An examination of the Severe Impairment Battery as a measure of cognitive decline in clients with Down syndrome

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Abstract

The present study examined the validity of the Severe Impairment Battery (SIB) as a tool for measuring cognitive decline in clients with Down syndrome. Two groups participated: 10 clients who showed behavioural decline over at least a 2 year period as measured by the Vineland Adaptive Behaviour Scales, and 14 clients who showed no decline on this measure over the same period. No differences were found between the two groups in relation to health or life factors which may have impacted on functional and cognitive decline. The deteriorating group were found to be significantly older than the non-deteriorating group. The comparison of the SIB scores indicated that the deteriorating group showed a significant decline between baseline and 12 months and baseline and 24 months on the orientation factor. By contrast, for the non-deteriorating group, significant increases were found for praxis, orientating to name and total scores.

Keywords cognitive decline; Down syndrome; Severe Impairment Battery; validity
Introduction

It is being increasingly documented that people with Down syndrome are at a greater risk of developing Alzheimer’s disease than the general population (Crayton and Oliver, 1993). This has led to the search for early indicators of cognitive and behavioural decline. The progression of Alzheimer’s disease in individuals with Down syndrome is similar to that of the general population in that it involves a decline in both cognitive and behavioural functioning (Hutchinson, 1999). Researchers have, however, also identified a number of differences relating to the course, progression and diagnosis of the disease in people with Down syndrome (Hutchinson, 1999).

First, the onset of Alzheimer’s disease in people with Down syndrome has been found to be earlier than in the general population, with studies placing the age of onset between 49 and 54 (Lai and Williams, 1989; Rasmussen and Sobsey, 1994). The progression is also more rapid, with a period of between 2 and 15 years and an average of 3 to 6 years from detection to death (Hutchinson, 1999; Kerr, 1997; Lishman, 1998). The initial signs of decline also appear to differ. The first indicator of Alzheimer’s disease in the general population is a deterioration in cognitive functioning, while the first sign of a developing dementia in people with Down syndrome tends to be a decline in behavioural functioning. This may be because early deterioration in the cognitive abilities of people with Down syndrome may simply go undetected in the presence of the pre-existing cognitive deficits associated with the intellectual disability (Lai and Williams, 1989).

Diagnosis of Alzheimer’s disease is also more difficult in people with Down syndrome as compared with the general population (Holland et al., 1993). Accurate diagnosis is hindered by a number of factors. General health conditions, e.g. depression, thyroid problems, can
mimic the behavioural and cognitive decline associated with Alzheimer’s disease (Dalton et al., 1993). People with an intellectual disability have been found to have increased health needs compared with the general population which can go undetected (Paxton and Taylor, 1998). Life events, such as loss and bereavement, may also have effects similar to those related to Alzheimer’s disease (Kerr, 1997). Any assessment of Alzheimer’s disease must therefore take into account the possible influence of such health and social factors on the client’s functioning.

There is also a lack of appropriate neuropsychological tests which have been designed and standardized for people with an intellectual disability. Particular difficulties include floor effects, i.e. the assessment items may not be achievable for clients (Crayton et al., 1998; Witts and Elder, 1998); a difficulty in determining whether individual variability in performance over time is indicative of cognitive deterioration or attributable to the person’s normal fluctuations (Burt et al., 1998); and difficulty in identifying a single assessment to aid diagnosis, as not all individuals will necessarily show the same initial signs of deterioration, course or progression of the disease (Burt et al., 1998). Assessments of cognitive decline which are currently available to aid diagnosis in the general population may therefore be ineffective at detecting early signs of cognitive decline in individuals with Down syndrome.

Early detection is important, not only to help future service planning, but also to ensure that a comprehensive package of care, designed to meet the needs of the individual, is in place. As a result, the search for an accurate measure of early cognitive decline continues, and the literature reflects a broad range of assessments that have been employed as measures of
cognitive decline with individuals with Down syndrome (Crayton et al., 1998; Hutchinson, 1999).

One assessment that appears to be increasingly used by clinicians is the Severe Impairment Battery (SIB) (Saxton et al., 1993). This was originally developed as a diagnostic tool to help in the diagnosis of dementia in the general population. It provides cut-off scores and is said to plot ongoing cognitive decline through repeated use. It consists of 39 items and nine subsections reflecting those areas that are identified as being subject to decline in the general population. These are: social interaction, e.g. holding a brief conversation; memory; orientation, e.g. awareness of time, place and date; language; attention, e.g. the ability to focus on information; praxis, e.g. putting instructions into action; visuospatial ability, e.g. copying and recognizing shapes; construction; and orientation to name. Previous research by Witts and Elder (1998) suggested that the SIB had adequate test–retest reliability and criterion validity, as measured against the Vineland Adaptive Behaviour Scales (Sparrow et al., 1984). This study, however, was not longitudinal and none of the participants had been diagnosed as having dementia. The usefulness of the SIB as an indicator of cognitive decline over time, therefore, requires further investigation. The present study therefore aims to do the following:

• To examine if significant differences exist in SIB scores at baseline and follow-up, for clients with Down syndrome showing decline in their adaptive skills, as measured by the Vineland Adaptive Behaviour Scales (Sparrow et al., 1984).

• To compare these results with a group of clients with Down syndrome who do not show behavioural decline, using the same assessments.
It is hypothesized that SIB scores for the group showing decline in their adaptive skills will decrease, while there will be no change or an increase in SIB scores of the group showing no behavioural decline.

**Method**

**Participants**

Twenty-four clients participated, as part of a larger clinical pathway screening for Alzheimer’s disease (McKenzie et al., 2000). All of the clients were followed up for at least 2 years, and some for up to 5 years. Ten clients were found to show a sustained decline in their adaptive behaviour as measured by the Vineland Adaptive Behaviour Scales. Of these seven met the criteria for probable Alzheimer’s disease and three met the criteria for possible Alzheimer’s disease (Aylward et al., 1995; McKhan et al., 1984). The mean age of this group was 51.9 years (SD = 6.4) and four were male and six were female. Fourteen clients showed no decline in adaptive skills. Of these three were male and 11 were female. The mean age of this group was 44.2 years (SD = 6.63). All clients were offered a health screen to identify and treat any medical cause for the deterioration other than Alzheimer’s disease.

In addition, information was obtained about any recent life events that may affect the clients’ functioning, e.g. a bereavement or change in residence. Health problems were experienced by 78 percent of the deteriorating group and 73 percent of the non-deteriorating group. A life event such as accommodation or staff changes or a bereavement had been experienced by 56 percent of the deteriorating group and 55 percent of the non-deteriorating group.

**Measures**

**Adaptive behaviour**
This was measured using the Vineland Adaptive Behaviour Scales (Sparrow et al., 1984). This tool is commonly used in the field of intellectual disability, and is reported as having sound psychometric properties. The assessment relies on carer reports of client functioning in the following domains: communication, daily living skills, socialization, motor skills and maladaptive behaviour.

**Cognitive decline**

This was assessed using the Severe Impairment Battery (Saxton et al., 1993). This assessment, as described above, has 39 items and is completed by the client. Research by Witts and Elder (1998) suggests that it is not prone to the floor effects commonly encountered with other neuropsychological assessments adapted for use with clients with an intellectual disability.

**Procedure**

Following a referral, information was obtained using the measures outlined above. Clients were then followed up every 12 months. Referrals were also made for a health screen and for follow-up medical treatment of identified problems, and to other team members as appropriate (McKenzie et al., 2000). Clients were assigned to either a ‘deterioration’ or a ‘no deterioration’ group, depending on the outcome of the Vineland Adaptive Behaviour Scales assessment. As clients were followed up over differing periods, and as deterioration in functional skills occurred at different periods, the present study adopted as baseline the first assessment prior to which deterioration was detected. This referred to those clients who showed a consistent decline in functional skills, as measured on the Vineland Adaptive Behaviour Scales, over at least a 2 year period (deteriorating group). The following comparisons were then made for the SIB scores: baseline and 12 months, baseline and 24
months, and 12 and 24 months. Equivalent comparisons were also made for the non-deteriorating group, who showed no behavioural decline over at least a 2 year period.

**Results**

An unrelated $T$-test illustrated that the ‘deterioration’ group were significantly older than the ‘non-deterioration’ group ($t = 2.838$, d.f. = 19.97, $p < 0.01$). A series of Wilcoxon signed ranks tests illustrated a significant decline in orientation scores for the deteriorating group between baseline and 12 months ($Z = -2.428$, $p < 0.01$) and between baseline and 24 months ($Z = -2.414$, $p < 0.01$). An examination of the individual items making up this score illustrated a significant decline in the ability of the client to name the city they lived in between baseline and 24 months ($Z = -1.667$, $p < 0.05$). No other significant differences were found for this group.

For the non-deteriorating group, significant increases were found for the following scores: total scores between baseline and 12 months ($Z = 2.241$, $p < 0.05$); praxis scores between baseline and 12 months ($Z = 1.697$, $p < 0.05$), 12 and 24 months ($Z = -2.09$, $p < 0.05$) and baseline and 24 months ($Z = -2.647$, $p < 0.005$); and orientating to name scores between baseline and 12 months ($Z = -2.07$, $p < 0.05$) and baseline and 24 months ($Z = -1.732$, $p < 0.05$).

**Discussion**

The present study aimed to see if significant differences were found in the cognitive functioning of clients with Down syndrome, as measured by the SIB, who were reported as consistently showing behavioural decline over at least a 2 year period. The study found a significant decline between baseline and 12 and 24 months on the orientation domain. This
domain incorporates only three items: client name, month of the year and city the client lives in. A further examination of these items found a significant decline in only one specific item, the ability of clients to name the city they lived in 24 months after baseline. By contrast, no such decline was found for the ‘non-deterioration’ group. In fact, this group showed significant improvement on a number of the domains over time.

A general limitation of the use of existing neuropsychological assessments with individuals with Down syndrome is that not all individuals will necessarily show the same initial indicators, course or progression of Alzheimer’s disease. Both Crayton et al. (1998) and Burt et al. (1998) emphasize the need for longitudinal assessments to allow for an examination of the significance of cognitive decline in comparison with that person’s previous performance. This would also allow for normal individual fluctuations in performance. The present study adopted this methodology and followed up individuals for at least two years from the point at which behavioural decline was first reported. While it is likely that individual patterns of cognitive decline occurred during this period, as a group decline was found in relation to the orientation factor, suggesting a useful area for further research.

A decline in orientation has been identified as one of the early indicators of Alzheimer’s disease in the general population and previous studies examining people with Down syndrome have also found a decline on this factor (Crayton et al., 1998). The results of the present study may indicate that the orientation domain of the SIB provides an early indicator of cognitive decline associated with Alzheimer’s disease, particularly as the same pattern was not found for the ‘non-deterioration’ group. If this were the case, the SIB may offer a relatively quick and non-threatening assessment which is applicable to the general population
and to individuals with an intellectual disability. Crayton et al. (1998) highlight the benefits of an assessment which is applicable to both groups, in particular for strengthening the research links and paradigms for both.

There are, however, some difficulties with the assessment. While it uses gestural prompts and simple language and takes place in the individuals’ own familiar environment (Witts and Elder, 1998), it remains difficult to use with clients with greater degrees of pre-existing cognitive impairment and limited verbal skills. Similarly, for clients with lesser degrees of intellectual disability a ceiling effect can occur. The results of the present study, however, suggest that the SIB would merit further investigation with clients with an intellectual disability who are at risk of developing Alzheimer’s disease.

However, there may be alternative explanations for the results of the present study. Cognitive deficits have been found in previous studies to increase with age (Crayton et al., 1998), and it remains unclear which aspects of decline are due to the normal ageing process and which are due to a dementia. In the present study the ‘deterioration’ group were found to be significantly older than the ‘non-deterioration’ group and the decline in orientation scores may have been partly attributable to this factor. Further research would require to match individuals in the ‘deterioration’ and ‘non-deterioration’ groups on age to address this question.

An additional factor which may have influenced the results is the life changes experienced by clients. Fifty-six percent of clients in the ‘deterioration’ group experienced life changes, including bereavement, staff and accommodation changes. It may be that these changes resulted in a decrease in orientation scores, particularly if the accommodation change resulted
in the person moving to a different city or town. No direct correspondence was found, however, between those who had moved accommodation and a decline in orientation scores. In addition, if the decline had been attributable to life changes, a similar pattern would have been expected for the ‘non-deterioration’ group, 55 percent of whom also experienced the life changes outlined above. In fact, this group experienced a significant increase over time on many of the domain scores, suggesting that the results of the study were not attributable to life changes per se.

In summary, the present study indicated that the orientation domain of the SIB may have discriminant validity as an early indicator of cognitive decline related to Alzheimer’s disease in people with Down syndrome. Further research is required to establish the impact of factors such as age on changes in scores in this area.
References


