



Dementia in Intellectual Disability Disorder

Diagnosis and management Information for GPs

Intellectual Disability Disorder (IDD, ICD-11, 2017) is the internationally agreed term that replaced “Learning Disabilities” and “Mental retardation”. It refers to 'a group of developmental conditions characterized by significant impairment of cognitive functions, which are associated with limitations of learning, adaptive behaviour and skills'

There is no definition of dementia that is specific for individuals with IDD.

Prevalence of Dementia in IDD

The overall prevalence rate of dementia in IDD population is comparable to that in the general population (6.1% in those aged 60 and older). In adults with Down syndrome, the rates are higher (22% for those aged 40 and older and 56% for those aged 60 and older). Individuals with IDD experience onset of the ageing process earlier than in the general population, and they are at risk of developing age related diseases (age is the strongest risk factor for dementia in IDD).

Clinical presentation of Dementia in IDD

- Dementia may present atypically in the IDD population. The perception of decline and the way it manifests will depend on the premorbid level of IDD, pattern of abilities, and the environmental demands placed on the person.
- Presentation of dementia may be subtle and may go unrecognised.
- Decline in cognitive function and behaviour in individuals with mild IDD who develop dementia can be very similar to that seen in the general population with Alzheimer's disease.
- Memory loss is an important feature but this may be masked.
- Loss of adaptive function may be the first sign of dementia and may be associated with slowing down in the daily function or the decrease of self-care skills in the home domain.
- A frontal lobe pattern of decline with apathy, poor motivation and behavioural changes may occur, but if there is no associated decline of previous level of

cognitive and independent level of day to day functioning, these changes in behaviour are not sufficient to make a diagnosis of dementia.

- The development of epilepsy is an important feature of dementia in Down syndrome that is not seen to the same extent in the general population.
- Progressive dysphagia and frequent choking may be observed in people with Down syndrome in the early stages of dementia.
- As the disease progresses there is loss of speech, altered gait, dysphagia, immobility and incontinence.

Suggested Screening for dementia in IDD for GP

A simple clinical assessment asking carers if there have been any changes in behaviour suggesting a deterioration of memory, communication skills, disorientation, decline in level of self-care, difficulties in following routine, epilepsy, dysphagia or falls should be sufficient to consider an assessment to exclude dementia.

Assessment

There is no definitive 'test' for dementia. Its presence is a matter of eliciting a clinical history suggesting dementia and establishing evidence of change in function from a known baseline and excluding other diagnoses that may mimic dementia.

- A thorough physical examination and relevant clinical tests are required at the time of initial assessment.
- Sensory screening for **vision & hearing** is important.



- Various medical and social problems may mask the diagnosis and careful evaluation is necessary to establish a diagnosis of dementia.

- Exclusion of obvious environmental issues such as change in accommodation, carers or co-residents as well as life events such as losses, depression or other intercurrent illness should complete the screening assessment at GP level before referral to secondary care.

It is recommended to assess every adult with Down syndrome by the age of 30 to establish a baseline against which to compare future suspected changes in functioning.

It is worth considering screening all adults with Down syndrome over 40 regularly because of the increased risk of dementia and the prevalence of undetected but treatable illnesses. This should link to the person's health action plan.

Differential Diagnoses.

- The common differential diagnoses for individuals with Down syndrome presenting with loss of skills are depressive illness, sensory impairments (hearing or visual), hypothyroidism, obstructive sleep apnoea and dementia.

- Sometimes conditions can co-exist.
- Do not forget about the following causes of apparent functional decline as these are often missed:
 - Iatrogenic causes of cognitive impairment particularly when the individual is taking multiple medications;
 - Impact of the environment particularly in relation to occupational deprivation and under stimulation; and
 - Impact of abuse on the individuals with IDD.

Where to refer: Referral will depend on availability of local services and clinical presentation.

Reference: CR196. Dementia and people with learning disabilities. Guidance on the assessment, diagnosis, treatment and support of people with learning disabilities who develop dementia. Co-published by The British Psychological Society and the Royal College of Psychiatrists.

<http://www.rcpsych.ac.uk/usefulresources/publications/collegereports/cr/cr196.aspx>

1. IDD services tend to have a dementia care pathway.
OR

2. Memory Clinics for the elderly assess patients with IDD.

3. A minority of patients can be referred to Neurology Services for diagnostic purposes but then the follow up and management is provided by IDD services.

What to expect of a specialist service

- Multi-disciplinary assessment is important.
- Assessment for other co-morbid conditions as well as signposting to other services that may be required.
- Pharmacological Treatment with cognitive enhancers; evidence suggests that it is difficult to draw conclusions about the effectiveness of any pharmacological intervention for cognitive decline in people with Down syndrome.
- Management of Epilepsy, Pain, infections, sleep difficulties, dysphagia, motor and gait disorders and falls.
- Follow up including dementia training specific for carers of people with IDD, support in facilitating capacity assessments and Best Interest Decisions (MCA 2005), as well as help in considering and completing care plans, Deciding Right & Emergency Health Care Plans (EHCP) which will include discussions about Do Not Attempt Cardio Pulmonary Resuscitation (DNACPR) decisions.
- There is a need for staff training involving dying, death and bereavement, communication with palliative care services, some aspects of palliative care such as communication, symptom management, psychological support, care after death, and the involvement of individuals and family members in decision-making.

Think about what the person is actually experiencing, and use that to inform the care and support that is required.