



Faculty of Medicine, The Department of
Developmental Disability Neuropsychiatry 3DN

Dementia in people with Intellectual Disability: Guidelines for Australian GPs.

Elizabeth Evans
Research Fellow
Department of Developmental Disability Neuropsychiatry
School of Psychiatry, Faculty of Medicine
University of New South Wales, Sydney
lizevans@unsw.edu.au

Professor Julian Trollor
Chair, Intellectual Disability Mental Health
Head, Department of Developmental Disability Neuropsychiatry
School of Psychiatry, Faculty of Medicine
University of New South Wales, Sydney
j.trollor@unsw.edu.au

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Summary of key recommendations

1. GPs need to be aware that people with intellectual disability (ID) are at increased risk of dementia.
2. 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.
3. 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.
4. 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.
5. Diagnosing dementia in people with ID requires establishing longitudinal declines in function across at least 3 sequential assessments. Standard tests used with the general population are unsuitable for this group.
6. 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.
7. Important principles for managing dementia in patients with ID include:
 - a. 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>.
 - b. Screen and examine for other potential causes of cognitive decline
 - c. 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.

Further resources and links are listed on page 17.

Short summary version:

Prevalence and incidence of dementia in Intellectual Disability

- 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 Zigman & Lott, 2007 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 (Dodd et al., 2015).

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.

Presentation of dementia in people with ID

- The full range of dementias may appear in people with ID, including mixed presentations (Strydom, Livingston, King, & Hassiotis, 2007). 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 (Adams & Oliver, 2010; Ball et al., 2006; Ball, Holland, Treppner, Watson, & Huppert, 2008; Deb, Hare, & Prior, 2007a; Holland, Hon, Huppert, & Stevens, 2000).

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 (Torr, 2016), across at least three longitudinal assessments (Burt and Alyward 2000).
- 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-DS ID:

- 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/ntq/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 (Torr, 2009); and adaptive behaviour (or Instrumental Activities of Daily Living).

Other investigations

- Medical examination including biochemical, haematological and thyroid function tests, should be performed as per recommendations for the general population (Tyrrell, Mulryan, & Dodd, 2014).
- Testing for the APOE $\epsilon 4$ 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 (Torr, 2009).

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 (Tyrrell et al., 2014).

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 and should also be 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.
- 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 at <https://www.alzheimers.org.uk/get-support/publications-and-factsheets/easy-read-factsheet-what-dementia>
 - 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.
- 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.

- Planning for declines: As dementia progresses, the care goal needs to shift from supporting independence towards providing care and eventually palliative care (Carling-Jenkins & Bigby; Jokinen, Janicki, Keller, McCallion, & Force, 2013).
 - 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.
- 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.
- Review mental health and behaviour: People with ID (without dementia) experiences 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 (Trollor, Salomon, & Franklin, 2016).
 - Commence medication at a lower dose with attentive follow-up (Trollor et al., 2016).
 - Be aware that extrapyramidal side effects may be difficult to recognise in people with ID (de Kuijper, Evenhuis, Minderaa, & Hoekstra, 2014; Lindsay, 2011).
- **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.

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 at http://dsagsl.org/wp-content/uploads/2012/11/ds_and_alzheimers1.pdf and <http://www.bild.org.uk/information/ageingwell/dementia/>.

Literature Review – Dementia in ID

Prevalence and incidence of dementia in ID.

People with intellectual disability (ID) are at increased risk of dementia compared with people without ID. It is well-known that Down syndrome carries a higher risk of dementia, and specifically Alzheimer's disease, with younger onset (see Zigman & Lott, 2007 for review). Around 50-70% of people with Down syndrome aged over 60 exhibit the clinical symptoms of dementia (Janicki & Dalton, 2000). There is far less research on dementia in people with ID from other causes (non-DS ID), and studies looking at the prevalence of dementia in this group show conflicting results. Some (e.g., Cooper, 1997) find the prevalence to be higher in non-DS ID compared with the general population, while others find no significant increase (e.g., Zigman et al., 2004).

Still others have found an increased incidence of dementia (that is, the number of *new* cases within a time period) coupled with more rapid declines and an earlier death (Strydom, Chan, King, Hassiotis, & Livingston, 2013). Shorter survival times could mask the true rate of dementia in studies looking only at prevalence. Furthermore, the onset of dementia in people with non-DS ID is on average around 10 years earlier than in the general population (Dodd et al., 2015). Therefore, it makes sense for primary care providers to be alert to the potential for dementia in patients with intellectual disability, irrespective of whether the person has Down syndrome.

Risk factors for dementia in people with ID

The strongest risk factor for dementia in people with ID is having Down syndrome. Other factors found to increase the risk of dementia in people with ID include:

- poor physical and mental health status (e.g., Cooper, 1997; Moss & Patel, 1997). In particular, depression (Evenhuis, 1997) and epilepsy (Torr, 2005). There is a high rate of undiagnosed health problems in people with ID (Beange, McElduff, & Baker, 1995).
- an adverse cardiovascular health profile. Poor cardiovascular health is common in this group, yet vascular dementia is often overlooked (Torr, 2005)
- a high rate of sensory impairments, including undiagnosed ones.

There are also a number of factors known to increase risk in the general population that are common in people with ID (see Evans et al., 2013 for a review). These include:

- poor diet and exercise
- poorer engagement in education, social activities, and employment
- head injury.

Within people with Down syndrome, the APOE ϵ 4 allele has been found to be associated with a higher risk of dementia, earlier onset, and more rapid decline (Prasher et al., 2008; Torr, 2016). It has also been found to increase risk of mortality in those with and without dementia (Prasher et

al., 2008; Zigman, Jenkins, Tycko, Schupf, & Silverman, 2005). However, findings are mixed regarding whether the APOE ϵ 2 allele is protective in people with Down syndrome as it is in the general population (Prasher et al., 2008; Tyrrell et al., 1998). It has been suggested that the triplication of the amyloid precursor protein (APP) gene on chromosome 21 overwhelms the potential protection from ϵ 2 for those with Down syndrome (Prasher et al., 2008). There is inadequate research in people with non-Down syndrome ID regarding the risk conferred by APOE genotype.

Presentation of dementia in people with ID

In people with Down syndrome, behavioural and personality changes and declines in executive function may appear before memory deficits become obvious (Adams & Oliver, 2010; Ball et al., 2006; Ball et al., 2008; Deb et al., 2007a; Holland et al., 2000). These early signs can resemble fronto-temporal dementia, which is usually followed by the memory and cognitive declines more typical of Alzheimer's disease. However, less common forms, such as Dementia with Lewy Bodies, have also been documented in this group (Holland, Hon, Huppert, Stevens, & Watson, 1998).

The presentation of dementia in non-DS ID is less researched. However, the full range of dementias may appear as per the general population, including mixed presentations (Strydom et al., 2007). Several research studies (e.g., Evenhuis, 1997; Johannsen, Christensen, & Mai, 1996; Zigman et al., 2004) have found a sizeable subgroup of people with ID who meet some, but not all, criteria for dementia.

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, since people with ID are likely to score poorly on tests of cognition even in the absence of dementia. Diagnosing dementia in people with ID therefore requires demonstrating a decline from baseline in cognition and functioning (Torr, 2016). However, in practice, many people will not have had a baseline assessment before declines are noted.

Moreover, the earliest signs of dementia can be easily overlooked by carers. There is a lack of information in the community regarding dementia in people with ID and misattributing signs of decline to 'ageing', 'behavioural issues', or the disability itself, is common. For these reasons, it is important for primary care providers to ask regarding changes in functioning, including more subtle signs such as coarsening behaviour and personality changes. Where concerns are noted, the GP has a vital role to play in coordinating more comprehensive investigations.

When to assess – Down syndrome

Given their high risk of dementia, adults with Down syndrome should be actively monitored for signs of dementia. This allows a baseline level of function to be established while the person is still healthy, leading to an earlier diagnosis where dementia does occur. Some literature recommends that people with Down syndrome receive a comprehensive assessment at around age 30 or 35, then are reassessed every 2 years in their 40's and annually from the age of 50 onwards (Dodd et al., 2015; Turk, Dodd, & Christmas, 2001). Others suggest baseline assessment when the person is in their 40's (e.g. Jokinen et al., 2013). Until further research clarifies, a practical approach would be to:

- arrange a comprehensive baseline cognitive assessment at around age 30
- 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 each year from the age of 30 onwards. This could include a carer-report checklist to facilitate discussion (see below)
- arrange a repeat baseline cognitive assessment at the age of 40 for those who appear to have no signs of functional decline and/or no concerns
- where concerns or changes are noted, arrange regular (e.g. annual) repeat assessments.

When to assess – non-Down syndrome ID

For those with ID from other causes, baseline cognitive testing of healthy individuals may not be indicated (Torr, 2016). However, GPs should still be attentive to the possibility of dementia, particularly of earlier onset. Here, a practical approach would be:

- ask questions regarding decline in function, changes in personality or behaviour, and cognitive slowing at the age of 40. A carer-report checklist could also be useful at this point (Jokinen et al., 2013) - see below
- repeat these questions and carer checklist at the age of 50, and each year thereafter.
- where a concern or change in cognition or function is noted, arrange regular (e.g. annual) comprehensive assessments.

Screening checklists

Screening checklists that are useful in the general population will not be of use with people with an ID, who are likely to obtain atypical scores on standardised measures of cognition even in the absence of dementia. Comparing the person's current function against prior reports of their capacity will be more informative. With the person's consent, asking for information from a family member, carer or keyworker can be useful for establishing a pattern of declines, but that informant should be someone who has known the person with ID for some time.

A number of carer-report checklists to screen for dementia in people with ID have been developed. One which is freely available is the Dementia Screening Questionnaire for Individuals with Intellectual Disability (DSQIID; Deb, Hare, Prior, & Bhaumik, 2007b), which has subsequently been adapted into the National Task Group Early Detection Screen for Dementia (NTG-EDSD, 2013). At present, the US National Task Group recommends using this as a qualitative clinical tool only. It is available in several languages and can be downloaded free from <http://aadmd.org/ntg/screening>.

Referring for a comprehensive cognitive assessment

A detailed assessment of cognition should be undertaken by a psychologist or psychiatrist with experience assessing adults with ID. Areas to assess include: memory, executive function, praxis, visual spatial skills, language including a sample of their writing and/or utterances, attention, and processing speed (Torr, 2009). In addition, adaptive behaviour or Instrumental Activities of Daily Living (IADLs) should be assessed. Both of these terms refer to the skills used in everyday life, such as self-care skills, managing finances, and communication. Disability workers are likely to be more familiar with the concept of adaptive behaviour, while those working in the aged care sector are more likely to use the term Activities of Daily Living.

At least three longitudinal assessments are required to establish the presence of dementia in someone with ID (Burt & Aylward, 2000). This is because test scores tend to be less stable in this population than in those without ID. Where the diagnosis is uncertain, it is better to wait and continue to investigate and address other causes of decline, rather than give a hasty diagnosis (Torr, 2009). Address any health problems or sensory impairments and review medications prior to the next cognitive assessment.

Neuroimaging

Neuroimaging may be required as part of clinical investigations in complex cases or to exclude other causes of symptoms. However, for people with moderate to profound ID, neuroimaging may require sedation, or sometimes a general anaesthetic, which carry risks in themselves. In the absence of sedation, an MRI can provoke a high degree of anxiety for the person with ID. Providing them and their carer with information regarding what to expect ahead of time can be beneficial. Some scanning services offer familiarisation programs prior to the actual scan.

The brains of people with ID, especially those with genetic syndromes, can be structurally different from the general population even in the absence of dementia, and this can complicate interpreting the results of imaging in these patients (Torr, 2009).

Other investigations

A thorough medical examination is an essential first step, and other investigations should follow the recommendations for the non-ID population. This includes biochemical, haematological and thyroid function tests (Tyrrell et al., 2014). Undiagnosed vision and hearing problems are common in this group so these should be investigated prior to cognitive assessment. This may require referral to a specialist optometrist or audiologist in the case of illiterate or non-verbal people. Testing for the APOE ε4 allele may determine the risk that declines are due to dementia.

GPs should be vigilant regarding the high rate of polypharmacy in adults with ID, and their propensity for adverse reactions. In the face of declining function or changes in behaviour, medications and doses should be reviewed. This has particular importance for drugs with anticholinergic effects, even if the person has been on that medication for some time (Torr, 2009).

Differential Diagnoses to consider

As in the general population, there are a number of medical conditions that can mimic dementia and that must be investigated and excluded. These include delirium, which is common in people with ID but often overlooked (Torr, 2016). Other differentials include depression, other mental health problems, thyroid disorders, infection, sensory loss, malignancy, nutritional deficiencies such as folate or vitamin B12, sleep disorders, and reactions to medications including polypharmacy. Non-medical considerations include grief and abuse (Tyrrell et al., 2014). Many of these conditions are treatable, and are more common in people with ID than the general population (Torr, 2009).

Communicating with the person with ID

Assessing any mental health condition in a person with ID requires time and often requires adapting one's style of communication. Helpful e-learning regarding assessing health and mental health in people with ID, is available at www.idhealtheducation.com.au.

Managing dementia in ID:

Primary care providers play an integral role in coordinating and managing the care of a patient with ID who has been diagnosed with dementia. Vigilant health monitoring with specialist referrals as appropriate can promote optimal health for as long as possible. Coordination may encompass services across three sectors (health, disability and aged care). The person remains eligible for disability-related supports but may also be eligible for aged care services. A person with younger-onset dementia may be eligible for some aged care services based on their diagnosis. The National Younger Onset Dementia Keyworker Program can be accessed even before a formal diagnosis is made.

Adults with ID frequently fall through the gaps between services, and their carers report difficulties navigating appropriate pathways to care (The Disability Investment Group, 2009). When making referrals, check service eligibility criteria and ensure that the referral specifies how the patient meets these.

Sharing the diagnosis

Communicating the diagnosis to the person with ID is important as it allows them to be as involved as possible in decisions regarding their care (Jokinen et al., 2013; Tyrrell et al., 2014). More information about core clinical competencies when working with a person with intellectual disability can be found in guidelines produced by Department of Developmental Disability Neuropsychiatry (2016), available for download at <https://3dn.unsw.edu.au/idmh-core-competency-framework>.

Sharing the diagnosis in a manner that the person can understand may require some planning. Ahead of this appointment, GPs could helpfully:

- Ask if they want a support person to be present
- Ask what communication aids they usually use, if any, and ensure these are available at the time
- Assemble resources such as easy-to-read fact sheets about dementia. An example is available for download at <https://www.alzheimers.org.uk/get-support/publications-and-factsheets/easy-read-factsheet-what-dementia>
- During the appointment:
 - Adapt your communication style to meet their needs
 - Gauge the person's capacity to understand the past, present and future, and the way they normally talk about these things. Tailor the way you share information about dementia accordingly
 - Break the information down into chunks and check their understanding as you go.
 - Communicate not only the diagnosis but also the sorts of supports available to them and any options regarding future care.

The diagnosis should also be communicated to the person's support network, provided consent to their involvement is given. It is helpful to also provide education regarding dementia and what to expect.

Risk Assessment

A risk assessment for dementia patients with ID should cover the same issues as for other dementia patients, but physicians should be mindful that the risk of abuse may be higher. Because there are fewer functional skills to lose, risks of things like wandering, getting lost, or choking, may present earlier in the illness. Furthermore, the rate of decline may be faster in this group (Strydom

et al., 2013) and so a risk assessment should be repeated more often. Staff in residential services should be encouraged to assess ways they can provide increased or closer supervision and support to manage risks.

Arranging supports to maintain function and optimise health

A range of allied health professionals may be involved in the care of someone with ID and dementia to promote their wellbeing, per the general population.

Planning for declines

As dementia progresses, the type of supports provided and the goal of that support will need to shift from supporting independence and promoting skill maintenance in the early stages, through to providing nursing and personal care, and eventually palliative care (Carling-Jenkins & Bigby; Jokinen et al., 2013). Many people with ID who live at home or in residential care express a desire to age in place, and in many cases their families and service providers are supportive of this. For how long this is feasible and appropriate depends on many factors, including the physical home environment, the capacity of staff or family to provide appropriate supports, and the needs of other residents. When the person's care requirements can no longer be met, options include transfer to an aged-care facility or to another, better equipped, disability service. Staff members working in disability group homes and family carers may need education about how to support someone with dementia. Meanwhile, if the person is transferred to an older aged service, that service may need information about supporting someone with ID.

Early discussions about the future and long-term planning for transitions are therefore important (Jokinen et al., 2013). Where possible, the desires of the person with ID should be ascertained soon after diagnosis. Ask staff and family carers to monitor changes over time, in order to track the progress of dementia and ensure any changes in care are well-timed and can be planned in advance.

Managing medical complications related to dementia in a person with ID is similar to that for any dementia patient. However, this is more likely to be complicated by multiple pre-existing physical health problems. Late-onset seizures are particularly common in people with Down syndrome and AD, and these patients should be monitored for seizures. In this group, death occurs on average just 2 years following the onset of seizures (McCarron, Gill, McCallion, & Begley, 2005; Torr, 2016).

Review mental health and behaviour

As for any dementia patient, there is a high potential for behavioural and psychological symptoms associated with disease progression. Depression and anxiety are commonly comorbid to dementia (Torr, 2016; Tyrrell & Dodd, 2003), as are behaviours such as aggression. People with ID (without dementia) experiences a high rate of behaviours of concern, and so there is risk of new or escalating behaviours being overlooked or misattributed. However, behavioural programs using incentives or punishment are inappropriate in the context of dementia (Carling-Jenkins & Bigby). People with ID and dementia may respond to the same types of treatments used in other dementia patients, such as music or light therapy, but the evidence base in this group is lacking.

If medication is required to manage behaviours associated with dementia, be sure to review existing medications first. This is particularly important for adults with ID who already have high rates of polypharmacy. Extrapyramidal side effects of antipsychotics may be difficult to recognise in people with ID (de Kuijper et al., 2014), and even symptoms of the potentially fatal malignant extrapyramidal syndrome can be overlooked (Lindsay, 2011). Some research suggests these side effects may present as increased behavioural problems in adults with ID (Valdovinos et al., 2005). Commencing medication at a lower dose with attentive follow-up is recommended (Trollor et al., 2016).

Cholinesterase inhibitors

Most trials of cholinesterase inhibitors to treat dementia have excluded people with intellectual disability. A handful of small studies have examined their use to slow decline or even improve cognition in people with Down syndrome and Alzheimer's disease, with mixed results regarding benefits (see de la Torre, 2012; Prasher, 2004; Torr, 2016, for reviews). Case reports have suggested they may be effective for some individuals (e.g. Kondoh et al., 2005). These medications do appear to be safe for people with Down syndrome who do not have bradycardia or cardiac problems (Torr, 2016). However, Prasher and colleagues (2003; 2002) found that side effects were relatively common in people with Down syndrome, including gastrointestinal side effects, agitation, sleep disturbances, and muscle weakness. People with ID show increased side effects from certain medications and so caution is advised. This is particularly true for people with Down syndrome, who have a high rate of cardiac problems, typically small stature, and differences in brain structures compared with the general population (Torr, 2016). People with Down syndrome have been reported to have increased serum levels of donepezil, a cholinesterase inhibitor, for a given dose (Kondoh et al., 2005; Torr, 2016). Commencing medication at a lower dose with frequent review and slow titration is recommended (Prasher, 2004; Torr, 2016).

Memantine

Although transgenic mouse studies showed promising results for the use of Memantine in Down syndrome, it has been found to be safe but ineffective in adults with Down syndrome (Hanney et al., 2012). However, only 35% of people in that trial had dementia. Further research is needed to establish the efficacy of this drug for people with Down syndrome.

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. Unlike many other dementia carers who are spouses or children of the patient, carers of adults with ID are frequently their parents. Receiving the diagnosis of dementia in their son or daughter can be devastating. With this can be the realisation that their child may predecease them, and that they themselves may be unable to continue to meet their son or daughter's care needs. Some carers will not be aware of the increased risk of dementia in people with ID and may require additional support and information about what to expect. Research has suggested that both respite and coping skills play an important role in mediating the impact of the caring role on a person's mental health.

Primary care providers have an important role to play in assessing carers' wellbeing and their experience of burden and its impact on their mental health (Parks & Novielli, 2000). Where needed, arranging access to mental health professionals and encouraging carers to access respite services can improve the quality of life for both the carer and the person with ID. Some useful resources specific to caring for a person with ID and dementia are available at http://dsagsl.org/wp-content/uploads/2012/11/ds_and_alzheimers1.pdf and <http://www.bild.org.uk/information/ageingwell/dementia/>.

Promoting Healthy Ageing in Patients with ID

Adults with ID face many risk factors for dementia, but some of these are modifiable. GPs can assist people with ID to reduce their risk of dementia by:

- Conducting annual health checks using a tool such as the Comprehensive Health Assessment Program (CHAP), for which there are specific Medicare item numbers - see <https://qcidd.centre.uq.edu.au/resources/chap> for further details.
- Assessing and addressing cardio-metabolic health risk factors, including nutrition and exercise.
- Giving the person with ID, and their carer where appropriate, information regarding the importance of social contact and engaging in meaningful educational, occupational and leisure activities.

List of resources:

<http://aadmd.org/ntg/screening>

http://www.bps.org.uk/system/files/Public%20files/rep77_dementia_and_id.pdf

<http://www.bild.org.uk/information/ageingwell/dementia/>

<https://www.alzheimers.org.uk/get-support/publications-and-factsheets/easy-read-factsheet-what-dementia>http://dsagsl.org/wp-content/uploads/2012/11/ds_and_alzheimers1.pdf

<http://www.bild.org.uk/information/ageingwell/dementia/>

<http://www.idhealtheducation.edu.au/>

<https://fightdementia.org.au/videos/living-with-intellectual-disability-and-dementia>

<https://qcidd.centre.uq.edu.au/resources/chap>

<https://our.learningdisabilitieshealthnetwork.org.uk/t/dementia-and-people-with-learning-disabilities-making-reasonable-adjustments/475>

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