

# Dementia in Down syndrome

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# What are we going to talk about today?

- What dementia is
- Why it is common in people with Down syndrome
- Signs of dementia in people with Down syndrome
- Preventing or delaying dementia
- What to do if someone shows signs of dementia
- What to expect from others
- Other considerations
- Resources

# What is dementia?

# Dementia is a syndrome:

- Many causes
- Usually progressive and irreversible
- Declines:
  - Thinking & memory
  - Functional skills
- Onset:
  - Younger: <65 years
  - Older: 65+ years

# Dementia is NOT 'normal' Ageing

Area	Normal Ageing	Abnormal Ageing
Forgetting	Occasional & temporary	Whole experiences
Thinking	A bit slower	All affected
Decision making	OK	Major trouble
Knowing surroundings	OK	Difficult
Recognising loved ones	OK	Difficult
Mental health	OK	Hearing voices, seeing things
Skills lost	Due to health conditions	Unrelated to conditions

# **“Dementia” vs “Major Neurocognitive disorder”**

➤ essentially the same thing.

# What is Mild Cognitive Impairment (MCI)?

= “Mild Neurocognitive Disorder”

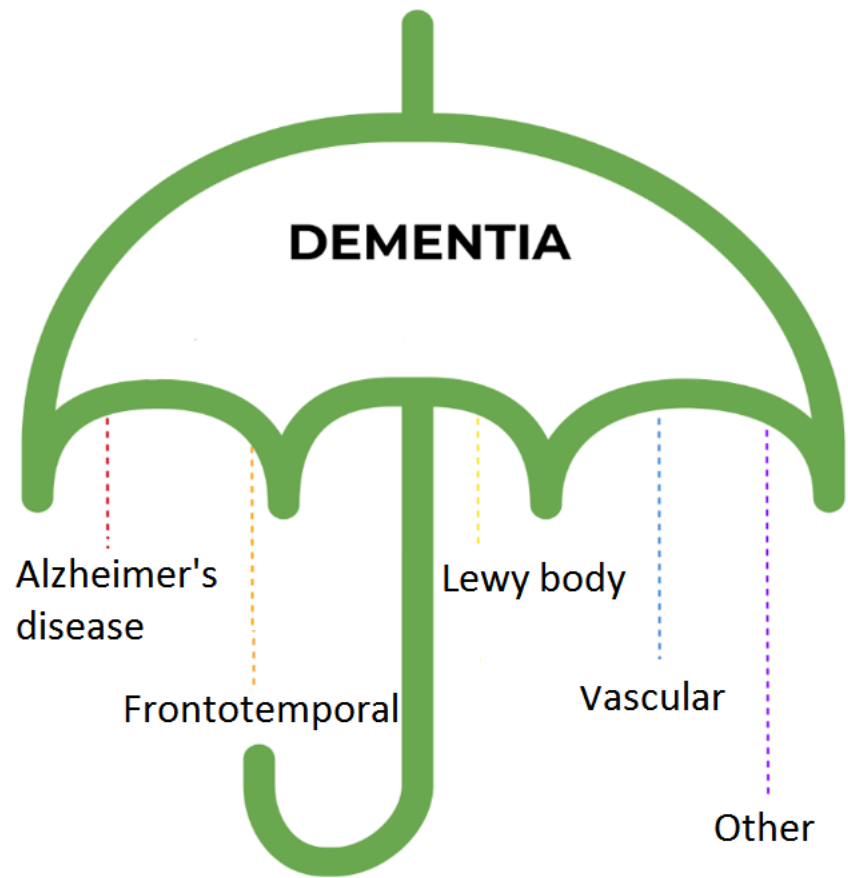
- Some declines in cognition
- Not dementia
  - but may progress to become dementia
- Many causes – some treatable

# Types of dementia <sup>1</sup>

Can be mixed

## In Down syndrome:

- Alzheimer's most common
- Others rarer
- "Dementia due to Down syndrome"

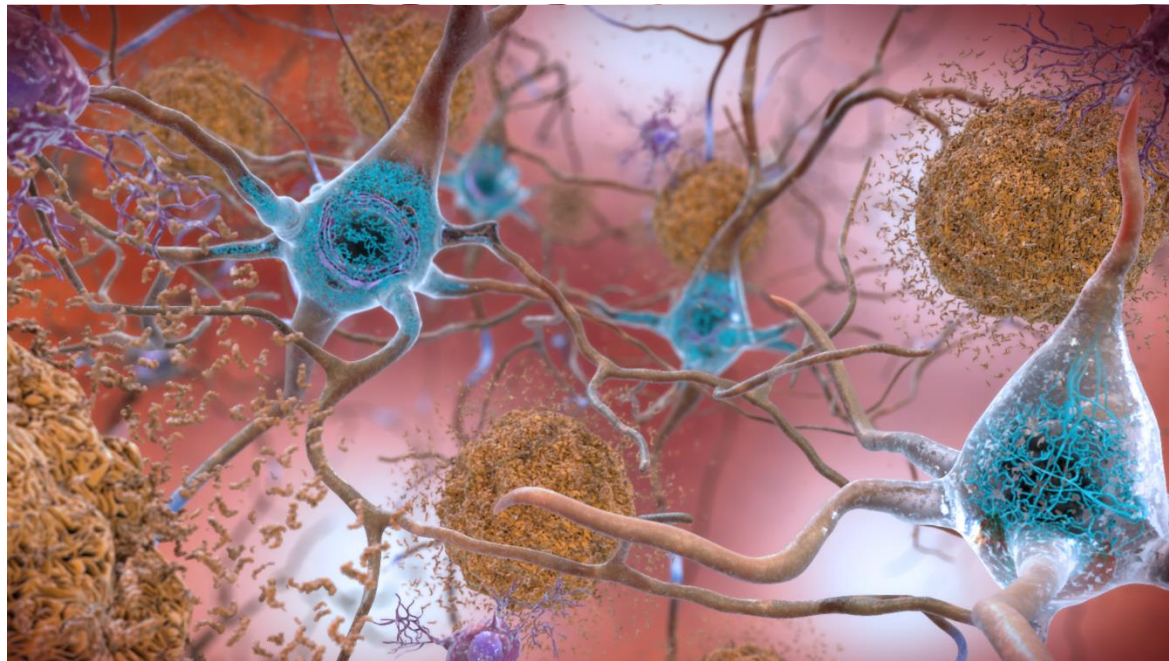


[1] <https://www.dementia.org.au/information/about-dementia/types-of-dementia>



# Brain changes in Alzheimer's disease

- Build up of proteins:
  - **Amyloid** 'plaques'  
└─┬─▶ **Chromosome 21**
  - **Tau** 'tangles'
- Shrinkage
- Inflammation



National Institute on Aging, NIH, US Department of Health & Human Services.

# Why is dementia more common in people with Down syndrome? <sup>1,2,3</sup>

- Poor physical health
- Sensory disabilities
- Obstructive sleep apnoea
- Lifestyle factors
- 'Cognitive reserve'

[1] Evans et al 2013; Wilcock et al 2016; Trollor et al 2016.

# How common is dementia in Down syndrome?

Research – variable rates: 1, 2, 3

Age	Rate
< 40	<10%
50's	30%
60+	50%
> 65	80%

Average age of diagnosis: mid 50's. <sup>3</sup>

[1] Sinai, et al. 2014; [2] Margallo-Lana et al 2007; [3] McCarron et al 2017.

# What are the signs of dementia in someone with Down syndrome?

# Early signs of Alzheimer's disease<sup>1</sup>

## Problems with:

- Memory
  - recent worst
- Visuo-spatial skills <sup>2</sup>
- Attention
- Planning
- Problem solving
- Judgement.

## Personality changes:

- Stubbornness
- Apathy
- Motivation
- ↓ social engagement

## Behaviour changes:

- Irritability, aggression, self-injury

[1] Ball et al 2008; [2] Strydom 2018

# Course of dementia

Memory

Recent

Distant

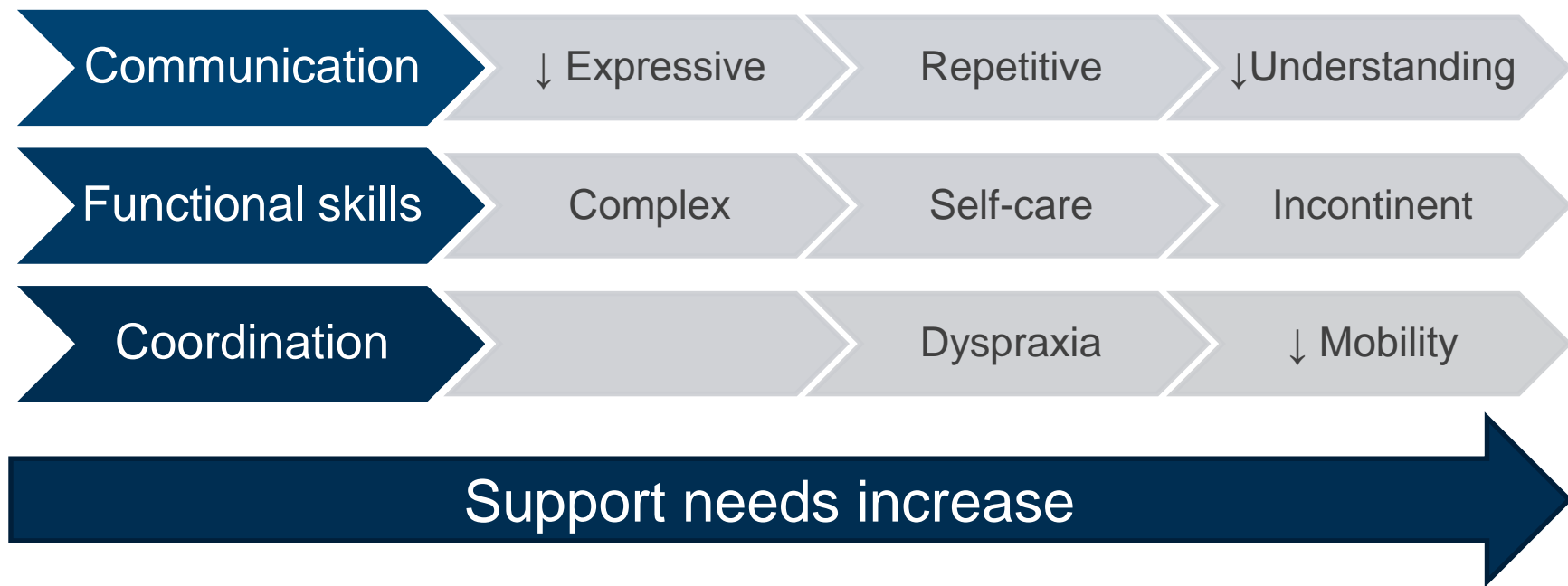
Orientation

? Day or time

? Familiar  
places

↓ Response  
to  
environment

# Course of dementia



# Seizures:

Alzheimer's disease: 10 – 22%. <sup>1</sup>

Down syndrome: <sup>2</sup>

- 50+ years: 46%
- Alzheimer's disease: 84%
- Childhood-onset = ↓ risk of dementia.

[1] Hauser et al 1986; [2] Mendez, M. 2005.



# Preventing or Delaying Dementia

# Preventing or delaying dementia - 1

Healthy lifestyle: <sup>1,2,3</sup>

- Diet & exercise
- Address sleep apnoea

Stay active: <sup>2</sup>

- Education
- Vocation
- Leisure
- Social

[1] Sisirak & Marks 2015; Reppermund & Trollor 2016; WHO 2000

# Preventing or delaying dementia - 2

Reduce inflammation: 4

- Infections
- Dental issues

**Regular medical & dental check-ups**


Review medications:

**Minimise anticholinergic load**

# Start monitoring

# Recording baseline function

- Free carer checklist
- 15 – 20 minutes
- Unscored
- Annual Health Checks:
  - Age 30 years
  - Regularly aged 40 +
  - Declines



## NTG-EDSD

v.1/2013.2

The NTG-Early Detection Screen for Dementia, adapted from the DQIID<sup>®</sup>, can be used for the early detection screening of those adults with an intellectual disability who are suspected of or may be showing early signs of mild cognitive impairment or dementia. The NTG-EDSD is not an assessment or diagnostic instrument, but an administrative screen that can be used by staff and family caregivers to note functional decline and health problems and record information useful for further assessment, as well as to serve as part of the mandatory cognitive assessment review that is part of the Affordable Care Act's annual wellness visit for Medicare recipients. This instrument complies with Action 2.B of the US National Plan to Address Alzheimer's Disease.

It is recommended that this instrument be used on an annual or as indicated basis with adults with Down syndrome beginning with age 40, and with other at-risk persons with intellectual or developmental disabilities when suspected of experiencing cognitive change. The form can be completed by anyone who is familiar with the adult (that is, has known him or her for over six months), such as a family member, agency support worker, or a behavioral or health specialist using information derived by observation or from the adult's personal record.

The estimated time necessary to complete this form is between 15 and 60 minutes. Some information can be drawn from the individual's medical/health record. Consult the NTG-EDSD Manual for additional instructions ([www.addmd.org/ntg/screening](http://www.addmd.org/ntg/screening)).

(1) File #: \_\_\_\_\_ (2) Date: \_\_\_\_\_

Name of person: (3) First \_\_\_\_\_ (4) Last: \_\_\_\_\_

(5) Date of birth: \_\_\_\_\_ (6) Age: \_\_\_\_\_

(7) Sex:

<input type="checkbox"/> Female
<input type="checkbox"/> Male

(8) Best description of level of intellectual disability

No discernible intellectual disability
Borderline (IQ 70-75)
Mild ID (IQ 55-69)
Moderate ID (IQ 40-54)
Severe ID (IQ 25-39)
Profound ID (IQ 24 and below)
Unknown

(9) Diagnosed condition (check all that apply)

<input type="checkbox"/> Autism
<input type="checkbox"/> Cerebral palsy
<input type="checkbox"/> Down syndrome
<input type="checkbox"/> Fragile X syndrome
<input type="checkbox"/> Intellectual disability
<input type="checkbox"/> Prader-Willi syndrome
<input type="checkbox"/> Other: _____

**Instructions:**  
For each question block, check the item that best applies to the individual or situation.

**Current living arrangement of person:**

- ☐ Lives alone
- ☐ Lives with spouse or friends
- ☐ Lives with parents or other family members
- ☐ Lives with paid caregiver
- ☐ Lives in community group home, apartment, supervised housing, etc.
- ☐ Lives in senior housing
- ☐ Lives in congregate residential setting
- ☐ Lives in long term care facility
- ☐ Lives in other: \_\_\_\_\_

# Other areas to assess – ideal scenario

- IQ
- Attention
- Processing Speed
- Visuospatial skills
- Language and communication
- Executive Function
- Fine motor

# Planning ahead

↑ Independence

+ Supports:

- For person
- For carers

Transitions

Memory-prompts:

- Memory boxes
- Communication books

# If someone shows signs of dementia



# 1. Where to start

GP

Consider other reasons:


- Physical health
- Depression
- Sensory
- Thyroid disorders
- Vitamin / mineral deficiencies
- Environmental stressors
- Medication reactions

? Treatable

## 2. Track changes

- NTG-EDSD
- Assessment of skills
- Clinical interview

v. 1/2013.2



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Male

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Intellectual disability
Prader-Willi syndrome
Other: _____

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- ☐ Lives in senior housing
- ☐ Lives in congregate residential setting
- ☐ Lives in long term care facility
- ☐ Lives in other: \_\_\_\_\_

# 3. Breaking the news

Supporting:

- the person with Down syndrome to understand
- Family & carers
- Others they love

Resources

# 4. Planning for changes<sup>1</sup>

- Focus: preserving & supporting skills
- Home modifications
- Risk assessments
- Allied health:
  - Falls prevention
  - Swallowing & speech

[1] Evans & Trollor 2018

# 5. Planning transitions

↑ supports needed

↑ time: ↑ choice:

- Carers & staff can prepare
- Discuss options and preferences early <sup>1</sup>
- Ageing in place vs change residence

[1] Tower et al 2014.

## 6. Maximise quality of life

- Healthy lifestyle
- Stay active & engaged: ↑ support
- Address:
  - Physical health
  - Sleep problems
  - Depression
- Use memory boxes & communication books
- Aids

# 6. Medications

- Research on Down syndrome <sup>1,3,2</sup>
  - results unclear
  - benefit some
  - side effects

Types:

- Memantine
- Cholinesterase inhibitors

[1] Courtenay & Eady 2014; [2] Eady et al 2018; [3] Torr 2016.

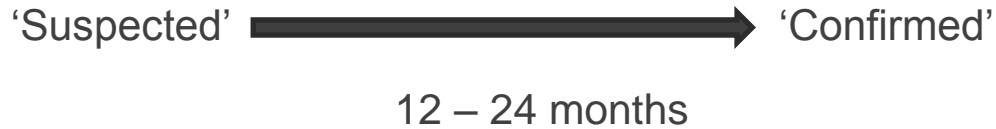
# 7. Address related symptoms

- Behavioural:
  - Common in dementia
  - Management:
    - » Environmental changes
    - » Medications
- Seizures:
  - Medications



# What to expect from others

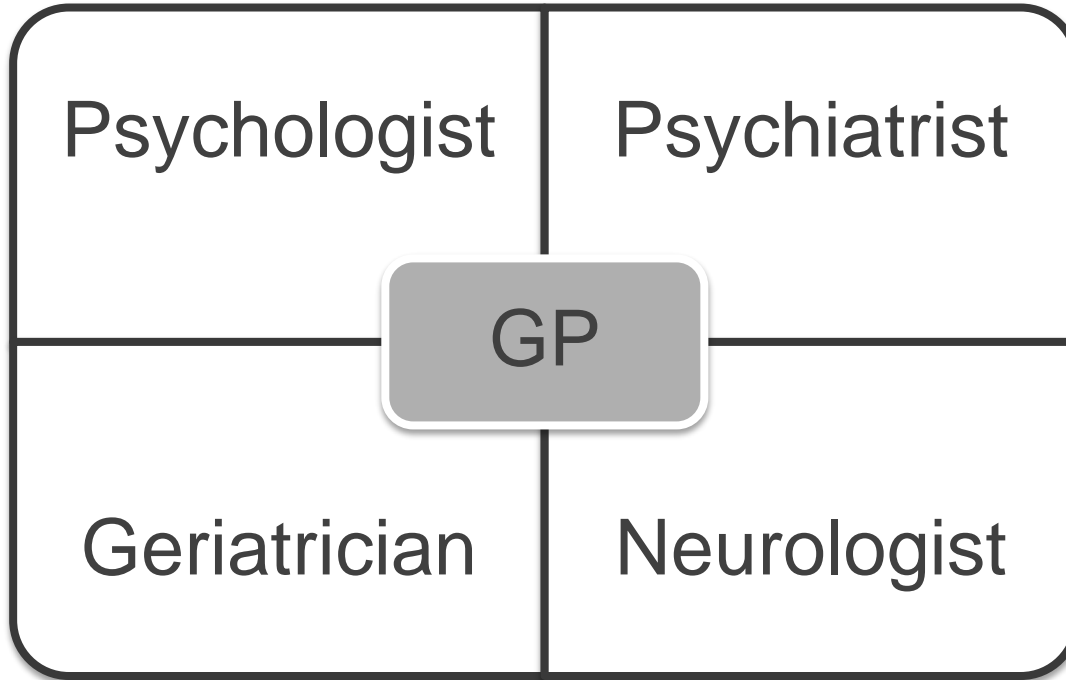
# Diagnosis – a lengthy process



# What should a GP do?

- Listen
- Comprehensive health check
  - Blood tests
  - Review medications
  - Refer - vision and hearing
- Monitor change:
  - Before & after diagnosis
- Ask about carers separately

# What should a specialist do



# NDIS

Supports related to disability:

- Onset < 65 years

From 1 October: specific health supports

# Aged care

Supports for disability:

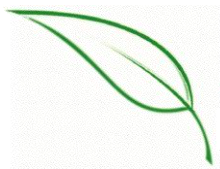
- onset > 65 years
- Dementia

# Younger onset dementia supports

Can be accessed:

- < 65 years
- Dementia:
  - Suspected
  - Confirmed

# What else can I do?



# The SAGE-ID Study

## Recruiting nationwide:

- People with ID aged 40+
  - with or without declines.

## More info:

- Call Liz on 02 9931 9160
- Email: [sageid@unsw.edu.au](mailto:sageid@unsw.edu.au)



# Carers

- Build & maintain your own supports
  - Better mental wellbeing
- Seeking volunteers:
  - Guide Australian adaptation of resource.
  - Contact Liz if interested.

# Advocacy

Services need improving: 1,2, 3

- Baseline assessments
- Pathways to diagnosis and care
- ↑ Supports to meet need
- Incremental steps

[1] Salvador-Carulla et al. (2012); [2]Carling-Jenkins et al. (2014); [3] Washko et al. (2012).

# Resources

# For people with intellectual disability:

- Easy read factsheet from Alzheimer's Society (UK):  
[https://www.alzheimers.org.uk/site/scripts/download\\_info.php?downloadID=1092](https://www.alzheimers.org.uk/site/scripts/download_info.php?downloadID=1092)
- *Jenny's diary* is a book made of a set of postcards, designed to facilitate conversations with people with ID about dementia. There is a section for talking with people who are friends with the person, as well as a section for talking with the person who has dementia.
- <https://www.stir.ac.uk/about/faculties-and-services/health-sciences-sport/research/research-groups/enhancing-self-care/jennys-diary/>
- *About my friend* by Karen Dodd, Vicky Turk and Michelle Christmas (2005). A resource to assist friends of people Down's syndrome and dementia.
- *Ann has dementia*. By Sheila Hollins, Noelle Blackman and Ruth Eley. Illustrated by Lisa Kopper. A story told in pictures, available from Books beyond words.  
<https://booksbeyondwords.co.uk/ebooks/ann-has-dementia?rq=dementia>

# For families and carers - Books

- *Intellectual disabilities and dementia: a guide for families* edited by Karen Watchman (2017)
- *Jenny's diary* is made of postcards, designed to facilitate conversations with people with ID about dementia in particular. There is a section for talking with people who are friends with the person, as well as a section for talking with the person who has dementia. Available here:

<https://www.stir.ac.uk/about/faculties-and-services/health-sciences-sport/research/research-groups/enhancing-self-care/jennys-diary/>

## ***Dementia Australia library service:***

- Books loaned by post 4 or 8 weeks
- Their list on dementia and Down syndrome is here:  
<https://www.dementia.org.au/files/library/Down%20Syndrome%20and%20dementia.pdf>

# For families and carers – websites:

- The **carer checklist** recommended by the US National Task Group: <http://aadmd.org/ntg/screening>
- Websites with information and resources:
  - Karen Watchman's website : [www.learningdisabilityanddementia.org](http://www.learningdisabilityanddementia.org)
  - The Down's Syndrome (UK) website [http://dsagsl.org/wp-content/uploads/2012/11/ds\\_and\\_alzheimers1.pdf](http://dsagsl.org/wp-content/uploads/2012/11/ds_and_alzheimers1.pdf)
  - The BILD (UK) website <http://www.bild.org.uk/resources/ageingwell/dementia/>
  - Down syndrome Australia: [https://www.downsyndrome.org.au/documents/resources/health/Down\\_syndrome\\_and\\_Alzheimers\\_disease.pdf](https://www.downsyndrome.org.au/documents/resources/health/Down_syndrome_and_Alzheimers_disease.pdf)
- Dementia Australia has made a video about dementia in people with intellectual disability, available through the “resource finder” section of their website.
- *Living with Dementia* booklet from Down syndrome Scotland. Includes practical environmental modifications. <https://www.dsscotland.org.uk/resources/publications/for-parents-of-adults/>

# For health & disability professionals

- The **carer checklist** recommended by the US National Task Group is available from this site: <http://aadmd.org/ntg/screening>. Other resources and webinars: <http://aadmd.org/ntg>
- 3DN resources @ <https://3dn.unsw.edu.au/content/health-mental-health-professionals> :
  - Dementia in people with Intellectual Disability: Guidelines for Australian GPs; Cardiometabolic Resources
  - E-learning modules for health & disability professionals: [www.idhealtheducation.edu.au](http://www.idhealtheducation.edu.au)
- Webinars & resources from US National Task Group: <http://aadmd.org/ntg>
- This is an Australian resource to facilitate conversations about end of life care for people with intellectual disability. <https://www.caresearch.com.au/TEL/>
- Jokinen, N., Janicki, M. P., Keller, S. M., McCallion, P., & Force, L. T. (2013). Guidelines for structuring community care and supports for people with intellectual disabilities affected by dementia. *Journal of policy and practice in intellectual disabilities*, 10(1), 1-24.
- Book: *Down's syndrome and dementia: a resource for carers and support staff*/ Dodd, Karen (2009) Website: 'Supporting Derek' (UK). Includes video and booklet resource for people working with people with intellectual disability and dementia. <https://www.jrf.org.uk/report/supporting-derek-new-resource-staff-working-people-who-have-learning-difficulty-and-dementia>

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# Key points

- ↑ Healthy lifestyle & engagement
- Health check-ups
- Establish baseline function & track changes
- Lengthy diagnosis time → investigate changes early
- Plan transitions early
- ↑ Carers' supports
- Medications may help
- Resources available