DEMENTIA IN PEOPLE WITH INTELLECTUAL DISABILITY

CONTENTS

This is an abbreviated form of similar guidelines, which are updated regularly and can be accessed here: http://unsw.to/iddementiaguidelines

and here https://3dn.unsw.edu.au/content/health-mental-health-professionals

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Disclaimer: This document is a general guide, to be followed subject to the clinician’s judgment and person’s preference, choices and decisions in each individual case.

1. Key Messages

- GPs need to be aware that people with intellectual disability (ID) are at increased risk of dementia.
- Alzheimer's disease, in particular, is very common in Down syndrome, is often of early onset, and typically begins with changes in personality and executive function. People with Down syndrome should receive a comprehensive baseline 'healthy' assessment at around 30 years of age, and again at 40. From 30 onwards, questions regarding signs of decline should be incorporated into annual health checks.
- In people with other forms of intellectual disability (ID), average dementia onset is approximately 10 years prior to that experienced by the general population. A practical approach would be to screen for evidence of decline at around the age of 40, by asking questions about decline and using a carer-report checklist. This checklist should be repeated at the age of 50 and each year thereafter. Those with signs of potential decline should receive a comprehensive assessment.
- After performing a standard dementia work-up, refer a person with suspected cognitive declines to an experienced psychologist or psychiatrist for a full cognitive assessment.
- Diagnosing dementia in people with ID requires establishing longitudinal declines in function, ideally across at least 3 sequential assessments. Standard tests used with the general population are unsuitable for this group [11].
- A number of screening checklists are available. The US National Task Group Early Detection Screen for Dementia (NTG-EDSD) is free to download from http://aadmd.org/ntg/screening and can be used qualitatively to examine declines.
- Important principles for managing dementia in patients with ID include:
    - Be equipped to manage mental disorders in people with ID. Recommended adjustments to practice can be found here https://3dn.unsw.edu.au/the-guide.
    - Screen and examine for other potential causes of cognitive decline

  - Use principles of dementia care applicable to people without ID, including communicating the diagnosis to the patient as early as possible in a manner they understand; seeking their preferences for care; coordinating services across relevant sectors (which may include the disability sector); and, where applicable, encouraging family carers to access emotional support and to make use of respite services

- There is general information about supported decision-making with people with dementia available here: https://cdpc.sydney.edu.au/research/planning-decision-making-and-risk/supported-decision-making/

- The Down Syndrome Association of Australia website has information on supported decision-making here: https://www.downsyndrome.org.au/news/supported_decision_making.html

2. Practice Points

a) Prevalence and incidence of dementia in Intellectual Disability (ID)

- People with intellectual disability (ID) are at higher risk of dementia than the general population.

- Down syndrome carries a higher risk of dementia, and specifically Alzheimer's disease, with younger onset (see 1 for review).

- Dementia in people with ID from other causes is less researched. Prevalence studies show conflicting results. However, research indicates that the onset of dementia in people with non-DS ID is on average 10 years earlier than in the general population [2].

b) Risk factors for dementia in people with ID

- Specific risk factors for dementia in people with ID include Down syndrome, poor physical and mental health, including undiagnosed health problems, and sensory impairments.

- Risk factors for dementia in the general population are also relevant to people with ID: poor diet and exercise, cardiovascular risk factors, poor engagement in education, social activities, and employment, head injury, and genetic factors such as APOE genotype.

c) Presentation of dementia in people with ID

- The full range of dementias may appear in people with ID, including mixed presentations [3]. A sizeable group of people with ID with suspected declines meet some, but not all, criteria for dementia.
- At least in people with Down syndrome, behavioural and personality changes and declines in executive function may appear before memory deficits [4-8].

d) Assessment of dementia in people with ID

- There is no gold-standard diagnostic test for dementia in people with ID. Assessment tools useful for the general population are not appropriate for this group. Diagnosing dementia in people with ID requires demonstrating a decline from baseline in cognition and functioning [9], across at least three longitudinal assessments [10].
- The earliest signs of dementia can be easily overlooked or misattributed by carers.
- Primary care providers should screen for dementia in people with ID. When this should occur depends on whether the person has Down syndrome.
- Comprehensive cognitive assessments should be conducted for those at high risk of dementia. This includes people with Down syndrome, and those with non-Down syndrome ID who show signs of slowing or declines.

  For People with Down Syndrome

  - provide education for the person with Down syndrome and their carers about the risk of dementia and the warning signs, at around 30 years of age
  - include questions about functional declines, cognitive slowing, or changes in personality or executive function in annual health checks from the age of 30 onwards. This could include a carer-report checklist to facilitate discussion (see below)
  - arrange a comprehensive baseline cognitive assessment at around age 30 to establish a ‘healthy baseline’. Repeat the baseline cognitive assessment

at the age of 40 for those who appear to have no signs of functional decline and/or no concerns

- arrange regular (e.g. annual) repeat assessments where concerns or changes are noted (whether before the age of 40 or afterwards).

For People with non-Down Syndrome Intellectual Disability

- ask questions regarding decline in function, changes in personality or behaviour, and cognitive slowing at the age of 40 and again at 50, and each year thereafter. A carer-report checklist could also be used at these points (see below).
- arrange regular (e.g. annual) comprehensive assessments once a concern or change in cognition or function is noted.
- A carer-report screening checklist is available in several languages downloadable from http://aadmd.org/ntg/screening. This is currently recommended as a qualitative tool only. It can facilitate a discussion with the person and their carer/s regarding potential declines. Where declines are noted, a person should be referred to a psychologist or psychiatrist for a comprehensive assessment.
- A comprehensive assessment should cover memory; executive function; praxis; visual spatial skills; language including a sample of their writing and/or utterances; attention and processing speed [11]; and adaptive behaviour (or Instrumental Activities of Daily Living).

e) Other investigations

- Medical examination including biochemical, haematological and thyroid function tests, should be performed as per recommendations for the general population [12].
- Testing for the APOE ε4 allele can help determine dementia risk
- Assess vision and hearing problems. This may require referral to a specialist optometrist or audiologist in the case of illiterate or non-verbal people.
- Neuroimaging can cause a high degree of anxiety for people with ID, and can require sedation for people with moderate to profound ID.

- Review medications and doses, particularly drugs with anticholinergic effects, even if the person has been on them for some time [11].

f) Differential Diagnoses are as per the general population

- Note the increased risk of undiagnosed medical or mental health problems, many of which are treatable.
- Also note the potential for grief and abuse to present as declines [12].

g) Managing dementia in ID Coordination of services

Adults with ID frequently fall through the gaps between services. When making referrals, first check service eligibility criteria.

- A person with ID who develops dementia remains eligible for disability-related supports but should also eligible for specific dementia care services.
- The National Younger Onset Dementia Keyworker Program can be accessed even before a formal diagnosis is made.
- A range of allied health professionals may be involved in the care of someone with ID and dementia to promote their wellbeing.

h) Sharing the diagnosis

Wherever possible, communicate the diagnosis to the person with ID in a manner they can understand.

- Establish ahead of time if they want a support person to be present, if they require communication aids, and assemble resources such as easy-to-read fact sheets about dementia. An example is available for download from the Alzheimer’s Society ((United Kingdom (UK)) website at https://www.alzheimers.org.uk/site/scripts/download_info.php?downloadID=1092
- Communicate both the diagnosis and the supports available to them and any options regarding future care. Ascertain their preferences for care options.
- The diagnosis should also be communicated to the person’s support network, once consent to their involvement is given. Provide education regarding dementia and what to expect.

i) Risk Assessment

- Do a standard risk assessment but repeat it more often as decline may be faster.
- Wandering, getting lost, or choking may present earlier. The risk of abuse may be higher.

j) Planning for declines

As dementia progresses, the care goal needs to shift from supporting independence towards providing care and eventually palliative care [13, 14].

- In many cases, the person with ID, their family and service providers want them to remain ageing in place. However, if and when their care requirements can no longer be met in their current place, options include transfer to an aged-care facility or to another disability service.
- Long-term planning for such transitions is important.

k) Managing medical complications

People with ID are more likely to have pre-existing physical health problems than the general population.

- Late-onset seizures are particularly common in people with Down syndrome and AD.

l) Review mental health and behaviour

People with ID (without dementia) experience a high rate of behaviours of concern, and so there is risk of new or escalating behaviours being overlooked or misattributed.

- Review existing medications before prescribing new ones to manage behaviours [15].
- Commence medication at a lower dose with attentive follow-up [15].
- Be aware that extrapyramidal side effects may be difficult to recognise in people with ID [16, 17].

m) Cholinesterase inhibitors and memantine

- Further research is needed to establish the efficacy of these medications in people with ID.

- Be aware of the increased potential for side effects, especially in people with Down syndrome who can often have cardiac problems and small stature.
- Commence at a lower dose, with slow titration and frequent review, particularly for those with Down syndrome.

n) Caring for a family carer of a person with ID and dementia

- Family carers of people with ID who develop dementia are a unique group of carers. Where needed, arrange access to mental health professionals and encourage carers to access respite services.
- Educational resources on caring for a person with ID and dementia are available from the Down's Syndrome (UK) website (https://dsagsl.org/wp-content/uploads/2012/11/ds_and_alzheimers1.pdf) and the BILD (UK) website (http://www.bild.org.uk/resources/ageingwell/dementia/)

3. List of Resources

The US National Task Group	Early Detection Screen for Dementia (NTG-EDSD) – http://aadmd.org/ntg/screening
The British Psychological Society	"Dementia and People with Intellectual Disabilities: Guidance on the assessment, diagnosis, interventions and support of people with intellectual disabilities who develop dementia" http://www.bild.org.uk/resources/ageingwell/dementia/
Bild (UK)	"Dementia Support" http://www.bild.org.uk/resources/ageingwell/dementia/
Alzheimer's Society (UK)	"Factsheet: What is dementia?" https://www.alzheimers.org.uk/site/scripts/download_info.php?downloadID=1092
Down's Syndrome Association (UK)	"Down's Syndrome and Alzheimer's Disease: A Guide for Parents and Carers" https://dsagsl.org/wp-content/uploads/2012/11/ds_and_alzheimers1.pdf
IDMH	Intellectual Disability mental health e-learning resources for Professionals http://www.idhealtheducation.edu.au/
Dementia Australia	"Living with intellectual disability and dementia" https://www.dementia.org.au/about-dementia/resources/videos/collections?playlist=Intellectual%20Disability
The University of Queensland	Comprehensive Health Assessment Program (CHAP) https://qcidd.centre.uq.edu.au/resources/chap

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