Dementia in Down syndrome

Julian Trollor & Liz Evans
3DN, UNSW
dddn@unsw.edu.au
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

<table>
<thead>
<tr>
<th>Area</th>
<th>Normal Ageing</th>
<th>Abnormal Ageing</th>
</tr>
</thead>
<tbody>
<tr>
<td>Forgetting</td>
<td>Occasional &amp; temporary</td>
<td>Whole experiences</td>
</tr>
<tr>
<td>Thinking</td>
<td>A bit slower</td>
<td>All affected</td>
</tr>
<tr>
<td>Decision making</td>
<td>OK</td>
<td>Major trouble</td>
</tr>
<tr>
<td>Knowing surroundings</td>
<td>OK</td>
<td>Difficult</td>
</tr>
<tr>
<td>Recognising loved ones</td>
<td>OK</td>
<td>Difficult</td>
</tr>
<tr>
<td>Mental health</td>
<td>OK</td>
<td>Hearing voices, seeing things</td>
</tr>
<tr>
<td>Skills lost</td>
<td>Due to health conditions</td>
<td>Unrelated to conditions</td>
</tr>
</tbody>
</table>
“Dementia” vs “Major Neurocognitive disorder”

- essentially the same thing.
What is Mild Cognitive Impairment (MCI)?

= “Mild Neurocognitive Disorder”

• Some declines in cognition
  - but may progress to become dementia

• Not dementia

• Many causes – some treatable
Types of dementia

Can be mixed

In Down syndrome:
- Alzheimer’s most common
- Others rarer
- “Dementia due to Down syndrome”

Brain changes in Alzheimer’s disease

- Build up of proteins:
  - **Amyloid** ‘plaques’
  - **Tau** ‘tangles’
- Shrinkage
- Inflammation

Why is dementia more common in people with Down syndrome? \(^{1,2,3}\)

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

How common is dementia in Down syndrome?

Research – variable rates: ¹, ², ³

<table>
<thead>
<tr>
<th>Age</th>
<th>Rate</th>
</tr>
</thead>
<tbody>
<tr>
<td>&lt; 40</td>
<td>&lt;10%</td>
</tr>
<tr>
<td>50’s</td>
<td>30%</td>
</tr>
<tr>
<td>60+</td>
<td>50%</td>
</tr>
<tr>
<td>&gt; 65</td>
<td>80%</td>
</tr>
</tbody>
</table>

Average age of diagnosis: mid 50’s. ³

What are the signs of dementia in someone with Down syndrome?
## Early signs of Alzheimer’s disease

**Problems with:**
- Memory
  - recent worst
- Visuo-spatial skills
- Attention
- Planning
- Problem solving
- Judgement.

**Personality changes:**
- Stubbornness
- Apathy
- Motivation
- ↓ social engagement

**Behaviour changes:**
- Irritability, aggression, self-injury

Course of dementia

Memory
- Recent
- Distant

Orientation
- ? Day or time
- ? Familiar places
- ↓ Response to environment
Course of dementia

Communication
- Expressive
- Repetitive
- Understanding

Functional skills
- Complex
- Self-care
- Incontinent

Coordination
- Dyspraxia
- Mobility

Support needs increase
Seizures:

Alzheimer’s disease: 10 – 22%. ¹

Down syndrome: ²
- 50+ years: 46%
- Alzheimer’s disease: 84%
- Childhood-onset = ↓ risk of dementia.

Preventing or Delaying Dementia
Preventing or delaying dementia - 1

Healthy lifestyle: $^{1,2,3}$
- Diet & exercise
- Address sleep apnoea

Stay active: $^2$
- Education
- Vocation
- Leisure
- Social

Preventing or delaying dementia - 2

Reduce inflammation:

- 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
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
3. Breaking the news

Supporting:

• the person with Down syndrome to understand

• Family & carers

• Others they love

Resources
4. Planning for changes

- 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 \(^1\)
• Ageing in place vs change residence

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 \(^{1,3,2}\)
  - results unclear
  - benefit some
  - side effects

Types:
- Memantine
- Cholinesterase inhibitors

7. Address related symptoms

• Behavioural:
  – Common in dementia
  – Management:
    » Environmental changes
    » Medications

• Seizures:
  – Medications
What to expect from others
Diagnosis – a lengthy process

'Suspected' → 'Confirmed'

12 – 24 months
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

Psychologist
Psychiatrist

Geriatrician
Neurologist

GP
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
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

Resources
For people with intellectual disability:


- *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/](https://www.stir.ac.uk/about/faculties-and-services/health-sciences-sport/research/research-groups/enhancing-self-care/jennys-diary/)


- *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](https://booksbeyondwords.co.uk/ebooks/ann-has-dementia?rq=dementia)
For families and carers - Books


• *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:
For families and carers – websites:

- The **carer checklist** recommended by the US National Task Group: [http://aadmd.org/ntg/screening](http://aadmd.org/ntg/screening)

- Websites with information and resources:
  - Karen Watchman’s website: [www.learningdisabilityanddementia.org](http://www.learningdisabilityanddementia.org)
  - The BILD (UK) website: [http://www.bild.org.uk/resources/ageingwell/dementia/](http://www.bild.org.uk/resources/ageingwell/dementia/)

- Dementia Australia has made a video about dementia in people with intellectual disability, available through the “resource finder” section of their website.

For health & disability professionals

- The **carer checklist** recommended by the US National Task Group is available from this site: [http://aadmd.org/ntg/screening](http://aadmd.org/ntg/screening).
  Other resources and webinars: [http://aadmd.org/ntg](http://aadmd.org/ntg)

- **3DN resources** @ [https://3dn.unsw.edu.au/content/health-mental-health-professionals](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](http://aadmd.org/ntg)


References


References


3dn.unsw.edu.au  @3DN_UNSW
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