

## Mental health

# A prospective 14-year longitudinal follow-up of dementia in persons with Down syndrome

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## Abstract

**Background** To examine dementia characteristics, age at onset and associated comorbidities in persons with Down syndrome.

**Method** Seventy-seven people with Down syndrome aged 35 years and older were followed longitudinally. The diagnosis of dementia was established using the modified International Classification of Diseases, Tenth Revision (ICD-10) criteria and a combination of objective and informant-based tests. Cognitive tests included the Test for Severe Impairment and the Down Syndrome Mental Status Examination; adaptive behaviour was measured using the Daily Living Skills Questionnaire. The Dementia Questionnaire for Mental Retarded Persons (DMR) was added to the test battery in 2005 and this study includes follow-up data for this instrument.

**Results** Over the 14-year period the average age of diagnosis at 55.41 years (SD = 7.14) was in the higher range of previously reported estimates (51–56 years) and a median survival of 7 years after

diagnosis. Persons with dementia in the sample were significantly older than persons without dementia.

The presence of dementia was also associated with epilepsy and sensory impairments. Among instruments the DMR appeared most sensitive to tracking change in symptoms over time before diagnosis.

**Conclusion** The previously reported high risk levels for dementia among people with Down syndrome was confirmed in these data as was the value of the instruments utilised in tracking decline and helping to confirm diagnosis even in persons with severe intellectual disability.

**Keywords** behavioural measurement methods, Down syndrome, intellectual disability, learning disability

## Background

Life expectancy in people with Down syndrome (DS) has increased dramatically in recent years from an average of 9 years in 1930s to age 60 years and beyond today (Bittles & Glasson 2004). There has been signification interest in understanding the ageing of this population and much research has focused on change in behaviours and global day to

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day functioning with increasing age (McCarron *et al.* 2011). It is generally agreed that dementia occurs more commonly in people with intellectual disability (ID) and particularly those with DS with prevalence rates estimated to be between 15% and 45% in those with DS over the age of 45 years (Prasher & Krishnan 1993). There is consensus that average age of onset among people with DS is between 51 and 56 years and duration is reported as averaging between 3.5 and 6 years, with individual cases as high as 21 years (Lai & Williams 1989; Prasher & Krishnan 1993).

There is also emerging consensus on prevalence rates of dementia in persons with DS by age cohort. Strydom *et al.* (2010) in a review of published studies (1997–2008) confirms both a higher prevalence of dementia in persons with DS, and accelerating rates with age with reports of 9% in those under 49 years (Coppus *et al.* 2006), 5.7–10.3% in ages 40–49 years (Holland *et al.* 1998; Tyrrell *et al.* 2001), 30.4–40% in ages 50–59 years (Holland *et al.* 1998; Tyrrell *et al.* 2001; Coppus *et al.* 2006) and 41.7–50% in those aged 60–70 years (Tyrrell *et al.* 2001) with one report of 100% prevalence rate in adults aged 65 years and over (Visser *et al.* 1997). Some of the variation noted reflects that there are many challenges in diagnosing and estimating the prevalence of dementia in this population including pre-morbid functioning and difficulties in using standardised tests, communication difficulties, improvised care environments, lack of base line performance data and the consequences of high staff turnover (McCallion & McCarron 2004).

The International Association for the Scientific Study of Intellectual Disabilities (IASSID) outline a model of best practice for dementia recognition and diagnosis in persons with ID (Burt & Aylward 2000); including (1) baseline assessment and annual follow-up of persons with DS > 35 years; (2) comprehensive diagnostic work-up; and (3) person centred approaches to care, including the need for staff training and service and policy re-design. However these recommendations are not yet embedded in health policy and it continues to be more likely that assessments are carried out in a crisis, with dementia often already at an advanced stage when best practice recommendations are for early testing and longitudinal follow-up to confirm diagnosis (Burt & Aylward 2000; Strydom *et al.*

2007; Coppus *et al.* 2008). Furthermore, a number of the published studies have methodological limitations including a reliance on cross-sectional designs (Zigman *et al.* 2002) with the few longitudinal studies usually of relatively short duration and with few points of data collection (see, for example, Holland *et al.* 2000; Margallo-Lana *et al.* 2007; McKensie *et al.* 1998) and there are also findings that longitudinal follow-up is not useful in people with severe ID (Margallo-Lana *et al.* 2007).

In the general population utilisation of memory clinics is viewed as an effective means of ensuring prompt and accurate diagnosis of dementia, as well as offering supports and appropriate referral for care and treatment (Gardner *et al.* 2004). Referral of people with ID to generic memory clinics appears to be fraught with difficulties, including lack of understanding of assessment procedures and of the recommended assessment tools for persons with ID among generic professionals, communication barriers and lack of experience in interviewing people with ID, interpreting decline in the context of pre-existing impairment, as well as difficulty in access and eligibility for services which are based on chronological age versus need (McCarron & Lawlor 2003). In response to these concerns some attempts have been made to develop memory clinics within ID services (McCreary *et al.* 1993; Chicoine *et al.* 1999; Hassiotis *et al.* 2003; Cahill *et al.* 2011). The present analysis results from the development of such a memory clinic within the Daughters of Charity Service in Ireland which primarily serves women with DS. This clinic provides a service to 200 people with DS over the age of 40 years. The data reported here examined changes from pre-morbid level of functioning over a 14-year period and assessed the usefulness of specific assessment instruments and the relationship between age and cognitive and functional decline, comorbidity and mortality risk in a convenience sample of 77 persons with DS who were aged 35 years and older at outset of the study.

## Methods

This is a prospective longitudinal study on symptom progression, dementia incidence, comorbidities and mortality in persons with DS reporting on a 14-year follow-up of 77 subjects.

### Sample

Seventy-seven women with DS, over the age of 35 years, that is, all women over 35 with DS at the Daughters of Charity Service were enrolled and screened in 1996 and then assessed for symptoms of dementia on an annual basis until death, with assessments completed in a specialist memory clinic. DS was established from records review. Following recommendations of Alyward *et al.* (1997), dementia diagnosis was confirmed using International Classification of Diseases, Tenth Revision (ICD-10) criteria. Institutional ethical approval for the study was received.

### Assessment protocol

At the time of entry into the study, each person received a complete comprehensive assessment including interviews with main caregivers/family; a detailed clinical screen and a medical records review to establish past and present physical and/or mental health disorders including cardiovascular health, lung disease, diabetes, epilepsy, depression, sensory impairments and gastric disease; and measures of cognitive and physical functioning. Chronic conditions were established in a records review and confirmed by a physician. Diagnosis of depression was by a psychiatrist familiar with the individual and who had the opportunity to interview the individual and key informants and to review all records. Pre-morbid level of ID was also obtained from the medical records/psychological reports, classified using the ICD-10 criteria (World Health Organization 1992). The comprehensive clinical screen and cognitive and physical functioning was then repeated annually during the years the subject was still alive.

### Cognitive and physical functioning measures

A combination of informant-based (Daily Living Skills Questionnaire – DLSQ – National Institute of Aging 1989) and objective-based (Down's Syndrome Mental Status Examination – DSMSE – Haxby 1989; Test for Severe Impairment – TSI – Albert & Cohen 1992) test instruments were administered. The utility of these instruments in measuring cognitive and functional decline in persons with DS have previously been reported

(Tyrrell *et al.* 1996, 2001; Cosgrave *et al.* 1998; McCarron *et al.* 2002). An additional informant-based questionnaire, the Dementia Questionnaire for Mentally Retarded individuals – DMR (Evenhuis 1990), was introduced in 2005 and then repeated each year.

### Diagnosis

A comprehensive diagnostic workup including a full physical examination, urinalysis, geriatric blood screen and mental health assessment, in line with recommendations from Alyward *et al.* (1997), helped in ruling out all other potential causes of decline. Only then was dementia diagnosis ascertained using the modified ICD criteria and after all materials were reviewed in a consensus meeting with members of the person's multi-disciplinary team including the clinical nurse specialist in dementia, caregivers/family, consultant psychiatrist and psychologist. Consistency in both study team and assessment protocol was purposefully maintained over the assessment period to overcome limitations reported in prior studies.

### Data analysis

After demographics of the sample were examined and the incidence of dementia over time plotted, the sample was divided into those with and those as yet without dementia and the two groups compared using *t*-tests for the continuous variables and Chi-square analysis for the categorical variables. In particular, patterns in comorbid health conditions were examined and compared for those with and without dementia diagnosis. Then, hierarchical linear modelling – HLM (Bryk & Raudenbush 1987; Bryk & Raudenbush 2001) was used to examine the personal characteristics that predicted change in the individual over the study period. Consistent with HLM requirements a baseline was established that described the status of the study cohort at time one as was a slope that described changes in functioning across subsequent time periods with the ability to model individual differences in trajectories of change. For the analysis here, to the extent possible, data points were graphed before dementia diagnosis for four measures (TSI, DLSQ, DSMSE and DMR) and for time points after diagnosis for

subjects for whom there occurred a confirmed dementia diagnosis. Similar time points were also graphed for persons who did not present with dementia over the study period.

Hierarchical linear modelling was particularly useful as an analysis approach as it does not require that all individuals have the same number of data points; an important consideration for this study given that some individuals have fewer data points than others given attrition over time through death ( $n = 44$ ) and that some subjects were not assessed at the same data points. A survival curve was used to plot the survival proportion as a function of time using the Kaplan–Meier method with a 95% confidence interval. Finally, logistic regression analysis was used to consider the respective contributions of age, presence of dementia and presence of epilepsy to observed mortality among the cohort.

## Results

### Incidence and risk of dementia

Over the 14-year follow-up period, 69 (89.6%) subjects developed dementia. Some individuals were found at baseline to have a diagnosis of dementia and others were followed for up to 14 years. The mean age of dementia diagnosis was 55.41 years ( $SD = 7.14$ ). Of those classified as having moderate ID 88.52% (54/61) had developed dementia by the end of the study period as did 93.3% (14/15) of those classified as having severe ID ( $n = 15$ ). There were no individuals in the sample with mild ID. Of those who developed dementia none had confirmed symptoms before the age of 40 years but based upon verified age of incidence data, 26.09% had developed dementia by age 50 years, 50.72% by age 55 years, 79.71% by age 60 years and 95.65% by age 68 years.

Based upon these incidence findings, the risk for developing dementia was also mapped by age. As can be seen in Fig. 1 a 20% risk was established at age 50 years; 45% at age 55 years and 80% risk at age 65 years.

### Comorbidities

The clinical characteristics and health comorbidities in persons with and without dementia are presented in Fig. 2.

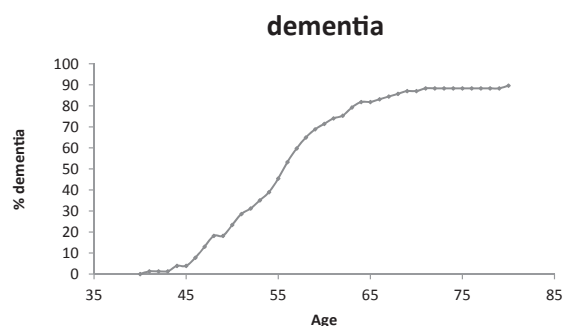


Figure 1 Cumulative risk of developing dementia by age.

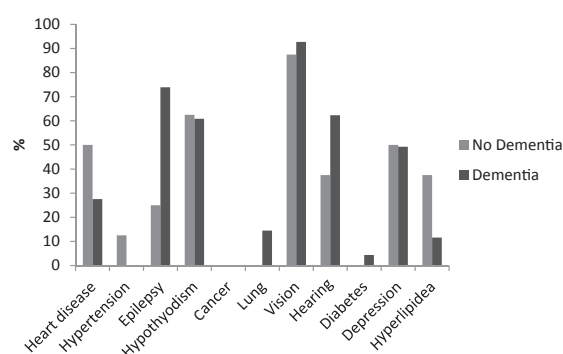
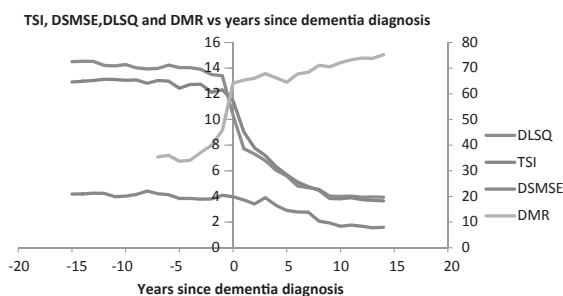


Figure 2 Prevalence of comorbidities.

Among comorbidities, epilepsy was significantly more common in subjects with dementia compared with subjects without dementia. Of those with dementia diagnosis, 73.9% (51/69) had epilepsy, compared with 25% (2/8) in those without a dementia diagnosis (Chi-square = 7.995;  $df = 1$ ;  $P = 0.0075$ ). Mean age at which people were diagnosed with epilepsy was 55.396 ( $SD = 9.92$ ) and diagnosis of epilepsy was on average 0.59 ( $SD = 6.18$ ) years after a diagnosis of dementia.

Depression was not significantly more common in subjects with dementia (Chi-square = 0.002;  $df = 1$ ;  $P = 0.969$ ). By 2011, among those with a dementia diagnosis 49.3% (34/69) were also diagnosed with depression, compared with 50% (4/8) for those without a dementia diagnosis. Vision impairments were reported in 87.5% of subjects without dementia and in 92.8% of subjects with dementia. Hearing impairments were reported in 37.5% of subjects without dementia and in 62.3% of subjects with dementia.



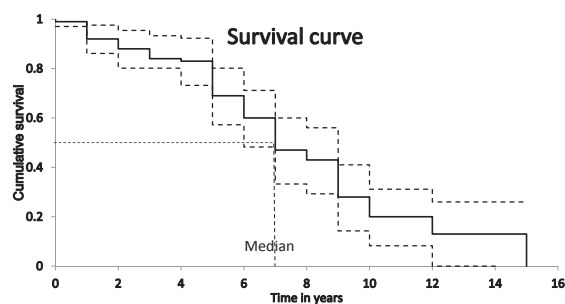
**Figure 3** Dementia scale scores by year before and after diagnosis. TSI, Test for Severe Impairment; DSMSE, Down's Syndrome Mental Status Examination; DLSQ, Daily Living Skills Questionnaire; DMR, Dementia Questionnaire for Mentally Retarded individuals.

### Cognitive and physical functioning measures

As can be seen in both Fig. 3 and Table 1, decline on the TSI and DSME was found to occur 1 year prior to diagnosis and the rate of decline on these scales after diagnosis was more gradual. The decline in DLSQ score was over 3–4 years and decline prior to diagnosis on the DMR (measured in increased scores), despite the shorter period of administration (only introduced in 2005), was pronounced and occurring over a period of 5 years. Rates of decline in scores in the 5 years before and after diagnosis for the respective measures in Table 1 are also broken out by moderate versus severe and profound levels of ID. There was no significant age difference in the moderate and severe ID groups.

### Mortality

Fifty-eight per cent ( $n = 45$ ) of the study cohort had died by 2011. Of these all but one died with a confirmed diagnosis of dementia. The last individual remained dementia symptom free and died age 52 years with a diagnosis of cancer. Of those who had developed dementia 63.8% (44/69) were dead by 2001 and duration of their dementia was 6.05 years ( $SD = 3.23$ ); if alive, as of 2011, it was an average of 6.24 years ( $SD = 4.1$ ). A significance difference ( $t = 2.181$   $P = 0.04$ ) was also noticeable in the duration of dementia in persons with moderate ID (mean = 5.32,  $SD = 4.11$ ) versus subjects with severe ID (mean = 9.17,  $SD = 2.14$ ) but not in the mean age of diagnosis of dementia, 55.56 ( $SD = 6.12$ ) for those with moderate ID and 53.71



**Figure 4** Dementia survival curve.

( $SD = 9.60$ ) for those with severe ID ( $t = 0.884$ ,  $df = 66$ ,  $P = 0.38$ ).

Figure 4 presents the survival curve for the 69 subjects with dementia using the Kaplan–Meier method with a 95% confidence interval. The survival curve plots the survival proportion as a function of time. Time zero represents the year a subject was diagnosed with dementia. The median survival time was 7 years, the time at which half the subjects have died and half are still alive.

Logistic regression examined the ability of the variables age, epilepsy and level of ID to predict onset of dementia. Epilepsy was found to be the only significant predictor of dementia ( $P$ -value = 0.023). Subjects with epilepsy were 7.24 (1.31, 40.0) times more likely to present with dementia than those without diagnosed epilepsy.

Further use of logistic regression examined age, epilepsy and dementia as predictors of death among the sample. Here, epilepsy was found to be the only significant predictor of mortality ( $P$ -value = 0.002). A subject with epilepsy was 6.39 (2.02, 20.19) times more likely to have died than a subject without diagnosed epilepsy.

### Discussion

The memory clinic approach recommended as best practice (Burt & Aylward 2000) and utilised here proved capable of tracking change suggestive of dementia and potentially supported earlier diagnosis. Also the risk for dementia established, 20% risk at age 50 years; 40% at age 55 years and 80% risk at age 65 years, was in line with many previous studies (Prasher 1995; Holland *et al.* 2000; Zigman *et al.*

Table 1 Measured scale scores by level of intellectual disability, before and after diagnosis of dementia

Diagnosis	TSI			DSMSE			DMR			DLSQ				
	Moderate	Severe		Moderate	Severe		Moderate	Severe		Moderate	Severe			
-5	15.8	(6.97)	2.62	(1.85)	4.84	(4.41)	0.92	(0.92)	29.40	(19.51)	14.32	(7.63)	13.23	(5.8)
-4	16.17	(7.13)	2.67	(2.06)	4.97	(4.39)	0.62	(0.62)	29.80	(19.06)	14.40	(7.8)	13.03	(5.86)
-3	16.14	(7.32)	2.82	(2.04)	4.84	(4.45)	0.69	(0.69)	33.20	(19.71)	14.34	(7.22)	12.64	(7.17)
-2	15.56	(7.15)	2.05	(1.84)	4.77	(4.63)	1.00	(1)	36.20	(18.77)	13.77	(7.57)	12.72	(5.49)
-1	15.6	(7.12)	2.69	(1.63)	4.83	(5.07)	1.94	(1.94)	42.60	(17.98)	13.77	(7.28)	12.38	(6.03)
0	14.85	(6.96)	1.83	(2.17)	4.80	(4.97)	1.59	(1.59)	60.32	(21.06)	10.57	(5.81)	9.58	(5.43)
1	11.65	(7.95)	0.98	(1.71)	4.47	(4.51)	1.59	(1.59)	60.69	(21.03)	8.37	(5.48)	5.72	(4.9)
2	10.07	(8.89)	0.73	(0.96)	3.96	(4.27)	1.82	(1.82)	61.73	(21.01)	7.85	(6.29)	5.64	(4.87)
3	9.27	(8.83)	0.75	(1.22)	4.62	(4.97)	1.82	(1.82)	64.10	(21.08)	6.92	(5.86)	6.43	(5.49)
4	7.99	(8.52)	1.22	(2.19)	3.84	(4.65)	1.73	(1.73)	62.02	(22.81)	6.18	(5.66)	5.61	(4.88)
5	7.43	(8.65)	0.32	(0.64)	3.38	(4.4)	1.55	(1.55)	59.64	(24.46)	5.47	(5.65)	6.00	(5.59)
6	6.68	(8.79)	0.33	(0.65)	3.15	(4.74)	1.73	(1.73)	64.60	(22.5)	4.78	(5.82)	4.92	(5.25)
7	6.26	(8.62)	0.17	(0.39)	3.17	(4.95)	1.64	(1.64)	64.97	(24.22)	4.53	(5.66)	5.17	(5.86)
8	5.86	(9.02)	0.17	(0.39)	2.34	(4.44)	1.27	(1.27)	68.30	(24.6)	4.59	(6.03)	4.42	(5.26)
9	5.03	(8.51)	0.17	(0.39)	2.09	(4.5)	1.45	(1.45)	68.00	(25.29)	4.16	(5.7)	3.58	(5.25)
10	5	(8.48)	0.17	(0.39)	1.81	(4.4)	1.27	(1.27)	69.30	(26.05)	4.14	(5.72)	3.58	(5.25)
11	5.11	(8.63)	0.17	(0.39)	1.94	(4.99)	1.27	(1.27)	70.00	(25.24)	4.16	(5.73)	3.58	(5.25)
12	4.92	(8.39)	0.17	(0.39)	1.81	(4.88)	1.27	(1.27)	70.35	(24.5)	4.05	(5.7)	3.58	(5.25)
13	4.84	(8.34)	0.17	(0.39)	1.66	(4.37)	1.27	(1.27)	70.13	(24.82)	4.08	(5.7)	3.58	(5.25)
14	4.78	(8.31)	0.17	(0.39)	1.71	(4.43)	1.27	(1.27)	72.13	(24.12)	4.05	(5.7)	3.58	(5.25)

TSI, Test for Severe Impairment; DSMSE, Down's Syndrome Mental Status Examination; DMR, Dementia Questionnaire for Mentally Retarded individuals; DLSQ, Daily Living Skills Questionnaire.



2002), but was lower than that reported by Visser *et al.* (1997), and higher than in at least one longitudinal study which reported a 50% risk at age 60 and above (Margallo-Lana *et al.* 2007). The average age of diagnosis at 55.41 years (SD = 7.14) was in the higher range of previously reported estimates (51–56 years) and duration after diagnosis, of 6.0–6.25 years was similarly at the high end of estimates (3.5–6.0 years) (Lai & Williams 1989; Prasher & Krishnan 1993). The survival curve reported here noted a median survival of 7 years and this may offer more useful guidance for clinical services, given prior reports of precipitous decline (Lai & Williams 1989); dementia is just as likely to be for an extended period. Understanding survival in persons with DS and dementia is of critical importance having major implications from a care and resource perspective in terms of environmental capability, staff competency and nursing related care needs.

A most interesting and previously unreported finding in this study was that for those with a pre-existing severe or profound level of ID, duration after diagnosis was for an average 9.17 years despite no significant difference in age of onset as compared with persons with DS at a moderate level of ID. This has not been examined or reported upon in other studies, largely because of difficulties in diagnosing dementia. This study has demonstrated that its measurement approach will identify and diagnose symptoms in persons with DS with a pre-existing severe or profound level of ID; additional studies using a similar assessment approach will be able to examine further this increased duration as compared with persons with DS at a moderate level of ID. The rigorous approach taken to diagnosis and the effort to include people with severe ID proved to be a strength of this study. The absence of similar findings in other studies probably reflects that sufficient people with severe ID were not included, the approaches to assessment and diagnosis were not sufficiently rigorous and/or the individuals were not followed for a sufficient period.

The DSMSE proved less sensitive than the other measures and although designed specifically for people with DS may be most useful for people with less severe levels of ID. This lack of sensitivity was not a concern for the other instruments

and it appears they were also useful in establishing decline in people with severe ID. This is an important contribution of the work here as prior studies have reported that other instruments were not useful for people with severe ID (see, for example, Margallo-Lana *et al.* 2007). Finally, the introduction of the DMR in 2005 helped establish the particular sensitivity of this instrument and some evidence that decline may be noted earlier when the DMR is utilised. Interestingly a gradual decrease in functioning prior to the onset of dementia was observed, suggestive perhaps of pre-clinical dementia in those with DS, and supporting the Coppus *et al.* (2008) recommendation on the need for more extensive longitudinal studies to better discriminate between normal ageing and dementia.

There are a number of methodological issues to be considered in the interpretation of these findings, not least that this study focused on women with DS only and represents a small and perhaps unique (one agency) sample. Also each assessment was by the same evaluator and subsequent findings might be more easily influenced by prior impression; however, each diagnosis was a consensus diagnosis by a multidisciplinary team. Future studies may benefit from having larger and more diverse samples and from including men with DS. However the duration of follow-up, prospective versus retrospective design, annual assessment and consistency in use of a memory clinical team and consensus diagnosis operationalised through a memory clinic offer guidance for the structuring of future studies and have demonstrated an ability to identify clinical symptoms of dementia even in persons with severe ID. The findings on survival also highlight that shorter survival reported in prior studies (Lai & Williams 1989; Prasher & Krishnan 1993) probably reflect the limitations of shorter follow-up periods and greater longevity may also reflect earlier diagnosis in this sample. Again these are issues worthy of further investigation.

The need for comprehensive assessment was also confirmed as over time the participants experienced a range of comorbidities. In particular the previously identified strong association between comorbid epilepsy and dementia in people with DS (see, for example, Simone *et al.* 2010; Lott *et al.*

**Table 2** Predictors of dementia

Dementia diagnosis*	Estimate	Std. error	Wald	df	Sig.	OR	95% confidence interval for OR	
							Lower bound	Upper bound
Age	0.092	0.065	2.004	1	0.157	1.097	0.965	1.246
ID (moderate)	-0.596	1.110	0.288	1	0.591	0.551	0.063	4.856
Epilepsy (yes)	-2.140	0.861	6.174	1	0.013	0.118	0.022	0.636

\* The reference category is: no dementia.  
ID, intellectual disability; OR, odds ratio.

**Table 3** Predictors of mortality

Dead y/n*	Estimate	Std. error	Wald	df	Sig.	OR	95% confidence interval for OR	
							Lower bound	Upper bound
Age	0.026	0.042	0.405	1	0.524	1.027	0.946	1.114
ID (moderate)	-1.868	1.173	2.538	1	0.111	0.154	0.016	1.537
Epilepsy (yes)	-1.854	0.587	9.976	1	0.002	0.157	0.050	0.495

\* The reference category is: not dead.  
ID, intellectual disability; OR, odds ratio.

2012) was supported here with almost 74% of those with a dementia diagnosis also having epilepsy and its onset frequently within the same time period as dementia decline. Indeed, the logistic regression findings further support that there may be a strong relationship between presence of epilepsy and the onset of dementia. That the effect of epilepsy was not overwhelmed by age in explaining onset of dementia (Table 2) and mortality (Table 3) helps illustrate the critical nature of the experience of epilepsy for people with DS. Documenting the clinical characteristics of epilepsy and its relationship to cognitive decline warrants further attention including greater understanding of the linkage with what has become more recently characterised as senile myoclonic epilepsy, one of the most common forms of progressive myoclonic epilepsy (Simone *et al.* 2010). The finding that epilepsy was a stronger predictor of mortality than dementia in persons with

DS also deserves further investigation with different and larger samples.

There have also been reports suggesting a strong association between dementia and depression in people with DS. For example, Burt *et al.* (1992) reported much higher levels of depression among people with DS with dementia than among those without dementia. Here, however, rates of depression were high for both groups (49.3%/50%). This may be an artefact of longitudinal follow-up as earlier in the study more distinct differences on depression were identified (Cosgrave *et al.* 2000); after 14 years most of the sample did present with dementia and it is possible that the remaining eight non-dementia participants may be at a pre-dementia stage for which depression may be a co-occurring condition. Understanding the relationship with depression will require further investigation.



## Conclusion

The limitations of a convenience sample notwithstanding, the results here indicate that the model of best practice recommended by Burt & Aylward (2000) when operationalised in a memory clinic was effective in tracking decline and diagnosing dementia in persons with ID. This is in keeping with previous recommendations and an emerging consensus in the field that dementia diagnosis of persons with ID should be based on longitudinal follow-up (Holland *et al.* 2000; Zigman *et al.* 2004). It is hoped that the demonstration of the success of longitudinal follow-up and the usefulness of particular instruments will facilitate the adoption for this approach and thereby early identification of dementia in persons with DS thus helping to ensure earlier access to appropriate treatments and programme redesign.

## Conflict of interest

There are no conflicts, financial or personal for the authors that may potentially bias the work.

## Acknowledgement

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## References

- Albert C. & Cohen C. (1992) The Test for Severe Impairment: an instrument for the assessment of patients with severe cognitive dysfunction. *Journal of the American Geriatrics Society* **40**, 449–53.
- Aylward E. H., Burt D. B., Thorpe L. U., Lai F. & Dalton A. (1997) Diagnosis of dementia in individuals with intellectual disability. *Journal of Intellectual Disability Research* **41**, 152–64.
- Bittles A. H. & Glasson E. J. (2004) Clinical, social, and ethical implications of changing life expectancy in Down syndrome. *Developmental Medicine and Child Neurology* **46**, 282–6.
- Bryk A. S. & Raudenbush S. W. (1987) Applications of hierarchical linear models to assessing change. *Psychological Bulletin* **101**, 147–58.
- Bryk A. S. & Raudenbush S. W. (2001) *Hierarchical Linear Models: Applications and Data Analysis Methods*. Sage Publications, Inc., Newbury Park, CA.
- Burt D., Loveland K. & Lewis K. (1992) Depression and the onset of dementia in adults with mental retardation. *American Journal of Mental Retardation* **96**, 502–51.
- Burt D. B. & Aylward E. H. (2000) Test battery for the diagnosis of dementia in individuals with intellectual disability. Working Group for the Establishment of Criteria for the Diagnosis of Dementia in Individuals with Intellectual Disability. *Journal of Intellectual Disability Research* **44**, 175–80.
- Cahill S., Moore V. & Pierce M. (2011) *Memory Clinics in Ireland*. DSIDC, Dublin, Ireland.
- Chicoine B., McGuire D. & Rubin S. S. (1999) Specialty clinic perspectives. In: *Dementia, Aging and Intellectual Disabilities: A Handbook* (eds M. P. Janicki & A. J. Dalton), pp. 278–93. Hamilton, New York.
- Coppus A. M. W., Evenhuis H. M., Verberne G. J., Visser F. E., van Gool W. A., Eikelenboom P. *et al.* (2006) Dementia and mortality in persons with Down's syndrome. *Journal of Intellectual Disability Research* **50**, 768–77.
- Coppus A. M. W., Evenhuis H. M., Verberne G. J., Visser F. E., Oostra B. A., Eikelenboom P. *et al.* (2008) Survival in elderly persons with Down syndrome. *Journal of the American Geriatric Society* **56**, 2311–16.
- Cosgrave M. P., McCarron M., Anderson M., Tyrrell J., Gill M. & Lawlor B. A. (1998) Cognitive decline in Down syndrome: a validity/reliability study of the Test for Severe Impairment. *American Journal on Mental Retardation* **103**, 193–7.
- Cosgrave M. P., Tyrrell J., McCarron M., Gill M. & Lawlor B. A. (2000) A five year follow up study of dementia in persons with Down's syndrome: early symptoms and patterns of deterioration. *Irish Journal of Psychological Medicine* **17**, 5–11.
- Evenhuis H. M. (1990) The natural history of dementia in Down syndrome. *Archives of Neurology* **47**, 263–7.
- Gardner I. L., Foreman P. & Davis S. (2004) Cognitive dementia and memory service clinics: opinions of general practitioners. *American Journal Alzheimer's Disease Other Dementias* **19**, 105–10.
- Hassiotis A., Strydom A., Allen K. & Walker Z. (2003) A memory clinic for older people with intellectual disability and Down syndrome. *Aging and Mental Health* **7**, 418–23.
- Haxby J. V. (1989) Neuropsychological evaluation of adults with Down's syndrome: patterns of selective impairment in non-demented old adults. *Journal of Intellectual Disability Research* **33**, 193–210.
- Holland A. J., Hon J., Hubbert A. J., Stevens F. & Watson P. (1998) Population-based study of the prevalence and presentation of dementia in adults with Down's syndrome. *British Journal of Psychiatry* **172**, 493–8.

- Holland A. J., Hon J., Huppert F. A. & Stevens F. (2000) Incidence and course of dementia in people with Down's syndrome: findings from a population-based study. *Journal of Intellectual Disability Research* **44**, 138–46.
- Lai F. & Williams R. S. (1989) A prospective study of Alzheimer disease in Down syndrome. *Archives of Neurology* **46**, 849–53.
- Lott I. T., Doran E., Nquyen V. Q., Tournay A., Movsesyan N. & Gillen D. L. (2012) Down syndrome and dementia: seizures and cognitive decline. *Journal of Alzheimer's Disease* **29**, 177–85.
- McCallion P. & McCarron M. (2004) Aging and Intellectual disabilities: a review of recent literature. *Current Opinion in Psychiatry* **17**, 349–52.
- McCarron M. & Lawlor B. A. (2003) Responding to the challenges of ageing and dementia in intellectual disability in Ireland. *Ageing and Mental Health* **7**, 413–17.
- McCarron M., Gill M., Lawlor B. & Begley C. (2002) Time spent caregiving for persons with the dual disability of Down syndrome and Alzheimer's dementia: preliminary findings. *Journal of Learning Disabilities* **6**, 263–76.
- McCarron M., Swinburne J., Burke E., McGlinchey E., Mulryan N., Andrews V. *et al.* (2011) *Growing Older with an Intellectual Disability in Ireland; First Results from the Intellectual Disability Supplement to the Irish Longitudinal Study on Ageing*. School of Nursing & Midwifery, Trinity College Dublin, Dublin.
- McCreary B. D., Fotheringham J., Holden J., Ouellette-Kuntz H. & Robertson D. (1993) Experiences in an Alzheimer clinic for persons with down syndrome. In: *Alzheimer Disease, Down Syndrome and Their Relationship* (eds J. M. Berg, H. Karlinsky & A. J. Holland), pp. 115–31. Oxford University Press, Oxford.
- McKensie K., Muray G., McKensie S. & Muir J. (1998) A prospective, longitudinal study of functional decline in individuals with Down's syndrome. *Journal of Intellectual Disabilities* **2**, 98–104.
- Margallo-Lana M. L., Moore P. B., Kay D. W. K., Perry R. H., Reid B. E., Berney T. P. *et al.* (2007) Fifteen-year follow-up of 92 hospitalized adults with Down's syndrome: incidence of cognitive decline, its relationship to age and neuropathology. *Journal of Intellectual Disability Research* **51**, 463–77.
- National Institute of Aging (1989) *The Daily Living Skill Questionnaire*. National Institute of Aging, Laboratory of Neurosciences.
- Prasher V. P. (1995) End-stage dementia in adults with Down syndrome. *International Journal of Geriatric Psychiatry* **10**, 1067–9.
- Prasher V. P. & Krishnan V. H. R. (1993) Age of onset and duration of dementia in people with Down syndrome: integration of 98 reported cases in the literature. *International Journal of Geriatric Psychiatry* **8**, 915–22.
- Simone R. D., Duig X. S., Lisse P. G., Crespel A. & Genton P. (2010) Senile myoclonic epilepsy: delineation of a common condition associated with Alzheimer's disease in Down syndrome. *Seizure* **19**, 383–9.
- Strydom A., Livingston G., King M. & Hassiotis A. (2007) Prevalence of dementia in intellectual disability using different diagnostic criteria. *British Journal of Psychiatry* **191**, 150–7.
- Strydom A., Shooshtari S., Lee L., Raykar V., Torr J., Tsiouris J. *et al.* (2010) Dementia in older adults with intellectual disabilities – epidemiology, presentation, and diagnosis. *Journal of Policy and Practice in Intellectual Disabilities* **7**, 85–154.
- Tyrrell J., Cosgrave M., McLaughlin M. & Lawlor B. (1996) Dementia in an Irish population of Down syndrome people. *Irish Journal of Psychological Medicine* **13**, 51–4.
- Tyrrell J., Cosgrave M., McCarron M., McPherson J., Clavert J., Kelly A. *et al.* (2001) Dementia in people with Down's syndrome. *International Journal of Geriatric Psychiatry* **16**, 1168–74.
- Visser F. E., Aldenkamp A. P., van Huffelen A. C. & Kuilman M. (1997) Prospective study of the prevalence of Alzheimer-type dementia in institutionalized individuals with Down syndrome. *American Journal on Mental Retardation* **101**, 400–12.
- World Health Organization (1992) *ICD-10: International Statistical Classification of Diseases and Related Health Problems*, 10th revision. World Health Organization, Geneva.
- Zigman W. B., Schupf N., Urv T., Zigman A. & Silverman W. (2002) Incidence and temporal patterns of adaptive behavior change in adults with mental retardation. *American Journal of Mental Retardation* **107**, 161–74.
- Zigman W. B., Schupf N., Devenny D. A., Miezieski C., Ryan R., Urv T. K. *et al.* (2004) Incidence and prevalence of dementia in elderly adults with mental retardation without Down syndrome. *American Journal on Mental Retardation* **109**, 126–41.

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