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50 years with Down syndrome: A longitudinal study

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Background: A population sample of people with Down syndrome, studied from infancy, has now been followed up at the age of 50 years. From the original sample of 54, there were 27 still in the study at the age of 50, all but four of the losses resulting from deaths.

Methods: Intelligence and language skills were tested and daily living skills assessed. Memory/cognitive deterioration was examined using two test instruments. Other aspects of the people's lives were examined via carers' reports.

Results: Scores on verbal tests showed little change. Those on a non-verbal test, on self-help skills and on both memory tests showed some decline, even when the scores of those already suffering from dementia were discounted.

Conclusions: At the age of 50, those not already diagnosed with dementia showed some decline on most tests. While this may include scores of people who subsequently develop dementia, it may also reflect the normal ageing process in this population.

KEYWORDS

50 years, ageing, dementia, Down syndrome, longitudinal

1 | INTRODUCTION

Life expectancy for people with Down syndrome has increased dramatically over the last century. In 1929, life expectancy was estimated as 9 years, rising to 12 years some 22 years later (Penrose & Smith, 1966). Since then, 60% of children born between 1940 and 1950, 80% born between 1950 and 1970, and 90% born between 1976 and 1985 reach at least their first birthday, and 42%, 71% and 79%, respectively, their fifth birthday (McGrother & Marshall, 1990). In 1988, it was estimated that nearly half (44%) of those born between 1952 and 1981 will survive to the age of 60 and 13.6% to the age of 68 (Baird & Sadovnik, 1988). This increased lifespan is attributed largely to the decline, due to heart surgery, in deaths brought about by heart defects (Zhu et al., 2013), supported by Englund, Jonsson, Zander, Gustafstasson and Anneren (2013) who cite pneumonia as now the main cause of death in people with Down syndrome. Similarly, the number living to be older adults has increased. Earlier, of 138 cases known to five local authorities, only 6% were over the age of 34 and none over 45 (Penrose, 1949); by 1975, the oldest person that could be traced was 63 (Carr, 1975); more than a decade later, a woman aged 75 was known (Demissie, Ayres, & Briggs, 1988), surpassed in her turn 2 years later by "three persons with DS 74, 75 and 86 years of age, respectively, presently alive and leading healthy lives with no apparent evidence of impairment or deterioration" (Dalton & Wisniewski, 1990). Indeed, "some 20%–30% of elder adults with Down syndrome might never show any, or at most mild, symptoms of AD" (Zigman, 2013). Welcome as is this increase in the lifespan of people with Down syndrome, it brings with it some concerns. First, parents who saw, as some of those of 6-week-olds in the present study did, some comfort in that "at least I know I can look after this baby for the whole of his/her life" can no longer do so. They now must face the prospect, when they are themselves unable to continue caring, of needing to look for alternative accommodation for their Down syndrome offspring, sometimes repeatedly, with the possibility of instability, of frequent turnover of carers, of unpredictable changes in the acceptability of the provision, and the consequent frequency of transitions to new placement. Second, the vulnerability of the group to dementia has become increasingly clear, leading to the current emphasis on diagnosis and the search for effective treatments.

Strydom et al. (2010), reviewing the published literature over the previous 11 years, found dementia reported in <10% of people with Down syndrome in their 40s, increasing to 30% in their 50s, and rates of prevalence varying in different studies of people aged 60+. McCarron et al. (2014) followed a group of people aged 35 and older for 14 years, finding the average age of diagnosis to be 55.4 years, rather older than had been previously reported, and considered the Dementia Questionnaire for Mentally Retarded Persons to be the most sensitive in tracking the progress of dementia symptoms. Holland, Hon, Huppert, Stevens and Watson (1998) used the Cambridge Examination for Mental Disorders (CAMDEX) with a population-based sample of older people with Down syndrome and found that with increasing age an increasing proportion were reported to have declines in memory and in two or more other areas, such as daily living skills or general mental functioning. However, the two oldest people, both over 60, showed no sign of deterioration in any area of functioning. McQuillan et al. (2003) suggested that, to monitor emergent signs of cognitive deterioration, all individuals with Down syndrome should be assessed at the age of 30 and the assessment repeated every 12–18 months, a similar viewpoint being put forward by Ghezzo et al. (2014).

Changes in a number of specific abilities have been identified as indicative of the development of dementia. Decline in language skills has been implicated: Dalton, Mehta, Fedor and Patti (1999) showed that declines on a language memory task preceded those on dyspraxia by two and a half years, suggesting that these changes were indicative of the early stages of dementia. Cooper and Collacott (1995) found language ability, in comprehension but not expression, to decline over age groups from 20 to 60+, becoming significant overall after 50. Some executive function tasks, in people with Down syndrome not currently diagnosed with dementia, were significantly associated with changes in behaviour and personality (Ball, Holland, Treppner, Watson, & Huppert, 2008), while some executive function tasks, those concerned with shifting and with verbal aspects of memory and inhibition, differentiated between people with Down syndrome and with Williams syndrome (Costanzo et al., 2013).

As they age, it becomes pertinent to enquire whether the declines shown are attributable to the effects of dementia or are due to those of normal ageing. Devenny et al. (1996) studied three groups aged 31–63 over 6 years, screened to exclude any suspected of decline in functioning and found the younger group to show an increase in a long-term memory test (part of the Buschke Selective Reminding Test), while the older group, of 27 people over age 50, mean age 55, showed "very slight decreases" in recent memory and coding which could be attributed to "normal if precocious" ageing. Crayton, Bradbury, Oliver, Hall and Holland (1998) studied 70 people with Down syndrome, also screened to exclude any with cognitive impairment, whether or not this was due to dementia. As expected, they identified none with neuropsychological deficits, but the older group, aged 50+, performed significantly worse on memory tests than the younger group, suggesting that these are good indicators of the early stage of dementia. Collacott and Cooper (1992) found deterioration in most domains of the Adaptive Behaviour Scale in a cohort aged 50–59 and in all domains in those aged 60+. Older people with Down syndrome have been shown to experience more residential relocations than do those with intellectual disability not due to Down syndrome (Patti, Amble, & Flory, 2010), although Woodman, Mailick, Anderson and Esbenson (2014) dispute this, finding adaptive behaviour, not type of disability, to be a predictor of number of residential placements.

Although most studies examining the differences in achievement of different age groups have been cross-sectional, a number of longitudinal studies have been carried out. Earlier, these were of very young children, examining the changes, in the event the declines, in measured abilities as the children grew older (Dameron, 1963; Dicks-Mireaux, 1966; Share et al., 1961; Stedman & Eichorn, 1964), and the differences in abilities between those brought up in or out of their own homes. Later attention turned to the possibility of enhancing the abilities by "early intervention," brought about by systematic and intensive teaching and stimulation, carried out by (usually) the mothers under instruction by a team of psychologists (De Coriat et al., 1967; Ludlow & Allen, 1979; Piper & Pless, 1980). In each of the two latter scenarios, the early evidence was of significant advantage, for, in the one case, those brought up at home, and in the other for those experiencing the stimulation regime. These advantages were not borne out in the longer term, in the former disappearing by 11 years (Carr, 1995) and in the latter by age 5, the intervention having been terminated after 2 years (Cunningham, 1987). More recently, the focus has been on the advent of dementia and on older groups the longitudinal research being mainly (and understandably) of quite short duration: of 3–5 years (Burt et al., 1995; Devenny et al., 1996; Oliver et al., 1998; Roeden & Zitman, 1997); a 7-year study was carried out by Kittler et al. (2004) and one of eight years by Dalton and Crapper-McLachlan (1984) while that by McCarron et al. (2014) continued for 14 years. A longer study, albeit not of older people, is that of Ludlow & Allen who, in an early intervention project, studied groups totalling 184 from 1 to 10 years and followed up about 100 some eight later, the data being reported by Cunningham (2006). The longest study previously recorded, albeit not prospective, is that of Couzens et al. (2012) reviewing test results from children from the age of four and followed up to adulthood at the age of 30. No previous study has exceeded 30-year duration.

The search for effective treatments has not to date produced much that is positive. In the general population, "although the German psychiatrist and neurologist Aloysius Alzheimer first identified Alzheimer's disease over a hundred years ago, we remain without effective treatments or a cure for this devastating disorder" (Kent, 2015). In regard to Down syndrome, the situation is similar, with one drug, Donepezil not being found to have resulted in benefits (Mohan, Carpenter, & Bennett, 2009), while another, memantine, which had been found to have beneficial effects in transgenic mice, failed to show the same effects in people with Down syndrome (Hanney et al., 2012).

The present study was designed to complete 50 years of research on one cohort of people with Down syndrome. The aim of the study was to describe the changes in cognitive ability and self-help skills of the whole cohort over the 50 years (carers were also interviewed about the people's lives, and this will be described in a companion paper).

TABLE 1 Number and mean IQs[a], 6 weeks to 50 years

Age	Number	Mean IQ
6 weeks	29[b]	73
4 years	54	44
11 years	44	37.2
21 years	41	41.9
30 years	38	42.1
35 years	37	41.3
40 years	33	42
45 years	30	41.3
47 years	29	37.9
50 years	27	36.4

[a]IQ tests do not cover all ages. The Bayley Scales of Infant Development were used up to 4 years; the Merrill Palmer Scales at 11 years; and the Leiter Scales from 21 years.
[b]Some children were referred later, the last at the age of 2 years.

2 | METHOD

2.1 | Participants

A population sample of babies with Down syndrome, born in one year (December 1963–November 1964) in the county of Surrey less the borough of Croydon, and area 7 of southeast London, then Camberwell and Lewisham, has been seen and tested by the first author when they were at the age of 6 weeks, at intervals to age 4 years, then at 21 and 30 years, thereafter at 5 year intervals to age 45, then at the age of 47 (Carr, 1975, 1995, 2000, 2003, 2012; Carr & Collins, 2014) and now at the age of 50. Fifty-four babies with Down syndrome, 45 reared at home and 9 out of home, were referred to the study. A control group of typically developing babies, matched individually for sex, age and social class with each home-reared baby with Down syndrome, was seen and given the same tests as were those with Down syndrome from the age of 6 weeks to 3 years. They were not seen after that because there was no overlap between the scores of the two groups, but the families were included up to the age of 45. By the age of 50, 27 of the original 54 participants with Down syndrome were still in the study, all but four of the losses being due to death (one of the four untraced participants was from a family who moved to Africa before the baby was 2 years old, and the other three were withdrawals).

The 27 people were retraced and seen in their homes. One woman died before she could be seen, and two men soon after. One woman who had been lost at the age of 40 (she was moved to yet another foster placement and could not at that time be traced) was found and seen again. Two women who were in the "dementia suspected" group at 47 were no longer included in it: each had a substantially increased score on one memory test and a non-verbal intelligence score that was only slightly below that at 47. Nine people, including the five now diagnosed as demented, these including for the first time one man, made no score on any test. Four of the five with dementia, all women, had previously been among the highest achievers in the whole group.

A third of the people still lived at home, most with a surviving parent but two with a sister and one with a brother; one woman was fostered, and the remaining 64% were in health, private/voluntary or social services facilities. Over half attended a day service, one also attended further education courses part-time and two also had part-time jobs although only one of these was paid.

2.2 | Measures

The same tests were employed as had been used from the age of 21 to 47—the Leiter International Performance Scale (LIPS, Leiter, 1980), the British Picture Vocabulary Scale (BPVS, Dunn, Dunn, Whetton, & Pintillie, 1982), the vocabulary scale from the Wechsler Pre-School and Primary Scale of Intelligence (WPPSI, Wechsler, 1967); and two tests of memory/dementia, the Rivermead Behavioural Memory Test for Children (RBMT-C, Wilson & Ivani-Chalian, 1995), which has been standardized on adults with Down's syndrome (Hon, Huppert, Holland, & Watson, 1988), and the Neurological Assessment of Dementia in Individuals with Intellectual Disability (NAID, Adams & Oliver, 2010), formerly known as the Oliver & Crayton Dementia Battery. It is recognized that some of these tests now have newer versions, but the tests used were deliberately not changed so as to ensure better comparability. Self-help skills (feeding, washing, dressing and toiletting) were again rated from assessments by carers, as in the previous studies, using the handicaps, behaviour and skills schedule (HBS, Wing, 1980), scores in this case being the sums of the raw scores as set out by Wing.

2.3 | Procedure

The study was submitted to the Social Care Research Ethics Committee (SCREC) and was approved by them after minor changes. Accessible information sheets and consent forms were used for those participants with Down syndrome who could consent. For those who could not consent for themselves, research consultees were sought, as required under the Mental Capacity Act 2005. All carer participants also received information sheets and consent forms.

The first author had stayed in touch with all families, through Christmas cards, and this meant that very few were lost to follow-up. Information sheets and consent forms were sent to home addresses, and capacity to consent was checked by the parent or carer who knew the person best. Most participants were seen in the parental home or in their residential home; a few were seen in their day services.

2.4 | Analysis

Means and standard deviations were calculated for all data as appropriate. The distributions of data for the various measures were examined for normality, and because of the distribution of data, as well as the relatively small numbers, non-parametric statistics were used. The Wilcoxon test was used for all repeated measure

comparisons (such as participants' changes in scores with age, for the whole group, and for men and women separately). In each of these analyses, the change between the peak score (at whatever age) and the score at the age of 50 years was analysed. The Mann–Whitney test was used for between-group comparisons (such as for differences between men and women, on particular tests at specific ages).

3 | RESULTS

In each analysis, all those who were tested were included, except where otherwise stated.

First, Table 1 shows the numbers of participants surviving and seen, with mean IQs at successive ages. As IQ tests do not cover all ages, the Bayley Scales of Infant Development were used up to 4 years; the Merrill Palmer Scales at 11 years; and the Leiter Scale from 21 years.

From this point on, only those people who featured at every stage of the study were included for analysis, that is those 27 who were in the study at every age from 21 up to 50 years. Of these 27, five had already been diagnosed with dementia by the age of 50 years and these five were unable to score on the psychometric tests, so for these tests n = 22 (unless otherwise stated). Where participants could generally score on the tests but had very occasional missing data, the mean for that person on the other occasions s/he was tested was used (this was carried out for the BPVS at 21 years for case 38; for the Leiter and BPVS for case 50 when she was untraced at 45 & 47 years; for the NAID at 50 years for case 9).

3.1 | Intellectual, verbal and self-help ability

3.1.1 | Intellectual ability

Average non-verbal IQs on the Leiter International Performance Scale, from the age of 21 to 50, are shown in Figure 1, for men and for women. The five people with dementia were excluded (so n = 22). Those who did not have dementia but were unable to complete any test items were included here but given the lowest possible score (so as not to inflate the average).

For those 22 people unaffected by dementia, the loss in IQ for the whole group, from the high point at 21 years, is 4.1 points, significant at $p < .02$ (on the Wilcoxon), but this fails to reach significance for either sex when these were examined separately. For the whole group, the loss remains only about a third of that expected of the non-disabled population over a similar period (Carr, 2005).

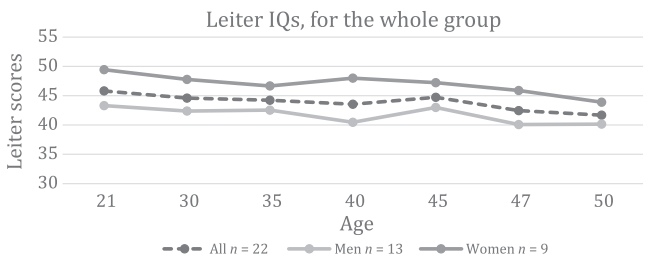

FIGURE 1 Leiter IQs, for the whole group, and for men and for women, excluding the five with dementia

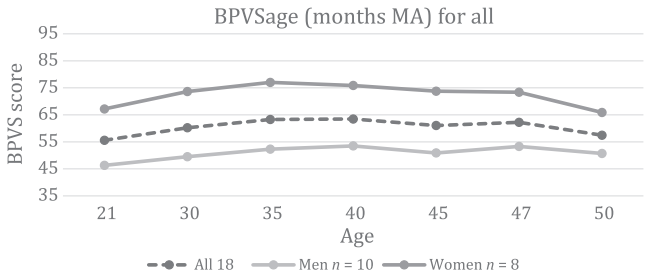

FIGURE 2 BPVSage (months MA) for all, and for men and for women. (Note: those affected by dementia are not included here, as they did not make a score at the age of 50)

3.1.2 | Verbal ability

Of the 22 (i.e., those who did not have dementia) who were tested on the BPVS, only 18 could score. Figure 2 shows scores on the BPVS, in age equivalents in months, for all 18, and for men and for women separately (n = 10 and 8, respectively).

Following a small rise from the age of 21 to 30 years, scores fluctuated by a few months, in both the men and the women and in the two groups combined. None of the changes is significant.

Of the 22 (i.e., those who did not have dementia) who were tested on the WPPSI, only 12 could score on at least five occasions (of the seven assessments). Figure 3 shows WPPSI age in months MA (on the vocabulary subscale) for all (n = 12), and for men and for women separately (n = 6 and 6, respectively).

Scores for the whole group of 12 fluctuated relatively little, with only a small drop (0.99) in the overall mean from the age of 21 to 50. This difference, which is not statistically significant, was similar for the men (a drop of 2.16), while the women's mean score increased very slightly (0.19).

In the results of all the tests shown here, the Leiter, BPVS and WPPSI, mean scores of the women are higher than those of the men. In the case of non-verbal ability (Leiter), the differences between men and women are not significant (using the Mann–Whitney) at any age. In the case of BPVS, the women score significantly better than the men at the ages of 21, 30, 35 and 45 years ($p < .05$ each time). As regards

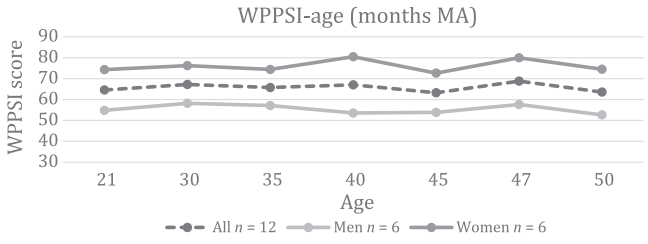

FIGURE 3 WPPSI age (months MA), for all, and for men and for women. (Note: those affected by dementia are not included here, as they did not make a score at the age of 50)

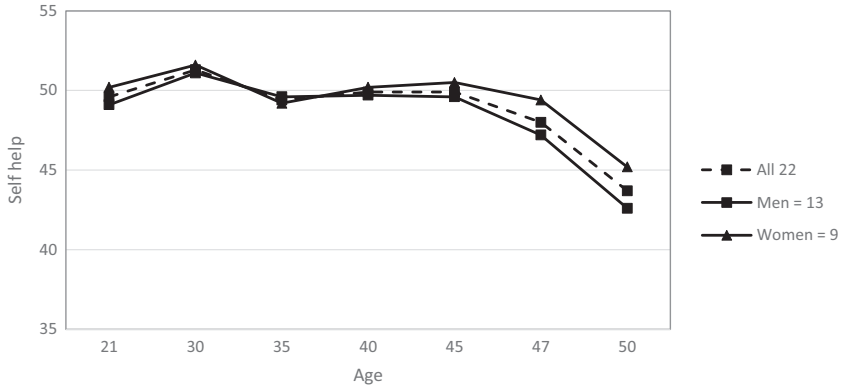

FIGURE 4 Total self-help skills for all, and for men and women, excluding the five with dementia. (Maximum = 58)

expressive language (WPPSI age), the differences between men and women are significant at every age except 21, 30 and 35, the level of significance varying from $p = .002$ at the age of 40, and $p < .05$ at the age of 45, 47 and 50, (again using the Mann–Whitney). The advantage to the women in verbal ability has been shown virtually throughout the study.

3.1.3 | Self-help skills

Figure 4 shows total self-help skills for all ($n = 22$), and for men and women separately ($n = 13$ and 9, respectively), excluding the five diagnosed with dementia. The score was the sum of the scores on each of the four items, and the maximum possible score was 58.

Setting aside the scores of the five with diagnosed dementia, the loss for the whole group from the maximum at the age of 30 was 7.6 points (Wilcoxon, $Z = -3.5$, $p = .0005$), that for the men 6.5 points, ($Z = -2.8$, $p = .004$) and that for the women 5.0 points (non-significant). Looking at the separate items, the difference from the age of 30 to 50 is significant for feeding ($Z = -3.45$, $p = .0006$), washing ($Z = -3.28$, $p = .001$) and for toiletting ($Z = -2.55$, $p = .1$). That for dressing is not significant.

3.2 | Tests of dementia

Nine people of the 27 (the five with diagnosed dementia and four with profound learning disabilities) were unable to score on either test, so are excluded.

Figure 5 shows results from the RBMT-C, for all ($n = 18$) and for men and women separately ($n = 11$ and 7, respectively). These figures show that the loss on the RBMT-C, from a peak at the age of 40, was, for the whole group, 14.7 points (Wilcoxon, $p = .001$); for the men 8.1 points ($p < .05$); and for the women 25.1 points ($p < .02$).

Figure 6 shows results from the NAID, for all ($n = 18$) and for men and women separately ($n = 11$ and 7, respectively). The loss at the age of 50 years from the peak figure attained at the age of 40 by the whole group was 7.8 points (Wilcoxon, $p < .05$); for the men, from a peak attained at the age of 35, was 5.0 points (non-significant); and for the women, from a peak attained at the age of 40, was 14.6 points ($p < .05$). Thus, even for those not currently with confirmed dementia, these tests show some falling off in performance.

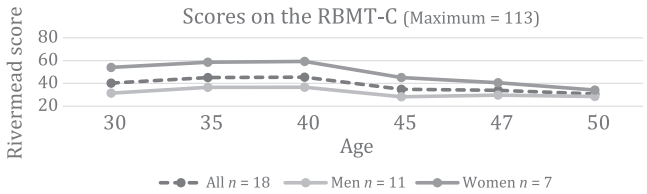

FIGURE 5 Scores on the RBMT-C, for all and for men and women. (Maximum = 113)

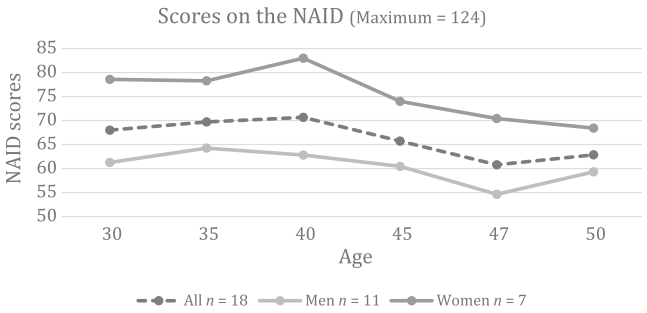

FIGURE 6 Scores on the NAID, for all and for men and women. (Maximum = 124)

4 | DISCUSSION

Half the original 54, seen for the first time as 6-week-old babies, survived to age 50, and are now well into what is for them middle age. All but four of the losses from the sample were brought about by deaths, so this may be thought of as a reasonably representative sample, albeit a relatively small one. Of the 27 seen at the age of 50, five (18%) were found to be in the strongly suspected/confirmed dementia group, placing them between the percentages reported by Strydom et al. of 10% affected by dementia in their 40s, and 30% in their 50s. Results from this latest phase of the study show that, even when those from the confirmed dementia group are discounted, many scores have declined: for the whole group, on non-verbal IQ, and more strongly on self-help and both memory tests, while receptive and expressive language skills seem to be protected from this general decline. On non-verbal IQ, the decline is significant for the whole group, and on self-help, it is significant for the whole group and for the men. On self-help, three of the four components of this

score, feeding, washing and toiletting, have declined significantly, and these may be adding to the task of caring for these people.

As for language, the decline was minimal and non-significant in receptive language, unlike in Cuskelly, Povey and Jobling (2016), who found receptive language declined after the age of 20 years, and Cooper and Collacott (1995), who also found lower comprehension in those over 40 years. The losses in expressive language skills were also minimal. Non-significance of these losses may be attributed at least partly to the very small numbers in this study who scored on this test (the WPPSI vocabulary test).

On the memory tests, the picture is more striking. Results for the whole (non-demented) group for the RMBT-C and the NAID show a continuous falling from the age of 40 to 50. What then is the import of this pattern of scores? Does it represent "the early stages of dementia" (Oliver et al. 1998) or the effects of normal ageing? In regard to the former, it would be unlikely to be the case for the whole group. Examination of the trajectory of scores for each individual reveals a similar downward slant for eight on the RBMT-C and four on the NAID: one man and one woman, not so far among the "suspected" group, feature on both these and on self-help, as do both women recently removed from that group, suggesting that these four might indeed be likely candidates for dementia and might properly be added to the previous five. One of the most striking aspects of this group has been that, until most recently, it has been entirely composed of women, the majority of whom were, before the advent of the condition, among the highest achievers of the whole cohort. This has been an unexpected finding, other studies suggesting that cognitive deterioration is less rapid in higher-functioning people, (Oliver et al., 1998; Temple, Josvai, Konstantareas, & Hewitt, 2001). However, the remaining 18 people (67%) currently show no sign of major deterioration; they may be seen as falling well within Zigman's (2013) 20%–30% who may never suffer from dementia. For this remainder of the group then, their scores may, for the time being, be taken as indicating the effects of normal ageing.

Turning to more general issues, a reservation commonly expressed regarding cross-sectional studies refers to the comparison necessarily made at different ages between groups of different individuals. Longitudinal studies, such as the present one, avoid that concern—the performance of each individual is compared with that of the same individual at each age. However, a hazard involved in such a longitudinal study, particularly in one of the current duration, where many of the target people move to live in care, is the turnover in carers, and hence in informants, leading to uncertainty as to the consistency of the information supplied. (This applies also of course in the case of cross-sectional studies.) In the present study for only three people was the informant the same person, a family member, throughout all ages, although for two more the informant was consistent from the age of 30 to 47. So for a number of informant-derived items, particularly those requiring a subjective judgement, the variations seen from one age to another may be due to the different perceptions of different informants (such variation not being apparent for more objective measures). It is, however, very difficult to see how this problem could be overcome.

At this culminating 50-year point of the present study, a concern for the first (and for 45 years only) author is to discern what of value it may have given to people with Down syndrome and to the families? What has it contributed to the body of knowledge of the condition? First, for the people themselves, the test sessions were seen as enjoyable, described as "all those games" by one man, others being said to look forward to the researcher's visit, while family members valued the long-term contact with the first author and the practical help and support it was occasionally possible to give. Second, in the context of the wide range of other research into the condition currently in existence, it may have set a precedent, being thought to be the longest known study of its kind; against that may be set its very small size, becoming smaller as time went on, precluding the use of many valuable statistical procedures to explore the data available (although there are studies reporting on considerably smaller numbers, Haxby, 1989; Fenner et al., 1987). Third, when the author(s) of cross-sectional research have expressed misgivings about the validity of their findings, because of the unknown effect of other putative factors, such as the effects of changes in educational practices in different cohorts (Crayton et al., 1998; Haxby, 1989), it has been possible to show that these are supported by the findings of the present longitudinal research. Fourth, the study was one of the first to show that the arrival of a Down syndrome baby did not herald disaster for the family (Carr, 1995). Fifth, it has been suggested that the present study demonstrates, through the very low dropout rate of participants, the usefulness of the Christmas card strategy, engaged in from the age of 4 to 50 (and which it is intended to continue indefinitely), although it has to be said that with the larger numbers advocated, such a strategy might well be impracticable. Finally, on a more personal note, there is no possible question that it has been, for the first author, a most rewarding 50 years.

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