Dementia in individuals with Intellectual disabilities - epidemiology

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Outline

• Background
• Dementia in individuals with Down syndrome
• Dementia in individuals with other intellectual disabilities
• Diagnostic criteria for dementia in ID
• Conclusions
Dementia in individuals with Down syndrome
PART 1.—ORIGINAL ARTICLES.


6. Many of the fifty-four are now dead. Phthisis caused a large majority of these deaths. In not a few instances, however, death was attributed to nothing more definite than general decay—a sort of precipitated senility.
Life expectancy of people with Down syndrome

Bittles et al, 2004
Aging DS population in UK

From: Wu and Morris, 2013
Ageing in adults with Down Syndrome

- Precocious ageing
- Hearing loss/ cataracts
- Depression
- Increased mortality
- Low rates of cardiovascular disease/ solid tumours
- High rates of Alzheimer’s disease
Age of dementia diagnosis in DS

Age at diagnosis (n= 339)

Median age = 55.47 years
IQR: 50.99 - 59.42 years
Age range 32.83 - 74.11 yrs

Median survival time (N = 278)
4.10 years (95% CI: 3.61 - 4.59; IQR: 2.34- 6.67)

(Data from the ADSID Collaboration)
Prevalence of dementia in DS  
- selected studies (%)  

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<tr>
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<tbody>
<tr>
<td>&lt; 40</td>
<td>0</td>
<td>3</td>
<td></td>
<td></td>
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<tr>
<td>40 - 49</td>
<td>16</td>
<td>10</td>
<td>6</td>
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<tr>
<td>50 – 59</td>
<td>38</td>
<td>40</td>
<td>30</td>
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<tr>
<td>50 – 54</td>
<td></td>
<td></td>
<td></td>
<td>18</td>
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<tr>
<td>55 - 59</td>
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<td></td>
<td>32</td>
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<tr>
<td>60 +</td>
<td></td>
<td></td>
<td>42</td>
<td>26</td>
</tr>
<tr>
<td>70 +</td>
<td></td>
<td></td>
<td>50</td>
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Incidence of dementia in DS

• Incidence increases steadily with increasing age
  – from 2.5 per 100 person years in those aged <50 to 13.31 per 100 person years in those aged 60 and older (Coppus et al., 2006).
Cumulative risk for dementia in DS

Figure 1  Cumulative risk of developing dementia by age.

McCarron et al, 2014
• But some seem to be protected despite having full trisomy 21 (Krinsky-McHale et al, 2008)
• AD is now the leading cause of death in DS (Englund et al, 2013; Zhu et al, 2012)
Dementia in other individuals with ID

• There are theoretical and medical reasons why dementia may be more common in ID
  – Cognitive reserve theory
  – Cardiovascular disorders
  – Genetic factors
• But may also be less common
  – Some risk factors less common, e.g. smoking
  – Healthy survivor effect
Comparison with population rates using Standardised Morbidity Ratios (SMR)

<table>
<thead>
<tr>
<th>Age band</th>
<th>Total N</th>
<th>Observed cases % (N)</th>
<th>Expected cases % (N)</th>
<th>SMR</th>
<th>Exact 95% Poisson CI</th>
</tr>
</thead>
<tbody>
<tr>
<td>60 plus</td>
<td>222</td>
<td>13.1% (29)</td>
<td>5.4% (11.9)</td>
<td>2.42</td>
<td>1.62 – 3.47</td>
</tr>
<tr>
<td>65 plus*</td>
<td>142</td>
<td>18.3% (26)</td>
<td>6.6% (9.4)</td>
<td>2.77</td>
<td>1.81 - 4.07</td>
</tr>
</tbody>
</table>

Reference prevalence rates from:

• Northern European Consensus rates (Ferri et al, 2005)  
  - for age 60 plus
• MRC CFA Study (Brayne et al 1998)  
  - for age 65 plus

Strydom et al, 2007
Dementia by severity of ID

• Prevalence rates do not appear to differ significantly between those with mild, moderate and severe ID
• But it is more difficult to diagnose dementia in those with more severe ID
Incidence of dementia in older adults with ID

- Overall, the incidence rate for dementia in those aged 60 and older is estimated to be 54.6/1000 person years.
- Highest incidence rate in the age group 70–74
  – (Strydom et al, 2013).
Comparison of dementia prevalence rates by age
Dementia criteria
<table>
<thead>
<tr>
<th>Impaired domain</th>
<th>DSM-IV</th>
<th>ICD-10</th>
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<tbody>
<tr>
<td><strong>Memory</strong></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Short and/or long term</td>
<td></td>
<td></td>
</tr>
<tr>
<td><strong>Decline in higher cortical functions</strong></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Executive function</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Thinking</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Judgement</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Information processing</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Aphasia/language skills</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Apraxia</td>
<td></td>
<td></td>
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<tr>
<td>Agnosia</td>
<td></td>
<td></td>
</tr>
<tr>
<td><strong>Change in behavioural and emotional function</strong></td>
<td></td>
<td></td>
</tr>
<tr>
<td><strong>Change from previous/decline in function</strong></td>
<td></td>
<td></td>
</tr>
<tr>
<td><strong>Duration of 6 months</strong></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Not caused by delirium</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Not caused by mental illness</td>
<td></td>
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**Key**

- Yellow: **OR**
- Red: **AND**
Dementia criteria in the general population

- **DSM-IV** is much more inclusive than ICD-10 *(DSM-IV dementia = 9.6% vs. ICD10 dementia = 3.1%)* (Erkinjuntti et al, 1997; Wancata et al, 2007)

- **ICD-10** less inclusive due to
  - 6 month criterion (Stevens et al, 2002)
  - Depends on reliable informants (Henderson et al, 1994; Riedel-Heller et al 2001)
  - Has more criteria, i.e. change in emotional control and behaviour (Erkinjuntti et al, 1997; Wancata et al 2007)


Dementia cases by diagnostic criteria in ID (excluding DS)

- DSM-IV prevalence: 12.2%
- DC-LD prevalence: 10.4%
- ICD-10 prevalence (aged 60 plus): 9.9%
Inter-rater reliability of dementia criteria in DS

(n=21)

<table>
<thead>
<tr>
<th></th>
<th>Kappa</th>
<th>Significance</th>
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<tbody>
<tr>
<td>ICD-10</td>
<td>0.901</td>
<td>&lt;0.001</td>
</tr>
<tr>
<td>DSM-IV</td>
<td>0.786</td>
<td>&lt;0.001</td>
</tr>
<tr>
<td>Clinical impression</td>
<td>0.809</td>
<td>&lt;0.001</td>
</tr>
</tbody>
</table>
Dementia diagnosis in DS

Clinical diagnoses vs. DSM/ICD over time

Sheehan et al, 2014
Conclusions

• Dementia is common in DS – 80% at risk
  – but not invariable
• Dementia also more common in other older service users with ID
• Prevalence rates influenced by
  – mortality,
  – diagnostic issues,
  – and diagnostic criteria used
Thank you
Any comments or questions?

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