Older adults with intellectual disabilities (ID) have become increasingly visible in community-based assessment and treatment settings over the course of the last 20 or so years. This is due partly to an increase in longevity for this group,\textsuperscript{1} as there has been improvement in early and ongoing treatment of medical problems associated with ID (\textit{e.g.}, congenital heart disease in Down Syndrome [DS]) superimposed on the increased life expectancy of the general population resulting from social and public health advances.

What has made this fact even more visible, though, is a trend towards de-institutionalization, which has increasingly moved people with disabilities of all kinds into community settings. Independent living programs, small and large group homes specialized for ID, sheltered workshops, generic private-care homes and nursing homes all are seeing more aging adults with ID in their clientele, and are striving to deal with increasing aging-related problems, such as dementia. The latter is particularly important, as DS, one of the most common forms of ID, is intrinsically linked to dementia because of the triplication of regions on chromosome 21, which are known to have etiological significance to Alzheimer’s disease (AD).

Primary care physicians and specialists are increasingly being called on to assess and manage patients with ID for possible dementia, and are struggling to adapt their skills to deal with this challenging group. This article is a brief introduction to this complex topic.

**Epidemiology**

How common is AD in adults with ID? It is clear that the rates are very high among those with DS. By the age of 40, all individuals with DS have characteristic neuropathologic findings of AD on autopsy,\textsuperscript{2} although this does not correlate clearly with clinical findings. For reasons not altogether understood, some people with DS reach old age with no evidence of cognitive or functional deterioration. There certainly is an age-related increase in the clinical and pathological evidence of dementia. Recently, Holland \textit{et al}\textsuperscript{3} published a dementia increase of 3.4\%, 10.3\% and 40\% in groups of DS-afflicted adults aged 30-39, 40-49 and 50-59, respectively.
These rates might have been higher if criteria for dementia other than the CAMDEX had been used.

Other chromosomal abnormalities also may be associated with increased rates of dementia, although this data is less conclusive.

**Clinical Problems Associated with the Assessment of Dementia in ID**

The diagnosis of dementia in patients with pre-existing ID is difficult for a number of reasons. First, environmental demands and stimulation of people with ID are reduced, especially in those living in protective environments, so early cognitive deterioration may not have the noticeable impact on the person's daily functioning that might prompt referral if it were more evident. Once referred, there is great heterogeneity seen among this group, which has cognitive and functional abilities ranging from mildly impaired to profoundly impaired. Instruments designed to measure abilities tend to be normed for shorter ranges of abilities, so more severely impaired individuals might exhibit a "floor effect" (i.e., function so low to start with that further loss is not apparent) or a "ceiling effect," in which the test is so easy that progressive dementia may not be apparent until it is in its final stages. In practice, there is no single test that can be used for all people with ID to measure the progression of a dementing illness accurately and reliably.

The testing process itself is subject to several confounders. Many adults with ID are awkward and shy with strange interviewers, and may not offer any verbal response on first visits despite having intact language skills. Of course, clear and sophisticated communication is not present in many people with ID. Moderately to severely impaired people with ID may have little or no communication skills, and standard cognitive tests are language-based, even if only to convey instructions for the administration of the test. As a result of this communication barrier, it may not be possible to administer tests for early dementia in ID patients, and losses may be obvious only in the latest stages (e.g., the loss of basic self-care, or the beginning of seizures, in DS patients with AD). This may be one of the reasons that, despite overwhelming neuropathologic evidence of brain changes, clinical evidence of dementia in older patients with DS is much weaker. There are some specialized computer-based tests designed to overcome this to some extent.

Classic behavior changes associated with dementia, such as mood and personality change, also are more easily masked by baseline abnormalities in ID or by overlapping psychiatric disorders, such as depression. Some physical disorders also are more common in ID, and might cause diagnostic difficulty. For example, hypothyroidism, hematologic malignancies, hearing impairment, language deficits and depression all are more common in DS, and might present as cognitive impairments. Sensory deficits of various types are very common in ID, and must be considered as a factor in any functional loss.

Seizure disorders are much more common in people with ID overall, and both the seizure disorder and its treatment can decrease various aspects of cognitive and behavioral functioning. Polypharmacy for the treatment of refractory seizure disorders, as well as for behavior problems such as aggression, is more common in those with ID, and adds significantly to the difficulty in making a diagnostic decision.

Although the above factors limit the usefulness of a one-time assessment of cognition and functioning in the diagnosis of dementia, retrospective longitudinal information often is not available, as many older people with ID do not have caregivers (or living parents) well-versed enough in their long-term functioning to provide this information reliably. Furthermore, the mainstay of the assessment of early dementia in a non-ID person is evidence of prospective longitudinal progression in the deficits, but this often is not practical when a swift diagnosis is needed to assess appropriate placement or decide on management strategies.

**Diagnostic Strategies**

What practical strategies, then, can be used in the office to make a determination of a possible dementia and then allow for follow-through by making reasonable therapeutic plans? The Canadian Consensus Conference on Dementia has published guidelines for...
the general population,\textsuperscript{13} which are useful stepping stones to the more complex scenario for patients with ID. The Canadian consensus Guidelines are applicable to ID patients, recognizing pre-existing deficits and judging all areas of decrements in comparison to previously attained levels. Conditions more commonly presenting comorbidly in ID are given more emphasis, as summarized below.

History is taken from an informed caregiver, with the goal of obtaining evidence of progressive symptom development in key dementia-related areas (Table 1). Brief notes on functional abilities in each of these areas should be made for later comparison. Objective functional losses, as shown by re-assignment of duties at a sheltered workshop, should be sought. If possible, staff should be given the Evenhuis Dementia Questionnaire for Mentally Retarded Persons (DMR) instrument\textsuperscript{14} to complete. This itemizes specific areas of abilities, and can be used as a comparator in follow-up. Information about previous illnesses, such as depression, thyroid treatment and sensory assessments, should be obtained, and medications that might impair cognition should be reviewed. Particular attention should be paid to anticholinergics, such as low-potency traditional antipsychotics (and atypical antipsychotics, such as olanzapine, with some anticholinergic activity) and tricyclic antidepressants, as well as anticonvulsants and sedative-hypnotics.

**Physical examination.** Of particular importance during the physical examination is the assessment of hearing and vision, as these are much more commonly impaired, particularly in DS. Other signs of premature aging, such as cataracts, should be sought.

**Psychometric testing.** Most people with ID are not able to perform at all on the Mini-Mental State Examination (MMSE),\textsuperscript{15,16} and there is no other single test suitable for easy office administration in all people with ID. Increasingly, tests are becoming available for neuropsychologists to use in this group,\textsuperscript{17} but these are not practical for use in the physician's office. Particularly on first visits, physicians often will be unable to obtain any meaningful information out of the interview, as patients tend to be fearful of the interview situation.

**Laboratory testing.** According to Canadian Consensus criteria,\textsuperscript{13} routine blood work includes:
- Complete blood count
- Thyroid-stimulating hormone (TSH)
- Electrolytes
- Calcium
- Glucose

This should be supplemented by anticonvulsant levels, where appropriate, and the other supplementary tests listed in the Canadian consensus Guidelines, as needed.

**Neuroimaging.** Due to non-compliance, it is very difficult to obtain computed tomography (CT) scans from some adults with ID. This necessitates significant tranquilization (which is not always innocuous) for accurate assessment. The decision to order a CT scan therefore becomes slightly more complex, and involves a careful cost-benefit analysis. Specifically in DS, the Canadian consensus Criteria for CT scanning should have the "onset under age 60" criterion omitted, because dementia Alzheimer's Type (DAT) develops in higher frequency, and earlier, in this group than in the general population. Most practitioners would order a CT scan in these cases only if there were other unusual features, such as localizing signs.

**Final assessment.** In the final analysis, the diagnosis of dementia in ID is a clinical one based on the best compilation of available information, and this diagnosis might change with repeated assessments of the patient.

**Management Strategies**

Once a diagnosis of AD is made, information can be provided to caregivers about the course of the disease and its associated behaviors, such as wandering, sleep change and incontinence. Environments must be made safe for wanderers, and work settings simplified to decrease demands and secondary agitation. Caregivers should know that there is a high rate of early seizures in people with DS and AD. Behavior change, however, often is the first sign of a dementia, with exacerbation of pre-existing traits, such as stubbornness and irritability, being a frequent reason for referral. In the early stages of dementia, people with ID fit better into the ID service system, where activities are geared towards lower-functioning people, and because they often are not accepted by other seniors in generic aging programs. By later stages of dementia, those with ID...
become hard to distinguish from the general population, and placement in dementia units of nursing homes often becomes necessary.

Medications like donepezil have not been tested in large studies with dementing adults with ID, although case reports exist. It is therefore not clear what role these medications have in ID and AD. Theoretically, there should be improvement with these medications, but the degree of functional and quality-of-life gain is little understood, as is any kind of cost-benefit assessment. There is no particular reason not to try this medication, but functional improvement—rather than Mini-Mental State testing—would have to be the measuring stick for patient response.

Treatment of agitation of dementing adults with ID is the same as for those in the general population, with the added proviso that brains with pre-existing impairment seem even more sensitive to adverse pharmacologic effects. Pharmacologically, risperidone has been best tested in large-scale, double-blind studies in dementia, and also in behavior disturbance with ID (data to be presented in the near future). Because of its lack of anticholinergic effects, risperidone is a natural choice of neuroleptic, although extrapyramidal symptoms tend to emerge in the elderly at doses over 2 mg. The usual starting dose is 0.5 mg, once daily, with increases occurring no faster than weekly due to the long half-life. No pharmacologic agent, however, is able to improve all the behavior problems of dementia significantly and reliably, so the mainstay of treatment remains environmental.

Conclusion
Dementia rates are increasing in those with ID, and this is especially marked in DS patients. Assessments must be tailored to baseline activities, and must rely largely on caregiver reports of functional abilities. Of particular importance is the assessment of sensory deficits, as these are very common in DS. Interventions for these patients are less studied than for the general population, but may have benefits (although many treatments may, themselves, further impair cognition). The provision of service relies on a combination of generic aging services and specialty ID services.

References