

Assessment and Diagnosis of Dementia in Individuals with Intellectual Disability: A Toolkit for Clinicians and Caseworkers

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Table of Contents

Introduction

Why is this important?

What is Dementia?

What is Alzheimer's Disease?

What is Intellectual Disability?

What is Down Syndrome?

Risk Factors for Alzheimer's Disease Among People With Developmental Disability

Common Signs of Dementia in ID

Diagnostic Challenges

Is There a Test for Alzheimer's Disease?

Diagnostic Criteria

Clinical Manifestations

Depression and Life Stress

Assessment-A Baseline Comparison Approach

Assessment Measures

Observer Rated Scales

Direct Neuropsychological Measures

Adaptive Behavior Scales

Adjustment, Emotional Functioning and Psychopathology

Summary

References and Resources

Introduction:

Individuals with intellectual disability (ID) are living longer and, as such, are at increased risk for age-related health conditions including dementia. Detecting dementia in intellectual disability populations is difficult due to their premorbid cognitive deficits, the tendency for dementia to be expressed differently in adults with ID and their susceptibility to other medical problems that can mimic dementia. Standard assessment measures used to detect dementia in the general population are inappropriate for use with this population as many ID adults lack the skills to perform these tasks at baseline. As such there is a strong need for enhanced diagnostic processes and assessment resources to improve diagnostic accuracy.

This tool kit was developed for clinicians and caseworkers who are concerned about the presence of dementia in their patients and clients with ID. It describes the clinical features of dementia in adults with ID, the diagnostic challenges faced in the clinical evaluation of dementia in this population, and the modifications to methods and measures needed to enhance accuracy of clinical diagnosis of dementia in ID. A number of neuropsychological assessment measures and methods that have been developed or adapted for use with ID adults are then reviewed. It is hoped that this resource will provide clinicians and caseworkers with the basic tools and technical information to recognize signs and symptoms of dementia in ID, and conduct good quality diagnostic assessments that ultimately lead to prompt and appropriate medical and care management.

Why is this important?

Individuals with ID are living longer. As they age they are at increased risk for age-related conditions such as dementia. As the disease progresses the need for support and supervision increases placing greater demands on a families, carers and support systems. With the rising life expectancy and growing population of individuals with ID, clinicians and caseworkers can expect to encounter increasing numbers of persons with ID who develop dementia. Development of specialist assessment skills in this area will be important to meet this growing demand.

What is Dementia?

According to the ICD-10 diagnosis criteria dementia refers to a collection of diseases of the brain that lead to deterioration in memory and thinking and ability to care for one's self. Loss of these abilities may be preceded or accompanied by changes in personality, mood, motivation or social behavior.

What is Alzheimer's disease?

Alzheimer's disease is the most common form of dementia. It is characterized by two signature brain changes: development of neuritic amyloid plaques and neurofibrillary tau tangles. Over time these pathological processes contribute to deterioration of synaptic networks, neuronal loss and gross brain atrophy, and ultimately, progressive loss of cognitive and functional abilities but with considerable variability in their clinical and behavioral manifestation.

Early symptoms of Alzheimer's disease in the general population often include:

- Language problems – difficulty with wording finding, coming up with names of familiar people, places or objects
- Short-term memory loss – forgetfulness for conversations and recent events; recall of remote events in the distant past is preserved
- Disorientation – confusion about date and day and how to get to familiar places
- Decline in activities of daily living
- Subtle personality changes – individuals may become quiet and withdrawn, increasingly restless, irritable and moody. The affected individual may become easily angered over little things.

In the early or onset stage of Alzheimer Disease (AD), symptoms often appear gradually and may at first be difficult to distinguish from normal aging. Often there is subjective awareness of subtle changes such as lapses in memory or word-finding difficulty but the individual may still be able to function independently and continue to drive, work and engage in social activity (although greater than normal memory or cognitive changes may be documented on in-depth neuropsychological evaluation). Eventually, persons with AD begin to experience short term memory loss, language problems and disorientation. The affected individual begins to experience impairment in functioning, particularly when attempting to perform tasks in social or work settings. As awareness of losses increases the affected person may react with anxiety, denial or compensation. Subtle personality changes emerge and affected persons may cope by becoming quiet and withdrawn, restless or irritable and short tempered, or defensive. Changes in the first or onset phase last for approximately five years.

The middle or progressive stage is marked by pronounced cognitive losses and functional declines. Problems with language abilities become more apparent. This is typically the most obvious sign of progression into this stage. Ability to name objects and maintain a logical conversation becomes affected along with ability to comprehend directions and instructions. There is frequent disorientation and confusion to time and place and people around them. Memory loss becomes more pronounced. Confusion and resulting frustration become evident. A loss of self-care skills starts to emerge including the ability to toilet one's self. Severe changes in personality may become obvious, and social behavior may be marked by suspiciousness and delusions. Changes in this stage evolve over approximately 12 years' time.

In the third or terminal stage, there is substantial impairment eventually leading to complete loss of function. Ability to recognize other persons and their environment is lost. Long term (remote) memory erodes. Basic skills, such as eating and drinking, deteriorate and substantial weight loss ensues. Imbalance, rigidity and weakness may lead to recurrent falls. With increasing immobility, affected individuals become effectively bedridden and inactive. At this stage, 24 hour care and total assistance is required. The terminal stage can often last up to 3-4 years. Typically death results from health complications such as infection, especially pneumonia.

Signs and Symptoms of Alzheimer’s Disease in the General Population

| EARLY STAGE | MIDDLE STAGE | LATE STAGE |
|---|--|---|
| <ul style="list-style-type: none"> • forgetfulness – recent memory loss • Cognitive decline • Subjective awareness of cognitive changes • Gradually evolving vocational/social dysfunction • Difficult to distinguish from normal aging • No distinct physical presentation | <ul style="list-style-type: none"> • Distinct problems with language • Pronounced loss of memory and cognitive abilities • Frequent confusion and disorientation • Loss of self-care skills • Personality and behavior change | <ul style="list-style-type: none"> • Global disorientation and confusion • Remote memory loss • Disordered and fragmented speech • Basic skills forgotten • Incontinence • Weight loss • General physical deterioration • Parkinsonian features • Immobility, rigidity and frequent falls • Totally dependent |

Approximately 5.3 million Americans have Alzheimer’s disease, 200,000 of which have early onset. By the year 2025 that number is expected to grow to 7.1 million and to nearly double to 13.8 by 2050. The average age of onset to Alzheimer’s disease is 75 years, and the average duration is 8-10 years with a range of anywhere from 3 to 20+ years, depending on the age of onset and the presence of other health conditions. Two-thirds of those affected by Alzheimer Disease are women. Dementia rates for the general population are listed below.

Dementia Rates in the General Population, Brookmeyer et al (2011)

| AGE | ALL DEMENTIA | ALZHEIMER’S DISEASE |
|-------|--------------|---------------------|
| 71-79 | 4.97 | 2.32 |
| 80-89 | 24.19 | 18.10 |
| 90+ | 37.20 | 29.6 |
| Total | 13.67 | 9.51 |

What is Intellectual Disability (ID)?

Mental retardation (ID) is a developmental disability characterized by significant sub-average general intellectual function with concurrent impairment in age appropriate adaptive functioning that occurs before the age of 22 and continues into old age.

What is Down Syndrome?

Down syndrome is the most common genetic cause of intellectual disability accounting for 1/691 live births each year in the USA. JL Down (1866) first described the syndrome characterized by classical physical stigmata associated with ID. It was not until 1959 when Lejeune uncovered the genetic basis of Down syndrome, a full or partial extra copy of Chromosome 21 (Trisomy 21). The additional genetic material alters the course of development and causes the characteristic features associated with the condition.

People with Down syndrome are at increased risk of certain medical conditions either as a child or as they age. Among these are congenital heart defects, vision and hearing problems, compromised immune systems with susceptibility to infection, hypothyroidism, blood disorders, hypotonia, digestive problems, celiac disease, atlantoaxial instability, disruptive sleep patterns and sleep disorders, gum disease and dental problems, epilepsy, mental and emotional problems, precocious aging and dementia. Many of these conditions are treatable, so individuals with Down syndrome can live healthy lives. However, health disparities in both health-related problems and health care delivery still persist. Despite these disparities, life expectancies for people with Down syndrome have increased dramatically in recent decades.

Average life expectancy of an individual with Down syndrome born in:

- Early 1900's – 9 years
- 1930 – 19 years
- 1983 – 25 years
- Today – 66 years

Risk factors for Alzheimer's disease or dementia among people with developmental disability

Individuals with developmental disability are at greater risk for developing dementia if the individual is:

- over the age of 40 and has Down syndrome
- if the individual is over the age of 59 and has an intellectual disability of another cause
- if the individual has some form of head injury, especially severe or multiple head injuries
- if the individual has a family history of Alzheimer's disease

People with Down syndrome are affected differently by dementia of the Alzheimer's type. They have higher rates of Alzheimer's disease when compared to the general population or to that of non-DS-ID. Neuropathological studies have shown that virtually all older adults with DS who come to autopsy show the characteristic brain changes associated with Alzheimer Disease. "Over expression" of the amyloid precursor protein (APP) gene is related to amyloid plaque deposits in the brain and is critical to the development of AD. Researchers have established a genetic link between AD and DS: The APP gene is located on chromosome 21 in an area that must be trisomic for full expression of the DS phenotype. Down syndrome persons also experience premature aging which means they experience age-related health concerns including dementia approximately 20 years earlier than other adults in the general population. Typically Down syndrome adults are in their late 40's or early 50's when symptoms of Alzheimer's disease first appear.

Syndrome of premature aging

- skin changes
- menopause
- osteoporosis
- osteoarthritis
- hypogonadism
- immunological changes
- senile cataracts
- atrophy and white matter lesions

Importantly, not all adults with DS will develop Alzheimer’s disease. Some will show no sign of dementia well into their 70’s and die of other causes. The dementia rates for Down syndrome are listed below.

Dementia Rates in Down Syndrome, Ball et al. (2006)

| AGE RANGE | PREVALENCE RATE RANGE |
|-----------|-----------------------|
| 30-39 | < 3% |
| 40-49 | 10-25% |
| 50-59 | 20-50% |
| > 59 | 30-75% |

The symptoms of Alzheimer disease may be experienced somewhat differently in adults with DS. Memory loss is not always the prominent or first noted symptom, and cognitive and executive function changes may be present but not readily apparent. Instead, personality and behavior changes may be some of the earliest signs. Men and women with Down syndrome appear to be equally susceptible to Alzheimer’s disease. The progression of the disease takes, on average, approximately eight years. This is somewhat less than the general population, likely due to the effects of precocious aging and perhaps a more aggressive form of the disease.

The rates of dementia in non-DS-ID are also higher than in the general population (but not as high as DS). However, the age of onset and course of dementia tend to approximate that of the general population.

Dementia Rates in Non-DS-ID, Strydom et al (2007)

| AGE | ALL DEMENTIA | ALZHEIMER’S DISEASE |
|------|--------------|---------------------|
| > 59 | 13.1 | 8.6 |
| > 64 | 18.3 | 12.0 |

As with Down syndrome, memory and cognitive loss may not be evident early on in non-DS-ID. Rather, behavioral and emotional changes may be the initial presenting symptoms, particularly that of deterioration in functional status and signs and symptoms of depression.

Common Signs of Dementia in ID

Signs that an older person with ID may be showing behavioral changes with Alzheimer’s disease include:

- a general loss of interest
- forgetting common day-to-day routines and places
- increasing depression
- changes in personality, behavior and mood

While adults with ID are at increased risk for developing dementia as they age, it is important to remember that many other disorders or conditions can result in symptoms similar to that of AD, some of which may be treatable and/or potentially reversible. When there is a suspicion about the presence of AD (or other dementia), medical follow-up is needed to insure a proper differential diagnosis. The sequence of dementia symptoms in ID adults (as summarized by Strydom et al, 2010) is detailed below.

Sequence of Dementia Symptoms in ID Adults

| | ADULTS WITH DS | NON-DSID |
|---|---|---|
| Trigger Symptoms | <ul style="list-style-type: none"> • Memory loss and disorientation • Deterioration in speech • Personality and behavior change • Functional deterioration • Neurologic symptoms – seizures, incontinence • Frontal lobe-related symptoms | <ul style="list-style-type: none"> • General deterioration in function • Behavioral and emotional change |
| Sequence of Memory and Cognitive Changes – Early and Middle Stage Dementia (in order of typical progression) | <ul style="list-style-type: none"> • Memory loss • Deficits in executive function • Complex cognitive functions • Visual organization • Verbal memory • Semantic and short-term memory • Dyspraxia | <ul style="list-style-type: none"> • Memory/cognitive decline less prominent early on |
| Sequence of Functional Decline, Personality and Behavior Change | <ul style="list-style-type: none"> • Decline in ADLs –successive deterioration in <ul style="list-style-type: none"> ○ Personal hygiene ○ Housekeeping skills ○ Dressing ○ Spatial orientation ○ Eating • Increasing number and more severe maladaptive behaviors <ul style="list-style-type: none"> ○ Irritability ○ Aggression ○ Self-injurious behavior ○ General slowness ○ Apathy ○ Loss of interest ○ Decreased social engagement | <ul style="list-style-type: none"> • Signs of depression – <ul style="list-style-type: none"> ○ Lack of energy ○ Low mood ○ Disturbed sleep • Psychosis – persecutory delusions; auditory hallucinations • More aggressive than DS counterparts (but higher prevalence of other behavioral change in DS) |
| | ADULTS WITH DS | NON-DSID |

| | | |
|--|---|---|
| Neurological and Physical Changes | <ul style="list-style-type: none"> • Epilepsy • Myoclonus • Pathologic reflexes • Brain atrophy associated with frontal lobe dysfunction • Rigidity • Postural abnormalities | <ul style="list-style-type: none"> • Late stage symptoms – <ul style="list-style-type: none"> ○ Urinary/fecal incontinence ○ Difficulty walking |
| End-Stage Symptoms | <ul style="list-style-type: none"> • Unresponsive to environment • Loss of ability to speak • Total dependence • Unable to walk • Incontinent • Parkinsonian features • Almost all have seizures | |

Diagnostic Challenges

The diagnosis of dementia relies on excluding other disorders and documentation of declines from previous levels of functioning. In the ID population the diagnosis of dementia is more complex for several reasons:

- Premorbid cognitive deficits – ID adults often lack the skills to perform common standardized diagnostic tests. The matter is compounded by the ID person’s limited ability to self-report. Often there is a lack of consistent and reliable documentation of premorbid function. The combination of these factors makes it difficult to detect declines until they become pronounced
- Atypical presentation – changes in adaptive skills and behavior may pre-date early impairments in memory in individuals with ID. Given the high prevalence of maladaptive emotional and behavioral problems in this population, especially depression, it can be difficult to differentiate changes related to dementia from new onset or recurrent behavior or psychiatric problems.
- Susceptibility to other causes of decline – differential diagnosis is made harder by susceptibility to other medical conditions common in individuals with ID. Associated disorders, such as thyroid abnormalities, arthritis, hearing and visual loss, vitamin deficiencies, depression, susceptibilities to the effects of systemic illness, infections, pain, as well as adverse effects of medications, all can have effects on memory, cognition, mood and behavior and lead to deterioration in function.

It is important to systematically consider these factors in your differential diagnosis to avoid misdiagnosis, to minimize pain and suffering and to identify areas that warrant further evaluation. Diagnosis can be greatly facilitated by having an established baseline of function before any problems become apparent. The AAMR/IASSID, a working group of experts in the field (Aylward et al., 1995) have recommended that assessments of baseline function be conducted at least once before the age of 36 in individuals with DS and by the age of 50 in non-DS-ID, and periodically thereafter to facilitate early detection. If individuals are screened regularly, other conditions that mimic dementia can be ruled out

and/or treated. Early detection is also important as it enables for care and treatment decisions to be adjusted over time to cope with increasing care needs.

Is There a Test for Alzheimer 's disease?

As with the general population, there is no single diagnostic test for Alzheimer's disease or dementia in the ID population. If suspected, a complete physical exam and more frequent medical, neurological and psychological evaluations are needed to establish the progressive nature of the symptoms and rule out other causes.

Components of a standard diagnostic workup include a detailed medical history presented by a family member, caregiver or someone else well-acquainted with the individual. A thorough physical and neurologic exam to include testing of sensorimotor systems, a psychiatric assessment to rule out the presence of a psychiatric disorder, particularly depression; neuropsychological testing to obtain in-depth measures of neurocognitive functions, including memory, orientation, language skills, intellectual abilities and perception; as well as routine laboratory tests including blood work, urinalysis, chest x-ray, EEG and electrocardiography as well as specialized tests as deemed appropriate. It is recommended, at minimum, that annual evaluations and rescreening be obtained to look for changes in existing skills and functioning.

In addition, clinicians should pay special attention to these conditions that occur commonly in individuals with intellectual disability.

- Depression or other mental impairments
- Sensory impairments
- Thyroid
- B12/folate deficiency
- Acute/chronic medical conditions – infection, pain, epilepsy, sleep apnea, cardiac abnormalities
- Adverse medication effects
- Major life stressors

Imaging techniques such as CT and MRI may be useful in ruling out structural brain abnormalities related to stroke, subdural hematoma, brain tumors and normal pressure hydrocephalus.

Diagnostic Criteria

Currently there are no diagnostic criteria specific for use with the ID population. The previously referenced working group (AAMR/IASSID) has proposed ICD-10 criteria as the most appropriate for use with individuals with ID. While not specific to the ID population, the ICD-10 criteria place greater emphasis on non-cognitive aspects of dementia which are often the early signs in individuals with ID, especially the severely cognitively impaired.

The ICD-10 diagnostic system also makes use of a two-step process to first establish a diagnosis of dementia and then subtype it. Because clinicians are well-aware of the strong associations between Down syndrome and Alzheimer's disease, the Work Group reasoned there may be a tendency for

evaluators to assume all cognitive declines represent Alzheimer's disease. Conversely, the clinician may attribute the development of non-cognitive behavior changes to dementia without evidence of cognitive or memory decline. This two-step process helps to avoid the pitfalls of preconception and confirmatory bias which can adversely influence clinical inference, by providing a built-in mechanism to consider other possible causes of cognitive decline.

Diagnostic Certainty

To make a probable diagnosis of dementia of the Alzheimer's type, a well-documented progression of symptoms needs to be established and other possible conditions or disorders ruled out. Complete evaluations must be performed periodically to establish decline.

A possible diagnosis of dementia is made when the presentation or course is somewhat aberrant or is made in the presence of secondary disorders that may produce dementia but are not considered a primary cause of dementia. ICD-10 criteria for dementia and Alzheimer's disease are outlined below.

ICD-10 CRITERIA FOR DEMENTIA AND ALZHEIMER'S DISEASE

ICD-10 Criteria for Dementia

1. *Decline in memory.* Most evident in the learning of new information, although in more severe cases the recall of previously learned information may also be affected. The impairment applies to both verbal and nonverbal material.
2. *Decline in other cognitive abilities.* Characterized by deterioration in judgment and thinking, such as planning and organizing, and in the general processing of information. Deterioration from a previously higher level of performance should be established.
3. *Awareness of the environment.* Absence of clouding of consciousness for a period of time sufficiently long to allow the unequivocal demonstration of decline in memory and other cognitive functions.
4. *Decline in emotional control or motivation, or a change in social behavior.* Changes are manifested in at least one of the following: (1) emotional lability, (2) irritability, (3) apathy, or (4) coarsening of social behavior.
5. *Duration.* Decline of memory and other cognitive functions must be present for at least six months.

IDC-10 Criteria for Alzheimer's Disease

1. *All criteria for dementia are met.*
 2. *Exclusionary criteria.* No evidence from the history, physical examination or special investigations for any other possible cause of dementia, a systemic disorder, or alcohol or drug abuse.
 3. *Onset and progression.* For a diagnosis of Alzheimer's disease, there must be evidence of gradual onset and continuing cognitive decline.
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Clinical Manifestations

Similar to the general population, the core symptoms of dementia in ID involve progressive loss of function in multiple cognitive domains. As noted, the clinical presentation of dementia in the ID population may be different as personality and behavior changes can mark the earliest signs especially

in lower functioning individuals. Changes in personality and behavior are often manifested by emotional lability, increased irritability, apathy/inactivity and stubbornness and coarsening of social behavior.

Individuals may become emotionally labile with crying spells, ease of upset and frequent mood changes. Irritability is often manifested by moodiness, low frustration tolerance or the ease of anger. Stubbornness is manifested by oppositional and resistive behaviors and uncooperative mood. Coarsening of social behavior may be seen by a loss in baseline social manners and verbal and/or physical aggression. Often there is marked apathy, inactivity and withdrawal characterized by a pervasive loss of interest and initiative in favorite foods and tv programs, pastimes, family activities, and social gatherings, coupled with a slowness affecting all aspects of functioning including walking, eating, speaking and general movements.

Cognitive Loss

The perception of cognitive loss will depend on the individual's premorbid level of function and the demands of everyday life. The first suspicions of decline are often based on changes in adaptive function, as these are more concrete and generally easier to document.

Corresponding to the personality and behavior changes that can be seen early on in the development of dementia in ID, there may be a decline in frontal/executive and related adaptive functioning **prior** to the onset of memory impairment or full blown dementia. This has led some investigators to conclude that a frontotemporal type of dementia may represent the pre-clinical stage of dementia, particularly in adults with DS.

Cognitive deficits in judgement planning and organization may be observed in persons with mild ID who can no longer make weather-appropriate clothing choices or plan or carry out non-routine tasks such as shopping or cooking. For those who normally only engage in routine tasks such as dressing, grooming, and toileting, etc., more fundamental cognitive deficits are observed typically with indications of apraxia, aphasia, alexia and agraphia.

For those individuals with mild to moderate ID (IQ = 40-70) who have relatively well-developed verbal skills, cognitive loss can be seen with word finding difficulty, dysnomia and diminished ability to follow commands. If the individual's premorbid verbal skills were relatively poor, decreased use of language progressing to total loss of verbal expression is seen. Often there is a corresponding loss of previously acquired skills to read, write, count, color and draw. For those previously able to perform these skills there is a decreased ability to carry out basic ADL's such as dressing, grooming, bathing, toileting and self-feeding. Inappropriate use of everyday objects may also be observed, e.g. using a toothbrush as a hair brush.

In individuals with severe to profound ID (IQ <40) a general slowing in all areas is present with greater impairments to attention and decreased temporal and spatial orientation. Difficulties distinguishing day and night and inability to locate rooms in the home may be seen.

Memory Loss

As with cognition, memory loss will depend upon the individual's premorbid level of IQ and the memory demands of everyday life. In ID adults, memory may not be the first or most prominent change early on. When memory loss does occur its progression is similar to that seen in the general population accepting those tasks too complex or difficult to begin with.

Memory loss in individuals with mild to moderate ID is frequently manifested as forgetfulness for names, recent conversations and events as well as the location of everyday items. Disorientation to time and the temporal sequence of events is present, as well as spatial disorientation to one's environment as evidenced by difficulties getting around the home, neighborhood or worksite. As memory loss progresses increasing reminders and prompting are required to carry out daily activities, and the affected individual may experience difficulty remembering the steps necessary to perform previously mastered tasks or directions.

The ability to assess memory loss in severe to profound ID adults depends upon their premorbid verbal skills and their ability to meet task demands. For individuals with profound ID (mental age of <2) their cognitive intellectual limitations may preclude detection on standardized tests. Greater reliance needs to be placed on informant report and the use of neurologic signs such as the presence of myoclonic jerks, seizure activity, abnormal posture/gait, rigidity and incontinence.

Common Physical Signs

Common physical signs can accompany the onset or progression of dementia in ID adults. Approximately 50% to 80% of ID adults develop late onset seizure or a new type of seizure after being diagnosed with dementia. Approximately 20% who develop dementia also show Parkinsonian features including slowness and shuffling of gait, rigidity and posture abnormalities. Pathological reflexes, urinary incontinence and myoclonic jerks also can be present.

Depression and Life Stress

Depression is common in ID adults. Prevalence rates at least parallel, if not exceed, that of the general population (6-13%) and are more common in Down syndrome ID than non-DS-ID individuals. In addition to mood changes, concurrent severe behavioral problems and psychotic features are common along with other coexisting mental disorders such as anxiety, OCD, schizophrenia, personality disorder and manic depression.

The significance of an undiagnosed and untreated mood disorder cannot be understated. In addition to the strong association between depression and problems with behaviors and aggression, depression is also associated with significant declines in intellectual, memory, language and adaptive function. There is also evidence to suggest depression may serve as a prodromal syndrome for a later developing progressive dementia, (Burt, D., 1999). Therefore, it is critically important to consider a diagnosis of depression before assuming the presence of Alzheimer's disease.

The diagnosis of depression in ID adults is more complex. Depressed affect and cognition are often expressed indirectly in this population. Traditional diagnostic criteria and assessment procedures rely heavily on self-reported subjective feelings, a task that is often beyond the capabilities of many ID adults. Depression and dementia share many overlapping symptoms, and depression may also coexist with dementia and other forms of mental illness. Comorbid medical conditions can also mimic depression.

While assessment of depression in ID adults can be challenging, McGuire & Chicoine (1996) note that differential diagnosis can be enhanced when:

- Diagnostic criteria are adjusted to account for the adaptive and expressive limitations of ID adults;
- Behavioral changes are emphasized as opposed to subjective feelings;
- Other medical problems are ruled out; and,
- By paying close attention to clinical course.

Generally speaking, individuals with depression will tend to show an up-and-down course with regard to cognitive and functional status, while individuals with dementia will eventually show progressive decline.

There are no widely accepted diagnostic criteria or scales for use in the diagnosis of depression in adults with ID. A number of diagnostic schemes have been proposed. One useful set of criteria relies on observable symptoms of depression that have been reported in the literature for adults with mental retardation and is based on a revised version of the Mood Assessment Scale for Demented Adults (Sunderland et al, 1988). The adaptive diagnostic criteria from this scale are as follows:

| OBSERVABLE SYMPTOMS OF DEPRESSION IN ADULTS WITH MENTAL RETARDATION | |
|--|---|
| Core symptoms suggesting depression | <ul style="list-style-type: none"> • Looks sad, down-cast, preoccupied or tearful • No longer initiates or participates in activities previously enjoyed • Gets irritable or upset easily, more than expected for a given situation • Looks restless or agitated |
| Additional symptoms | <ul style="list-style-type: none"> • Looks sleepy or drowsy during the day • Has trouble falling asleep, staying asleep or waking too early • Has had a decrease or increase in eating • Complains about feeling bad or needing to go to the doctor • Looks anxious or concerned about something • Has a low energy level |

| | |
|--|--|
| | <ul style="list-style-type: none"> • Had reduced physical and facial expressiveness to external stimuli • No longer enjoys previously enjoyed activities • Frequently asks for assistance regardless of need for help • Speech has changed in terms of tone, rate, spontaneity, loudness • Has noticeable difference in mood throughout the day • Repeatedly does something to hurt him/herself • Sits and stares or sits stiffly • Looks slowed down • Physically or verbally aggressive • Loss of daily living skills • Looks fearful |
|--|--|

Recommended criteria for diagnosis of depression in ID is the presence of at least one core symptom, plus 4 additional symptoms present for a minimum of 2 weeks, resulting in significant impairment in daily function, and with other causes being ruled out.

Life Stress

Loss and life stress are common triggers for depression. Carers often do not believe individuals with ID understand the concept of death and minimize its effects. Typically ID adults report distress over sadness, even if they do not understand the concept of death completely (Burt D, 2008). Changes in residence, routine or support systems also trigger depression. It is important to ask what changes or stressors have been present in the individual’s life to determine if stress and loss may be factors in the individual’s mood or behavioral presentation.

Assessment-A Baseline Comparison Approach

The diagnosis of dementia in individuals with intellectual disability requires a change in status from baseline functioning, not a change from a normal level of functioning (as with the general population). Longitudinal assessment that documents baseline or best level of functioning in addition to changes in cognitive and behavioral functioning over time is necessary before sufficient information can be obtained to form a diagnosis of dementia.

Baseline functioning in individuals with intellectual disability is significantly more heterogeneous than the general population and individuals can have wide-ranging and varying baselines of strengths and weaknesses even within the same range of intellectual disability. In addition, people with intellectual disability can have a wide range of behavior problems typical for them. Assessing the clinical significance of such behaviors requires a comparison with typical adulthood functioning. For these reasons, a working group of dementia experts (Aylward et al, 1995) recommended use of a baseline

comparison approach to diagnosis of dementia in ID. ID persons are screened when healthy to obtain a “best level” of baseline performance and then followed longitudinally, with periodic re-screenings to assess for change in status. The baseline comparison approach incorporates use of a combination of direct assessment measures and caregiver reports. Direct assessments are needed to document changes in neuropsychological status as caregivers are often not reliable in reporting memory and cognitive functioning. Conversely, caregiver reports are required to document that any declines in memory and cognitive function are significant enough to affect daily functioning.

A number of direct assessment measures have been developed or adapted for use with the ID population. While these instruments can provide objective evidence of current cognitive function, they are not without their limitations. Even though the measures have been developed for use with the ID population, floor effects are present to varying degrees and the instruments may be of limited use with individuals with severe intellectual disability or limited verbal ability. In these cases, collateral reports take precedence.

A number of indirect measures have also been adapted for use with the ID population. However, these measures must also be interpreted with caution as aging informants may be developing cognitive difficulties themselves, limiting accuracy, or caregivers may know the subject too well or not well enough to be objective. It is recommended that multiple informants across several different settings be utilized. In addition, while many observer-rated indirect measures include coverage of cognitive domains of function in their measures, indirect assessments do not assess cognition directly. Ultimately, a combination of direct and indirect assessment measures is likely to provide the highest sensitivity and specificity.

There are a variety of different tools used for the assessment of dementia in ID adults, but as Zeilinger et al (2013) found a number of the measures were neither designed for assessment of dementia nor for persons with ID! It is therefore recommended that clinicians select only those instruments designed or adapted for use in detecting dementia in ID and that the measures are valid and reliable, sensitive to change early and throughout the course of dementia and measure a wide range of neuropsychological domains. See Elliott-King J. et al (2016) for a critical review of numerous instruments used in the diagnosis of dementia in adults with ID.

Assessment Measures

Below are listed some direct and indirect assessment measures for use with the ID population.

Observer-rated scales observed below include:

- Dementia Scale of Down Syndrome (DSDS)
- Dementia Questionnaire for Mentally Retarded Persons (DMR)
- Camdex-DS
- Dementia Screening Questionnaire for Individuals with Intellectual Disabilities (DSQIID)

Dementia Scale for Down Syndrome:

The Dementia Scale for Down Syndrome (Gedye, 1995) is an informant-based instrument designed to aid in the diagnosis of dementia in adults with ID, especially DS. It is also used to establish a baseline for

those at risk. The items are grouped into early, middle, late and very late stages of dementia. The informant is asked to classify features as not applicable, absent or present and, if present, if the feature is typical or atypical of the adult. New signs are tallied for each stage along with a separate tally of cognitive signs and then compared to criteria for diagnosis and staging. The DSDS provides a differential diagnosis screening questionnaire section to address other potential causes for dementia. The DSDS has been found to have good sensitivity and specificity. It is comprehensive but contains no measure of general disability. It correlates well with other observer rated instruments. Unfortunately, it has use restrictions limited to psychologists and psychometrists with two years of experience in dementia evaluation. **Order forms for the DSDS booklets and manual can be obtained at <http://www.gedye.ca/>**

DMR:

The Dementia Question for Mentally Retarded Persons (Evenhuis, 1992) is an English translation of the work of Heleen Evenhuis' who is an ID physician in the Netherlands. It was designed to facilitate a diagnosis of dementia in ID. It is described as a "screening instrument" for selection of persons for further specialist diagnostic assessment.

The DMR is based on observation of caregivers over the previous two months. It consists of 50 items and 8 subscales divided into 2 subcategories. The cognitive subcategory covers short-term and long-term memory, as well as spatial and temporal orientation, while the social subcategory covers speech, practical skills, mood, activities, interests and behaviors. There are three response categories, 0 (no deficient) to 2 (severe deficit). It provides a measure of general disability. It is generally quick and easy to use (15-20 minutes) and there is a short form available. It does, however, have floor effects with advanced dementia and is not considered an appropriate instrument for single application. It is applicable for individuals with mental ages of 2-10 but is not appropriate for individuals with profound or severe ID with severe comorbid physical, motor or hearing impairments. The DMR does not have use restrictions or specialized administrative training required. **To obtain test materials go to www.harcourt.nl**

Camdex-DS:

The Camdex-DS (Hon et al (1999); Ball et al (2006) is a modified version of the Cambridge Exam for Mental Disorders of the Elderly used to document increasing prevalence with age. The author notes that the aims (in part) were to incorporate in a single schedule all the information necessary to enable an accurate clinical diagnosis of dementia with people with ID, but caution that it is not to be viewed as a substitute but rather an aide to the diagnostic process. The Camdex-DS is designed for use in the community by trained healthcare professionals. It is considered a test battery for comprehensive assessment that can be somewhat time-consuming. It is structured about areas of function likely to change with the onset of dementia. The measure contains informant interview and direct cognitive assessment (the CAMCOG-to be detailed later), patient interview and a standardized schedule for recording observations, physical exam and information on labs. It also collects information on cognitive and functional decline, current mental and physical health and best level of functioning. It has generally good reliability and predictive validity but is susceptible to floor effects in more significantly impaired individuals. **Test materials available at Cambridge University Press. (Cambridge.org)**

DSQIID:

The Dementia Screening Questionnaire for Individuals with Intellectual Disabilities (Deb et al. 2007) was designed as a user friendly, observer rated dementia screening developed to overcome the floor effects of existing dementia screening scales. It was derived from interviews with caregivers of adults with DS and dementia. It was then validated on a large sample. It consists of 53 items in 3 parts. Part 1 assesses “best” ability. Part 2 assesses behaviors and symptoms suggestive of dementia on the basis of a 4-point rating scale. Part 3 consists of 10 comparative questions answered on a yes/no basis. The DSQIID has very good sensitivity and specificity. It is quick and easy to score in any setting. Its single fixed cutoff may limit usefulness in adults with more advanced stages of dementia and with varying degrees of baseline intellectual disability. **A copy of the test form and administrative instructions can be found in the original journal article. See reference list.**

Direct Neuropsychological Measures

Direct neuropsychological measures commonly used for cognitive assessment and intellectual ability in the ID population include the:

- Test of Severe Impairment (TOSI)
- CAMCOG-DS
- Institute for Behavioral Research Evaluation of Mental Status (IBREMS)

Test of Severe Impairment:

The Test of Severe Impairment (Albert and Cohen, 1992) was originally designed as a downward extension of the MMSE to objectively assess patients with severe cognitive dysfunction (MMSE < 11/30) in the general population and was subsequently validated in persons with ID (Cosgrove et al, 2000). It assesses a range of skills involving motor, language, memory, conceptualization and general knowledge functions. Only eight questions require a verbal response. It is generally brief and easy to use and yields a range of scores that enables the measure to be applicable across a range of ID. Its psychometric properties are generally good and have been correlated with tools that measure functional decline (DLSQ) at baseline and diagnosis. Its degree of difficulty is appropriate for moderate to severe intellectual disability to score on unless they are in advanced stages of dementia. **A copy of the test protocol is available in Prasher (2009).**

CAMCOG-DS:

The CAMCOG-DS (Hon et al, 1999; Ball et al., 2006) is the self-contained neuropsychological component of the CAMDEX that was validated in subjects with DS. It is made up of seven subscales that measure orientation, language, memory, attention, praxis, abstraction and perception, functions that are known to decline in dementia. The authors state that the CAMCOG-DS enables examination of patterns and profiles of cognitive performance through subscale analysis. As a revised version it has expanded remote recall and executive function measures involving ideational fluency and visual reasoning. The authors report that it has few floor effects, and scores correlate with age. **Available at Cambridge University Press. Cambridge.org.**

IBREMS:

On the IBR Evaluation of Mental Status (Wisniewski and Hill, 1985) adults respond to 37 items assessing orientation, language, short-term and long-term memory, writing, drawing and general knowledge. Recall and recognition format is provided. If an IQ is available when the individual was healthy, the IBREMS enables a cut-off score for assessment at a single point in time, but the IQ must be > 25. Caution is advised as when using this cut-off Wisniewski et al. (1985), found a rather high rate of misclassification of non-demented subjects as demented patients(20%). The IBREMS has good sensitivity and specificity, otherwise. **The test protocol and administration instructions are appended to the original journal article by Wisniewski and Hill (1985). See reference list.**

Adaptive Behavior Scales

A number of adaptive behavior scales have been used for use in documenting declines in everyday function. Two commonly used instruments include the Adaptive Behavior Dementia Question (ABDQ) and the Daily Living Skills Questionnaire (DLSQ).

Adaptive Behavior Dementia Questionnaire (ABDQ):

The ABDQ (Prasher et al., 2004) contains 15 items from the Adaptive Behavior Scales that were found to be strongly linked between declines in adaptive skills and aging and dementia in older adults with DS. Items are rated much worse, worse, the same, better, much better than normal. Weighted scores are summed and compared to cut-off. The ABDQ has good sensitivity and specificity. It is user friendly. It can be done on older DS adults regardless of ID or cooperation level, but it does not provide a measure of general disability. **Test items and administration/scoring instructions are listed in the original journal article by Prasher et al (2004). A copy of the test materials is contained in Prasher, (2009).**

Daily Living Skills Questionnaire:

On the DLSQ (National Institute on Aging, 1989) informants provide information concerning a variety of ADL's including dressing, grooming, eating, manual dexterity and geographical orientation. No significant floor effects were found. It has a high positive predictive value and correlates strongly with direct cognitive tests. **The DLSQ can be found at NIA.NIH.gov**

Indirect and Direct Screening Measures for Dementia

| CLASSIFICATION | MEASURE | ADVANTAGES | DISADVANTAGES | SENSITIVITY/SPECIFICITY FOR AD IN DS | REFERENCE |
|-------------------------------|-----------|--|---|--------------------------------------|------------------------------------|
| Observer-rated measures | DSDS | <ul style="list-style-type: none"> Comprehensive Measures change through course of dementia Includes differential diagnosis screening scale No significant floor effects Allows for staging of dementia | <ul style="list-style-type: none"> No measure of general disability Use restrictions Lengthy administration Reduced sensitivity with mild-moderate ID Scoring system is not simple | 89% / 85% | Gedye 1995 |
| | DMR | <ul style="list-style-type: none"> Applicable to mild, moderate and severe ID Includes a general measure of disability No restrictions on use Brief administration | <ul style="list-style-type: none"> Not applicable to severe ID with other severe disabilities Requires repeat administration for valid results Ceiling/bottom effects with profound/ v. mild ID | 92% / 92% | Evenhuis (1992) |
| | CAMDEX-DS | <ul style="list-style-type: none"> Comprehensive Includes a measure of general disability Incorporates 3-step assessment process that facilitates formulation of differential diagnosis | <ul style="list-style-type: none"> Floor effects Lengthy administration | 88% / 94% | Ball et al. (2004) |
| | DSQIID | <ul style="list-style-type: none"> Validated in a large sample Brief administration Excellent IRR | <ul style="list-style-type: none"> Single fixed cut-off limits usefulness in more advanced stages of dementia and varying degree of baseline ID | 92% / 91% | Deb et al. (2007) |
| Neuropsychological tests | TOSI | <ul style="list-style-type: none"> Assess range of skills Only 8/24 items require a verbal response Brief/easy to administer Range of scores makes applicable across all levels of ID IRR good | <ul style="list-style-type: none"> No measure of general disability Ceiling effect in mild ID | NA / NA | Albert & Cohen (1992) |
| | CAMCOG-DS | <ul style="list-style-type: none"> Assesses wide range of cognitive domains including remote recall and executive function Subscale profile analysis Scores correlate with age Few floor effects | <ul style="list-style-type: none"> Limited diagnostic value at a single point in time. Not as sensitive with severe ID, severe sensory impairment or already advanced dementia | See CAMDEX-DS | Ball et al. (2004) |
| | IBREMS | <ul style="list-style-type: none"> Brief, easy to administer Enables cut-off score if IQ available when healthy. | <ul style="list-style-type: none"> Concentration subscale too difficult for many | 90% / 89% | Wisniewski and Hill (1985) |
| Measures of adaptive function | ABDQ | <ul style="list-style-type: none"> Can be completed on all ID adults regardless of level of ID or cooperation User friendly Brief administration Screens for DAD in DS | <ul style="list-style-type: none"> No measure of general disability Cognition not assessed Utility with non-DSID unclear | 89% / 94% | Prasher et al. (2004) |
| | DLSQ | <ul style="list-style-type: none"> Correlates well with direct cognitive measures No floor effects | <ul style="list-style-type: none"> No assessment of cognition No measure of general disability | NA / NA | National Institute of Aging (1989) |

Screening Measures

In addition to the above noted indirect and direct assessment measure for diagnosing dementia in ID, one additional screen measure is worth noting. The National Task Group – Early Detection Screen for Dementia (NTG-EDSD), Esralew et al (2013) offers carers and staff a resource to record changes in cognitive and adaptive function known to be associated with dementia. It is not an assessment or diagnostic instrument but rather an “administrative screen” that provides information to begin conversations with healthcare providers. It was adapted for use from the DSQIID (Deb et al, 2007). The EDSD gathers information on relevant demographics and ratings of health, mental health and life stressors. It reviews multiple domains of function and includes surveys of chronic health conditions. It also lists signal items that may reflect early signs of dementia. The EDSD can be helpful in training caregivers and staff to become good observers and reporters of information. It serves as running record of health and function to be reviewed annually. In addition, it serves as a tool accessible to caregivers who are not trained to do assessment but who have valuable information about change in daily function.

Measures of Adjustment, Emotional Functioning and Psychopathology

A number of psychopathology scales have been developed for adults with ID. These instruments allow for making normative comparisons with what symptoms fall outside the typical range and, in some cases, permit comparisons between self-report and informant ratings. These scales were not typically designed for use with individuals with possible comorbid dementia but may be used to document baseline symptoms (if screened early) and may be clinically useful to document change in status over time with serial reassessments. Psychopathology scales listed below include:

- Emotional Problems Scale (Prout and Strohmer, 1991)
- Reiss Screen for Maladaptive Behavior (Reiss, 1988)
- DASH-2 (Matson, Gardner, Coe and Sovner, 1991; Sevin, Matson, Williams & Kirkpatrick-Sanchez, 1995)
- Anxiety, Depression and Mood Scale (ADAMS) (Esbensen, Rohahn, Aman & Ruedrich, 2003)
- Psychopathology Instrument for Mentally Retarded Adults (PIMRA) (Senatore, Matson & Kazdin, 1984)

Emotional Problems Scale

The Emotional Problems Scale (Prout and Strohmer, 1991) consists of two complimentary instruments designed for use as part of a comprehensive clinical evaluation of individuals aged 14 years and older with mild intellectual disability or borderline intelligence. The 135 item 4-point behavioral rating scale is used to indicate how often a client has exhibited specific behaviors during the previous 30 days including those reflective of thought/behavior disorder, verbal aggression, physical aggression, sexual maladjustment, distractibility, hyperactivity, somatic concerns, depression, withdrawal and low self-esteem. Broader scales assess externalizing problems and internalizing problems.

The 147 item self-report inventory uses a yes/no format and is written at a third grade reading level. The inventory is read to the individual and the authors claim that it can be completed by persons with very low reading levels and non-readers. The self-report inventory yields scores for positive impression, thought/behavior disorder, impulse control, anxiety, depression, low self-esteem and total pathology.

Normative data were obtained on 673 and 704 individuals respectively for each of the two instruments. Results of the ratings are tallied and presented in normalized T-scores and percentile scores. Internal consistency and reliability coefficients ranged from .9 to .97 and from .77 to .96 respectively. The authors claim the EPS has been shown to be related to a variety of relative clinical variables. **The EPS is available at: integratedassessments.com**

Reiss Screen for Maladaptive Behavior

The Reiss Screen for Maladaptive Behavior (Reiss, 1988) screens for mental health problems in adolescents and adults with intellectual disabilities. Its empirically derived scales evaluate the probability that a person has an aggressive disorder, autism, avoidant disorder, dependent personality disorder, depression, paranoia and psychosis. The instrument also evaluates the probability of drug abuse, overactivity, self-injury, sexual problems, stealing and suicidal tendencies.

The Reiss Screen is completed by caregivers, teachers, work supervisors or parents. Respondents rate the extent to which each of 38 symptoms is no problem, a problem or a major problem in the person's life. Each item is defined in non-technical language and includes concrete examples. The test is normed for adolescents and adults aged 16 and up with all levels of severity of intellectual disability.

The test screens for psychiatric disorder in three different ways: severity of challenging behavior, psychiatric diagnosis and rare but significant symptoms such as suicidal behavior. The authors report the total score that is generated reflects a measure of severity of disorder that is well-suited to assess progress over time. The authors report the total scores are a valid indicator of whether or not a person with an intellectual disability also has a mental health problem. The eight scale scores also were reported to have a high degree of validity as well. **Test materials may be obtained at: www.idspublishing.com.**

Diagnostic Assessment for the Severely Handicapped Scale -2 (DASH-2)

The DASH-2 (Matson et al., 1991) was designed as a multi-dimensional instrument to assess the severity of individual symptoms as well as frequency and duration covering 13 major psychiatric disorders. It is intended to address psychiatric problems for profoundly and severely mentally retarded persons. Items comprising the DASH were derived from DSM-3-R and previously published studies and instruments and selected according to appropriateness for subjects with intellectual and adaptive abilities in the severe to profound ID levels and comprehensibility to informants without training in psychiatric assessment. A total of 83 items was included. The separate dimensions of behavior were selected for rating frequency, duration and severity and are scored on one of three levels: 0, 1 or 2. Ratings are requested based upon the subject's behavior during the previous two weeks. Subscales included anxiety, mood disorder/depression, mood disorder/mania, pervasive developmental disorder- autism, schizophrenia, stereotypies/tics, self-injurious behavior, elimination disorders, eating disorders, sexual disorders,

organic syndromes, impulse control and miscellaneous behavior problems. Data is collected on symptom frequency, duration and severity in individual interviews with direct care staff. Interrater reliability was generally good. **Test materials are available at:** www.disabilityconsultants.org.

Assessment Instrument for Anxiety, Depression and Mood Scale (ADAMS)

The ADAMS (Esbensen et al, 2003) was designed as a comprehensive screening measure for anxiety and depression in persons with mental retardation. The measure consists of 55 items generated from DSM-4 criteria, previous assessment instruments and clinical experience that tap aspects of anxiety, fear, compulsiveness, appetite, communication, concentration, depression, energy level, mood, physical state, sleep disturbance and social interaction. Items are rated on a scale of 0 “not a problem” to 3 “severe problem” indicating the frequency and severity with which a particular behavior or symptom is present. Factor analysis indicated that it is generally a measure of manic/hyperactive behavior, depressed mood, social avoidance, generalized anxiety and compulsive behavior. Internal consistency of the subscales and retest reliability for both the total scale and subscales was high. Interrater reliability was satisfactory. The validity of the ADAMS was assessed with a clinical sample of 129 individuals with mental retardation who were seen in a psychiatric clinic, and this provided additional support for the subscales. The ADAMS was proposed as a psychometrically sound instrument for screening anxiety, depression and mood disorders among individuals with mental retardation.

Publisher: Anna Esbensen, Ph.D.
University of Wisconsin-Madison
1500 Highland Avenue
Madison, WI 53705
608-263-5609

Psychopathology Instrument for Mentally Retarded Adults (PIMRA)

The PIMRA (Senatore et al., 1985) was developed as a screening instrument to aid in the differential diagnosis of individuals with intellectual disability. The PIMRA includes a respondent version and an informant version. There are 56 items on the Informant version representing 7 classes of psychopathology including schizophrenia, affective disorders, psychosexual disorder, adjustment disorder, anxiety disorder, somatoform disorder and personality disorder based on DSM-III criteria. There is one additional subscale measuring inappropriate adjustment. Parents/caretakers familiar with the individual rates each item. The respondent version is adapted from the informant version and uses simpler, more concrete language. Both versions contain the same 8 scales. Test-Retest reliability was acceptable for the total scale but low to acceptable for the eight subscales. Validity was not well established for 5/8 subscales. **The PIMRA may be obtained at:** www.disabilityconsultants.org.

| PSYCHOPATHOLOGY INSTRUMENTS | | | | |
|----------------------------------|--|--|---|---|
| MEASURE | POPULATION/LEVEL OF ID | INFORMANT SOURCE | DOMAINS ASSESSED | COMMENTS |
| EPS Prout and Strohmer (1991) | <ul style="list-style-type: none"> Borderline to M – Mod ID Age 14 and older | <ul style="list-style-type: none"> Caregiver & Self-Report Scales | <ul style="list-style-type: none"> Behavioral Rating Scale (BRS) <ul style="list-style-type: none"> Caregiver ratings of frequency of 10 specific behavior dimensions past 30 days Two broader scales assessing internalizing/ externalizing behavior Self-Report Inventory (SRI) <ul style="list-style-type: none"> Yielding scales for positive impression plus 5 areas of psychopathology plus a total pathology score Behavioral dimensions include thought/behavior disorder, verbal aggression, physical aggression, sexual maladjustment, distractibility, hyperactivity, somatic concerns, depression, withdrawal and low self-esteem | <ul style="list-style-type: none"> Yields profile of normalized T-scores allowing comparisons against a reference group SRI can be completed by individuals with very low reading levels and non-readers |
| Reiss (1988) | <ul style="list-style-type: none"> All levels of severity of ID Age 16 and older | <ul style="list-style-type: none"> Caregiver ratings 2 raters required | <ul style="list-style-type: none"> Caregivers rate the presence of 38 maladaptive behaviors during the past 3 months Ratings then averaged and compared to norms for commonality It produces 8 subscales: aggressive behavior, autism, psychosis, paranoia, depression--behavioral & physical signs, dependent personality disorder and avoidant personality disorder | <ul style="list-style-type: none"> Uses cut-off scores to determine if results positive for dual diagnoses signaling need for further mental health evaluation No data on TRR Criteria validity established |
| DASH-II MATSON ET AL (1991) | <ul style="list-style-type: none"> Severe to profound ID adults | <ul style="list-style-type: none"> Caretaker ratings | <ul style="list-style-type: none"> Multidimensional instrument to assess severity, frequency and duration of symptoms covering 13 major psychiatric disorders—impulse control, organic problems, anxiety, mood disorder, mania, PDD/autism, schizophrenia, stereotypies/tics, self-injurious behavior, elimination disorder, eating disorder, sleep disorder, sexual disorders | <ul style="list-style-type: none"> Useful for identifying syndrome patterns Provides systematic means of assessing psychopathology with severe to profound ID with good reliability |
| ADAMS Esbensen et al (2003) | <ul style="list-style-type: none"> All ranges of ID | <ul style="list-style-type: none"> Caregiver | <ul style="list-style-type: none"> Comprised of 5 subscales: manic/hyperactive behavior, depressed mood, social avoidance, generalized anxiety, and compulsive behavior 28 items of the scale are rated on 0 to 3 point-scale from “not a problem” to “severe problem” | <ul style="list-style-type: none"> Psychometrically sound overall Narrow score limits usefulness for anxiety, mania and depression Valid screening instrument for bipolar disorder, depression and OCD but anxiety scale warrants further study—may not be independent construct due to overlap with other subscales |
| PRIMA Senatore et al (1985) | <ul style="list-style-type: none"> Adults with full range of ID | <ul style="list-style-type: none"> Respondent and informant version | <ul style="list-style-type: none"> Informant version contains 56 items derived from DSM-III classifications of psychopathology Respondent version adopted from informant version using simpler concrete language Each version consists of 8 subscales: schizophrenia, affective disorder, psychosexual disorder, adjustment disorder, anxiety disorder, somatoform disorder, personality disorder and poor mental adjustment | <ul style="list-style-type: none"> Test/Retest reliability acceptable for total scales but low to acceptable for 8 subscales Further research needed to establish validity of some subscales |

Summary

Individuals with intellectual disability (ID) are living longer and, as such, are at increased risk for age-related health conditions including dementia. With the rising life expectancy and growing population of individuals with ID, clinicians and case workers can expect to encounter increasing numbers of persons with ID who develop dementia.

Detecting dementia in intellectual disability populations is difficult due to their premorbid cognitive deficit, tendencies for dementia to present atypically in this population and their susceptibility to other medical problems that mimic dementia. Thus, there is a strong need for enhanced diagnostic processes and development of specialist assessment skills to meet this growing clinical demand.

Personality and behavior changes often predate the onset of memory or other cognitive loss in ID dementia. However, there are no criteria specific to the diagnosis of dementia in ID. Criteria commonly used in the general population do not take into account the unique non-cognitive features of dementia in ID. Use of ICD-10 dementia diagnostic criteria is recommended as these criteria place greater emphasis on the non-cognitive aspects of dementia and employ a 2-step differential diagnostic process.

Standard assessment measures used “at a single point in time” to document declines in function in the general population presume a normal level of premorbid functioning and are, thus, inappropriate for use with this population as many ID adults lack the skills to perform these tasks at baseline. Longitudinal assessment with measures appropriate for use with ID adults is required to document changes in status from a baseline or “personal best” level of function. Baseline screenings should ideally be done when healthy and periodically repeated. Comprehensive workups should be initiated when change is detected.

A number of neuropsychological assessment measures have been developed to aid in the clinical diagnosis of dementia in adults with ID. Combining direct assessment measures and observer-rated skills is likely to provide the highest sensitivity and specificity.

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