

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

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PART I.—ORIGINAL ARTICLES.

Kalmuc Idiocy: Report of a Case with Autopsy. By JOHN FRASER, M.B. *With Notes on Sixty-two Cases.* By Dr. ARTHUR MITCHELL, Commissioner in Lunacy.

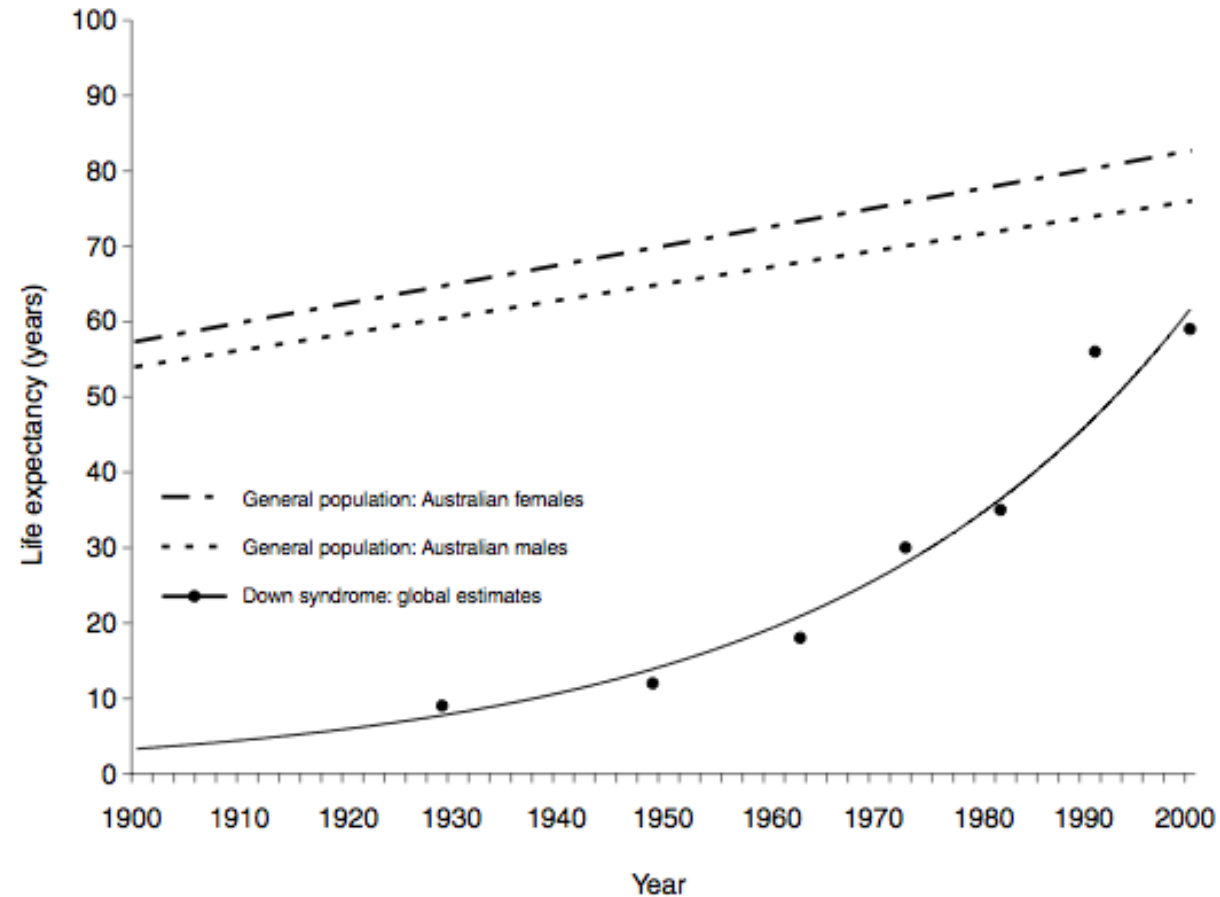


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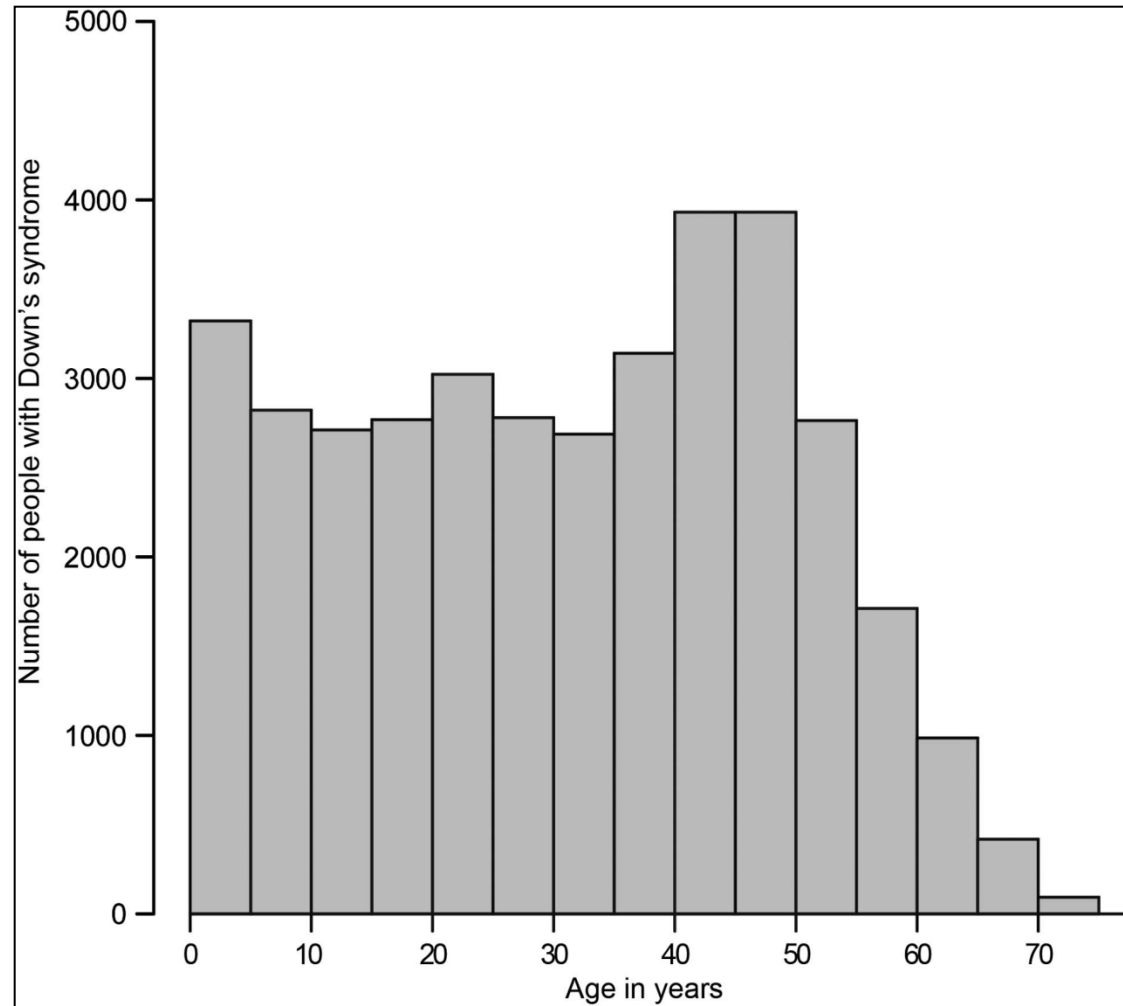
6. Many of the fifty-four are now dead. Phthisis caused a large majority of these deaths. In not a few instances, however, death was attributed to nothing more definite than general decay—a sort of precipitated senility.

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

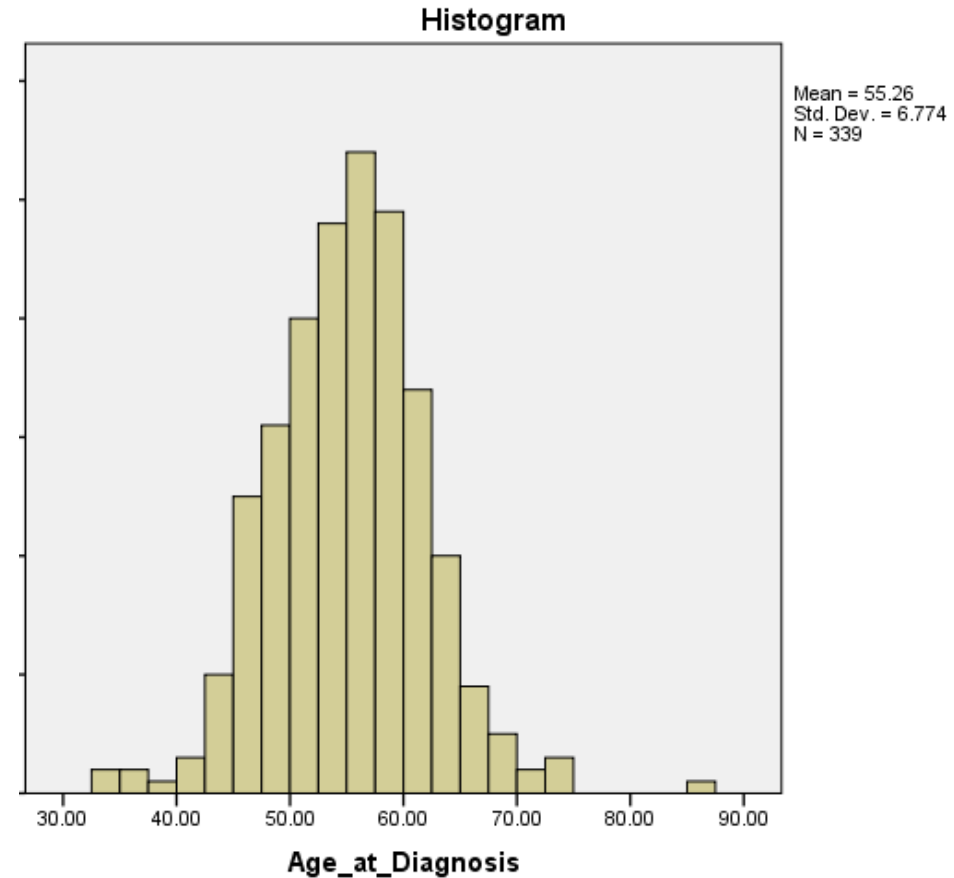
IQR: 50.99 - 59.42 years

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

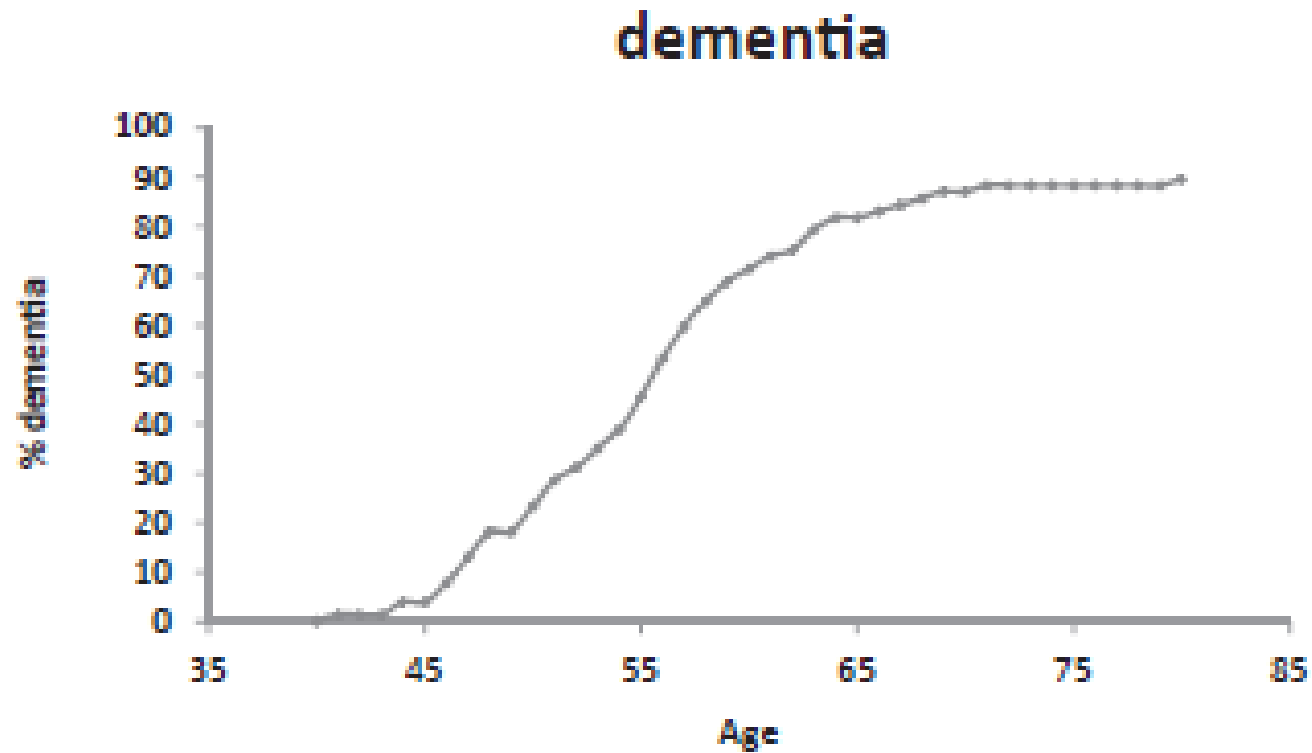
- selected studies (%)

Age range	Sikijima, 1998 Japan N = 106	Holland, 2000 UK N = 75	Tyrrell, 2001 Ireland N= 285	Coppus, 2006 NL N = 506
< 40	0	3		
40 - 49	16	10	6	
50 – 59	38	40	30	
50 – 54				18
55 - 59				32
60 +			42	26
70 +			50	

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).

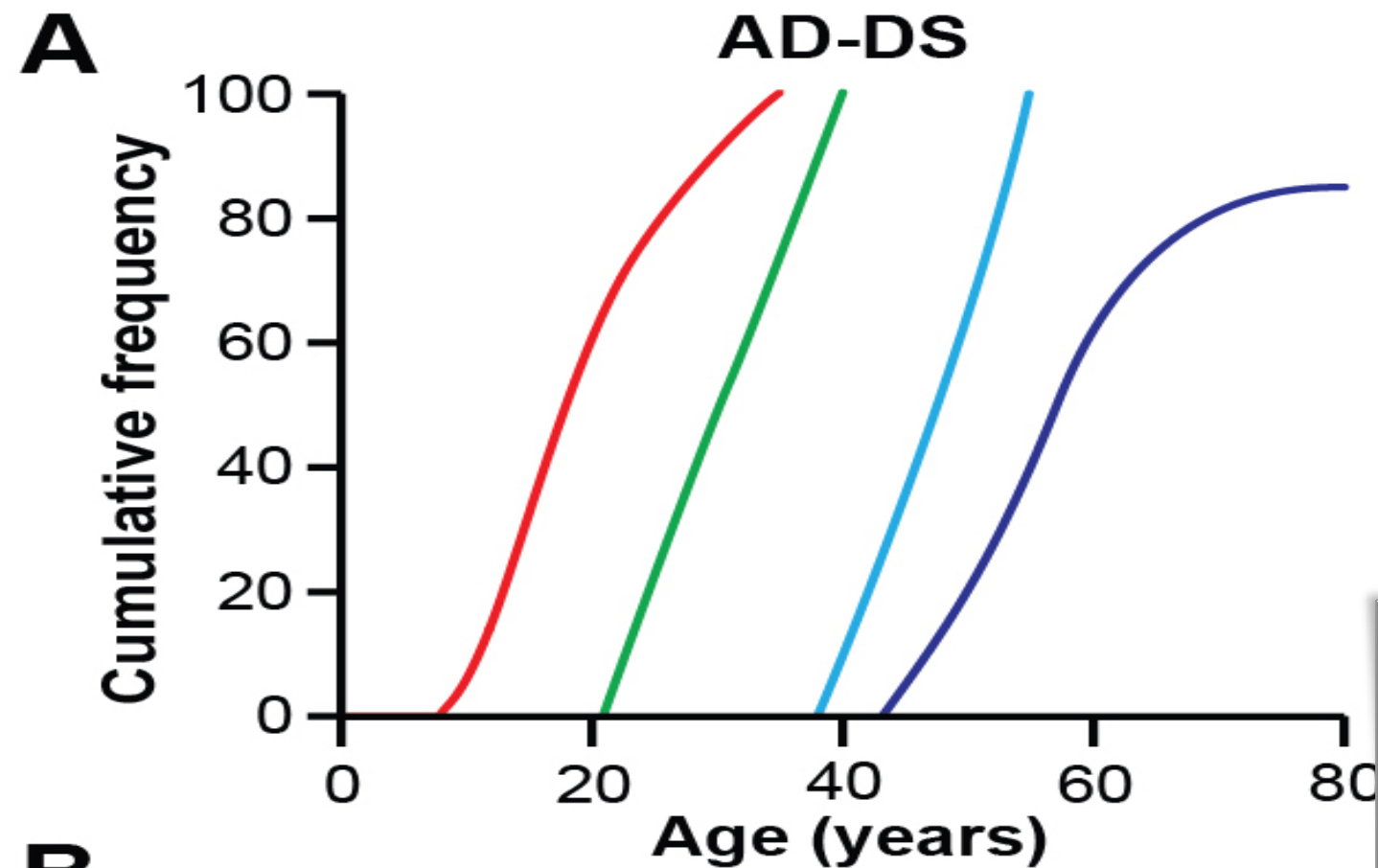
Cumulative risk for dementia in DS



McCarron et al, 2014

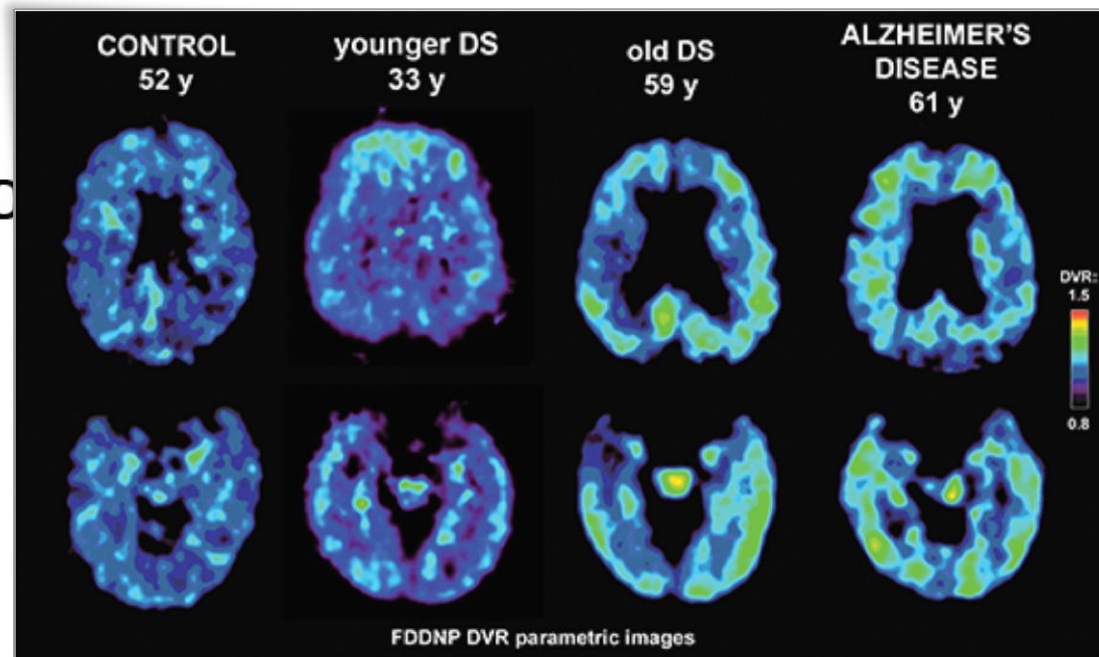
Figure 1 Cumulative risk of developing dementia by age.

- 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)



- Amyloid
- PiB positive
- NFT
- Dementia

B



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)

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

Reference prevalence rates from:

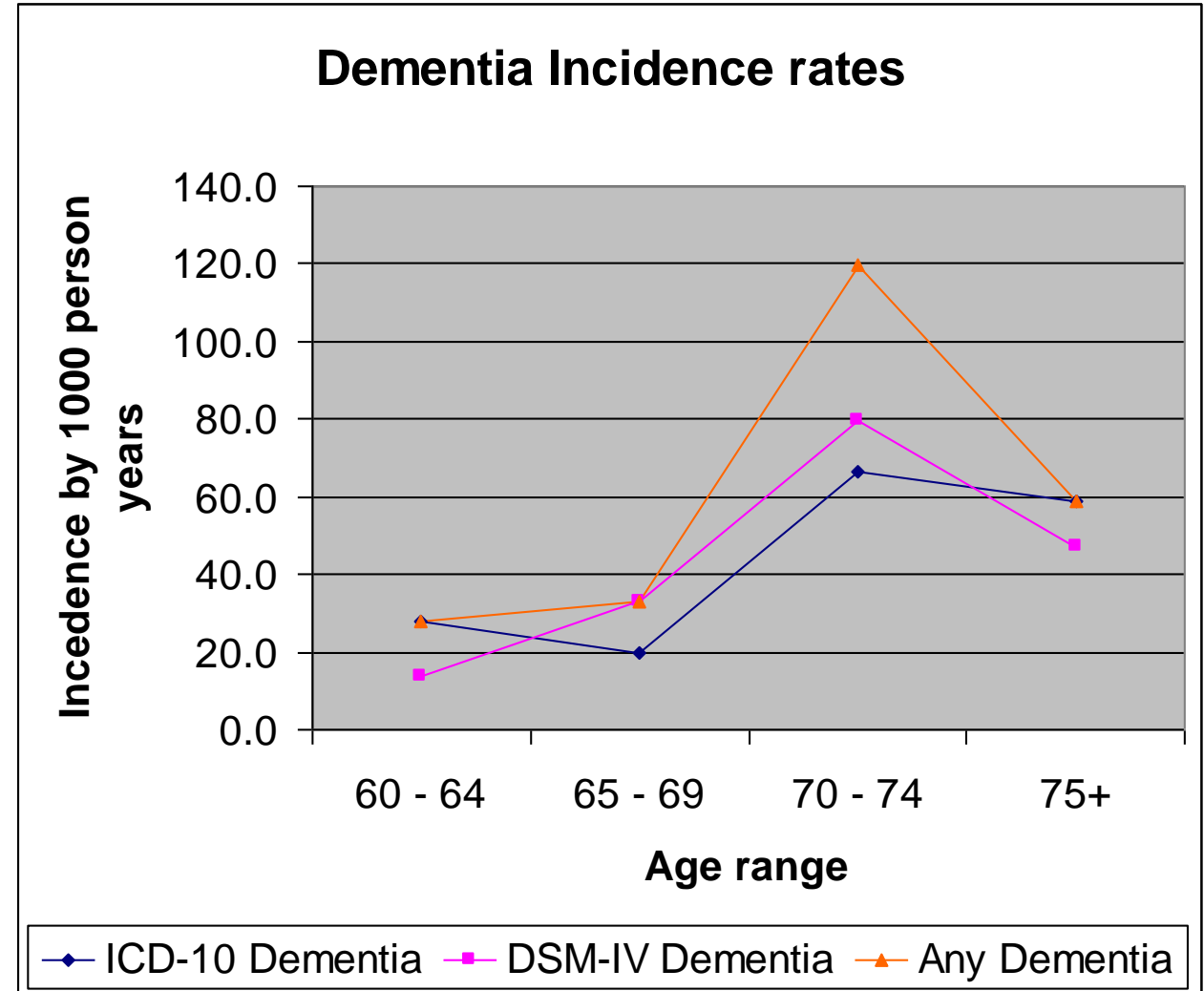
- Northern European Consensus rates (Ferri et al, 2005)
 - for age 60 plus
- MRC CFA Study (Brayne et al 1998)
 - for age 65 plus

Dementia by severity of ID

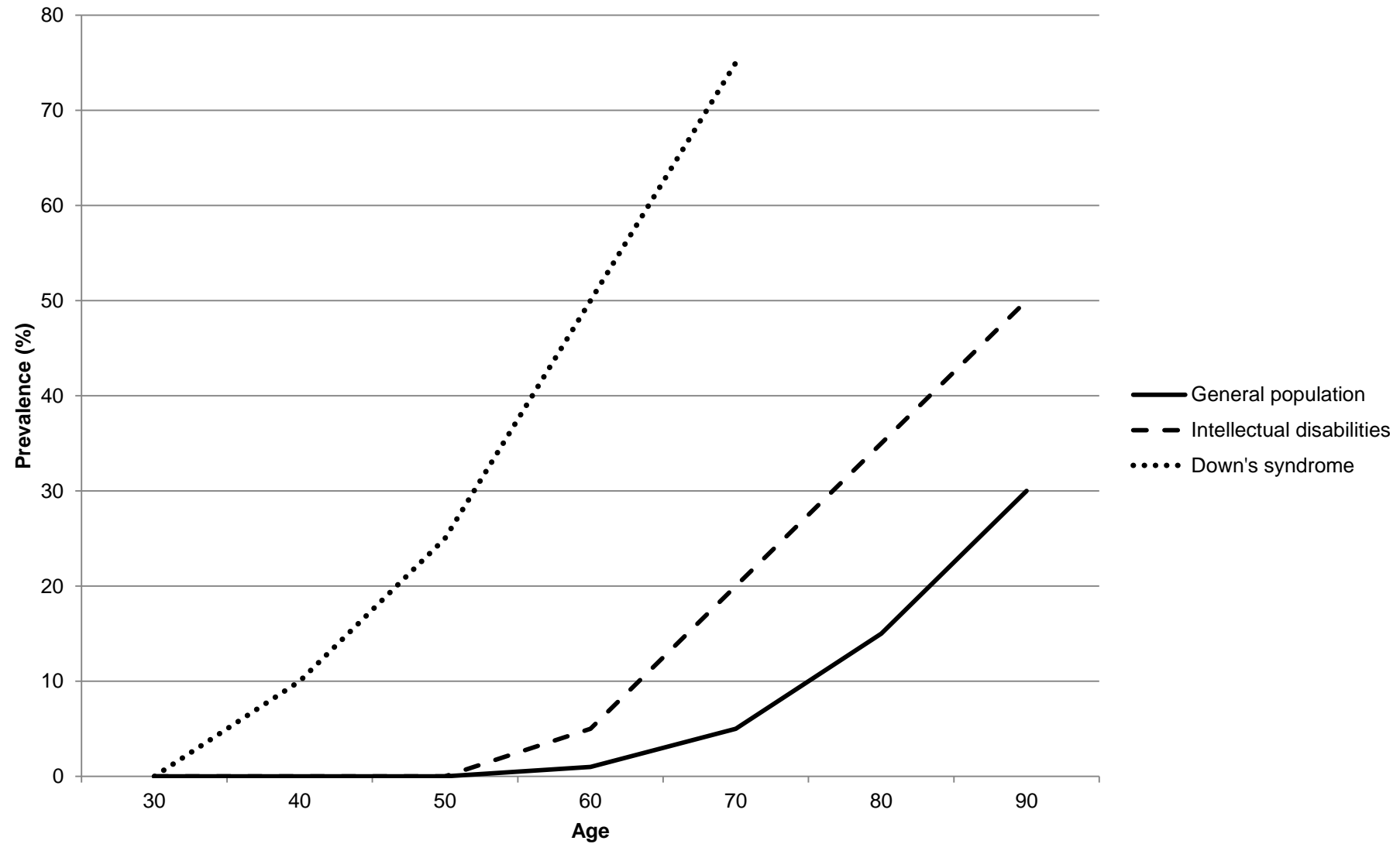
- Prevalence rates do not appear to differ significantly between those with mild, moderate and severe ID
- But it is more difficult to diagnose dementia in those with more severe ID

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

Impaired domain	DSM-IV	ICD-10
Memory		
Short and/ or long term		
Decline in higher cortical functions		
Executive function		
Thinking		
Judgement		
Information processing		
Aphasia/ language skills		
Apraxia		
Agnosia		
Change in behavioural and emotional function		
Change from previous/ decline in function		
Duration of 6 months		
Not caused by delirium		
Not caused by mental illness		

Key	
	OR
	AND

Dementia criteria in the general population

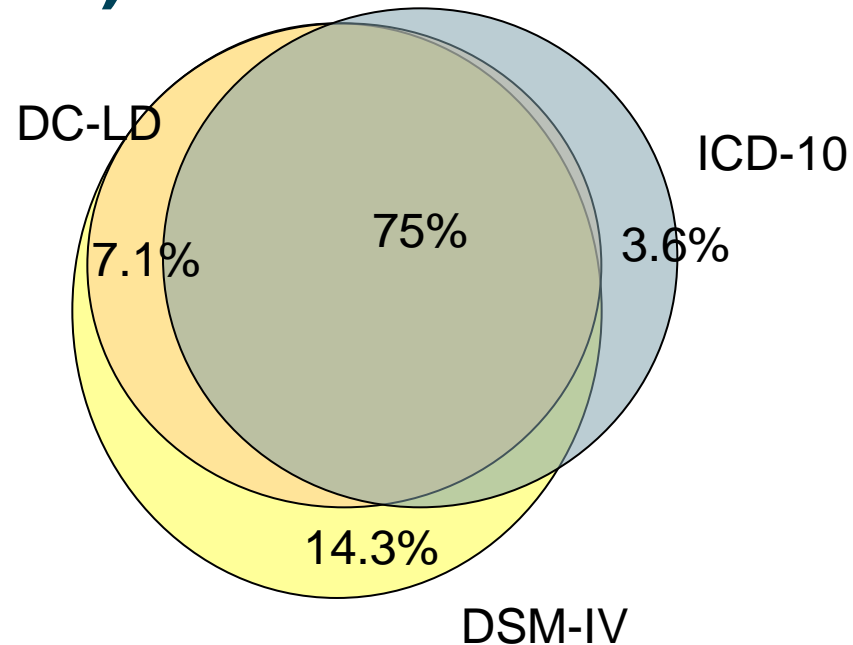
- DSM-IV is much more inclusive than ICD-10 (*DSM-IV* dementia = 9.6% vs. ICD10 dementia = 3.1%) (Erkinjuntti et al, 1997; Wancata et al, 2007)
- ICD-10 less inclusive due to
 - 6 month criterion (Stevens et al, 2002)
 - Depends on reliable informants (Henderson et al, 1994; Riedel-Heller et al 2001)
 - Has more criteria, i.e. change in emotional control and behaviour (Erkinjuntti et al, 1997; Wancata et al 2007)

Stevens T, Livingston G, Kitchen G, et al: Islington study of dementia subtypes in the community. *Br J Psychiatry* 2002; 180:270–276

Wancata J, Borjesson-Hanson A, Ostling S, et al: Diagnostic criteria influence dementia prevalence. *Am J Geriatr Psychiatry* 2007; 15:1034–1045

Erkinjuntti T, Ostbye T, Steenhuis R, et al: The effect of different diagnostic criteria on the prevalence of dementia. *N Engl J Med* 1997; 337:1667–1674

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%

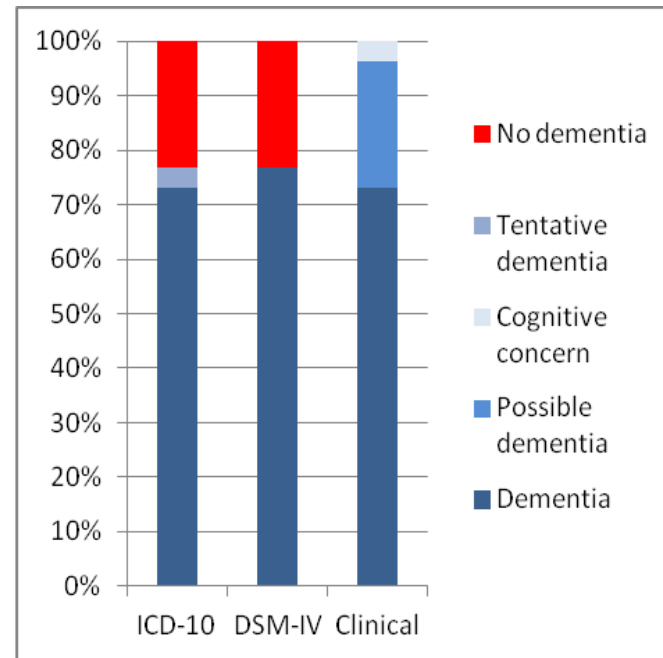
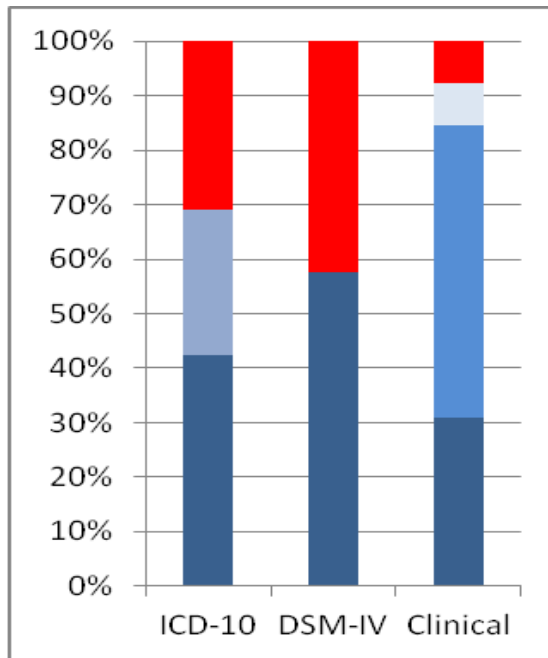
Inter-rater reliability of dementia criteria in DS

(n=21)

	Kappa	Significance
ICD-10	0.901	<0.001
DSM-IV	0.786	<0.001
Clinical impression	0.809	<0.001

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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