The assessment of behavioural decline in adults with Down syndrome

Karen McKenzie, Cyan Harte, Shona Patrick, Edith Matheson, George C Murray

Abstract

The present study examines two methods of using the Vineland Adaptive Behaviour Scales as a measure of behavioural change in people with Down syndrome who are at risk of developing Alzheimer's disease. The first method uses the Vineland scales as the basis of a semi-structured interview and notes all areas of behavioural change identified by staff; the second method scores the Vineland scales using the basal rule outlined in the manual. The comparison of these two methods illustrated that using the second method highlighted a significant decline in scores for the group meeting the criteria for 'probable Alzheimer's disease' on a number of domains between baseline and 12–24 months. However, this scoring method also appeared to miss more subtle changes in behaviour, which may be important early indicators of Alzheimer's disease, which were picked up by the first method. The implications of the study are discussed.

Keywords: Alzheimer's disease; Down syndrome; Vineland scales

Introduction

The lifespan of people with Down syndrome is increasing and as a result clients are now more susceptible to age related health and cognitive changes (Hutchison, 1999). In particular, people with Down syndrome have been found to be at an increased risk of developing Alzheimer's disease, with an increase in prevalence as they reach age 50 and above (Hutchison, 1999). A number of studies have found that, while many people with Down syndrome aged over 40 show the neuropathological features of Alzheimer's disease, these are not always accompanied by signs of behavioural or clinical change (Devenny et al., 1996; Evenhuis, 1990). As a result, many authors have highlighted the need for prospective, longitudinal studies as an essential aid to understanding the patterns of cognitive and behavioural decline associated with the onset of Alzheimer's disease.

A general pattern has begun to emerge, with the early stages of Alzheimer's disease being characterized by difficulties in carrying out day -to- day tasks, memory problems and disorientation. In the later stages individuals may become incontinent, immobile, totally dependent on others for care and susceptible to epilepsy (Lishman, 1998; McCarron, 1999; McKenzie et al., 1998).Thus, while the initial sign of Alzheimer's disease in the general population is a change in cognitive skills, the first observable signs of a developing dementia in people with Down syndrome tends to be a reduction in adaptive skills. This change is also the most common factor triggering a referral to specialist health services (Janicki and Dalton, 1993).

For some clients with a more limited repertoire of skills, such changes may go unnoticed until a much later stage in the disease. Clinicians and researchers alike, therefore, continue to search for tools that will pick up the earliest indicators of the onset of the disease. This would facilitate early and appropriate service planning to meet the changing needs of the client, although Zimbleman and Wilson (2000) highlight some of the ethical issues related to direct client testing.

One indirect assessment measure of adaptive behaviour is the Vineland Adaptive Behaviour Scales (Sparrow et al., 1984).This assessment is reported to be able to assess the personal and coping skills of the individual from birth to adulthood and it has norms for people with a learning disability. It has two formats, a survey form with 297 items, which provides a general assessment of adaptive behaviour, and an expanded form. The latter tool has 577 items and provides a more detailed and comprehensive assessment. The Vineland scales are commonly used in learning disability services (Burt et al., 1998) and are reported to have good psychometric properties (Sparrow et al., 1984).The assessment has been used in a number of studies examining the relationship between Down syndrome and Alzheimer's disease (Aylward et al., 1995; Burt et al., 1992; Cooper, 1997; Crayton et al., 1998; Hon et al., 1998) and it has been recommended as an assessment tool in good practice guidelines developed by Aylward et al. (1995).

One potential difficulty with the use of the Vineland scales in identifying subtle changes in behaviour which may indicate the onset of Alzheimer's disease relates to the scoring method. The manual outlines a basal rule whereby, if a client achieves a maximum score on two consecutive item clusters, all items prior to them are also assigned maximum points. This

may result in an individual being given credit for some skills which he or she may not possess. Subsequent identified changes in skills may not be based on an accurate baseline assessment. Many of the previous studies highlighted above, which have used the Vineland scales as a means of determining behavioural change due to Alzheimer's disease, have not outlined the scoring method used. It must therefore be assumed that the assessment has been scored according to the basal rule outlined in the manual. This raises the possibility that the actual abilities of the client are not always being accurately measured.

The present study aimed to explore this possibility by comparing two methods of using the Vineland Adaptive Behaviour Scales with clients with Down syndrome who were at risk of developing Alzheimer's disease. The first method was to use the Vineland scales as the basis of a semi –structured interview during which all identified skills of the client were noted. The second method was to score the assessment according to the basal rule outlined above. Here a client achieving a maximum score on two consecutive item clusters would be given credit for all items preceding them, irrespective of whether they have the skill or not. It was hypothesized that, if the Vineland Adaptive Behaviour Scales scoring method was valid for the use of identifying behavioural decline due to Alzheimer's disease, then:

1. The group meeting the criteria for 'probable Alzheimer's disease' (Aylward et al., 1995), as identified by sustained behavioural decline reported by staff in relation to the client's actual skills (method 1),would show a decline in Vineland scores in that same area when the assessment was scored according to the manual (method 2).

2. The group who did not meet any of the criteria for Alzheimer's disease, i.e. no evidence of behavioural or cognitive decline as reported by staff (method 1), would show no change or an increase on their Vineland scores (method 2).

Method

Participants

Forty-four clients with Down syndrome 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, with some clients being followed up for up to 5 years. Twenty-three clients were found to show a sustained decline in at least one area of their adaptive behaviour over at least 1 year, as measured by the Vineland Adaptive Behaviour Scales. The mean age of this group was 52.4 years (SD = 7.14); 10 were male and 23 were female. Twenty-one showed no decline in adaptive skills; of these, eight were male and 13 were female. The mean age of this group was 43.5 years (SD = 6.13). All clients were offered a health screen to identify and treat any medical cause for the deterioration other than Alzheimer's disease.

Measures:

Adaptive behaviour

This was measured using the Vineland Adaptive Behaviour Scales (Sparrow et al., 1984) as outlined above. The assessment relies on carer reports of client functioning in the following domains: communication (receptive, expressive and written), daily living skills (personal, domestic and community skills), socialization (interpersonal relationships, leisure and coping skills), motor skills (gross and fine skills) and maladaptive behaviour (part 1, part 2 and total). The assessment was used in two ways:

• Method 1: as a semi-structured interview where the client's actual skills were identified.

• Method 2: using the basal rule as outlined in the scoring manual.

Procedure

Following a referral, information was obtained using the measure 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. Clients were assigned to either a 'deterioration' or a 'no deterioration' group, depending on the outcome of the Vineland Adaptive Behaviour Scales assessment (method 1). 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 (method 1) over at least a 2 year period (deteriorating group). Comparisons were then made between the following Vineland scores using method 2: 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

Deterioration group

Assessment method 1: Table 1 illustrates the areas of behavioural deterioration between baseline and 12 months and between 12 and 24 months for the 'deterioration' group identified using assessment method 1.

Assessment method 2: A series of Wilcoxon signed ranks tests was used to determine those domains for which a significant decline in scores was found for the 'deterioration' group using the Vineland manual scoring method (method 2).Table 2 illustrates the domains for which a significant reduction in scores was found. A significant increase in scores was also found for maladaptive scores (part 1) between baseline and 12 months ($Z = -2.434$, $p < 0.05$)

and between baseline and 24 months (Z = -1.811, p < 0.05), and for total maladaptive scores between 12 and 24 months (Z = -2.06, p < 0.05).

No deterioration group

A series of Wilcoxon signed ranks tests found only one significant difference in scores for the group who showed no behavioural deterioration, using scoring method 2.This was in the personal skills domain (Z = 1.769, p < 0.05). Scores were found to decrease between 12 months and 24 months on this domain.

Discussion

The present study aimed to determine if the use of the basal scoring rule outlined in the Vineland Adaptive Behaviour Scales manual (Sparrow et al., 1984) would invalidate the use of the tool as a measure of behavioural change in those clients with Down syndrome at risk of developing Alzheimer's disease. It was hypothesized that the measure, if valid, would pick up a significant reduction in scores for those clients showing cognitive and behavioural decline consistent with a diagnosis of 'probable Alzheimer's disease' in the same areas reported by staff when the tool was used to identify all of the client's skills. It was also hypothesized that the tool would show no change or an increase in scores for clients who showed no deterioration, according to staff reports of actual client skills.

In terms of staff reports, behavioural decline was consistently reported for over half of the clients in all areas with the exception of motor skills and community skills. McKenzie et al. (1998) found, for a group of clients with Down syndrome living in a hospital setting, that the initial phase of Alzheimer's disease was marked by some degree of disorientation, a decline in self-care skills, increased overall dependency on others and reduced communication and

interaction with others. In the later stages, there was increased disorientation, an almost total decline in self-care skills and passivity. The results of the present study would appear to be consistent with these findings and those of previous researchers (Rasmussen and Sobsey, 1994).

When the Vineland scales were scored according to the basal rule a number of domains also showed a significant decline. These included a consistent decline in receptive communication, community and personal skills between baseline and 24 months. A decline was also found in leisure skills between baseline and 12 months; however, this decline was not sustained at 24 months. Similarly, a decline was found in relation to domestic skills between 12 and 24 months but not between baseline and 12 or 24 months. This would initially suggest that the skills measured in this domain are not affected until the later stages of Alzheimer's disease. However, when examining staff reports according to method 1, 57 percent of the group were reported as showing some decline in this area between baseline and 12 month follow-up. The most common areas were reduced motivation to carry out domestic tasks, the need for increased prompting, and difficulty using the washing machine. This may suggest that scoring the Vineland according to the manual may not always be able to pick up more subtle changes in behaviour, such as degrees of change.

The study also found a significant increase in challenging behaviours, as measured by the 'maladaptive behaviour' domain scores. These increases were sustained from baseline to 24 months. The most common change was an increase in verbal and/or physical aggression. This is of concern, as previous research has indicated that aggression can lead to the breakdown

of family and community placements (Attwood and Joachim, 1994; Tausig, 1985).

The domains in which a significant reduction in scores was found were broadly consistent with those identified by staff when the Vineland was used purely as a semi-structured interview. However, as noted above, some areas were identified where changes were not readily picked up by scoring method 2.These included more subtle changes, for example where a client who had previously needed some prompting or assistance to complete a task now needed even more help. While these subtle changes in degree can easily be identified in staff reports, they may be missed when the scoring procedures are applied. One example was in relation to the communication domain. While a significant reduction in scores was found for receptive communication, this was not the case for the expressive communication domain. Despite this, eight clients (35 percent) were reported as having a reduced vocabulary, particularly in relation to asking questions. Such potentially important early signs may be missed if there is a reliance on scores alone. Similarly, in relation to socialization, while staff reported that five clients had increasing difficulty with their emotional control, e.g. increased

tearfulness or anger, no significant change in scores was found on the coping skills subdomain. These differences cannot be attributed to a lack of reliability in staff reports, as the same staff reports are simply being used in two different ways. This would suggest that, in some cases, using the basal scoring method for the Vineland may lead to subtle behavioural changes being missed.

Broadly, however, using the scoring method for the Vineland still identified significant reductions in scores for clients who met the criteria for 'probable Alzheimer's disease'.

However, it was also hypothesized that, for those clients who were not reported as deteriorating, scoring the Vineland would result in no significant changes or an increase in scores. In fact, a significant decrease was found for this group in respect of their personal skills, between 12 and 24 months. This result is puzzling, and may have a number of possible explanations.

It may be that the result is due to the basal scoring method used. For example, it would only require an individual to no longer be able to do one item within a cluster to shift the basal lower. This would mean that all items which were previously given maximum scores now had to be scored according to whether the individual actually had the skill in his or her repertoire or not. If maximum scores had been assigned to skills the client did not have, this would lead to a reduction in scores over time and an apparent decline in skills where none existed. In fact, while only one result was found to be significantly lower, in all but the written communication domain, overall scores were found to be lower for the non-deterioration group between baseline and 24 months.

Another alternative explanation is that the results are due to a natural fluctuation in the individual's skills. If this were the case, however, it would be expected that these fluctuations would be picked up using assessment method 1 where the client's actual skills are recorded, particularly as both methods of using the assessment tool are based on the same information from the same informant, but simply utilized in a different manner. A third possible explanation could be that the 'non-deterioration' group are in fact themselves undergoing the beginning of a dementing process, and the reduction in their Vineland scores reflects this

early process. This is unlikely, however, as again it would be expected that such deterioration would be apparent from staff reports of the client's actual skills.

In conclusion, one possible implication of the study is that researchers and clinicians need to be aware of the impact that scoring the Vineland according to the manual may have when the assessment is used to determine behavioural decline over time in adults with Down syndrome at risk of developing Alzheimer's disease. It may be more prudent to examine clients' actual skills. An alternative may be to score the Vineland without applying the basal rule and to note any subtle changes which are not directly covered by the questions, e.g. in relation to degree of change separately. Similarly, researchers may wish to be explicit about the way in which

they scored and utilized the information from the Vineland to help to build a clearer picture of whether changes are due to a dementing process or simply to the scoring method employed in the study.

References

Attwood, T. & Joachim, R. (1994) 'The Prevention and Management of Seriously Disruptive Behaviour in Australia', in N. Bouras (ed.) Mental Health in Mental Retardation: Recent Advances and Practices. Cambridge: Cambridge University Press.

Aylward, C. H. , Burt, D. B. , Thorpe, L . U., Lai, F. & Dalton, A. J. (1995) Diagnosis of Dementia in Individuals with Intellectual Disability. Washington, DC: American Association on Mental Retardation.

Burt, D. B. , Loveland, K. A. & Lewis, K. R. (1992) 'Depression and the Onset of Dementia in Adults with Mental Retardation', American Journal of Mental Retardation 96: 502-11.

Burt, D. B. , Loveland, K. A. , Primeaux-Hart, S . , Chen, Y., Phillips, N. B., Cleveland, L . A. , Lewis, K. R. , Lesser, J. & Cummings, E. (1998) 'Dementia in Adults with Down Syndrome: Diagnostic Challenges', American Journal on Mental Retardation 103 (2): 130-45.

Cooper, S. A. (1997) 'High Prevalence of Dementia among People with Learning Disabilities Not Attributable to Down's Syndrome', Psychological Medicine 27: 609-16.

Crayton, L . , Oliver, C. , Holland, A. , Bradbury, J. & Hall, S. (1998) 'The Neuropsychological Assessment of Age Related Cognitive Deficits in Adults with Down's Syndrome', Journal of Applied Research in Intellectual Disabilities 11 (3): 255-72.

Devenny, D. A. , S I Lverman, W. P. , Hill, A. L . , Jenkins, E . , Sersen, E . A. & Wisniewski, K. E. (1996) 'Normal Ageing in Adults with Down's Syndrome: A Longitudinal Study', Journal of Intellectual Disability Research 40 (3): 208-21.

Evenhuis, H. (1990) 'The Natural History of Dementia in Down Syndrome', Archives of Neurology 47: 263-7.

Hon, J. , Huppert, F. A. , Holland, A. J. & Watson, P. (1998) 'The Value of the Rivermead Behavioural Test (Children's Version) in an Epidemiological Study of Older Adults with Down Syndrome', British Journal of Clinical Psychology 37: 15–29.

Hutchinson, N. J. (1997) 'Association between Down's Syndrome and Alzheimer's Disease: Review of the Literature', Journal of Learning Disabilities for Nursing, Health and Social Care 3 (4): 194-203.

Janicki, M. P. & Dalton, A. J. (1993) 'Alzheimer's Disease in a Select Populationof Older Adults with Mental Retardation', The Irish Journal of Psychology 14 (1): 38-47.

Lishman, W. A. (1998) Organic Psychiatry: The Psychological Consequences of Cerebral Disorders, 3rd edn. London: Blackwell.

Mccarron, M. (1999) 'Some Issues in Caring for People with the Dual Diagnosis of Down's Syndrome and Alzheimer's Disease', Journal of Learning Disabilities for Nursing, Health and Social Care 3 (3): 123-9.

Mckenzie, K. , Murray, G. C., Mckenzie, S. & Muir, J. (1998) 'A Prospective, Longitudinal Study of Functional Decline in Individuals with Down Syndrome', Journal of Learning Disabilities for Nursing, Health and Social Care 2 (2): 98-104.

Mckenzie, K. , Paxton, D. , Matheson, E . , Patrick, S. , Hamilton, L. & Murray, G. C. (2000) 'Pathways to Success', Learning Disability Practice 3 (1): 16-19.

Rasmussen, D. E. & Sobsey, D. (1994) 'Age, Adaptive Behaviour and Alzheimer's Disease in Down Syndrome: Cross-Sectional and Longitudinal Analyses', American Journal on Mental Retardation 92 (2): 151-65.

Sparrow, S . S . , Balla, D. A. & Cicchetti, D. V. (1984) Vineland Adaptive Behaviour Scales: Interview Edition, Expanded Form. Circle Pines, MN:American Guidance Service.

Tausig, M. (1985) 'Factors in Family Decision Making about Placement for Developmentally Delayed Adults', American Journal of Mental Deficiency 89: 352-61.

Zimbleman, K. & Wilson, C. (2000) 'Baseline Evaluations of Intellectually Disabled Clients Who Are at Risk for Dementia', Clinical Psychology Forum 146: 26–9.

Table 1 **Areas of behavioural deterioration between baseline and 12 months and between 12 and 24 months for the 'deterioration' group identified using assessment method 1**

Area of decline	*Baseline to 12 months*			*12 to 24 months*		
	no.	*%*	**Examples**	*no*	*%*	**Examples**
Communication	14	61	Increased difficulty following instructions, needs increased prompting, reduced vocabulary; especially questions	16	70	Increased forgetfulness and confusion, clarity of speech deteriorating, writing deteriorating, increasingly repetitive, increasing difficulty following instructions,
Personal skills	13	57	Increased prompting and assistance with eating, bathing and dressing	14	61	Increased assistance and prompting required, less meticulous, increasing incontinence, slower in all tasks
Domestic skills	13	57	Increased prompting needed, difficulty using the washing machine, decreased motivation	13	57	Reduced motivation, no longer helps with certain tasks, unable to use washing machine, does not initiate domestic tasks
Community skills	10	43	Confusion about days of the week, needs more assistance to cross roads, confusion about daily routine	9	39	No road safety skills, confused over days and time, confusion about daily routine, less aware of danger within the home, unable to answer phone/take messages
Socialisation skills	14	61	Decrease in emotional control, decrease in hobbies/activities	16	70	Withdrawn, unwilling to join in, difficulty controlling emotions, reduced confidence, less polite and sensitive
Motor skills	6	26	Slowing down, increased loss of balance	8	35	Slowing down, less motivation to move about, increasingly unsteady, less able to walk long distances
Maladaptive behaviour	19	83	Erratic sleep pattern, increase in aggression, reduction in established challenging behaviour	14	61	Increased aggression, erratic sleep pattern, increased hoarding, talking to self, incontinence.

Table 2 Adaptive behaviour domains for which a significant reduction in scores was found

Domain	Baseline to 12 months		Baseline to 24 months		12 to 24 months	
	Z-score	Significance level	Z-score	Significance level	Z-score	Significance level
Receptive communication	-1.841	$p < 0.05$	-2.201	$p < 0.05$	-2.032	$p < 0.05$
Personal skills	-1.782	$p < 0.05$	-2.18	$p < 0.05$	-2.673	$p < 0.05$
Community skills	-2.207	$p < 0.05$	-2.375	$p < 0.05$	_	_
Domestic skills	_	_	_	_	-2.375	$p < 0.05$
Leisure skills	-1.753	$p < 0.05$	-	-	-	-