Diagnosis of Dementia 
In Individuals with 
Intellectual Disability 

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Report of the AAMR-IASSID Working Group for the 
Establishment of Criteria for the Diagnosis of Dementia 
in Individuals with Intellectual Disability
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Diagnosis of Dementia in Individuals with Intellectual Disability

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Report of the Working Group for the Establishment of Criteria for the Diagnosis of Dementia in Individuals with Intellectual Disability\textsuperscript{1} under the auspices of the International Association for the Scientific Study of Intellectual Disability (IASSID) and the American Association on Mental Retardation (AAMR)

Abstract: This report proposes a set of standardized criteria for the diagnosis of dementia in individuals with intellectual disability (ID) and a standardized procedure for determining whether or not criteria are met in individual cases. It is the authors' intention that these criteria be appropriate for use by both clinicians and researchers. The need for standardized criteria and diagnostic procedures was identified by a group of clinicians and researchers attending the International Colloquium on Alzheimer Disease and Mental Retardation\textsuperscript{2}, as one of the foremost impediments to progress in the understanding and treatment of this condition. The diagnostic criteria and procedures outlined in this report are endorsed by those who attended the International Colloquium.

\textsuperscript{1} For Working Group participants and affiliations, see page 15.

\textsuperscript{2} The International Colloquium on Alzheimer Disease and Mental Retardation was held in Minneapolis, Minnesota, on July 28-29, 1994, with support from the National Institute on Aging and the National Institute for Disability and Rehabilitation Research (P.I.: M. Janicki) (1-3).
INTRODUCTION

Because of medical advances and improved living circumstances, individuals with ID are living longer (4-5). As age is the strongest risk factor for developing dementia, the likelihood that individuals with ID may develop dementia is increasing. It is known, for example, that all individuals with Down syndrome (DS) develop the characteristic neuropathological brain lesions of Alzheimer disease (AD) (i.e., neuritic plaques, granulo-vacuolar changes, cerebral vascular amyloidosis, Hirano bodies and neurofibrillary tangles) by the age of 40 years (6), although many do not demonstrate clinical features of dementia. There is increasing interest in accurate detection of dementia in these individuals for practical purposes, such as program planning and clinical care. There is also growing awareness of the need for accurate methods of differentiating individuals who have progressive, irreversible dementia (e.g., AD) from those who have cognitive decline due to treatable conditions.

Interest in accurate diagnosis is also increasing within the research community, especially as research methodologies become available to answer questions regarding factors associated with dementia, including genetic and neuroanatomical abnormalities. Unfortunately, lack of standard diagnostic procedures has limited meaningful communication among research laboratories. For example, some published studies involving individuals with ID and dementia do not report how a diagnosis of dementia was made. In other studies, all individuals with DS over the age of 40 have been assumed to be demented, even when currently accepted standards of practice for the diagnosis of AD (e.g., medical workup, longitudinal confirmation of deterioration, exclusion of other causes of impairment) have not been applied.

Investigators who do specify the method for diagnosis have employed a variety of both published and unpublished criteria for diagnosing dementia (e.g., NINCDS-ADRDA criteria for Alzheimer Disease (7), CERAD (Consortium to Establish a Registry for Alzheimer's Disease) criteria (8)), modified in a variety of different ways. This has led to large differences among research groups regarding such basic findings as incidence and prevalence figures (see Zigman et al (9)). Until a standardized set of criteria is accepted by researchers in this field, we will be unable to determine whether the brain changes of AD in individuals with DS are even relevant to the diagnosis of dementia. Furthermore, we obviously cannot expect to make progress regarding more complicated aspects of dementia in individuals with ID without first establishing agreement on what constitutes dementia in this population. Generally accepted criteria for the diagnosis of dementia in individuals with ID are also essential for therapeutic trials, evaluation of new diagnostic laboratory tests, and studies regarding the pathogenesis of the disease.

Although current diagnostic systems address both ID and dementia, they do not address the issue of diagnosis of dementia in adults with ID. The Diagnostic and Statistical Manual of Mental Disorders (DSM-IV) (10) mentions mental retardation in regard to "differential diagnosis" of dementia, stating that "Mental retardation is characterized by significantly subaverage current general intellectual functioning, with
concurrent impairments in adaptive functioning and with an onset before age 18 years. Mental retardation is not necessarily associated with memory impairment. In contrast, the age of onset of dementia is usually late in life" (pp. 138-139). DSM-IV provides further discussion regarding the diagnosis of dementia in individuals with ID under the age of 18, but does not provide any information regarding a dual diagnosis of ID and dementia in older individuals. The International Classification of Diseases (ICD-10) (11) does not mention intellectual disability in regard to differential diagnosis of dementia.

Consistent with both ICD-10 and DSM-IV definitions of dementia, the proposed diagnostic criteria for dementia in individuals with ID require documentation of cognitive decline that causes impairment in social or occupational functioning. Definitions of dementia in individuals without ID emphasize a change in status, usually from having a skill or behavior to its disappearance. Our definition for dementia in individuals with ID is similar, with the understanding that the observed change is a change from baseline, not a change from a "normal" level. Consistent with DSM-IV, the term "dementia," as used in this article, carries no connotation concerning prognosis. Although most cases of dementia that are identified by the criteria outlined here will be progressive and irreversible, we are not ruling out cases that are static or reversible. For dementias that are very slowly progressive, we acknowledge that suitable documentation of further decline may be difficult. Evidence of decline over time on measures of cognitive abilities, behavior, and/or adaptive functioning should be obtainable, however, for most individuals with ID.

ICD-10 criteria for dementia are proposed as the most appropriate for individuals with ID. In comparison with DSM-IV criteria, ICD-10 criteria place more emphasis on "noncognitive" aspects of dementia (e.g., emotional lability, irritability, apathy, coarsening of social behavior). It is in these noncognitive realms that the first signs of dementia are often observed in individuals with ID, especially for those who are more severely cognitively impaired (12-14). Furthermore, ICD-10 criteria are designed to first establish a diagnosis of dementia and then to differentiate AD from other forms of dementia, whereas DSM-IV provides separate criteria for individual subtypes of dementia. Because clinicians are familiar with the fact that the brain changes of AD are common in older individuals with DS, there has been a tendency to assume that all cognitive decline in these individuals represents AD. Furthermore, a common error is to ascribe the development of noncognitive behavior changes to dementia without evidence of cognitive decline. By advocating the use of a two-step process (i.e., diagnosing dementia and then subtyping), we are encouraging clinicians to consider other possible causes of cognitive decline (including those that are treatable). Finally, in order to facilitate international communication among researchers, we agreed that criteria based on the ICD-10 model might be more acceptable, and thus more widely adopted, outside of the United States than criteria based on DSM-IV. Our criteria for diagnosis of dementia in individuals with ID are, however, compatible with DSM-IV criteria.

CRITERIA
Following the model of ICD-10, we will first discuss criteria for a diagnosis of dementia, and then discuss additional criteria for the diagnosis of AD in ID.

Criteria For Diagnosis of Dementia
The major adaptation necessary for applying ICD-10 criteria for dementia to individuals with ID involves comparison of current level of functioning to previous level of functioning, not to a "normal" level of functioning. In individuals with ID, it cannot be assumed, of course, that impaired performance reflects dementia. Documentation of decline is essential. This section describes how each of the ICD-10 criteria can be interpreted for application to individuals with ID. Following this section, we will discuss the procedure for determining whether or not diagnostic criteria are met in individual cases.

Memory Decline:
ICD-10 criteria for dementia require a "decline in memory, which is 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 non-verbal material" (p. 29). Criteria for determining whether decline in memory is mild, moderate, or severe, are then presented.

The manifestation of memory loss in adults with ID will depend, in most cases, upon the premorbid level of intellectual functioning and pattern of memory strengths, and upon the memory demands required in the everyday life of the individual. To be indicative of dementia any memory changes over time must be greater than those related to normal aging. Memory changes related to normal aging in adults with DS have been demonstrated, for example, in verbal recall (15) and in ability to commit new information to long-term memory (16). For many higher functioning adults with ID, everyday memory losses will be similar to those observed in the general population (except for memory functions involved in tasks that are too complex or abstract for them to perform initially). For adults performing in the lower ranges, memory loss could be manifested quite differently.

Mild memory loss in adults functioning in the range of mild and moderate ID (IQ range of 40-70), for example, would be indicated by decline in ability to remember the following: (a) social arrangements, such as planned outings and dates made with friends, (b) the location of recently-placed objects, and (c) information imparted by family members or care providers (e.g., commands, chores to do, where care provider has gone for brief periods of time). Similarly, moderate memory loss in adults with mild to moderate ID would be indicated by decline in ability to (a) report the day's events (e.g., what they had for breakfast, where they had been), (b) remember the names of family members, and (c) find their way around their own neighborhood or house. Finally, severe memory loss would be indicated by (a) increasing need for prompting to remember the steps needed to perform previously acquired tasks, and (b) increasing failure to recognize friends and family.

Regarding performance on laboratory tests of memory, premorbid memory functioning for adults with mild to moderate ID would be impaired relative to that in the general population (i.e., their baseline level of performance will be lower) (17-19). In addition, healthy adults with DS often have reduced memory functioning relative
to equally-retarded adults whose ID is due to other causes (20-21). Finally, there can be considerable premorbid intra-individual variability with regard to verbal versus visual memory skills. Losses in memory over time can be demonstrated, however, on measures of both verbal and nonverbal recall and recognition.

For adults functioning in the severe to profound ranges of ID (IQ < 40), premorbid levels of everyday memory function will, of course, be more impaired relative to the general population than for adults in the mild to moderate ranges. Regarding performance on laboratory tests, the ability to assess memory loss will depend upon premorbid verbal skills and/or the ability to meet the demands of memory tasks (e.g., learning to criteria). Memory loss can generally be documented on nonverbal tests (e.g., Dalton (22)), even for those with severe and profound cognitive impairment. For adults performing at the lowest range of profound ID, memory functioning may be so impaired as to preclude detection of any decline on standardized tests. In such cases, change in memory will have to be based on caregiver report.

Decline in Other Cognitive Functions:

ICD-10 criteria for dementia require "a decline in other cognitive abilities characterized by deterioration in judgement 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" (p. 29). Criteria for determining whether decline in other cognitive functions is mild, moderate, or severe, are then presented. DSM-IV enumerates "other cognitive disturbances" as aphasia, apraxia, agnosia, or disturbance in executive functioning. DSM-IV criteria furthermore require that memory and cognitive deficits cause "significant impairment in social or occupational functioning and represent a significant decline from a previous level of functioning." The caregiver's first suspicion that an individual with ID is becoming demented will often be based on changes in adaptive behavior skills. Documentation of decline in adaptive functions, because they are more concrete (e.g., "Can the individual still do his/her own laundry?"), will generally be easier to obtain than documentation of changes in cognitive and memory skills (e.g., "Has the individual's verbal memory declined?").

As is the case for assessment of memory decline, perception of decline in other cognitive functions will depend on the individuals' premorbid level of ID. In order to be indicative of dementia, changes must also be clearly greater than those associated with normal aging. At this time longitudinal evidence suggests that nondemented older adults with DS (> 40 years) maintain function in several areas for relatively long periods (i.e., receptive and expressive language, short term memory, nonverbal reasoning, fine motor, perceptual motor, and sometimes visual spatial skills) (15,16,23). In contrast, slight, gradual decreases (< 1% per year) have been demonstrated in verbal, long-term memory for adults with DS older than 50 (15). Older healthy adults with ID due to causes other than DS have been found to maintain function for six years in short- and long-term memory, speeded psycho-motor function, and visuospatial organization (15).

A decline in abstract reasoning skills, such as judgment, thinking, planning
and organization, may be observed in those with milder ID. For example, decline in judgment would be demonstrated by an individual who can no longer select clothing that is appropriate for the weather. Regarding the areas of planning and organizing, one needs to consider whether the individual was previously able to carry out non-routine tasks (e.g., shopping or complete meal preparation). If the individual was never able to carry out non-routine tasks, documentation of declines in planning and organizing may be extremely difficult. In those who engage only in routine tasks (e.g., dressing, grooming and toileting, setting the table, social greetings), more fundamental cognitive deficits, such as apraxia and aphasia, may be the first deficits to be apparent. For the most severely impaired individuals, decline may be observed primarily as general slowing in all areas and greater impairments in attention. It is also important to note that for all individuals, declines in the ability to focus, maintain, and/or shift attention may mimic or compound impairments in memory and other cognitive functions.

Apraxia may be manifested as impaired ability to dress, self-feed, and brush teeth in individuals who previously demonstrated these skills. Aphasia will be easiest to recognize in those whose premorbid language skills were relatively well developed. In those with poorer verbal skills, evidence of aphasia will range from a gradual decrease in the use of language to the total loss of all verbal expression. Agraphia, alexia, acalculia (losses of ability in writing, reading, and calculation skills, respectively) can be diagnosed in those with the prior requisite skills in these areas. Agnosia may be manifested as inappropriate use of everyday objects (e.g., brushing hair with toothbrush).

In those with more severe ID, increasing temporal or spatial disorientation (e.g., inability to distinguish between day and night or inability to find the bedroom or kitchen) may be more salient than other cognitive deficits. For this reason, assessment of orientation to familiar environments is particularly important in these individuals. Of course, visual and auditory impairment, which occur frequently in older individuals (and particularly often in those with DS) will affect the assessment of orientation, and must be considered.

Awareness of Environment (Delirium): ICD-10 criteria for diagnosis of dementia require an awareness of the environment (i.e., 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. The diagnosis of dementia should be deferred during superimposed episodes of delirium. Diagnosis of delirium in individuals with ID should be made according to the same criteria used for the general population.3 Clinicians should be particularly aware, however, of the possibility of pharmacotoxic reactions that may cause delirium in individuals with ID (25) and pre-existing CNS abnormalities that may predispose these individuals to delirium (26).

Emotional Control, Motivation, or Social Behavior: ICD-10 criteria for dementia require a "decline in emotional control or motivation, or a change in social behavior manifested in at least one of

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3 See Jarvik et al. (24) for a thorough discussion of the diagnosis of delirium in elderly individuals, as well as DSM-IV and ICD-10 criteria.
the following: (1) emotional lability, (2) irritability, (3) apathy, or (4) coarsening of social behavior” (p. 30).

Although cognitive declines are essential for a diagnosis of dementia, changes in other aspects of behavior are highly significant. For individuals with ID, especially those who are very low functioning, these may be the symptoms of dementia that are first noticed by caregivers, as baseline abilities in other areas may be so low that early changes are imperceptible. Individuals with ID and dementia are known to present with a range of psychiatric and behavioral symptoms similar to those of individuals with dementia in the general population (e.g., sleep difficulty, hypersomnia, irritability, loss of interest) (27).

Evaluation of the clinical significance of emotional, motivational, and social symptoms depends upon accurate knowledge of pre-existing behavioral and intellectual functioning. There is a high baseline incidence of behavioral abnormalities (e.g., talking to oneself) in some individuals with ID. Behaviors which commonly occur in individuals with ID, therefore, may be misperceived as being clinically significant, when a carefully taken history would indicate that such behaviors have always been typical for a given individual. Conversely, abnormal behaviors may predate neurological changes of dementia and "overshadow" the true clinical syndrome. That is, all abnormal symptoms may be attributed to ID alone, rather than to a progressive dementing illness. This overshadowing is particularly problematic when professionals are not familiar with the behavior problems that commonly occur in individuals with ID, or when longitudinal information on behavioral progression is unavailable.

**Duration:**
For a confident clinical diagnosis of dementia, ICD-10 requires that decline of memory and other cognitive functions be present for at least six months. If the period since the manifest onset is shorter, the diagnosis must be tentative. Because of the greater degree of variability of cognitive functioning across time for individuals with ID, a diagnosis based on even a six-month decline may be premature. In addition, it may be difficult, in some cases to identify caregivers who have known the individual for a sufficient length of time to warrant a definite diagnosis.

**Criteria for Diagnosis of Alzheimer Disease**

**Exclusionary criteria.**
Consistent with DSM-IV and ICD-10, the proposed criteria for diagnosis of Alzheimer disease in individuals with ID require that the individual meet all criteria for dementia, with evidence for ruling out other specific causes of dementia. ICD-10 states that the diagnosis of AD requires "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" (p. 31). Conditions particularly common in ID should be explored most thoroughly (e.g., hypothyroidism, hearing loss and depression in individuals with DS, folic acid abnormalities in patients on anticonvulsants, and cognitive deterioration secondary to prescribed or non-prescribed medications in all individuals with ID).

Differentiating between depression and AD in individuals with DS is a
particularly important diagnostic challenge, because many symptoms associated with AD in this population are also associated with depression (30-33). Clinical studies (32, 34-35) have found that depressive affect and cognitions are often indirectly expressed by individuals with DS, with the most common symptoms including aggressive acting-out, withdrawal, somatic complaints, increased dependency, irritability, and disturbances of vegetative function. Historically, individuals with DS have sometimes received diagnosis of AD, but when successfully treated for depression have resumed their premorbid levels of cognitive functioning (32). (See NIH Consensus Conference (36) for a more detailed discussion of causes of dementia that can be arrested or reversed.)

Onset and progression.
In addition, for a diagnosis of AD, there must be evidence of gradual onset and continuing cognitive decline. Thus, whereas a diagnosis of dementia can be made for an individual with a static or remitting course, a diagnosis of AD requires a progressive decline. ICD-10 criteria distinguish between early- and late-onset AD (before versus after age 65 years). The average age of onset for clinical manifestations of AD in individuals with DS, according to prospective studies, is between 51 to 54 years of age (37), with no evidence for a bimodal distribution of age of onset. (Although this is an average age of onset, evidence of decline may, of course, appear much earlier.) Age of onset for individuals with ID due to causes other than DS tends to be over 65 (38). There is currently no evidence regarding early and late-onset of AD for individuals with ID due to causes other than DS.

ICD-10 criteria for diagnosis of early-onset AD require evidence of a relatively rapid onset and progression, and evidence of aphasia, agraphia, alexia, acalculia, or apraxia. For a diagnosis of late-onset AD, ICD-10 criteria require evidence of a very slow, gradual onset and progression, with a predominance of memory impairment over intellectual impairment. Further research will be necessary to determine whether individuals with ID whose onset of dementia is before age 65 differ from those with onset after age 65 on rapidity of onset, progression, and pattern of cognitive decline, and thus whether the early- versus late-onset dichotomy is valid in this population.

Definite, probable, possible diagnoses.
ICD-10 and NINCDS-ADRDA criteria for Alzheimer Disease (7) state that a diagnosis of definite AD requires histopathologic confirmation. As stated previously, the histopathologic markers of AD are observed in all individuals with DS over the age of 40 (6). It is currently not possible to distinguish the histopathologic abnormalities in those individuals with DS who demonstrate the clinical signs of dementia from those without the clinical signs (39). For this reason, we are not advocating the confirmation of a diagnosis of AD in individuals with DS through the use of histopathological analysis now in routine usage. Until research provides us with descriptions of histopathological abnormalities specific to those individuals with DS who have clinical features of dementia, we recommend that the diagnosis of AD in this group be based solely on clinical features. It is

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5 We are not, however, suggesting, that clinicians and investigators discontinue histopathological analysis of brains from individuals with DS. We recommend the continued collection and storage of samples of brain tissue in order to allow further investigations regarding the association between clinical and biological findings.
reason able to assume, however, that currently-accepted histopathological evidence of AD is still valid for individuals with ID without DS.

NINCDS-ADRDA criteria for Alzheimer disease (7) state that a diagnosis of probable AD can be made if there is a typical insidious onset of dementia with progression and if there are no systemic or brain diseases that could account for the progressive memory and other cognitive deficits.” The criteria outlined above for the diagnosis of AD in individuals with ID are, therefore, consistent with the NINCDS-ADRDA criteria for probable AD.

Possible AD is diagnosed in the general population according to NINCDS-ADRDA criteria, when "presentation or course is somewhat aberrant" or in the presence of secondary disorders that may produce dementia, but are not considered to be the primary cause of the dementia. It is recommended that these criteria for diagnosis of possible AD be applied in individuals with ID as well.

CLINICAL EVALUATION

The diagnosis of dementia in an individual with ID relies heavily on the clinical evaluation and caregiver interview. Mental status exams that are commonly used to assess dementia in the general population (e.g., the Mini-Mental Scale (40)) are usually inappropriate for individuals with ID because they were designed for individuals whose previous level of cognitive function was assumed to be normal. At this time, there is no mental status exam or neuropsychological instrument that can assess the presence of dementia in an individual with ID based on a single administration. In addition, although neuropsychological testing can be very useful in documenting functional decline over time, the optimal battery for detecting dementia in individuals with ID has not yet been validated.

In practice, the first evidence of decline is usually noticed by the caregiver, who often initiates the referral. It is also the caregiver who usually provides most of the information for the detailed assessment. Unfortunately, caregivers vary in how well they know the individual with ID, how long they have known the individual, and in how well they are able to objectively describe changes in his or her functioning. Family members may describe the individual’s behavior quite differently from a non-family caregiver, and information obtained from even carefully selected informants who are familiar with the individual will vary depending on the nature of their interaction with the patient. For example, informants who see the individual in challenging work environments could report small functional changes as very apparent, whereas others who observe the individual in less demanding leisure environments may not notice similar or even larger declines. Individuals with ID might also display quite different behavior in different situations. It is not uncommon that certain behaviors, such as aggression, occur exclusively or at far greater frequency in a particular environment.

The setting of the assessment itself may affect the results. Settings in which the individual with ID feels comfortable may be more conducive to an optimal assessment. For example, individuals examined in a hospital or clinic setting may be more inhibited and less likely to attempt to respond to questions than those
examined in their home environment. Assessment of some neuropsychological functions (e.g., orientation to place) will obviously yield better performance and more validly reflect actual level of functioning if conducted in an environment that is familiar to the individual with ID.

Ideally, therefore, an assessment for dementia should involve multiple informants, from a variety of situations, and should consider the informant's level of familiarity with the individual being assessed and the nature of the informant's relationship with him or her. At minimum, information from both a work or occupational setting and from a home or social setting should be obtained. Information gained prospectively is ideal, but retrospective information from previous assessments is also invaluable. Finally, it is very important that the examiner be aware of the cultural background of the individual being assessed and his or her family, because what is typical or expected for an individual with ID may vary among cultures. Standardized instruments that are useful for eliciting information from caregivers include the Dementia Questionnaire for Mentally Retarded Persons (DMR (41,42)) and the Dementia Scale for Down Syndrome (43).

**Initial Assessment.**

At an initial assessment, the diagnosis of dementia relies heavily on the retrospective report of the caregiver. In order to make the diagnosis of dementia more feasible (and perhaps at earlier stages), the Working Group recommends that all adults with ID be evaluated using the recommended procedures at least once in early adulthood (by age 25) to establish a record of baseline functioning. With further validation of dementia screens and neuropsychological instruments, it will be possible to establish criteria for diagnosing dementia without relying so heavily on retrospective reports. However, baseline evaluations and periodic screens are still recommended, as it is best for the individual to serve as his or her own point of reference.

All procedures involved in the assessment of dementia in the general population (history, physical, and further investigations) should be followed as closely as possible, with necessary modifications to accommodate the individual with ID. Additionally, the location of the assessment and characteristics of the informant should be recorded. The family history should include queries about intellectual disability, psychiatric illness, and dementing illnesses. Personal history should include results of chromosomal testing (if available), episodes of head-trauma, stereotypic behavior (especially head-banging), seizures, and metabolic dysfunction (especially in persons with DS). A chronological account of the individual’s current problems and psychosocial situation (e.g., current residential placement, work setting, social contacts) should be included. Current and past medication use must be described.

**Cognitive capabilities.**

As mentioned previously, there has not yet been any test developed that can, on the basis of a single administration, diagnose dementia in individuals with ID. Longitudinal administration of tests that assess level of impairment in individuals with ID is, therefore, an absolute necessity before sufficient information can be obtained to make a diagnosis of dementia. Although documentation of decline on cognitive tests is important

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6 See Jarvik et al. (24) for discussion of such an assessment.
in making and confirming a diagnosis of dementia, it must be accompanied by documentation of changes in adaptive functioning before the decline can be considered diagnostic.

Several mental status examinations have been developed for or adapted for use with individuals with ID. The Down Syndrome Mental Status Examination (DSME (44)) assesses a broad range of skills and is easy to administer. Other mental status examinations that have been developed for individuals with ID include those described by Wisniewski & Hill (45) and Thase et al. (46) (see also Davina et al. (47)). Although not specifically designed for individuals with ID, the Test For Severe Impairment (48) is appropriate for individuals who are functioning at a very low level and assesses cognitive functions that are important in the diagnosis of dementia. The standardized administration of a mental status instrument is preferable to less formal assessments of cognitive functioning because it allows confident comparison of results over time. Common use of standardized instruments across research groups will also greatly enhance communication among interested individuals and research centers and promote more valid reporting of study results.

Although performance on standardized instruments can be compared across time in an effort to document cognitive decline, it is not yet possible to state the minimum number or percentage of test "points" that must be lost over a specific time period to warrant a diagnosis of dementia. This is particularly true in the assessment of individuals with ID, whose low level of premorbid functioning may preclude observation of substantial decline. The authors of this article are currently developing a battery of tests7, with a focus similar to that of the CERAD battery (8), which was developed for diagnosing AD in the general population. The battery, which assesses general cognition, memory with and without delay, orientation, expressive and receptive language, fine motor speed, perceptual motor abilities, and adaptive behavior will be administered to a large multi-site sample on an annual basis.

Not surprisingly, it can be challenging to administer tests to individuals with ID, especially those who are becoming demented. As a result, many special accommodations are necessary that may not be required for the general population. Cognitive testing should, therefore, be performed by a psychologist who is familiar with the special testing needs of individuals with ID and who is thoroughly trained regarding the principles of psychometric instrumentation so that he or she can determine what accommodations are appropriate and valid for any individual subject. When possible, of course, it is desirable to have the subsequent testing administered by the same psychologist who performed the initial evaluation.

Functional and vocational abilities. Documentation of dementia in individuals with ID requires that cognitive decline interferes with previous level of social or occupational functioning. Therefore, ability to perform activities of daily living, and changes from previously

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7 A list of tests included in this battery is available from the senior author. Our eventual goal is to determine which of the tests is most useful in detecting decline and the amount of decline on test scores that corresponds to a clinical diagnosis of dementia in adults at each level of intellectual impairment.
higher levels of functioning must be documented. Caregivers should be questioned about the individual's self care skills (grooming, dressing, bathing, feeding, toileting), domestic skills (making the bed, household chores, laundry, meal preparation), work (attention to task, ability to learn and remember new tasks, productivity), travel skills (using the bus, walking to work), and money management (knowing the value of money, making change). As with assessment of cognitive skills, it is recommended that a standardized instrument be used. Available instruments that are appropriate for individuals with ID include the Scales of Independent Behavior (49), Vineland Adaptive Behavior Scales (50), the Adaptive Behavior Scale--Residential and Community (51), and the Disability Assessment Schedule (52). As with the tests of cognitive functioning, these instruments must be administered on a longitudinal basis to document change. When no baseline administration of these instruments has been performed, the clinician will have to rely on retrospective observations by the caregiver regarding changes in adaptive functioning over time. Although such retrospective reports are valuable, clinicians must be aware of their potential lack of reliability.

Behavioral/psychiatric characteristics.
A behavioral/psychiatric assessment is conducted to detect declines in emotional control or motivation, or a change in social behavior, and to identify psychiatric disorders that could affect functioning. Because behavioral/psychiatric changes may be the first indication of dementia, especially in individuals with moderate to severe ID, these must be carefully evaluated. Particularly important are changes from earlier baselines. As in the non-ID population, behavioral/psychiatric symptoms may be misperceived and overly personalized by the caregiver, leading to subjectivity in the description of these symptoms. Thus, direct observation and/or evaluation of the individual with ID is an essential component of the behavioral/psychiatric assessment. Several instruments are available for the assessment of behavioral problems, including the Scales of Independent Behavior (49), Vineland Adaptive Behavior Scales (50), the Adaptive Behavior Scale--Residential and Community (51), the Disability Assessment Schedule (52), and the Aberrant Behavior Checklist (53). (See Aman (54) for a detailed review of the validity, reliability, and appropriate use of these instruments.)

To determine whether behavior problems are severe and/or pervasive enough to indicate the presence of a psychiatric disorder several instruments are available. The Reiss Screen for Maladaptive Behavior (55) is considered an appropriate caregiver-report instrument to screen for psychopathology in individuals with all levels of ID (54). If psychiatric problems are indicated, it is recommended that the individual with ID be referred for complete evaluation and treatment by a psychiatrist/psychologist who has experience working with individuals with ID. It is particularly important that treatable psychiatric disturbance, especially depression (which often mimics the early symptoms of dementia), be diagnosed and properly managed. To establish classical categorical diagnoses the following instruments, considered most suitable for individuals with mild to moderate ID, are useful for obtaining information from the caregiver and individual with ID: the Emotional Problems Scale (56) and the Psychopathology Inventory for
Mentally Retarded (57). Semi-structured clinical interviews of both individuals with ID and their caregivers could also be conducted using the Psychiatric Assessment Schedule for Adults with a Developmental Disability (ICD 10 Version (58)). Finally, the Diagnostic Assessment for the Severely Handicapped II (59) was recently developed to aid in assessing psychopathology in adults with severe to profound levels of impairment. (See Aman (54) for a review of these instruments.)

General physical health/laboratory findings. The physical examination is conducted to detect causes of dementia and coexisting conditions that may contribute to the cognitive decline (36). Physical examination and laboratory studies should meet nationally accepted standards for the evaluation of dementia in the general population (8,24,36,60,61). In addition, supplemental tests should be performed to detect abnormalities that are more common in individuals with ID (e.g., EEG for detection of seizures, and cervical spine X-rays for individuals with DS to rule out atlanto-axial instability). It is particularly important to screen for new-onset hearing and visual impairments, which individuals with ID may be unable to report.

Although standard tests of vision and hearing can be used with individuals who are mildly or moderately cognitively impaired, special procedures will be necessary for more severely impaired individuals. Vision tests applicable for individuals with limited cognitive abilities include adaptations of the Snellen test (62,63), as well as resolution tests (64,65). Hearing in individuals who cannot be tested with behavioral audiometry may be evaluated with acoustic-immittance measures to determine middle ear function, and with auditory evoked potentials to estimate threshold levels of hearing (66,67). Otacoustic emissions is a new technique that may soon be available to augment the current assessment battery (68), but at present its reliability as a method for hearing screening in the ID population has not been established. Additional diagnostic tests may be indicated, based on clinical presentation (e.g., if multi-infarct dementia is suspected, administration of the Hachinski scale (69), followed by CT or MRI if appropriate; blood medication levels for patients on anticonvulsants, lithium, digoxin, or other medications that may affect cognition).

Neurological status. Although the most important part of the neurological examination for individuals suspected of having a dementia is the mental status exam (discussed previously), it is also very important to complete the standard neurological examination (see Adams & Victor, (70)) for localizing signs, paying particular attention to those that are frequently associated with dementia in individuals with ID (12-14). Neurological abnormalities that are more likely to be indicative of dementia in adults with DS include late-onset (i.e., not present during development) seizures (71), pathological reflexes, myoclonus, and gait disturbance (12-14). It is important for the clinician to recognize that the prevalence of neurological abnormalities in nondemented individuals with ID is higher than in the general population (e.g., Habbak et al. (72)), and thus may not necessarily be indicative of dementia. Again, a comparison to pre morbid status is essential.

Establishing diagnostic criteria. The table summarizes the diagnostic criteria and the evaluation procedures that are proposed to determine
whether or not each criterion has been met. Tests listed in the table are being used by at least one of the Working Group members. They are in varying stages of development for use in the population with ID, and thus they do not all have established levels of reliability and validity. Furthermore, it has not yet been established what level of decline in performance is clinically significant, and it should be noted that a statistically significant decline may not correspond to clinically significant decline. The instruments listed in the table do represent the current state of the art and are identified as a starting point for clinicians and researchers, who are encouraged to further explore the utility of these and other instruments for the particular group of individuals with whom they are working.

CONCLUSIONS

We have proposed a standardized set of criteria and procedures for the diagnosis of dementia in individuals with ID. The criteria and procedures were developed by an international group of clinicians and researchers who have experience following elderly individuals with ID. In addition, input was solicited from experts in the fields of aging and dementia in the general population, as well as in ID. Although based on extensive experience and discussion among members of the Working Group, the criteria and procedures should be considered provisional until longitudinal research is able to establish their usefulness.

We urge the adoption of these criteria and procedures for future research and clinical practices, and solicit input from those who use them. Consistent application of the proposed criteria by clinicians and researchers will provide a basis for systematic comparison of observations and ultimately yield improved understanding of the clinical course and natural progression of dementia in this at-risk population. By allowing increased communication among researchers and clinicians, use of these criteria may assist in advancing our understanding of possible risk factors, early signs and symptoms, stages, subtypes, clinical course, and symptom patterns of dementia in individuals with ID, and ultimately improve standards of practice and care for this population.

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