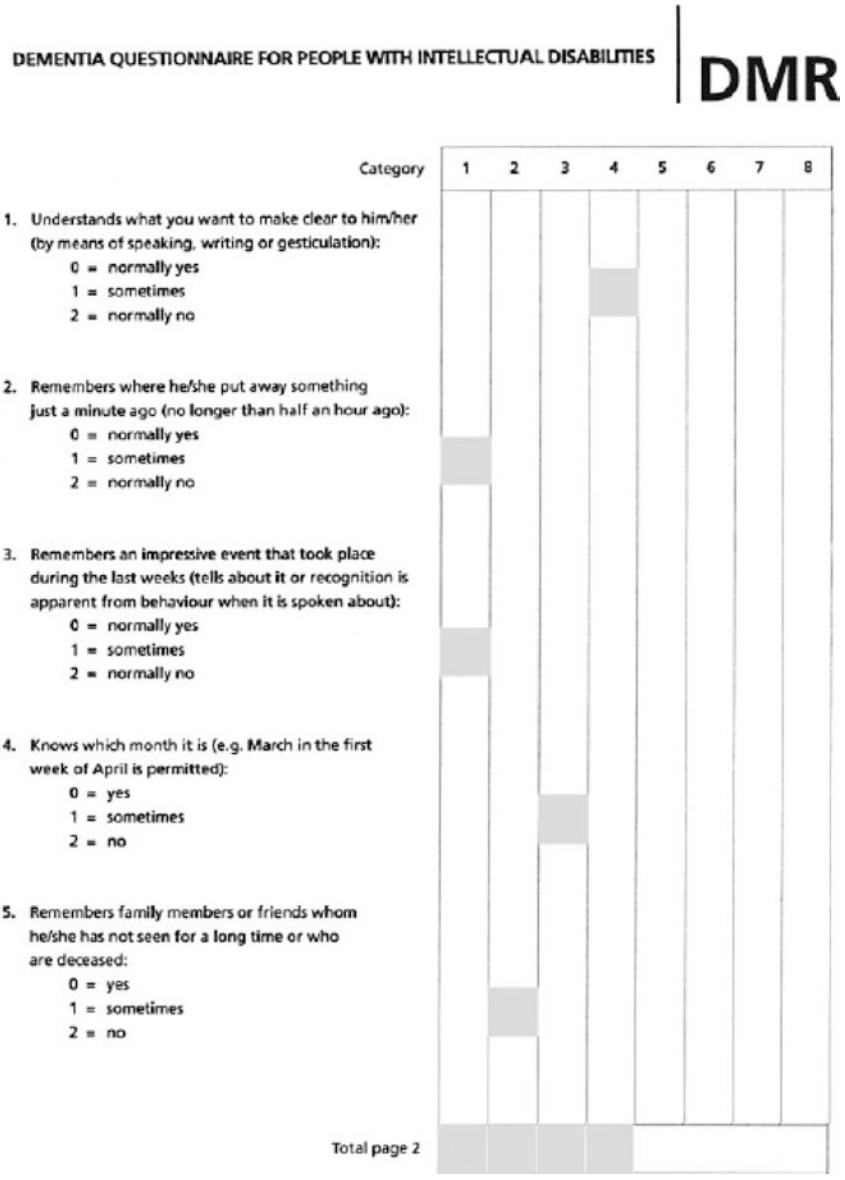
- Is a particular scale or technique designed to aid in formulating a diagnosis of dementia (e.g., dementia scales), or is the technique designed to determine whether one of many diagnostic criteria have been met (e.g., memory tests, adaptive behavior scales)?
- Does the scale clearly indicate performance that would be indicative of dementia vs. performance that would not?

V.P. Prasher (ed.), *Neuropsychological Assessments of Dementia in Down Syndrome and Intellectual Disabilities*, DOI 10.1007/978-3-319-61720-6

- Does the scale provide a dichotomous classification (dementia vs. no dementia) or does it provide an indication of mild neurocognitive disorder or possible dementia?
- Is the technique designed to detect all types of dementia or just progressive dementia like that associated with Alzheimer's disease?
- Has the scale been evaluated to see if it is a good indicator of early signs of dementia, as in a dementia screening test? Or is the scale better suited as a confirmation that dementia is present? Perhaps the scale is designed to gather information pertinent to dementia diagnosis, but is not a diagnostic instrument.
- If the scale indicates stages of dementia, to what extent were such stages validated (i.e., all adults pass through the same stages, only adults with certain types of dementia pass through them, only adults with certain etiologies of ID or premorbid levels of functioning pass through them)?
- Is there a series of questions or techniques designed for purposes of differential diagnosis (e.g., to identify untreated thyroid disease, depression [1])?
- If the test is an assessment of one area of functioning, has it been evaluated as part of a larger battery to see if sensitivity, specificity, or predictive value are improved [2].
- If the scale or technique involves the combination of several skills into one test (e.g., a test with memory and other cognitive components) are scores for each component skill available?
- Have statistical vs. clinically significant differences in functioning been differentiated?[1]
- Can the test be applied in different countries with appropriate translation, or are the items culture specific? Has the technique been evaluated across sites, languages, and cultures?
- If the technique is an adaptation of one used in the general population, has it been modified appropriately so that it is applicable to adults with ID?

[1] Perhaps in a research setting, it has been determined that a decline on a memory test of two items is outside the range expected with normal aging. Would all adults with such a decline have dementia? With such a small indication of decline, any adult who does not recall two items when healthy could not meaningfully be assessed on the test. For this reason, tests with a large range of performance across all adults with ID are often recommended. With memory tests, for example, repeated presentation of test stimuli usually results in higher performance and a larger range of scores [3–5]. This makes the test more useful for dementia assessment. The use of memory cues has also been found to improve performance [5].

Appendix B: Dementia Questionnaire for People with Intellectual Disabilities



Appendix C: Scoring Sheet for the Dyspraxia Scale for Adults with Down Syndrome

DYSPRAXIA SCALE FOR ADULTS WITH DOWN SYNDROME

SCORING SHEET

NAME_____ **SEX** _____ **DOB** _____ **DATE EXAM** _____

LOCATION _____ **AGE** _____ **EXAMINER** _____

Part 1: Psychomotor Skills

While standing

No.	Item Score	4	3	2	1	0
1	Walking					
2	Standing					
3	Look up					
4	Bend your head					
5	Bow from waist					
6	Clap hands					
7	Lift one arm					
8	Lift other arm					
9	Turn head one side					
10	Turn head other side					
11	Lift one leg					
12	Lift other leg					
13	Sitting					

While seated

15	Draw straight line					
16	Clip two sheets					
17	Cut paper sheet					
18	Three coins (one hand)					
19	Coins (other hand)					
20	Put on cap/take off					
Subtotals						
Part 1 Total						

Part 2: Apraxia

No.	Item Score	4	3	2	1	0	
21	Make a fist						
22	Salute						
23	Wave good-bye						
24	Scratch your head						
25	Snap your fingers						
26	Close your eyes						
27	Sniff a flower						
28	Use a comb						
29	Use a toothbrush						
30	Use a spoon						
31	Use a hammer						
32	Use a key						
33	Open a jar						
34	Close a jar						
35	Put on right glove						
36	Put on left glove						
37	Unlock padlock						
38	Lock padlock						
39	Fold sheet of paper						
40	Fold sheet again						
Subtotal							
Part 2 Total							

Part 3: Body Parts/Coin Task

No.	Item Score	4	0
41	Point to your ear		
42	Point to your nose		
43	Point to your eye		
44	Point to your chest		
45	Point to your neck		
46	Point to your chin		
47	Point to your thumb		
48	Point to your ring finger		
49	Point to your index finger		
50	Point to your little finger		
51	Point to your middle finger		
52	Point to your right ear		
53	Point to your right shoulder		
54	Point to your left knee		
55	Point to your left ankle		
56	Point to your right wrist		
57	Point to your left elbow		
58	Point to your right cheek		
59	Give me a penny		
60	Give me a nickel		
61	Give me a quarter		
62	Give me a dime		
Part 3 Total			

Appendix D: Part One Domain I. Independent Functioning

PART ONE
DOMAIN I.
Independent Functioning

A. Eating

ITEM 1 — **Use of Table Utensils** (Circle highest level)

- Uses table knife for cutting or spreading 6
- Feeds self neatly with spoon and fork (or appropriate alternate utensil, e.g., chopsticks) 5
- Feeds self causing considerable spilling with spoon and fork (or appropriate alternate utensil, e.g., chopsticks) 4
- Feeds self with spoon—neatly 3
- Feeds self with spoon—considerable spilling 2
- Feeds self with fingers 1
- Does not feed self or must be fed 0 ☐

ITEM 2 — **Eating in Public** (Circle highest level)

- Orders complete meals in restaurants 3
- Orders simple meals like hamburgers or hot dogs 2
- Orders single items, e.g., soft drinks, ice cream, donuts, etc. at soda fountain or canteen 1
- Does not order in public eating places 0 ☐

ITEM 3 — **Drinking** (Circle highest level)

- Drinks without spilling, holding glass in one hand 3
- Drinks from cup or glass unassisted—neatly 2
- Drinks from cup or glass unassisted—considerable spilling 1
- Does not drink from cup or glass unassisted 0 ☐

ITEM 4 — **Table Manners** (Circle all answers)

If these items do not apply to the individual, e.g., because he or she is bedfast and/or has liquid food only, place a check in the blank and mark "Yes" for all statements. ____

	Yes	No
Throws food	0	1
Swallows food without chewing	0	1
Chews food with mouth open	0	1
Drops food on table or floor	0	1
Does not use napkin	0	1
Talks with mouth full	0	1
Takes food off others' plates	0	1
Eats too fast or too slow	0	1
Plays in food with fingers	0	1

B. Toilet Use

ITEM 5 — **Toilet Training** (Circle highest level)

- Never has toilet accidents 4
- Has toilet accidents only at night 3
- Occasionally has toilet accidents during the day 2
- Frequently has toilet accidents during the day 1
- Is not toilet trained at all 0 ☐

ITEM 6 — **Self-Care at Toilet** (Circle all answers)

	Yes	No
Lowers pants at the toilet without help	1	0
Sits on toilet seat without help	1	0
Uses toilet tissue appropriately	1	0
Flushes toilet after use	1	0
Puts on clothes without help	1	0
Washes hands without help	1	0 ☐

C. Cleanliness

ITEM 7 — **Washing Hands and Face** (Circle all answers)

	Yes	No
Washes hands and face with soap and water without prompting	1	0
Washes hands with soap	1	0
Washes face with soap	1	0
Washes hands and face with water	1	0
Dries hands and face	1	0 ☐

ITEM 8 — **Bathing** (Circle highest level)

- Prepares and completes bathing unaided 6
- Washes and dries self completely without prompting or helping 5
- Washes and dries self reasonably well with prompting 4
- Washes and dries self with help 3
- Attempts to soap and wash self 2
- Cooperates when being washed and dried by others 1
- Makes no attempt to wash or dry self 0 ☐

ITEM 9 — **Personal Hygiene** (Circle all answers)

If these items do not apply to the individual, e.g., because he or she is completely dependent on others, place a check in the blank and mark "Yes" for all statements. ____

	Yes	No
Has strong underarm odor	0	1
Does not change underwear regularly by self	0	1
Skin is often dirty if not assisted	0	1
Does not keep nails clean by self	0	1 ☐

ITEM 10 — **Toothbrushing** (Circle highest level)

- Cleans dentures appropriately 5
- Applies toothpaste and brushes teeth with up and down motion 5
- Applies toothpaste and brushes teeth with sideways motion 4
- Brushes teeth without help, but cannot apply toothpaste 3
- Brushes teeth with supervision 2
- Cooperates in having teeth brushed 1
- Makes no attempt to brush teeth 0 ☐
- Does not clean dentures 0

D. Appearance

ITEM 11 — **Posture** (Circle all answers)

If these items do not apply to the individual, e.g., because he or she is bedfast or non-ambulatory, place check in the blank and mark "Yes" for all statements. ____

	Yes	No
Mouth hangs open	0	1
Head hangs down	0	1
Stomach sticks out because of posture	0	1
Shoulders slumped forward and back bent	0	1
Walks with toes out or toes in	0	1
Walks with feet far apart	0	1
Shuffles, drags, or stamps feet when walking	0	1
Walks on tiptoe	0	1 ☐

Appendix E: Memory and Orientation

B MEMORY AND ORIENTATION

Memory

42	a)	Does he/she have difficulty remembering recent events e.g. when he/she last saw you or what happened the day before? *Examples of change:*	Yes 1 No 0 DK 8 N/A 9	→	b)	Is this a deterioration?	Yes No 0 DK 8 N/A 9	→	Slight deterioration 1 Great deterioration 2	
43	a)	Does he/she often have difficulty remembering where he/she has left things? *Examples of change:*	Yes 1 No 0 DK 8 N/A 9	→	b)	Is this a deterioration?	Yes No 0 DK 8 N/A 9	→	Slight deterioration 1 Great deterioration 2	
44	a)	Does he/she have difficulty remembering what has been said and repeat the same question over and over? *Examples of change:*	Yes 1 No 0 DK 8 N/A 9	→	b)	Is this a deterioration?	Yes No 0 DK 8 N/A 9	→	Slight deterioration 1 Great deterioration 2	
45	a)	Does he/she have difficulty in remembering short lists of items, e.g. shopping? *Examples of change:*	Yes 1 No 0 DK 8 N/A 9	→	b)	Is this a deterioration?	Yes No 0 DK 8 N/A 9	→	Slight deterioration 1 Great deterioration 2	
46	a)	Does he/she have difficulty remembering significant events from his/her past? *Examples of change:*	Yes 1 No 0 DK 8 N/A 9	→	b)	Is this a deterioration?	Yes No 0 DK 8 N/A 9	→	Slight deterioration 1 Great deterioration 2	

Appendix F: Case Studies

Case Study 1: John

Background/Best Level of Functioning

- John is 54 years old and lives in a group home with eight other residents.
- Has attended a day center for the last 10 years and used to help in the kitchen at lunchtimes.
- He is active socially, attending church, a social club, and going to the pub on a weekly basis.
- He has a group of friends and a best friend.
- He likes to watch Startrek on the TV and enjoys coloring.
- He never learned to read and has only ever been able to copy his name.
- At his best level of functioning, he was able to prepare meals and carry out housework independently, and took care of grooming, dressing, and personal hygiene without assistance.

Changes Reported by Carer

Everyday Skills

- He used to help in the kitchen at his day center but is unable to do this anymore because he gets muddled.
- He now needs much more prompting to perform everyday household chores and needs reminding how to do things.
- He used to be able to cook independently but only helps out now and can only prepare simple snacks independently, e.g., toast.
- He needs prompting at each stage to make a cup of tea. This is a big change.

- He used to go shopping in a group but now requires one-to-one supervision as he needs prompting as to what to buy, and will pick up items that he does not need.
- These changes have occurred gradually over the last **6 months**.

Other Cognitive Skills

- He finds it much more difficult to concentrate on things. He needs prompting halfway through activities.
- His thinking processes seem to have slowed down and his thinking seems muddled.
- He has word finding difficulties and often muddles words up that he used to know, using the wrong word.
- He has difficulty following instructions, particularly if too many instructions are given at once, and needs prompting at each stage.
- He has difficulty completing complex sequences of actions, e.g., household chores, cooking, dressing.
- He has difficulty planning ahead. He used to pack his bag for day center the night before but no longer thinks ahead and bag is packed for him.
- He has difficulty making decisions, e.g., what he would like to eat for dinner. He just says "don't know" or "don't mind" when given a choice.
- These changes have occurred gradually over the last **12 months**.

Personality/Behavior

- He shows inappropriate behavior in public, e.g., shaking hands with strangers. This is a change from usual behavior, since he used to be quite shy.
- He shows less concern for others, e.g., not noticing when other residents are unwell.
- Has become more stubborn, awkward, and uncooperative, particularly when reminded to do things he used to be able to do.
- He has started to repeat the same phrase over and over again.
- Gradual changes over last **12 months**.

Memory

- He has much greater difficulty remembering recent events, e.g., if asked what he did yesterday, he wouldn't remember that he went swimming. This is a major change.
- He forgets what has been said and repeats the same question over and over again. This has become much more noticeable recently.
- He has great difficulty in remembering short lists. He used to remember a short list of items to buy at the shop but can no longer do so.

- He often loses his glasses or other personal items. This is a slight change from previous behavior.
- He has become confused about what day it is. He used to remember which day he went to his social club but doesn't anymore.
- Sometimes he thinks it is morning in the middle of the night (for the last 3–4 months).
- He is aware of his memory problem and often frustrated by it.
- These changes have happened gradually over the last **12 months**, escalating more rapidly over last **6 months**.

Self-Care

- He has difficulty dressing, gets things in the wrong sequence and often forget items. He now requires a moderate degree of assistance but used to be completely independent.
- He now wets and soils himself. Always used to get to toilet on time.
- He needs much more prompting when combing hair and shaving. Moderate assistance required now but used to be independent.
- He needs to be washed, as can now only wash hands and face. He used to be independent in the bath.
- Gradual changes over **6 months**.

Mental Health

- He talks more slowly than he used to.
- No other signs of depression or other mental illness.

Physical Health

- No sensory difficulties.
- No other physical health problems.
- Doesn't take any medication.

Case Study 2: Michael

Background/Best Level of Functioning

- Peter is 52 years old and lives in supported housing with one housemate.
- He attended special school until the age of 15 and since then has held a number of jobs including working in a shoe factory, making tea and washing up at a

residential home for older people, cleaning at a pub, and a work placement at a furniture project. He had also received training in essential skills at college.
- He has never been able to read. He is able to write his name and copy.
- He has a very active social life, attending a drama group, a social club, and going to the pub at least once a week. He has many hobbies, listening to music, snooker, darts, dominos, gardening, and DIY. He has many friends without learning disabilities.
- At his best level of functioning he has been able to shop independently for small purchases, perform light housework tasks, prepare snacks, and heat up meals. He is independent in grooming, dressing, and personal hygiene.

Changes Reported by Carer

Everyday Skills

- He requires more prompting to carry out his activities at work and on placements. He has less confidence in his abilities and has become very sensitive about errors he makes.
- He requests more supervision/prompting to perform everyday household chores, which he used to do independently, and in his hobbies—gardening and DIY.
- These changes have occurred gradually over the last **18 months**.
- He is able to shop independently for small purchases, dial a few well-known numbers on the telephone, prepare snacks, and heat things up in the microwave. There is no decline in these abilities.

Memory

- There has been no change in his ability to remember recent events.
- He asks the same question again but only when he doesn't understand the answer!
- Has never been able to remember lists.

Other Cognitive Skills

- He finds it much more difficult to concentrate on things and gets easily distracted. He needs much more prompting for things he used to do independently.
- His thinking appears to be slower and more muddled than it used to be.
- He has difficulty making decisions and changes his mind a lot. He did not used to do this.

- He is less confident about making decisions/solving problems by himself—he used to do gardening, and DIY by himself but now asks for more supervision.
- These changes have occurred gradually over the last **18 months**.

Personality/Behavior

- He has become much more sensitive, e.g., if he does something wrong he has tearful moments—this has been noticed at his place of work (on a furniture project).
- He has become more changeable in mood.
- He has become more irritable and angry, particularly toward his housemate.
- He shows less concern for others' feelings.
- He has become more stubborn. Won't be hurried into doing things.
- He no longer enjoys colouring or cross-stitch (which he now says is a woman's activity!).
- He repeats the same phrase over again "Where you been?" (6 months)
- Gradual changes over last **18 months**.

Self-Care

- He has no difficulty in dressing, but requires prompting to change his clothes.
- There has been no change in grooming or other self-care skills

Mental Health

- Sometime he cries for no apparent reason. This is a change.
- He finds it more difficult to concentrate that usual for him.
- Gradual change over **18 months**.
- NB: He is currently on antidepressant medication.
- He complains unjustifiably of being picked on, but this has always been the case.
- Has always talked to himself and seen visions of animals such as rats. He does not believe these are real.

Physical Health

- He has a hearing problem, but this is corrected with a hearing aid.
- He has no other physical health problems and doesn't take any medication.

Case Study 3: Mary

Background/Best Level of Functioning

- Mary is 51 years old and lives in a residential home with five other residents (has been there for last 3 years).
- She did not attend school and entered institutional care at age 10. She is severely learning disabled and has only ever had very limited speech. She has never been able to read and write. She has attended college for a sensory music and movement group in the past.
- She is not involved in many activities outside the home. She occasionally goes shopping—in a wheelchair. She has always been "in her own little world." However, she enjoys singing and has a best friend who she lives with.
- At her best level of functioning she was able to use the toilet independently and walk independently. She was not able to perform any household tasks, and has been dependent on others for major assistance with dressing, bathing, and grooming.

Changes Reported by Carer

Everyday Skills

- She used to love singing and knew the words to songs. Does not know the words anymore and sings less often.
- She no longer attends the sensory, music, and movement course due to difficulties attending. She used to go on the bus but now has problems with mobility and takes no more than a few steps, supported by another person.
- Change has occurred gradually over the last **12 months**.
- She has never been able to perform any household tasks.

Memory

- Her memory has always been poor. Also, it is very difficult to judge how much she remembers, due to limited speech. She has never used people's names.
- However, she has much greater difficulty knowing where she is/interpreting surroundings—she used to recognize when she was in the bathroom, but now she does not.
- She has much greater difficulty finding her way round the home.
- She forgets what time of day it is and gets up during the night.
- These changes have been gradual over the last **6 months**.

Other Cognitive Skills

- Her thinking seems to have slowed down though it has always been muddled.
- She talks much less than she used to.
- These changes have occurred gradually over the last **12 months**.

Personality/Behavior

- She has become much quieter and more withdrawn.
- She has also become much more stubborn, e.g., refuses to go in the bath.
- She has lost interest in things going on around her and does not show as much emotion as she used to.
- Gradual changes over last **12 months**.

Self-Care

- She used to be able to feed herself independently, but now needs to be fed.
- She now wets herself quite often (more than once a week). This did not used to happen.
- These changes have occurred gradually over the last **6 months**.

Mental Health

- She shows a lack of interest in things in general.
- She has lost her appetite and lost weight in the last 6 months.
- She is less sociable than she used to be.
- She sleeps a lot by day.
- Gradual change over **12 months**.
- No other signs of depression or other mental illness.

Physical Health

- She has cataracts.
- She has suffered from seizures for the last 2 years.
- She is taking antiepileptic medication.

Appendix G: Test for Severe Impairment

NAME_____ ID Number_____
DATE_____ AGE _____

Write down all responses verbatim that are different from those on the sheet. If ***S*** does not hear a question or is distracted, you may repeat the question up to three times in order to engage their attention.

Motor Performance

Comb
"Show me how you would use this comb"
Hand **S** comb
Correctly demonstrates combing 1___
Pen and Top
"Can you put the top on the pen?"
Remove the top from the pen in full view of **S**
Hand the pen and top to **S**
Correctly puts top on pen (not on bottom of pen) 1___
Pen and Paper
"Write you name"
Hand **S** pen (without the top) and place paper on table in front of **S**
Writes name correctly (first or last name legible) 1___
TOTAL 3 ___

Language—Comprehension

"Point to your ear"
Correctly points to ear 1___
"Close you eyes"
Correctly closes eyes 1___
Pens—Red, Blue, Green
"Show me the red pen,… the green pen"
Place the three pens on the table spread out so that they have some space between them
Correctly points to red pen 1___
Correctly points to green pen 1___
Total 4 ___

Language—Production

"What is this called"
Point to your nose
Correctly names nose 1___
Pens—Red, Green
"What colour is this pen"
One at a time hold up a (red, green) pen in front of ***S***
Correctly names red 1___
Correctly names green 1___
Key
"What is this called"
Show ***S*** the key
Correctly names key ___

Total 4 ___

Memory—Immediate

One large paperclip
"Watch carefully"
Place clip in your hand so ***S*** can see
Hold hands out to ***S***
With hands open
"Which hand is the clip in?"
Correctly points to clip 1 ___
With hands closed
"Which hand is the clip in?"
Correctly point to hand with clip 1 ___
Move hands behind back
"Watch carefully. Which hand/side is the clip in/on?"
Correctly points to hand with clip 1___

Total 3 ___

General Knowledge

"How many ears do I have?"
Correctly states two 1___
"Count my fingers and thumbs"
Place hands in front of ***S*** with fingers pointing up, palms toward ***S***. Credit given even if no 1 to 1 correspondence between fingers and numbers. If ***S*** only gives final answer, ask, "Can you count to 10 starting at 1?"
Correctly counts to 10 1___
"How many weeks are there in a year?"
Correctly states 52 1___

"I'm going to sing a song. If you know the words
I want you to sing along with me." Softly sing "Happy Birthday."
Sings most of the words 1___

Total 4 ___

Conceptualization

Two large paperclips, one pen
"Which one of these is different form the other two"
Spread objects on the table.
Correctly points to pen or states pen 1___
Two red pens, one green pen
"Put this next to the pen that is the same colour"
Place 1 red and 1 green pen spread out on the table
Hand **S** the other red pen
Correctly places the red pen next to the other red pen 1___
One large paperclip
Place hands out in front of **S**
Alternate the clip between the opens hands 4 times
"Watch me move the paperclip
Which hand will I put the clip in next?" 1___
After **S** responds, place clip in correct hand
If **S** is incorrect say "I'd put the clip in this hand"
Then say, "Which hand will I put it in next?"
Correctly points to correct hand 1___

Total 4 ___

Memory—Delayed

Thread, key, paperclip
"Which one of these haven't we done something with
while you were here with me?"
Place objects spread out on table
Correctly points to thread 1___

Total 1 ___

Motor Performance

"Thank you for spending time with me"
Extend hand to shake hands
Correctly shakes hands 1___
Total 1 ___

Tsi Total Score = 24 ___

Appendix H: The Adaptive Behavior Dementia Questionnaire (ABDQ)

Name:_____ Date of Birth:_____

	Question		Answer		
1	Are they able to dress themselves?	Better than normal	Same as normal	Worse than normal	Much worse than normal
2	Can they use their hands to do things?	Better than normal	Same as normal	Worse than normal	Much worse than normal
3	Is their ability to buy things?	Better than normal	Same as normal	Worse than normal	Much worse than normal
4	Are they able to have a conversation?	Better than normal	Same as normal	Worse than normal	Much worse than normal
5	Is their awareness of time?	Better than normal	Same as normal	Worse than normal	Much worse than normal
6	Do they help to prepare food?	More than normal	Same as normal	Less than normal	Much less than normal
7	Do they help to clear the table?	More than normal	Same as normal	Less than normal	Much less than normal
8	Are they able to perform simple jobs?	Better than normal	Same as normal	Worse than normal	Much worse than normal
9	Can they initiate things/ activities?	More than normal	Same as normal	Less than normal	Much less than normal
10	Is their ability to persist in doing things?	Better than normal	Same as normal	Worse than normal	Much worse than normal
11	Can they take care of their belongings?	Better than normal	Same as normal	Worse than normal	Much worse than normal
12	Do they cooperate with requests?	More than normal	Same as normal	Less than normal	Much less than normal
13	Do they carry out simple commands?	Better than normal	Same as normal	Worse than normal	Much worse than normal
14	Do they participate in group activities?	More than normal	Same as normal	Less than normal	Much less than normal
15	Is their ability to do things independently?	Better than normal	Same as normal	Worse than normal	Much worse than normal

Appendix I: NTG-EDSD

NTG-EDSD

v.1/2013.2

The **NTG-Early Detection Screen for Dementia**, adapted from the DSQIID*, can be used for the early detection screening of those adults with an intellectual disability who are suspected of or may be showing early signs of mild cognitive impairment or dementia. The NTG-EDSD is not an assessment or diagnostic instrument, but an administrative screen that can be used by staff and family caregivers to note functional decline and health problems and record information useful for further assessment, as well as to serve as part of the mandatory cognitive assessment review that is part of the Affordable Care Act's annual wellness visit for Medicare recipients. This instrument complies with Action 2.B of the US National Plan to Address Alzheimer's Disease.

It is recommended that this instrument be used on an annual or as indicated basis with adults with Down syndrome beginning with age 40, and with other at-risk persons with intellectual or developmental disabilities when suspected of experiencing cognitive change. The form can be completed by anyone who is familiar with the adult (that is, has known him or her for over six months), such as a family member, agency support worker, or a behavioral or health specialist using information derived by observation or from the adult's personal record.

The estimated time necessary to complete this form is between 15 and 60 minutes. Some information can be drawn from the individual's medical/health record. Consult the NTG-EDSD Manual for additional instructions (www.aadmd.org/ntg/ screening).

---

(1) File #: _____ (2) Date: _____

Name of person: (3) First_____ (4) Last: _____

(5) Date of birth: _____ (6) Age: _____

(7) Sex:

	Female
	Male

Instructions:
For each question block, check the item that best applies to the individual or situation.

(8) Best description of level of intellectual disability

	No discernible intellectual disability
	Borderline (IQ 70-75)
	Mild ID (IQ 55-69)
	Moderate ID (IQ 40-54)
	Severe ID (IQ 25-39)
	Profound ID (IQ 24 and below)
	Unknown

(9) Diagnosed condition (*check all that apply*)

	Autism
	Cerebral palsy
	Down syndrome
	Fragile X syndrome
	Intellectual disability
	Prader-Willi syndrome
	Other:

Current living arrangement of person:
- □ Lives alone
- □ Lives with spouse or friends
- □ Lives with parents or other family members
- □ Lives with paid caregiver
- □ Lives in community group home, apartment, supervised housing, etc.
- □ Lives in senior housing
- □ Lives in congregate residential setting
- □ Lives in long term care facility
- □ Lives in other: _____

(10) General characterization of current physical health:

	Excellent
	Very good
	Good
	Fair
	Poor

(11) Compared to one year ago, current physical health is:

	Much better
	Somewhat better
	About the same
	Somewhat worse
	Much worse

(12) Compared to one year ago, current mental health is:

	Much better
	Somewhat better
	About the same
	Somewhat worse
	Much worse

(13) Conditions present (*check all that apply*)

	Vision impairment
	Blind (very limited or no vision)
	Vision corrected by glasses
	Hearing impairment
	Deaf (very limited or no hearing)
	Hearing corrected by hearing aids
	Mobility impairment
	Not mobile – uses wheelchair
	Not mobile – is moved about in wheelchair

(14) Significant recent [in past year] life event (*check all that apply*)

	Death of someone close
	Changes in living arrangement, work, or day program
	Changes in staff close to the person
	New roommate/housemates
	Illness or impairment due to accident
	Adverse reaction to medication or over-medication
	Interpersonal conflicts
	Victimization / abuse
	Other:

(15) Seizures

	Recent onset seizures
	Long term occurrence of seizures
	Seizures in childhood, not occurring in adulthood
	No history of seizures

If MCI or dementia is documented complete 16, 17, &18

(16) **Diagnostic History**

Mild cognitive impairment [MCI] or dementia previously diagnosed (Dx)?:

[ ] No

[ ] Yes, MCI

Date of Dx:

[ ] Yes, dementia

Date of Dx:

Type of dementia:

Diagnosed by:
□ Geriatrician
□ Neurologist
□ Physician
□ Psychiatrist
□ Psychologist
□ Other:

(17) Reported date of onset of MCI/dementia
[When suspicion of dementia first arose]
Note approximate year and month:

(18) Comments / explanations about dementia suspicions:

[Check column option as appropriate]

	Always been the case	Always but worse	New symptom in past year	Does not apply
[19]**Activities of Daily Living**				
Needs help with washing and/or bathing				
Needs help with dressing				
Dresses inappropriately (e.g., back to front, incomplete, inadequately for weather)				
Undresses inappropriately (e.g., in public)				
Needs help eating (cutting food, mouthful amounts, choking)				
Needs help using the bathroom (finding, toileting)				
Incontinent (including occasional accidents)				
[20]**Language & Communication**				
Does not initiate conversation				
Does not find words				
Does not follow simple instructions				
Appears to get lost in middle of conversation				
Does not read				
Does not write (including printing own name)				
[21]**Sleep-Wake Change Patterns**				
Excessive sleep (sleeping more)				
Inadequate sleep (sleeping less)				
Wakes frequently at night				
Confused at night				
Sleeps during the day more than usual				
Wanders at night				
Wakes earlier than usual				
Sleeps later than usual				
[22]**Ambulation**				
Not confident walking over small cracks, lines on the ground, patterned flooring, or uneven surfaces				
Unsteady walk, loses balance				
Falls				
Requires aids to walk				

	Always been the case	Always but worse	New symptom in past year	Does not apply
[23]**Memory**				
Does not recognize familiar persons (staff/relatives/friends)				
Does not remember names of familiar people				
Does not remember recent events (in past week or less)				
Does not find way in familiar surroundings				
Loses track of time (time of day, day of the week, seasons)				
Loses or misplaces objects				
Puts familiar things in wrong places				
Problems with printing or signing own name				
Problems with learning new tasks or names of new people				
[24]**Behavior and Affect**				
Wanders				
Withdraws from social activities				
Withdraws from people				
Loss of interest in hobbies and activities				
Seems to go into own world				
Obsessive or repetitive behavior				
Hides or hoards objects				
Does not know what to do with familiar objects				
Increased impulsivity (touching others, arguing, taking things)				
Appears uncertain, lacks confidence				
Appears anxious, agitated, or nervous				
Appears depressed				
Shows verbal aggression				
Shows physical aggression				
Temper tantrums, uncontrollable crying, shouting				
Shows lethargy or listlessness				
Talks to self				
[25] **Adult's Self-reported Problems**				
Changes in ability to do things				
Hearing things				
Seeing things				
Changes in 'thinking'				
Changes in interests				
Changes in memory				
[26]**Notable Significant Changes Observed by Others**				
In gait (e.g., stumbling, falling, unsteadiness)				
In personality (e.g., subdued when was outgoing)				
In friendliness (e.g., now socially unresponsive)				
In attentiveness (e.g., misses cues, distracted)				
In weight (e.g., weight loss or weight gain)				
In abnormal voluntary movements (head, neck, limbs, trunk)				

[Check column option as appropriate]

	[27]Chronic Health Conditions*	Recent condition (past year)	Condition diagnosed in last 5 years	Lifelong condition	Condition not present
	Bone, Joint and Muscle				
1	Arthritis				
2	Osteoporosis				
	Heart and Circulation				
3	Heart condition				
4	High cholesterol				
5	High blood pressure				
6	Low blood pressure				
7	Stroke				
	Hormonal				
8	Diabetes (type 1 or 2)				
9	Thyroid disorder				
	Lungs/breathing				
10	Asthma				
11	Chronic bronchitis, emphysema				
12	Sleep disorder				
	Mental health				
13	Alcohol or substance abuse				
14	Anxiety disorder				
15	Attention deficit disorder				
16	Bipolar disorder				
17	Dementia/Alzheimer's disease				
18	Depression				
19	Eating disorder (anorexia, bulimia)				
20	Obsessive-compulsive disorder				
21	Schizophrenia				
22	Other:				
	Pain / Discomfort				
23	Back pain				
24	Constipation				
25	Foot pain				
26	Gastrointestinal pain or discomfort				
27	Headaches				
28	Hip/knee pain				
29	Neck/shoulder pain				
	Sensory				
30	Dizziness / vertigo				
31	Impaired hearing				
32	Impaired vision				
	Other				
33	Cancer – type:				
34	Chronic fatigue				
35	Epilepsy / seizure disorder				
36	Heartburn / acid reflux				
37	Urinary incontinence				
38	Sleep apnea				
39	Tics/movement disorder/spasticity				
40	Dental pain				

*Items drawn from the Longitudinal Health and Intellectual Disability Survey (University of Illinois at Chicago)

(28) **Current Medications**

Yes	No	Indicate type
□	□	Treatment of chronic conditions
□	□	Treatment of mental health disorders or behavior problems
□	□	Treatment of pain

For reviews, attach list of current medications, dosage, and when prescribed

□ List is attached for reviews

(29) **Comments related to other notable changes or concerns:**

(30) **Next Steps / Recommendations**

□ Refer to treating physician for assessment

□ Review internally by clinical personnel

□ Include in annual review / annual wellness visit

□ Repeat in _____ months

Form completion information

(31) Date completed	(32) Organization / Agency
Name of person completing form	
Relationship to individual (staff, relative, assessor, etc.)	
Date(s) form previously completed	

Acknowledgement: Derived from the DSQIID (*Dementia Screening Questionnaire for Individuals with Intellectual Disabilities; Deb, S., 2007) as adapted into the Southeast PA Dementia Screening Tool (DST) – with the assistance of Carl V. Tyler, Jr., MD – and the LHIDS (Longitudinal Health and Intellectual Disability Survey; Rimmer & Hsieh, 2010) and as further adapted by the National Task Group on Intellectual Disabilities and Dementia Practices as the NTG Early Detection Screen for Dementia for use in the USA.

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Dr. Arthur Dalton Obituary [1]

Arthur J. Dalton (1937–2013)

Dr. Arthur Dalton, 76, died June 17, 2013, after a brief illness. He was born in 1937 in Alberta, Canada, and spent his early childhood in Edmonton. Dr. Dalton received his undergraduate and graduate degrees in psychology from McMaster University in Hamilton, Ontario, Canada. His Ph.D. was in behavioral psychology. Prior to his retirement from the New York State Office for People with Developmental Disabilities, he was the Director of the Center for Aging Studies at the New York State Institute for Basic Research in Developmental Disabilities (IBR/DD) on Staten Island, New York.

Dr. Dalton had been involved in research on Alzheimer's disease for more than 25 years, focusing on the connection with Down syndrome, the development of behavioral and biological markers, and treatment methods. Before moving to New York in 1990, he was based at Surrey Place Centre in Toronto, Ontario, and held an appointment at the University of Toronto's Department of Physiology. In Toronto, he pioneered the study of aging and Alzheimer dementia in people with Down syndrome. He established a mobile dementia clinic for adults with Down syndrome, and set a precedent for monitoring individuals longitudinally to check for signs of neurocognitive or neurobehavioral decline, with each person serving as his or her own control.

Dr. Dalton's ability to develop dementia screening instruments has had great impact in the Alzheimer's disease field generally, as well as specifically with Down syndrome. While at Surrey Place, Dr. Dalton developed the Dyspraxia Scale for Adults with Down Syndrome, which has gained international use in clinical screening for early signs of dementia. Between 1985 and 1991, he was the co-principal investigator, along with Drs. Donald McLachlan and Theo Kruck, of a Surrey Place Centre—University of Toronto phase II clinical trial of desferrioxamine, given by intramuscular injection, in people with moderate Alzheimer's disease. While in Toronto, Dr. Dalton was the visionary behind a plan to develop an institute dedicated to dementia research. With Dr. McLachlan, his plan took root and bore fruit in the form of the internationally renowned University of Toronto Tanz Centre for Research in Neurodegenerative Diseases. When in Toronto, Dr. Dalton also gave much to the community, including being one of the founders of the Alzheimer Society of Canada, of which he was a President.

Dr. Dalton's impact in the field of intellectual disabilities continued after relocating to New York. As well as conducting administrative and research activities at IBR/DD, he served for several years on the board of directors for the National Down Syndrome Congress. He was the author of numerous journal articles and book chapters and was the co-editor of a seminal text "Aging, Dementia, and Intellectual Disabilities: A Handbook." In his most recent activity, he served as the principal investigator (along with Drs. Paul Aisen and Mary Sano) of "Vitamin E in Aging Persons with Down Syndrome," a National Institutes of Health-funded multi-year international, multicenter, randomized, double-blind, and placebo controlled clinical trial designed to determine whether the administration of vitamin E would slow the rate of cognitive and functional decline in older adults with Down syndrome.

Since the mid-1990s, he directed the Center for Aging Studies at the New York State Institute for Basic Research in Developmental Disabilities (IBR/DD), which is affiliated with the New York State Office for People with Developmental Disabilities. The Center was charged with the development, conduct, analysis, and reporting of multidisciplinary research into age-associated conditions affecting older individuals with intellectual and/or developmental disabilities. Among its activities, the Center was involved in the development of tests of cognitive abilities to evaluate changes associated with the onset of dementia, collaborated with universities with respect to evaluating residential settings for persons with dementia and developmental disabilities, assessed the nature of dementia care procedures, and conducted surveys of the prevalence of dementia diagnoses in New York State facilities that provided services for older adults with developmental disabilities.

Dr. Dalton was married to Alexandra Kopinets and lived in Mahopac, New York. He had many hobbies, but his love for astronomy was most prominent. He enjoyed travel and had many colleagues across the world. He was an accomplished researcher, author, and educator—and most significantly a strong advocate for people with Down syndrome.

Reference

1. https://aadmd.org/articles/dr-arthur-dalton-obituary. *Written by* Matthew Janicki, Ph.D. NTG Co-chair.

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V.P. Prasher (ed.), *Neuropsychological Assessments of Dementia in Down Syndrome and Intellectual Disabilities*, DOI 10.1007/978-3-319-61720-6