Dementia in intellectual disability: a review of diagnostic challenges

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Abstract
The evaluation of dementia in individuals with intellectual disability (ID), which will guide subsequent intervention, care and management depends on the systematic review of a number of factors: (1) the individual historical context, obtained from multiple sources, (2) evaluation of the pre-existing cognitive, behavioural, psychiatric, medical and adaptive skill profile, (3) the constellation, and pattern of evolution, of presenting signs and symptoms, (4) results of focused investigations, and (5) refinement of the differential diagnosis. In patients with ID, standard clinical methods need to be supplemented by careful, longitudinal behavioural observations, and individually tailored assessment techniques. Co-morbidity, multiple biological, psychological and socio-environmental factors, and complex interactions among events, are the reality for many ageing people with ID. Determining the various influences is often a formidable clinical task, but should be systematically carried out using medical, cognitive, behavioural, neuropsychiatric and psycho-social frameworks.

Key words: Dementia; Intellectual disability; Mental retardation

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Introduction

People with intellectual disability (ID) [this term is used in preference to mental retardation (MR)] are prone to developing dementia in later life. This is particularly important in the context of rising life expectancies in this population. The average life expectancy of adults with ID (in developed countries) is 66 years and increasing.[1,2] As these individuals age, they present with increased rates of physical, sensory and cognitive impairments. This results in increasing health and social care needs, with resultant pressures on carers and support services. Clinicians can expect to come across increasing numbers of people with ID who develop dementia as they grow older. Whilst mental health resource priorities in developing countries may be focused elsewhere, people with ID still form a significant, and particularly vulnerable, proportion of our patient population, and have been historically neglected in many respects. As identification, evaluation, diagnosis, and management of such patients can be challenging, familiarity with the key issues is essential for all involved in their care.

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Epidemiology

Dementia can occur at any age, but is largely a disorder of later life. The prevalence of dementia in the general population rises markedly with increasing age: from 1-2% in people aged 65-69 years, to 16-25% in those over 80 years.[3] Among people with ID, dementia is liable to strike more often, and earlier. A number of community-based studies of ID populations have yielded age-matched prevalence rates of about 14% in people over 59 years, and 22% in those 65 years and over.[4] The comparative rates among same-age adults with Down syndrome are much higher, reaching over 40% in those over 50 years, and over 70% for ages above 60 years.[5]

Diagnostic criteria

The diagnosis of dementia in people with ID presents a number of challenges[6]:

- Assessment techniques and tools in common clinical use for the non-disabled population may not be appropriate, especially for individuals with more severe pre-existing levels of intellectual impairment. Diagnosis in the ID population requires a change in status from baseline functioning, not a change from a ''normal'' level.[7]
- Pre-existing cognitive impairments and disturbances in behaviour, personality and emotional control may conceal the often subtle and insidious emergent symptoms of dementia.[8,9]

- Some signs of dementia (such as dysphasia, dyspraxia and agnosia) are often difficult to recognize in people with ID, particularly in the early stages of the illness. These criteria have therefore been excluded from some ID diagnostic schemes.[10,11]
- Changes in repertoire of adaptive skills or behaviours may predate any demonstrable early memory impairments in ID patients.
- The perception, manifestation and degree of functional impairment caused by the dementia syndrome largely depends on pre-morbid intellect, level of education and training, individual life circumstances, and individual ability to compensate for newly acquired deficits.[7]
- To be indicative of dementia, any changes over time must be greater than those related to normal aging in adults with ID.[7]
- Neither the ICD-10 nor the DSM IV-TR diagnostic criteria for dementia make any specific mention of ID/MR.

To date, there is a lack of consensus on a standardized approach to the diagnosis of dementia in ID. As a step towards such standardization, diagnostic criteria have been proposed by the Working Group for the Establishment of Criteria for the Diagnosis of Dementia in Individuals with Intellectual Disability [under the auspices of the International Association for the Scientific Study of Intellectual Disability (IASSID) and the American Association on Mental Retardation (AAMR)].[10] These are largely based on ICD-10 diagnostic criteria (with its relatively heavier emphasis on non-cognitive changes in dementia). These authors strongly recommend that dementia should only be diagnosed when longitudinal data demonstrate clinically significant declines in functioning. If some, but not all, of the criteria are met, a diagnosis of ''possible dementia'' is appropriate. This group has further proposed a working battery of tests (for both informants and patients) for the diagnosis of dementia in adults with ID.[8,10] As in the general population, diagnostic certainty will be enhanced if decline is observed on many tests and across longitudinal assessments.

Clinical features

The clinical presentation of dementia among adults with ID (at most levels of disability) is similar to that of adults in the general population. Nonetheless, there are a number of factors that impact on the clinical evaluation of dementia in the context of ID[6,12,13,14], including:

- The lack of consensus on what constitutes normal age-related decline in people with ID, and how to distinguish this from dementia.
- The lower the pre-existing intellectual and adaptive level of functioning, the more difficult it becomes to evaluate change and document progression.
- Pre-existing cognitive and psycho-social deficits may mask signs of deterioration.
- Early signs of dementia (cognitive, neuropsychiatric or adaptive skills decline) are often subtle and difficult to detect.
- There is a bias amongst many health professionals and carers towards aging people with ID in expecting a deteriorating course, leading to delays in diagnosis and treatment.
- People with ID have a predisposition to co-morbidity, which is often severe or frequent enough to influence the evaluation and interpretation of their cognitive impairment.
- The effects of chronic medication on cognitive function are often overlooked in clinical practice (especially the use of multiple agents).
- There is often a lack of consistent and reliable documentation of prior cognitive and adaptive skill levels, making it difficult to detect decrements until they become pronounced.

A comprehensive and focused history from both the patient and multiple informants/sources (e.g. family members, carers, medical records) is usually the most valuable diagnostic tool for dementia in ID. The information-gathering exercise aims to: (1) systematically document details of symptoms and functionality, (2) identify treatable causes of dementia, and aid in clarifying a working differential diagnosis, and (3) conceptualise the current problems within a broader pre-morbid and psychosocial setting, in order to guide appropriate and effective management. It is imperative to obtain collateral information from people who are familiar with the individual's past behaviour and level of functioning, as well as her/his present performance. A clear history of declining function may be difficult to elicit in people with more severe ID, where levels of ability may be significantly impaired in the first place, or where impairments in communication may mask early evidence of decline. Specific enquiry about the presence of risk factors (e.g. family history of dementia, a previous head injury, or vascular morbidity) is equally important. A family history of dementia, ID, and neuropsychiatric problems requires special attention. The history should pay particular attention to previous diagnoses and treatments, especially the use of medication. Careful attention should also be paid to the timing of onset, pattern and progression of symptoms. Symptoms often have a subtle and insidious evolution (a fact usually more easily appreciated by patients and carers retrospectively). Symptoms may, however, present abruptly, especially in dementias with potentially treatable aetiologies. There may be identifiable precipitating factors e.g. a relatively minor physical illness, a change in environment or following bereavement. The history should screen for the presence of pre-existing or co-morbid medical conditions (and their respective treatments). Physical symptoms and signs may also be due to adverse effects of medication. It is well recognized that people with ID have a significant predisposition to sensory impairments, especially hearing and visual impairments that can be mistaken for, aggravate, be co-morbid with, or mask an underling dementia.[15,16]

Cognitive changes

The core symptoms of dementia in people with ID are of multiple, and usually progressive, deficits in a number of areas of cognitive function, similar to those in the general population. A history of memory impairment is often the

earliest and most prominent feature (particularly with the cortical dementia syndromes, e.g. Alzheimer's disease). Early changes may be subtle or intermittent, with non-specific complaints of forgetfulness, absent mindedness, difficulty concentrating or general mental fatigue, leading to difficulty with novel or more complex tasks, progressing to more mundane, everyday tasks. Memory decline most commonly manifests as impairment of recent memory and a reduced capacity for new learning. This is often noticed by others first, but it is usually difficult to date the onset with accuracy. There may be difficulty remembering events of the day, recently held conversations, or the names of acquaintances. Patients may have word finding problems, forget the location of everyday items, or perhaps require more prompting than usual to complete everyday tasks. They may have difficulty remembering the steps required to perform previously mastered tasks, or following direction. Carers may report that information needs to be presented more slowly, or in a more simplified format, and patients may require more frequent reminding. Eventually, memory impairment may become severe, with only the earliest or most remote information remembered. This may present as perseveration on the distant past, and may progress to become quasi-delusional, where the person experiences the present as if it is the past. Intellectually disabled individuals are particularly vulnerable to such memory impairments, as previously limited adaptive skills and coping strategies undergo even further decline. Patients may become even less able to communicate effectively e.g. to explain personal concerns and needs to others, or to socialize and interact adequately. Peers, acquaintances and carers may feel socially unsatisfied, or estranged, with regards to the person developing dementia. Patients themselves often grow increasingly frustrated, irritable, confused or depressed by the changing circumstances.

Orientation also becomes progressively more impaired, and may be more salient than other cognitive deficits. Disorientation for time is usually an early feature, and often presents as difficulty in temporal sequencing. This may upset personal, family, social or institutional routines e.g. arriving late for meals or missing social engagements. Spatial disorientation is also common, with people having difficulty getting around the home (e.g. returning to the bedroom from the bathroom) or the neighbourhood (e.g. getting lost whilst returning from the local shop). This can be a particular problem in unfamiliar environments or following unexpected changes in routine. Spatial disorientation may impact on everyday skills (e.g. placing the body in an appropriate position at the table for meals), requiring increasing levels of support from carers. Visual and hearing impairments, which occur frequently in older individuals with ID, may influence the assessment of orientation.[7,15,16]

Dementias that primarily affect the neo-cortex (e.g. Alzheimer's disease, fronto-temporal dementia) tend to affect language function. This may present as language use that is uncharacteristically vague, stereotyped, clichéd, repetitive, circumstantial, or impoverished. Word finding difficulties are often early manifestations. There may be increasing paraphrasic errors, a tendency to relate events in inappropriately minute or irrelevant detail, or perseveration. Language dysfunction may present as more specific deficits, such as receptive or expressive dysphasia. In the later stages of illness, speech becomes increasingly disorganized, progressing to incoherence or muteness. In those with poorer pre-existing verbal skills, evidence of language impairment will range from subtle, barely perceptible changes or decreases in the use of language to total loss of verbal expression.[7] The pre-existence of communicative impairments makes it more likely that dementia will be well advanced before it is clinically recognized.[9] As with many other symptoms and signs, it is the change from pre-morbid patterns and levels that is the crucial factor.

A range of other cognitive impairments may be present. There may be evidence of varying degrees of dyspraxia, typically presenting as difficulties with grooming, dressing, self-feeding, cooking or similar everyday motor tasks. Various agnosia's may occur e.g. in the visual (e.g. prosopagnosia) or tactile (e.g. astereognosis) sensory modalities. For individuals with moderate to severe levels of ID, dyspraxia's and agnosia's may be the first deficits to be apparent. Agraphia, alexia and acalculia may be diagnosed in those with prior requisite skills in these areas.[7] There is also usually a history of impairment in complex cognition (e.g. executive function) and motivational behaviour. Even mild executive dysfunction may significantly impair the capacity to fulfil basic needs (e.g. providing adequate self-care), perform everyday tasks (e.g. preparing a meal, or handling money), interacting with others in a socially acceptable manner, or regulating emotions and behaviour appropriately. Such changes in complex cognitive function are, relative to pre-morbid levels, and may be more difficult to recognize in people with more moderate or severe levels of ID.[7] Patients with a higher baseline of intellectual functioning may appear to lose more cognitive skills over time.[17] The pattern, rate and degree of decline, however, are highly variable from one individual to another, and are not easily predictable

Behavioural and psychiatric presentation

Changes in behaviour and psychiatric symptoms are common in ID patients with dementia. The Working Group of the International Association for the Scientific Study of Intellectual Disabilities (IASSID) recommends greater emphasis be placed on behavioural and personality changes (together with evidence of functional decline) in the diagnostic evaluation.[7,18,19,20] Behavioural changes present in a variety of ways, ranging from patterns of excitement, escalation and disinhibition, to slowing and apathy. They are also of variable severity, often being subtle at first and then progressing in tandem with progression of the underlying dementia. Restlessness, impulsivity and agitation are common, as are wandering and inappropriate motor behaviours (e.g. trying to leave home, or trailing of carers for no apparent reason). Psychomotor agitation can progress to overt hostility and aggression, often with unpredictable and unprovoked explosions of primitive affect. Such behaviours are a frequent cause of hospitalisation and institutionalisation.[21] A decrease in motor or social spontaneity is also common, and can occur at any stage of the dementing process. Patients may display psychomotor retardation or diminished spontaneous movement, remaining

sedentary for increasing periods of time. Social reciprocity and volition may be affected, with reduced initiative or drive for activities and interests, diminished interaction, and emotional restriction. Distinct nocturnal worsening of behavioural presentation in the evenings ('sundowning') may also be seen. The impact of such behavioural changes on all aspects of home, social or occupational life should not be underestimated. It should also be remembered that behavioural changes might be secondary to underlying, and potentially reversible, medical conditions, environmental factors (e.g. changes in routine or living situation), psychosocial stressors (e.g. interpersonal conflict or bereavement) or co-morbid psychopathology (e.g. depression or psychosis).

Mood (especially depressive) and anxiety symptoms are commonly encountered in patients with ID and dementia. Older adults with ID and dementia are known to be more susceptible to depression (either as a first presentation or at any stage in the course of dementia) than ageing individuals in the general population.[13] Other mood-disordered states (e.g. elation, hypomania, emotional disinhibition, irritability, mood lability, mania, adjustment reactions) do occur, but less frequently than depression.[22] Associated disturbances of neurovegetative features (e.g. insomnia, hypersomnia, reversed sleep-wake cycles), somatization (e.g. hypochondriasis), self-harming or suicidal ideation, or signs of anxiety or distress may be present. Many restless, agitated, or aggressive patients are, in fact, anxious or mood-disordered. This is particularly important in those with more severe levels of ID, and those with poor verbal skills, who may not be capable of effectively communicating subjective states of emotional distress. Management of the underlying psychopathology should attenuate or resolve the behavioural difficulties.

Psychosis is an important component of the dementia syndrome in some patients, with approximately 28% of intellectual disabled people with dementia experiencing at least one psychotic symptom at some stage.[4,23] Delusions are most commonly of the persecutory type, and are usually simple, unsystematized and crude. Hallucinations are usually of the auditory or visual variety, the latter a frequent occurrence in patients with Lewy body dementia. Other perceptual disturbances that may occur include illusions and misidentification phenomena.

Changes in personality invariably occur with the development of dementia, especially in the types of dementia that have a predilection for the frontal and temporal lobes. There may be an accentuation of pre-existing personality traits. Patients may become more apathetic, passive, socially withdrawn, disengaged, anxious, fearful, or unconcerned. Similarly, interpersonal styles of interactions may change to being more disinhibited, egocentric, hostile, irritable, unpredictable, or aggressive.

Adaptive function

In addition to deficits in cognition and associated behavioural and psychiatric features, dementia is always associated with a decline in adaptive functionality. The onset of dementia in individuals with low pre-existing levels of intellectual functioning is often reflected initially in changes adaptive behaviour, rather than memory loss or other obvious cognitive decline. All types of previously learned skills of daily living may be affected. These include impairments in: self-care, home-living, communication, functional academic skills, occupational performance, financial skills, social and leisure skills, capacity to use community resources, health and safety skills, and motor skills.

The assessment of adaptive behaviour and living skills is often difficult. Although there are a number of instruments available for this purpose[6,10,24], most of these are informant-based reports, which are not always reliable, particularly in institutional settings where staff may not be sufficiently familiar with the patient. Functional decline may impact significantly on the patient's capacity to cope with basic environmental, social or occupational demands. The decline usually starts with subtle and easily avoided skills (e.g. using a telephone properly). Increasingly, the ability to function in ordinary life is lost, with decline in basic and essential skills, such as those related to self-care, dressing, feeding and toileting. The degree and rapidity of decline depend on, amongst other things, the individual's level of pre-morbid intellect, the type and severity of the dementia, the presence of co-morbid illness, and the timing and efficacy of treatment. Individuals with ID, who may already be disadvantaged by significant pre-existing impairments in cognitive, communicative and adaptive skills, are particularly vulnerable to profound decrements in their quality of life. Further diminution of skills and abilities may also impact significantly on the resources of a care system that is often stretched to begin with.

Examination

Although the Mental State Examination (MSE) has some limitations in the quantification of intellectual impairment, it should provide, in conjunction with a detailed and focused history and physical examination, useful information for the formulation of a sound initial clinical opinion. The specific order and emphasis of the MSE may differ from patient to patient (or from interview to interview), and certain aspects may need modification, or may be inappropriate, for some individuals with ID. Early identification and management of physical symptoms and signs is important in the ID population, who are particularly prone to physical ill health and complications, and generally more sensitive to adverse effects of medication than others. Although physical, medical and neurological complications tend to be late features in the majority of patients, these may occur at any stage of the illness. People with ID and dementia are susceptible to developing a number of medical problems[9,14,15,16,25,26,27], including: incontinence (urinary or faecal; usually nocturnal); gastrointestinal problems (e.g. chronic constipation; diarrhoea); malnutrition and dehydration; infections (e.g. infections of the urinary or lower respiratory tract; bed sores); metabolic and endocrine disturbances (e.g. glucose or electrolyte imbalances); cardio-pulmonary complications (e.g. heart failure; atherosclerosis; hypertension; syncope; postural hypotension; chronic obstructive pulmonary disease); neurological problems (e.g. seizures; sensory impairments; dyskinesia's; spastic paresis); musculoskeletal and mobility difficulties (e.g. gait and balance disturbances; falls); haematological problems (e.g. anaemia); inflammatory conditions and neoplastic disease.

Investigation

All patients suspected of developing dementia should be investigated for disorders that could cause, exacerbate, complicate or be confused with dementia. The choice of initial investigations is determined to a large extent by the information already gathered on history and examination. Whilst not always necessary, neuroimaging investigations can be helpful, particularly in excluding some differential diagnostic options. CT scanning often reveals non-specific cortical atrophy and ventricular dilatation; MRI is useful where vascular dementia is suspected or for white matter pathology. Electro-encephalographic (EEG) findings of generalized slow wave activity are suggestive. Interpretation of results in some people with ID may be difficult: a relatively high proportion of abnormal neuroimage or EEG findings may be related to the underlying cause of ID, rather than the onset of dementia. It is particularly helpful therefore to compare results with previous investigations. Neuroimage and EEG investigation can be helpful in patients with more severe levels of ID, in whom options for formal neuropsychological testing are often limited. When neuropsychological testing is possible, valuable information on the pattern and extent of cognitive impairment can be obtained, which will help clarify the diagnosis, guide management planning, monitor progression and assist with prognostication. This notwithstanding, a significant obstacle is the fact that many standard neuropsychological tests are not sensitive or specific enough to be used reliably to measure early or subtle cognitive impairments in people with pre-existing intellectual impairments.

Differential diagnostic issues

It is essential to actively exclude delirium as the cause of the presenting symptoms and signs. Distinguishing between delirium and dementia may be difficult. The hallmark of delirium is disturbance of attention, usually of recent onset. There is alteration in the level of consciousness or arousal, which is typically fluctuating in nature. There are often associated disturbances in other areas of cognitive function, perception, mood, and/or behaviour. There may also be nocturnal exacerbation of symptoms, and sleep-wake cycle disturbances. To make things more difficult, delirium-like presentations form part of some dementia syndromes (e.g. Lewy body dementia), and delirium may be co-morbid with dementia. Patients with ID have higher rates of pre-existing medical and neurological abnormalities, and are known to be more susceptible to the effects of systemic illness and to the adverse effects of medication. This places such people at higher risk of developing delirium.[7,13]

Age-related cognitive decline may be associated with mild memory decline, slightly shorter attention span, slower cognitive processing, and a slightly reduced capacity to perform complex (especially novel) cognitive tasks. There may also be subtle personality changes (e.g. reduced drive, diminished interest in novelty, or conversely, heightened preference for routine and structure), mild sensory impairment (usually diminished sensory acuity), sleep-wake cycle disturbances (e.g. more nocturnal awakenings), and motor disturbances (e.g. postural changes). As people age, though, there should be no significant disturbances of orientation, comprehension, language, general knowledge, or overall executive function, nor should there be any marked decline in levels of adaptive functioning. Unsurprisingly, it is often very difficult to distinguish between age-related cognitive decline and dementia, particularly in people with ID, especially at lower levels of intellectual disability or in the presence of multiple sensory or physical impairments.[12] The literature on non-pathological ageing in people with ID is scant. A significant difficulty relates to the fact that this is a highly heterogeneous population, with individuals differing markedly in terms of underlying aetiology, intellectual profile, nature and extent of impairments, personalities and social background.[20] A number of proposals have been put forward to help define and understand the ageing process in ID[6,12,17,20,28]:

- General intellectual capacity in individuals with mild to moderate levels of ID without Down syndrome seems to remain intact until about 65 years of age (similar to the general population). Early or marked cognitive decline is usually related to specific medical, iatrogenic or environmental factors. The degree of age-related cognitive decline in people with severe to profound ID remains equivocal.
- Deficits on IQ tests are often exacerbated by more pervasive problems in cognitive functioning (e.g. difficulties with attention, arousal or motivation).
- Decline in cognitive skills, especially complex and abstract skills, may not directly parallel changes in adaptive functional skills.
- People with initially lower cognitive and adaptive levels may appear to decline sooner with advancing age.
- Age-related changes in adaptive function are observed in ID populations (as in the general population), though the issue is controversial.
- Older adults living in institutional settings display more severe decline in adaptive skills than those living in community settings.
- Mobility and motor functioning appear to decline consistently with age in this group.

Natural history and prognosis

There is considerable individual variability in the evolution of the dementia syndrome. The dementia may be progressive, relapsing-remitting or relatively static. The mode of onset and subsequent course are dependent on the underlying aetiology, as well as the pre-existing and co-morbid clinical profile of any given individual. This variability notwithstanding, the pattern of clinical evolution of dementia is commonly described using a three-stage model, similar to that seen in the general population: (1) in the early stages, cognitive, behavioural and psychiatric symptoms are often subtle or episodic. Such changes may be difficult to detect in people with ID. A decline of 8-10 IQ points per year on adult intelligence test scores (e.g. the Wechsler Adult Intelligence Scale – WAIS) has been reported[9]; (2) the middle stages present with progressively worsening cognitive impairment in a number of domains (e.g. memory, language, praxis, complex cognition), associated with significant and disabling decline in adaptive

functioning in important areas of daily living. Mood, perceptual, behavioural and personality changes become increasingly prominent, and there may be some neurological fallout; and (3) the late stages, during which patients eventually become significantly impaired in cognitive, psychiatric, physical and adaptive skill domains. Patients become dependent on others to meet even the most basic of needs, and may become bed-bound. Physical and medical difficulties become more disabling and severe, with terminal events usually related to cachexia, infections or cardiovascular aetiologies.

A number of prognostic factors can be identified among people with ID who develop dementia.[9,12,13,17] These include a family history of dementia; Down syndrome; early onset of symptoms, and late diagnosis or treatment; low pre-existing intellectual capacity or level of adaptive skills; severe cognitive or behavioural symptoms and signs; co-morbid medical or neurological illness; iatrogenic factors (e.g. chronic, multiple medication usage); and poor psychosocial support systems (e.g. patients in institutional settings with no family contact).

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