Down’s Syndrome and Dementia

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Figure Prevalence of Dementia in Down Syndrome

Haveman et al. + Schupf et al. + Lai & Williams ▼ Zigman et al.
Possible reasons for the discrepancy

- 100% NP
- Qualitative difference in NP
- Threshold of NP
- Cortical distribution
- Neurochemistry
- Tangles and plaques are part of a cascade
- Age of onset issue
- Different risk factors
- Difficulty in diagnosis
- Sampling bias
- Bias in data collection
- Different instruments used
- Different diagnoses
Dementia in Down syndrome (ICD-10)

- Impaired memory.
- Impairment of other cognitive functions.
- Unimpaired consciousness.
- Reduced emotional control or motivation or change in social behaviour.
- Six-month duration.
- Exclusion of other diagnoses
Dementia in Down syndrome
(Differential diagnosis)

• Hypothyroidism.
• Depression.
• Sensory impairment.
• Age-related decline.
• Environmental factors.

• Non-AD dementia.
• Other psychiatric/medical conditions.
• Epilepsy.
• Drugs.
Dementia in Down syndrome

- Patient interview.
- Patient examination.
- Information from carers.
- Investigations.
Dementia in Down syndrome (Scales)

- Neuropsychological tests.
- Observer-rated scales.
- ADL scales.
Dementia in Down syndrome
(Neuropsychological tests)

• Modified MMSE
• One-word Picture Vocabulary
• Modified Verbal Fluency
• Modified Buschke (Selective Reminding Test)
• Delayed Match-to-Sample
• Modified Purdue Pegboard
• Block design/picture completion
• Brief Praxis Test
• Spatial Recognition Span/copy simple drawing
Deb S. & Braganza J.  
(JIDR, 1999, 43, 5, 400-407)

- 26 = dementia and DS
- 36 = DS without dementia

- Clinicians’ diagnosis
- DSDS
- DMR
- MMSE
Deb & Braganza 1999

- MMSE 34/ 62 (55%)
- MMSE score (<24) 30/ 34 (95.2%)
- No dementia 23/ 30 (77%)
Dementia in Down syndrome (Observer-rated scale)

- DMR
- DSDS
- MOSES
- Early Signs of Dementia Checklist
- Test for Severe Impairment (TSI)
- Adaptive Behaviour Scale
- Maladaptive behaviour scale
DSQIID


S Deb
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DSQIID (Deb et al, 2007a, b)

- Qualitative study (24 carers of DS + AD adults)
- DS+AD=49 & Non-AD DS=68 from 28 centres in the UK
- 53 items Observer rated scale (4 points)
- (>=20) Sensitivity=0.92; Specificity=0.97
- Internal Consistency: Cronbach’s Alpha 0.91
- Reliability: Test retest (ICC=0.95); Inter-rater (ICC=0.9)
- Factor analysis
  - Memory/ confusion
  - Feelings of insecurity
  - Sleep problems
  - Behaviour problems
<table>
<thead>
<tr>
<th></th>
<th>Always been</th>
<th>Always, but worse</th>
<th>New symptom</th>
<th>Does not apply</th>
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<tbody>
<tr>
<td>1) Does not initiate conversation</td>
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<td>2) Cannot find words</td>
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<td>3) Cannot follow simple instructions</td>
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<td>4) Cannot follow more than one instruction at a time</td>
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<td>5) Stops in the middle of a task</td>
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<td>6) Cannot read</td>
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<td>7) Cannot write (including printing own name)</td>
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</tbody>
</table>
Dementia in Down syndrome
(Other scales)

- Adaptive Behaviour Dementia Questionnaire (ABDR; Prasher et al, 2004; Birmingham)
- Prudhoe Cognitive Function Test (PCFT; Margallo-Lana, 2005; Newcastle)
- CAMDEX-DS (Holland & Ball, 2006; Cambridge)
- Screening Checklist for Referral (Whitwham & McBrien; Plymouth)
- Caregiver Activity Survey-Intellectual Disability (CAS-ID; McCarron, Gill, Lawlor, Begley, 2002; Dublin)
- Assessment for Adults with Developmental Disabilities (AADS, Kalsy, Oliver et al; Birmingham)
- PIMRA, Reiss Screen, DASH-II, PAS-ADD, Depression Scales
Domains of changes

- Forgetfulness (STM problem)
- Confusion
- Instruction
- Slowness
- Speech problems
- Loss of skills
- Socialization problem
- Lack of confidence
- Lack of initiative

- Obsession
- Sleep problem
- Balance problem
- Emotional problems
- Sensory problems
- Loss of interest
- Covering up for the memory loss
- Hypochondriasis
- Change in personality

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