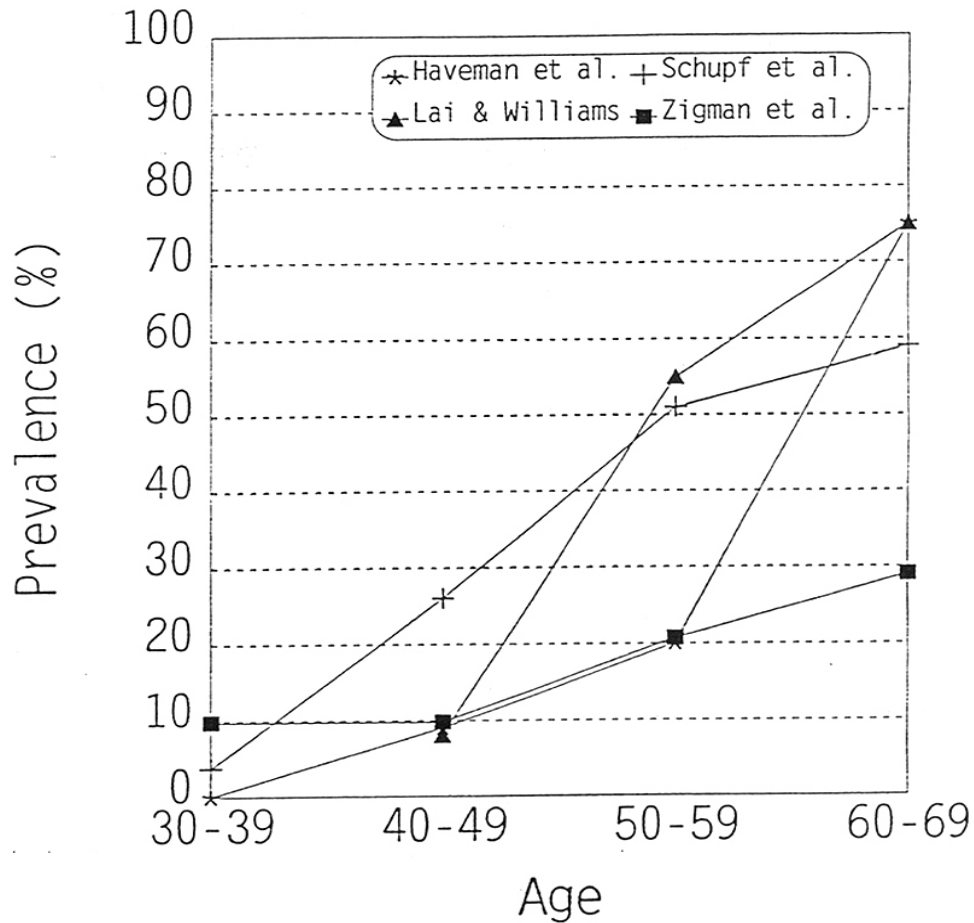


Down's Syndrome and Dementia

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Figure Prevalence of Dementia in Down Syndrome



EPIDEMIOLOGY OF ALZHEIMER DISEASE IN MENTAL RETARDATION - RESULTS AND RECOMMENDATIONS FROM AN INTERNATIONAL CONFERENCE (REPORT OF THE AAMR/ASSID WORKGROUP ON EPIDEMIOLOGY AND ALZHEIMER DISEASE)

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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 Purdoe 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

Deb S., Hare M., Prior L. & Bhaumik S. (2007) Dementia Screening Questionnaire for Individuals with Intellectual Disabilities (DSQIID). *British Journal of Psychiatry*, **190**, 440-444.

Deb S., Hare M. & Prior L. (2007) Symptoms of dementia among adults with Down's syndrome: a qualitative study. *Journal of Intellectual Disability Research*, **51**, 9, 726-739.

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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

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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