The National Task Group on Intellectual Disabilities and Dementia Practices Consensus Recommendations for the Evaluation and Management of Dementia in Adults With Intellectual Disabilities

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Abstract

Adults with intellectual and developmental disabilities (I/DD) are increasingly presenting to their health care professionals with concerns related to growing older. One particularly challenging clinical question is related to the evaluation of suspected cognitive decline or dementia in older adults with I/DD, a question that most physicians feel ill-prepared to answer. The National Task Group on Intellectual Disabilities and Dementia Practices was convened to help formally address this topic, which remains largely underrepresented in the medical literature. The task group, comprising specialists who work extensively with adults with I/DD, has promulgated the following Consensus Recommendations for the Evaluation and Management of Dementia in Adults With Intellectual Disabilities as a framework for the practicing physician who seeks to approach this clinical question practically, thoughtfully, and comprehensively.

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The National Task Group on Intellectual Disabilities and Dementia Practices (NTG) was formed as a response to the National Alzheimer's Project Act, legislation signed into law by President Barack Obama. One objective of the NTG is to highlight the additional needs of individuals with intellectual and developmental disabilities (I/DD) who are affected or will be affected by Alzheimer's disease and related disorders. The American Academy of Developmental Medicine and Dentistry, the Rehabilitation Research and Training Center on Aging With Developmental Disabilities—Lifespan Health and Function at the University of Illinois at Chicago, and the American Association on Intellectual and Developmental Disabilities combined their efforts to form the NTG to ensure that the concerns and needs of people with intellectual disabilities and their families, when affected by dementia, are and continue to be considered as part of the National Plan to Address Alzheimer's Disease[1] issued to address the requirements of the National Alzheimer's Project Act.

Among the NTG's charges were (1) the creation of an early detection screen to help document suspicions of dementia-related decline in adults with intellectual disabilities, (2) the development of practice guidelines for health care and supports related to dementia in adults with intellectual disabilities, and (3) the identification of models of community-based support and long-term care of persons with intellectual disabilities affected by dementia. In 2012, the NTG issued "'My Thinker's Not Working': A National Strategy for Enabling Adults With Intellectual Disabilities Affected by Dementia to Remain in Their Community and Receive Quality Supports."[2]

A subgroup of the NTG was formed to focus specifically on health practices. The guidelines and recommendations outlined in this document represent the consensus reached among said specialists at 2 plenary meetings and ongoing discussions that followed, informed by a review of the current literature and drawn

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from each specialist's clinical practice; thus, they meet level 2 (case-controlled studies) and level 3 (observational studies) for evidence in clinical application. These guidelines are a suggested starting point as we develop more formal methods to determine best practices of evaluation of dementia in this population.

BACKGROUND

Adults with I/DD are now regularly living into old age, with many surviving into their 70s, 80s, and beyond. Dementia is among the most clinically challenging co-occurring conditions of aging in a select group in this population (ie, adults with Down syndrome and those with brain injury) considering that the approach to evaluation, diagnosis, treatment, and management of dementia in adults with I/DD remains largely undefined in the literature.

It is well established that adults with I/DD experience poorer health outcomes compared with the general population, a trend seen in mortality, morbidity, and quality of life.[3,4] The cause of this disparity is complex and multifactorial, but poor training and preparedness of health care professionals nationwide ranks among the key contributing factors. Formal didactic training regarding adults with I/DD throughout the life span is not routinely incorporated into US medical school or residency training.[5]

Recognizing these existing disparities in medical training and health care services for adults with I/DD, the NTG organized a targeted effort to help address this gap. The goal of this article is to clarify key principles of evaluation and management of dementia in adults with I/DD (≥20 years old) on the basis of evidence-based research and consensus among experts in the NTG.

PREVALENCE OF DEMENTIA IN PATIENTS WITH INTELLECTUAL DISABILITY

In Down syndrome, one of the most common forms of intellectual disability, the underlying genetic link between trisomy 21 and Alzheimer's disease has been convincingly established.[6,7] By age 40 years, all adults with Down syndrome exhibit some degree of neuropathologic defects postmortem that meet the criteria for Alzheimer's disease.[8,9] Despite these neuropathologic changes, the development of clinical Alzheimer's disease is variable. Although specific prevalence estimates may vary, it is generally accepted that at least 50% of adults aged 60 years and older will have clinical evidence of dementia.[10-12] Thus, the development of clinical Alzheimer's disease is not inevitable in aging adults with Down syndrome, although risk increases incrementally with age.

In these practice guidelines, we take an encompassing approach and generalize recommendations for the broad population with I/DD, recognizing that distinct genetic and neurologic factors associated with specific conditions may compromise these generalizations.

SPECIFIC CHALLENGES IN THE AGING POPULATION WITH I/DD

One of the hallmark features of all causes of dementia is a decline from the baseline level of function and performance of daily skills. Although this is usually relatively straightforward to establish in the general population, it can be a much more complicated task in adults with intellectual disabilities because of variance in cognitive functioning. This is particularly true for the current generation of older adults with I/DD owing to a variety of factors, including poor record keeping from childhood, lack of ongoing involvement of family and involvement of multiple staff members (often due to a high degree of turnover), and inconsistencies in the physician-patient relationship that obviate knowing the person throughout his or her life span.[13]

In the absence of a personal historian who can accurately and comprehensively attest to an individual's baseline level of functioning, the assessment of a reported or observed change may be exponentially more complicated. The early signs of dementia in adults with I/DD can be subtle and often require an astute observer to identify these changes proactively. Often individuals with I/DD are served by numerous caregivers throughout their lifetime, and often newly involved caregivers will presume that their current level of ability represents their baseline level of functioning and, thus, miss signs of early decline.[14]

DOWN SYNDROME AND OTHER FORMS OF INTELLECTUAL DISABILITY

Down syndrome is the only genetically inherited form of intellectual disability that has been indisputably linked to the risk of early development of Alzheimer's disease. The leading explanation for this link is tied to the triplication

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of chromosome 21 (trisomy 21) and the overexpression of the gene coded on this chromosome for amyloid precursor protein. The excessive production of β-amyloid as a result is key to the pathogenesis of Alzheimer's disease. In addition, other genes coded on chromosome 21 are also theorized to potentially contribute to the early emergence of dementia and the phenomenon of accelerated aging seen in adults with Down syndrome.[15,16] Down syndrome has the most robust body of literature regarding the correlation to dementia in persons with I/DD in older age, although many unanswered and underexplored questions still remain. Comparatively, for nearly all other forms of I/DD, there is a widespread scarcity of life span research.[17]

ASSESSMENT GUIDELINES

The NTG recommends the following procedural steps for the accurate assessment of health and function regarding the identification and validation of symptoms of Alzheimer's dementia and related disorders. The steps include evaluation, diagnosis, treatment, and follow-up. As noted in the National Plan to Address Alzheimer's Disease, physicians and other health care professionals need information on how to implement the "detection of any cognitive impairment" requirement in the Medicare Annual Wellness Visit included in the Affordable Care Act.[18,19] The following guidelines also address this process and offer suggestions on how to meet this requirement.

Evaluation

The history is the cornerstone of a dementia diagnosis. It is important that a thorough and comprehensive history be obtained to compile evidence consistent with an emerging dementia while probing for other features or patterns that might suggest other contributing factors. Pertinent historical information is particularly useful from personal accounts of caregivers and family members who have known the individual for several years. In addition, other sources of information, such as previous neuropsychological testing or school Individualized Education Program information, can greatly assist in accurately characterizing an individual's baseline. One model for obtaining such a history is detailed in a separate NTG publication regarding community supports.[20] To perform the evaluation process, we recommend the following 9-step approach.

Step 1: Gather a Pertinent Medical and Psychiatric History. A thorough history should include, of course, a review of the medical and psychiatric history, with particular attention to issues of the patient's personal health that could potentially influence the likelihood of development of a premature dementia. These issues include a history of cardiovascular disease; a history of cerebrovascular disease or stroke; known underlying neurologic structural abnormalities; a history of head injury, concussion, or loss of consciousness; and poorly treated sleep disorders or thyroid disease, vitamin B_{12} deficiency, or metabolic syndrome (obesity, diabetes, hypertension).

Step 2: Obtain a Historical Description of Baseline Functioning. As discussed earlier, a dementia diagnosis requires evidence of change from a previous level of functioning, and this baseline is quite individualized in people with I/DD. This description is highly dependent on the historian, and, thus, a family member or caregiver who knows the individual well should be present for this interview. Supplemental Table 1 (available online at http://www.mayoclinicproceedings.org) outlines the key factors to review when constructing a historical baseline for an individual undergoing assessment for dementia.

Step 3: Obtain a Description of Current Functioning and Compare With Baseline. Information gathering regarding current functioning is obtained in a similar manner as was done for the historical baseline, allowing a side-by-side comparison of the scope and degree of change over time. This practice helps systematically assess for the key criteria of dementia, specifically, "dementia is diagnosed when there are cognitive or behavioral symptoms that 1) interfere with the ability to function at work or at usual activities; and 2) represent a decline from previous levels of functioning and performing."[21] Key features to look for include reported memory loss or impairment, marked changes in personality, disorientation, and decreasing performance in expected tasks/skills.

Step 4: Perform a Focused Review of Systems. A focused review should take an inventory of common issues that are seen with increasing age and also with possible emerging

TABLE 1. Common Medication Classes Associated With Possible Worsening of Cognitive Function in Patients With Dementia

Medication class	Examples	Comments
Antihistamines, especially first generation	Diphenhydramine Hydroxyzine Promethazine	Anticholinergic adverse effects, urine retention, confusion, sedation
Bladder agents	Oxybutynin Tolterodine	Anticholinergic adverse effects, urine retention, confusion, sedation
Certain pain medications	Meperidine Propoxyphene	Meperidine: increased risk of seizures with renal impairment
Tricyclic antidepressants	Amitriptyline Clomipramine Doxepin	Risks and benefits of this medication class should be guided by a psychiatrist with familiarity with patients with I/DD
Certain antipsychotics	Chlorpromazine Clozapine Pimozide	Sedation, mental sluggishness. Atypical antipsychotics have been associated with increased mortality when used to treat behavioral problems in elderly patients with dementia, but no such studies have been conducted in Down syndrome or I/DD in general
Long-acting benzodiazepines	Clonazepam Temazepam Diazepam	Very sedating; caution for gait impairment, dizziness If a benzodiazepine is required for anxiety, consider short-acting agents (appropriately dosed): alprazolam, lorazepam

I/DD = intellectual and developmental disabilities.

dementias. Supplemental Table 2 (available online at http://www.mayoclinicproceedings.org) summarizes the system areas that should be highlighted in an assessment of an adult with I/DD.

Step 5: Review the Medication List Thoroughly. Medications require specific focus, as the risk associated with polypharmacy and the involvement of multiple prescribing physicians increases with advancing age. Special attention should be given to all newly added medications, particularly those that are psychoactive, antiepileptic, or anticholinergic, and those with sedating properties. Common adverse effects and drug-drug interactions should be reviewed, with attention given to nonspecific signs and symptoms that might suggest a drug adverse effect, such as somnolence, gait instability, or urinary retention. Table 1 lists commonly used medications with potential deleterious effects on cognition in this population.

Step 6: Obtain a Pertinent Family History. A family history is important primarily for detecting a history of dementia in first-degree relatives, particularly if the disease presented prematurely (generally <50 years old, except in adults with Down syndrome), suggesting a stronger possible familial predisposition. Additional factors of note include a history of cerebrovascular disease, stroke, diabetes, heart disease, and rheumatoid arthritis or systemic lupus erythematosus in first-degree relatives.

Step 7: Assess for Other Psychosocial Issues or Changes. Aging adults with I/DD often confront a variety of potentially destabilizing life events merely by virtue of growing older. These events may include leaving the family home; witnessing the declining health or death of a parent, loved one, or friends/housemates; decline in one's own health, functionality, or employment status; or frequent turnover/departure of caregivers. The cumulative impact of these events in an adult with I/DD who may have limited coping skills or emotional maturity cannot be overstated. A careful assessment for these factors could identify certain triggering events and frequently also help in the identification of other coexisting mood disorders, such as untreated anxiety or depressed mood, which could be strongly influencing an individual's cognitive and functional performance.

Psychiatric illness may present atypically in adults with I/DD, and there is a common pitfall of diagnostic overshadowing, in which features truly related to an underlying mental illness are instead attributed to the individual's intellectual disability. The method of history taking described previously herein helps account for changes in baseline mood, behavior, or personality, which should raise red flags for

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a possible emerging psychiatric/mental health disorder. Pay close attention to the review of systems to highlight other potential neurovegetative signs of appetite changes, weight loss, and change in sleep patterns.

In patients with Down syndrome and dementia, depression and other prefrontal lobe symptoms may be more common, including signs and symptoms of indifference, uncooperativeness, apathy, and socially deficient communication or impaired adaptive functioning in general.[22] For a review of features of mental disorders that may present atypically in adults with I/DD, the *Diagnostic Manual—Intellectual Disability* is a particularly helpful desk reference.[23]

Step 8: Review Social History, Living Environment, and Level of Support. Assessment of current living conditions and level of support is increasingly critical when an emerging dementia is suspected, as evaluation of safety concerns and appropriateness of one's current placement is a priority. Even if current supports seem adequate, it is important to think proactively in the setting of a suspected dementia, knowing that there will be ever-changing and increasing needs.

Step 9: Synthesize the Information. Because the history and all the supporting data are obtained in a stepwise manner, the interviewer is advised to continually mentally cross-reference the information with the criteria for a dementia diagnosis, building evidence for or against the diagnosis. By the end of the history taking, a fairly strong level of suspicion should already be built.

Physical Examination

The physical examination should be comprehensive, with specific attention to physical findings that may suggest underlying medical issues that may be contributory. Supplemental Table 3 (available online at http://www.mayoclinicproceedings.org) summarizes the key components of focus during an office examination for suspected dementia.

Cognitive Assessment

Currently, there is no generally accepted criterion standard for memory screening or assessment in adults with I/DD. Assessment tools used in the general population (eg, the Folstein Mini-Mental State Examination and the Montreal Cognitive Assessment; see also the study by Cordell et al[24] for a recent compendium of assessment tools appropriate for the Annual Wellness Visit under the Affordable Care Act) have not been normed for adults with intellectual disabilities, and, thus, results cannot be interpreted meaningfully in adults with I/DD. The NTG has made the following recommendations for a general framework of cognitive assessment.

Inclusion of at least 1 standardized tool for cognitive assessment is recommended because it generates a score that can be tracked over time and provides an additional rigid data point that can be repeated on subsequent encounters. Many instruments have been developed and validated for the diagnosis of dementia in this population, including the Dementia Scale for Down's Syndrome,[25] the Dementia Questionnaire for People With Learning Disabilities,[26,27] the Cambridge Examination for Mental Disorders of Older People With Down's Syndrome,[28] the Down's Syndrome Mental State Examination,[29] and the Test for Severe Impairment.[30] Regardless of the clinician's choice of instrument, the focus should be on recognizing change and decline in relation to a premorbid baseline.

In addition, the initial interview and examination can be adapted to assess performance in a variety of cognitive domains. This could be performed in the office or at the bedside if questions are flexible enough to be appropriate for the individual's baseline intellectual ability. To further enhance the information provided by the test batteries suggested previously herein, physicians are encouraged to include additional questioning to assess the individual's general orientation, reading/writing/math skills (if applicable), naming abilities (body parts, common objects), basic motor skills, general knowledge (counting, days of the week), language comprehension, and recall of newly learned information. All these components can be fairly easily added to a physician's questioning repertoire and can help point to changes if abilities decay over subsequent visits.

Diagnosis

There exists great heterogeneity in adults with I/DD, and, thus, there is no basis for comparison

TABLE 2. Common Contributors to Memory Changes in Adults With I/DD

Condition	Presentation
Sensory deficits	Hearing loss Vision loss, low vision, depth perception changes
Metabolic disturbances	Electrolyte abnormalities Hypoglycemia/hyperglycemia B_{12} or folate deficiencies Undetected thyroid dysfunction Anemia Toxic levels of antiepileptic or psychoactive medications Toxic adverse effects of certain medications (eg, hyperammonemia in chronic valproic acid use)
Coexisting mood disorder	Either newly detected or subacute worsening of baseline mood disorder Note: Depression can cause symptoms that seem similar to dementia and can co-occur with early dementia
Pharmacologic concerns	Polypharmacy, drug-drug interactions, and altered pharmacokinetic properties
Sleep problems	Sleep apnea and other undetected sleep disorders
Seizures	Undetected or worsening seizure disorders
Pain	Undiagnosed pain or undertreated pain
Mobility problems	Mobility disorders and loss of functionality
Psychosocial or environmental stressors	Changes in routines, death or impairment of family members or close acquaintances, new regimen at home or in the workplace, reactions to threatening situations
Others	Conditions that may be associated with cognitive deficit (chronic subdural hematoma, brain tumors, multiple sclerosis, human immunodeficiency virus, and cryptococcal infection)
Additional considerations: prevalent conditions in adults with Down syndrome	Vision impairment due to early development of cataracts and increased risk of keratoconus Hearing loss due to conductive hearing deficits Thyroid dysfunction, particularly hypothyroidism Obstructive sleep apnea Celiac disease Atlantoaxial instability and other cervical spine disorders, including osteoarthritis and spinal stenosis Osteoarthritis and associated pain and mobility limitations

I/DD = intellectual and developmental disabilities.

against any other set of peer-based percentiles, standards, or generalized sets of expectations when assessing an individual for dementia. Assessment of decline should always be individualized and patient specific, with judgments made on the basis of deterioration from the patient's own individual baseline level of intellectual disability, function, and achievement (ie, their personal best).

A dementia diagnosis should never be arrived on prematurely, without a proper investigation into other contributing factors that are potentially correctable. There are several co-occurring issues that frequently masquerade as dementia or negatively compound the effects of early dementia. Table 2 summarizes these common conditions, which should be assessed during a general evaluation in an effort to identify contributors and, more importantly, to look for elements that can be treated or improved.

The workup after an office cognitive assessment should be customized to the specific signs and concerns of each individual patient. Further assessment of 1 or more of the previously noted components may comprise the "homework" that a patient's caregiver or referral agency may be instructed to complete after the first assessment. Laboratory testing is justifiable in nearly every patient seeking cognitive assessment because blood work has a potentially high yield for uncovering or ruling out multiple common conditions that might influence cognition. A complete metabolic panel, thyroid function tests, B_{12} measurement, folate measurement, liver function tests, and a complete blood cell count should routinely be performed. Use of neuroimaging is recommended on a case-by-case basis, and features that should prompt consideration of brain imaging include focal findings on neurologic examination,

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TABLE 3. Cholinesterase Inhibitor Use in Adults With Down Syndrome With and Without Dementia

Reference, year	No. of patients	Study type	Results
Kishnani et al,[36] 1999	4	Case reports	Improvement
Prasher et al,[37] 2013	23	Case control	Nonsignficant improvement
Heller et al,[38] 2003	6	Case reports	Improvement in language
Johnson et al,[39] 2003	19	Randomized controlled trial	Improvement in language
Prasher et al,[40] 2002	27	Randomized controlled trial	Nonsignificant improvement
Lott et al,[41] 2002	15	Case control	Significant improvement

underlying known structural abnormalities without recent imaging, suspicion of or risk factors for cerebrovascular disease, rapid and sudden neurologic/cognitive deterioration, a history of recent fall with head injury, or a history of repeated head trauma. Particularly in patients in whom the history is fragmented, treatable causes of functional decline, such demyelinating disease, infection, and intracranial mass, may be missed if imaging is not considered. Thus, proceeding with magnetic resonance imaging or noncontrast computed tomography is recommended, particularly if select brain features may lead to a differential diagnosis of the type of dementia.

Recommendations for additional testing or procedures are patient specific, on the basis of the components listed previously herein regarding suspicion for other contributing factors. These procedures may commonly include, but are not limited to, ophthalmology testing, cerumen disimpaction, audiology testing, referral for sleep studies, consideration of an antidepressant drug trial (if symptoms are present), referral to a psychiatrist, therapist, or behaviorist, electroencephalography, echocardiography (if an arrhythmia or dysautonomia is suspected), or additional recommendations to minimize polypharmacy.

Treatment

Treatment of dementia involves a pharmacologic and a nonpharmacologic approach. The pharmacologic treatment of dementia may include medications that are meant to slow the progression of cognitive decline, as methods of neuroprotection and curative treatment are not yet available. Medications may also be used to help with challenging behaviors in adults with I/DD and dementia. Treating affective behavior and psychotic behavior is a challenge that may improve the individual's function, but the benefits must be carefully weighed against potential adverse effects, and one may consider partnering with a psychiatrist for further expert guidance in this situation.

Data specifically focused on pharmacologic treatment interventions for adults with I/DD (and Down syndrome specifically) are limited, with many studies flawed by small sample sizes, nonblinded study design, and variable inclusion criteria for dementia. The current Food and Drug Administration–approved medications for Alzheimer's disease either raise the levels of acetylcholine (donepezil, rivastigmine, and galantamine) or block the activity of the neurotransmitter glutamate in the brain (memantine). In 2009, the Cochrane Collaboration[31-34] conducted reviews of donepezil, rivastigmine, galantamine, and memantine and their use in treating adults with Down syndrome and found that there is a dearth of rigorous data in this field, as only 1 study was identified that met the criteria for review. Furthermore, a 2011 study that focused on memantine therapy in adults with Down syndrome provided discouraging results, with no significant improvement noted in the treatment group vs the placebo group at 1-year follow-up.[35] Table 3 summarizes some of the limited studies of the effect of cholinesterase inhibitors on Down syndrome.

There is little evidence in the current literature about the efficacy, safety, and tolerability of pharmacologic interventions for dementia in adults with I/DD. Current practice is largely variable and is typically extrapolated from standard treatment principles applied to the general population with dementia. The question of efficacy and response to cholinesterase inhibitor and memantine treatment in the general population is challenging in and of itself, with outcomes often reflected as a modest

improvement in cognitive subscale values and measurements of functionality.

Other treatment options are dictated by what is uncovered in the workup of other contributing factors. Treatment of these issues may be quite gratifying, as some underlying issues may be highly improvable, such as correction of vision impairment; cerumen disimpaction; amplification for hearing deficits; initiation of antidepressant drug therapy; initiation or adjustment of thyroid supplementation; adjustment, elimination, or dose reduction of problematic medications; initiation of continuous positive airway pressure or other noninvasive measures for improvement of sleep disorders; behavioral or environmental modifications; and treatment for suspected underlying pain, discomfort, and mobility difficulties. Occupational and physical therapy consults should be considered in helping to enable the individual and care staff to help with current difficulties in activities of daily living and in trying to sustain current levels of function.

Pharmacologic treatment of dementia composes only a portion of the treatment plan. Most of the treatment approach should be nonpharmacologic, via communication and environmental and behavioral strategies. This topic is addressed and outlined in rigorous detail in the article by Jokinen et al,[20] another publication by the NTG.

Follow-up

At the conclusion of the initial assessment, it is recommended that a follow-up visit be planned to review the results of requested studies and to assess response to any recommended interventions. If there is any question, it is often prudent to suspend any formal diagnosis of dementia until at least the second meeting to allow for proper investigation into other contributing factors in the intervening months.

There is no consensus or formal framework by which a physician treating an individual with I/DD can judge the response to dementia treatment. Outcome measures used in drug studies for adults with Down syndrome and dementia typically include measures of cognition, neuropsychiatric features, adaptive behavior, and scores on subsequent standardized testing. Certainly, assessment of adherence to and tolerance of these medications should be a priority in all first follow-up visits. Thereafter, response to medication and decision making for continuing treatment is judged primarily on the basis of subjective and objective findings during the interview and follow-up memory testing. Because the impact of these drugs is often subtle and the theoretical mechanism is to slow the progression of disease, if the patient is tolerating the medication, it is often generally justifiable to continue treatment until at least moderate-stage disease or beyond.

During the disease course, regular support and education are critical. It is helpful to provide a general estimate of the stage of the disease (early, middle, or late stage) to the caregiver to help provide stage-specific education and expectations. Early-stage disease warrants counseling regarding communication strategies, modification of expectations at home/day/work programs, safety concerns, behavior or personality changes, and adapting the level of supervision and support to account for short-term memory loss.

Anticipatory guidance is necessary to prepare caregivers for changes and for setting realistic expectations going forward. In the early stages of disease, it is recommended to educate caregivers on certain features that commonly accompany progression to midstage, including dysphagia, mobility impairments, new or worsening urinary incontinence, and seizures. When the individual begins to exhibit changes consistent with midstage disease, anticipatory guidance should shift to more goal-directed discussions about future planning, advance directives, and goals of care. Having this discussion proactively allows the caregiver to consider scenarios that may be encountered in late-stage disease while avoiding having to make decisions in a crisis situation.

One important complicating factor that adds a considerable layer of complexity to this discussion when caring for individuals with I/DD is the issue of guardianship or proper identification of a decision maker. The discussion described previously herein would ideally be held in-person during an office encounter with the health care proxy or guardian present. It is common for some older adults with I/DD to have a court-appointed guardian or health care proxy who may be a family member or someone else. Thus, engaging the decision maker in this important discussion may often

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be easier said than done but is nonetheless extremely important.

NEED FOR FUTURE RESEARCH AND STUDY

As more individuals with I/DD regularly reach old age, the need for further life span research in this field cannot be overstated. There is a dearth of syndrome-specific information that can provide insight into common coincident conditions encountered as individuals progress into adulthood and old age. There remains conflicting information and uncertainty about the relative risk of dementia for adults with all forms of intellectual disability, and this is a major barrier to proper assessment, risk stratification, and guidance. The emerging field of adult developmental medicine has extensively fertile grounds for research in basic science and clinical areas, especially clinical trials. This field remains small in size but large in passion and commitment. Consensus guidelines are necessary to help standardize assessment practices among health care professionals performing memory assessments in this growing population, and this is one of the primary aims of the NTG. In addition, continued training on life span issues for adults with I/DD needs to be incorporated into the general medical training for providers of adult medicine, and there are other national efforts under way in this regard. Last, the efforts of specialists currently working in this field need to be harnessed in a formal manner and supported through funding opportunities so that efforts such as those undertaken by the NTG can be continued.

RECOMMENDATIONS

Similar to other dementia guidelines, the National Task Group Consensus Recommendations emphasize a stepwise and comprehensive assessment of suspected cognitive decline. However, in adults with I/DD, history is of paramount importance, given the great heterogeneity of individual baseline or premorbid functioning. In addition to interviewing the patient, health care professionals are recommended to engage multiple informants who know the individual well and to use additional collateral historical information as available. Finally, the NTG urges health care professionals to arrive on the diagnosis of dementia systematically and thoughtfully, so as not to prematurely close a window of opportunity to discover potentially modifiable and treatable coexisting conditions.

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SUPPLEMENTAL ONLINE MATERIAL

Supplemental material can be found online at http://www.mayoclinicproceedings.org.

Abbreviations and Acronyms: **I/DD** = intellectual and developmental disabilities; **NTG** = National Task Group on Intellectual Disabilities and Dementia Practices

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REFERENCES

1. US Department of Health and Human Services. National plan to address Alzheimer's disease. http://aspe.hhs.gov/daltcp/napa/NatlPlan.pdf. Accessed June 13, 2013.
2. National Task Group on Intellectual Disabilities and Dementia Practices. "My thinker's not working": a national strategy for enabling adults with intellectual disabilities affected by dementia to remain in their community and receive quality supports. http://aadmd.org/ntg/thinker. Accessed June 13, 2013.
3. Krahn GL, Hammond L, Turner A. A cascade of disparities: health and health care access for people with intellectual disabilities. *Ment Retard Dev Disabil Res Rev*. 2006;12(1):70-82.
4. Perkins EA, Moran JA. Aging adults with intellectual disabilities. *JAMA*. 2010;304(1):91-92.
5. Krahn GL, Drum CE. Translating policy principles into practice to improve health care access for adults with intellectual disabilities: a research review of the past decade. *Ment Retard Dev Disabil Res Rev*. 2007;13(2):160-168.
6. Janicki MP, Dalton AJ, eds. *Dementia, Aging, and Intellectual Disabilities: A Handbook*.. Philadelphia, PA: Brunner-Mazel; 1999.
7. Lott IT, Head E. Down syndrome and Alzheimer's disease: a link between development and aging. *Ment Retard Dev Disabil Res Rev*. 2001;7(3):172-178.
8. Hof PR, Bouras C, Perl DP, et al. Age-related distribution of neuropathologic changes in the cerebral cortex of patients with Down syndrome: quantitative regional analysis and comparison with Alzheimer's disease. *Arch Neurol*. 1995;52(4):379-391.

9. Mann DM, Yates PO, Marciniuk B. Alzheimer's presenile dementia, senile dementia of Alzheimer type and Down syndrome in middle age form an age-related continuum of pathological changes. *Neuropathol Appl Neurobiol*. 1984;10(3):185-207.
10. Lai F, Williams RS. A prospective study of Alzheimer disease in Down syndrome. *Arch Neurol*. 1989;46(8):849-853.
11. Prasher VP. Age-specific prevalence, thyroid dysfunction and depressive symptomatology in adults with Down syndrome and dementia. *Int J Geriatr Psychiatry*. 1995;10(1):25-31.
12. Visser FE, Aldenkamp AP, van Huffelen AC, Kuilman M. Prospective study of the prevalence of Alzheimer-type dementia in institutionalized individuals with Down syndrome. *Am J Ment Retard*. 1997;101(4):400-412.
13. Test DW, Flowers C, Hewitt A, Solow J. Statewide study of the direct support staff workforce. *Ment Retard*. 2003;41(4):276-285.
14. Aylward EH, Burt DB, Thorpe LU, Lai F, Dalton A. Diagnosis of dementia in individuals with intellectual disability. *J Intellect Disabil Res*. 1997;41(pt 2):152-164.
15. Prasher V. Genetics, Alzheimer's disease, and Down syndrome. In: *Down Syndrome and Alzheimer's Disease: Biologic Correlates*. London, UK: Radcliffe Publishing; 2006:37-57.
16. Zigman WB, Lott IT. Alzheimer's disease in Down syndrome: neurobiology and risk. *Ment Retard Dev Disabil Res Rev*. 2007;13:237-246.
17. Janicki MP, Henderson CM, Rubin L. Neurodevelopmental conditions and aging: report on the Atlanta Study Group Charette on Neurodevelopmental Conditions and Aging. *Disabil Health J*. 2008;1(2):116-124.
18. Department of Health and Human Services, Centers for Medicare & Medicaid Services. Providing the annual wellness visit (AWV). http://www.cms.gov/Outreach-and-Education/Medicare-Learning-Network-MLN/MLNProducts/Downloads/AnnualWellnessVisit-ICN907786.pdf. Published July 2012. Accessed June 13, 2013.
19. Borson S, Frank L, Bayley PJ, et al. Improving dementia care: the role of screening and detection of cognitive impairment. *Alzheimers Dement*. 2013;9(2):151-159.
20. Jokinen N, Janicki MP, Keller SM, et al; National Task Group on Intellectual Disabilities and Dementia Care Practices. Guidelines for structuring community care and supports for people with intellectual disabilities affected by dementia. *J Policy Pract Intellect Disabil*. 2013;10(1):1-24.
21. McKhann GM, Knopman DS, Chertkow H, et al. The diagnosis of dementia due to Alzheimer's disease: recommendations from the National Institute on Aging and the Alzheimer's Association Workgroup. *Alzheimers Dement*. 2011;7(3):263-269.
22. Ball SL, Holland AJ, Hon J, Huppert FA, Treppner P, Watson PC. Personality and behaviour changes mark the early stages of Alzheimer's disease in adults with Down's syndrome: findings from a prospective population-based study. *Int J Geriatr Psychiatry*. 2006;21(7):661-673.
23. Fletcher R, Loschen E, Stavrakaki C, First M, eds. *Diagnostic Manual—Intellectual Disability (DM-ID): A Textbook of Diagnosis of Mental Disorders in Persons With Intellectual Disability*. Kingston, NY: NADD Press; 2007.
24. Cordell CB, Borson S, Boustani M, et al; Medicare Detection of Cognitive Impairment Workgroup. Alzheimer's Association recommendations for operationalizing the detection of cognitive impairment during the Medicare Annual Wellness Visit in a primary care setting. *Alzheimers Dement*. 2013;9(1):1-10.
25. Gedye A. *Dementia Scale for Down's Syndrome Manual*. Vancouver, BC, Canada: Gedye Research and Consulting; 1995.
26. Evenhuis HM. Evaluation of a screening instrument for dementia in ageing mentally retarded persons. *J Intellect Disabil Res*. 1992;36(pt 4):337-347.
27. Evenhuis HM, Kengen MMF, Eurlings HAL. Dementia questionnaire for people with learning disabilities (DLD): UK adaptation 2007. http://www.pearsonclinical.co.uk/SearchResults.aspx?keywords=evenhuis. Accessed June 13, 2013.
28. Ball SL, Holland AJ, Huppert FA, Treppner P, Watson P, Hon J. The modified CAMDEX informant interview is a valid and reliable tool for use in the diagnosis of dementia in adults with Down's syndrome. *J Intellect Disabil Res*. 2004;48(pt 6):611-620.
29. Haxby JV. Neuropsychological evaluation of adults with Down's syndrome: patterns of selective impairment in non-demented old adults. *J Ment Defic Res*. 1989;33(pt 3):193-210.
30. Albert M, Cohen C. The Test for Severe Impairment: an instrument for the assessment of patients with severe cognitive dysfunction. *J Am Geriatr Soc*. 1992;40(5):449-453.
31. Mohan M, Carpenter PK, Bennett C. Donepezil for dementia in people with Down syndrome. *Cochrane Database Syst Rev*. 2009;(1):CD007178.
32. Mohan M, Bennett C, Carpenter PK. Rivastigmine for dementia in people with Down syndrome. *Cochrane Database Syst Rev*. 2009;(1):CD007658.
33. Mohan M, Bennett C, Carpenter PK. Galantamine for dementia in people with Down syndrome. *Cochrane Database Syst Rev*. 2009;(1):CD007656.
34. Mohan M, Bennett C, Carpenter PK. Memantine for dementia in people with Down syndrome. *Cochrane Database Syst Rev*. 2009;(1):CD007657.
35. Hanney M, Prasher V, Williams N, et al. Memantine for dementia in adults older than 40 years with Down's syndrome (MEADOWS): a randomised, double-blind, placebo-controlled trial. *Lancet*. 2012;379(9815):528-536.
36. Kishnani PS, Sullivan JA, Walter BK, Spiridigliozzi GA, Doraiswamy PM, Krishnan KR. Cholinergic therapy for Down's syndrome. *Lancet*. 1999;353(9158):1064-1065.
37. Prasher VP, Sachdeva N, Adams C, Haque MS. Rivastigmine transdermal patches in the treatment of dementia in Alzheimer's disease in adults with Down syndrome: pilot study. *Int J Geriatr Psychiatry*. 2013;28(2):219-220.
38. Heller JH, Spiridigliozzi GA, Sullivan JA, Doraiswamy PM, Krishnan RR, Kishnani PS. Donepezil for the treatment of language deficits in adults with Down syndrome: a preliminary 24-week open trial. *Am J Med Genet A*. 2003;116A(2):111-116.
39. Johnson N, Fahey C, Chicoine B, Chong G, Gitelman D. Effects of donepezil on cognitive functioning in Down syndrome. *Am J Ment Retard*. 2003;108(6):367-372.
40. Prasher VP, Huxley A, Haque MS; Down syndrome Ageing Study Group. A 24-week, double-blind, placebo-controlled trial of donepezil in patients with Down syndrome and Alzheimer's disease: pilot study. *Int J Geriatr Psychiatry*. 2002;17(3):270-278.
41. Lott IT, Osann K, Doran E, Nelson L. Down syndrome and Alzheimer disease: response to donepezil. *Arch Neurol*. 2002;59(7):1133-1136.