Neuropsychological Test Battery to Detect Dementia in Down Syndrome

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Abstract

There is no consensus among clinicians about what instruments best detect dementia in Down syndrome. The aim of this study was to evaluate the efficacy of various neuropsychological instruments for this purpose. The test battery consisted of: Information and Orientation Questions, Block Design Test, Fuld Object Memory Evaluation, Grocery List, Boston Naming Test, Peabody Picture Vocabulary Test - Revised, Test of Apraxia, and the Dementia Scale for Down Syndrome. The tests were administered to 35 people with Down syndrome to compare the group performance of older people with dementia (age 40-59) with older people without dementia (age 40-66) and younger people without dementia (age 28-39). Significant differences were found in performance on tests of information and orientation, immediate and delayed memory, and verbal learning. The two groups without dementia performed better on these tests than the group with dementia. This suggests that the Information and Orientation Questions and the Fuld Object Memory Evaluation are sensitive measures and should be included in neuropsychological test batteries assessing dementia in Down syndrome.

Dementia of the Alzheimer type (DAT) is the most common form of neurodegenerative disease affecting adults with Down syndrome (DS). The reported frequency of clinical features of (DAT) in DS varies somewhat, but has been reported to increase with age from 8% in those between 35 and 40 years (Lai & Williams, 1989; Wisniewski, Silverman & Wegiel 1994) to approximately 22% for those aged 40+, and approximately 60% in the 60+ age group (Janicki & Dalton, 2000). In individuals with mild-to-moderate levels of cognitive delay, the average age of onset of DAT is approximately 55 years of age. The pattern of cognitive decline related to DAT is similar in DS and the general population (Oliver, Crayton, Holland, Hall & Bradbury, 1998). Affected members in both populations exhibit changes in

orientation, learning and memory, confrontation naming, visuospatial skills, praxis, and skills of daily living.

The diagnosis of DAT in people with DS has not been easy. Clinicians often have difficulty distinguishing signs of DAT from age-associated cognitive decline or underlying cognitive deficits resulting from DS. The diagnosis of DAT in DS is further complicated by lack of premorbid (prior to apparent onset of dementia) baseline measures of cognitive functioning and age-appropriate test norms for adults with DS. In view of these difficulties, some clinicians (e.g., Deb & Braganza, 1999) have suggested that observer-rated scales, rather than neuropsychological instruments, may be more useful for the diagnosis of DAT in people with intellectual disabilities. Others (e.g., Burt & Aylward, 2000) have recommended specific neuropsychological tests to detect DAT in such adults, but have not provided data on how well these tests distinguish individuals with no signs of dementia from individuals who exhibit clinical signs of dementia.

The aim of the present study was to evaluate the diagnostic sensitivity of a neuropsychological test battery in detecting DAT in a DS population. The performances of older and younger people without dementia, and older people with dementia were compared on seven neuropsychological tests designed to assess memory, learning, orientation, aphasia, and apraxia. These domains of cognitive functioning are known to change with aging and dementia, both in people with DS and in members of the general population (Oliver et al., 1998).

Method

Participants and diagnostic procedures

The clinical records of thirty-five men and women with DS were selected from the Neuropsychological Baseline Assessment Service database compiled in a nonresidential treatment centre (Surrey Place Centre, Toronto) for people with developmental disability. All testing took place between 1995 and 2000 in the context of regular baseline and diagnostic assessment service. Anonymity and confidentiality were assured through coding clinical records. Written consent for using clinical data was obtained from each client or legal guardian.

The Dementia Scale for Down Syndrome (DSDS) (Gedye, 1995) was used to detect the presence or absence of dementia. The DSDS criteria for the clinical diagnosis of dementia are compatible with those of the Diagnostic and Statistical Manual of Mental Disorders, 4th edition (1994) (DSM-IV), although the DSM-IV does not provide information regarding dual diagnosis of developmental disability and dementia (for review see Aylward, Burt, Thorpe, Lai & Dalton, 1995). According to the DSM-IV, the diagnostic criteria for dementia requires documentation of cognitive

decline that causes impairment in social and occupational functioning. The DSDS is a 60-item caregiver instrument that inquires into cognitive and behavioural signs that are characteristic of "early," "middle," and "late" stages of dementia, and yields CCS (cognitive cutoff score) values specific for each stage. The DSDS also allows for a differential diagnosis of dementia with conditions that are common in DS, such as hearing and visual impairment, thyroid dysfunction, and depression. When compared with a clinical diagnosis of dementia of the Alzheimer type, the DSDS showed 58% sensitivity and 96% specificity at baseline. A two-year follow-up demonstrated 75% sensitivity and 96% specificity (Huxley, Prasher & Haque, 2000).

The 35 individuals with Down syndrome were administered the DSDS in the context of a regular clinical assessment service to determine whether or not they exhibited signs of DAT. Twelve of these (mean age=50.9, SD 5.6) were placed in a DAT group. The remaining 23 people were classified as showing no dementia. Because the age range here was wide, they were divided into two groups: nine younger individuals with no dementia (YND) (mean age=33.4, SD=3.4), and 14 older individuals with no dementia (OND) (mean age=46.2, SD=5.6). The efficacy of this grouping procedure was tested using one-way analysis of variance (ANOVA). A main effect for age was found ($F(2,32)=33.5, p<.05$), and post hoc comparisons indicated that the YND were significantly younger than both the OND and the DAT individuals, which did not differ in age.

To test for possible differences in cognitive functioning, the Peabody Picture Vocabulary Test-Revised (PPVT-R) (Dunn & Dunn, 1981) was administered. The PPVT-R is a measure of receptive vocabulary and is regarded as a good indicator of verbal intelligence (Das, Divis, Alexander, Parrila & Naglieri, 1995; Lezak, 1995). Scores were contrasted by one-way ANOVA and no group differences were found ($F(2,32)=2.69, p>.05$). This indicated that participants in all three groups fell within the same general range of verbal ability.

All individuals had a physical examination by their family physicians within six months prior to neuropsychological testing, which included a routine laboratory blood test to identify thyroid dysfunction, folate, and Vitamin B12 deficiency. At the time of testing, none of the participants had active major medical disorders (such as heart disease, infections, lung disease, liver disease, kidney disease, or diabetes), seizure disorder, uncorrected thyroid problem, or depression. All participants had hearing tests prior to the assessment.

Neuropsychological testing

In addition to the DSDS and the PPVT-R, the participants were administered the following battery of cognitive tests by a clinical psychologist or psychometrist:

1. Information and Orientation Questions (IO): Fourteen-item scale designed to assess the individual's ability to tell his/her name, age, date and place of birth, current address and telephone number, mother's name, current date, day of the week, season of the year, and orientation to time and place.

2. Block Design Test (BD) from WISC-R (Wechsler Intelligence Scale for Children-Revised) (Wechsler, 1974). This test of visuo-constructional praxis requires individuals to assemble red and white blocks to replicate the constructions made by the examiner or to reproduce designs printed in a smaller scale.

3. Fuld Object Memory Evaluation (FULD) (Fuld, 1978; 1980). The FULD measures immediate memory (IM) and delayed memory (DM) of objects. The test material consists of 10 common objects (ball, button, bottle, card, cup, key, matches, nail, ring, scissors) contained in a bag. Following tactile identification and visual processing of the objects, there is a 60-second distractor interval filled with a rapid semantic retrieval task (first names). Next, there is a recall trial and four additional learning trials separated by 30-second rapid semantic retrieval trials. After each recall trial, the subject is reminded of the omitted item(s). The number of items recalled on the fifth trial is a measure of verbal learning (VL). The final recall trial is administered after a 15-minute interval during which other tests are performed. The three-choice recognition part of the test was not administered because, in general, individuals with DS exhibit a strong preference for the last item.

4. Grocery List (GL). This 60-second test of category fluency requires rapid retrieval of items found in a supermarket or grocery store, in response to the examiner asking, "Tell me all the things you can buy in a grocery store."

5. Boston Naming Test (BNT) (Kaplan, Goodglass & Weintraub, 1983). This test of expressive vocabulary contains sixty line drawings of items ranging in familiarity from common objects such as a bed, to rare ones such as an abacus. Stimulus and/or phonetic cues are provided when subjects are unable to name an item spontaneously.

6. Test of Apraxia (PX). This 24-item test was designed to assess the ability to produce buccofacial, upper limb, lower limb, and whole body movements on command or by imitating the examiner.

Results

Multivariate Analysis of Variance was performed on scores of IO, BD, IM, DM, VL, GL,BNT, and PX. Significant between-subject effects ($F(16, 50) = 5.44, p < .001$) were found for the IO, IM, DM and VL variables.

Multiple comparison using the Bonferoni correction (a criterion of $p<.01$ to be significant) revealed that both the YND and OND groups exhibited better performance on tests of IO, IM, DM and VL than the DAT group. The performance of the YND group did not differ significantly from that of the OND group on these measures. However, mean scores for these variables were generally lower for the OND group than for the YND group. Results suggest an age-related decline in memory and verbal learning ability.

Likewise, there was a trend toward lower test performance on the BNT and BD for the OND and DAT groups compared to the YND group. Mean IO, BNT, GL, PX and BD scores for the three groups are depicted in Figure 1. Measures of IM, DM and VL are plotted in Figure 2. As shown in Figures 1 and 2, the difference between the DAT and OND mean scores were most noticeable on the IO test and measures derived from the FULD. Thus, the IO and the FULD appear to be the most sensitive tests in the battery, differentiating the DAT group from the group of older people without dementia.

Figure 1. Mean Information and Orientation (IO), Boston Naming Test (BNT), Category Fluency (GL), Praxis (PX) and Block Design (BD) scores.

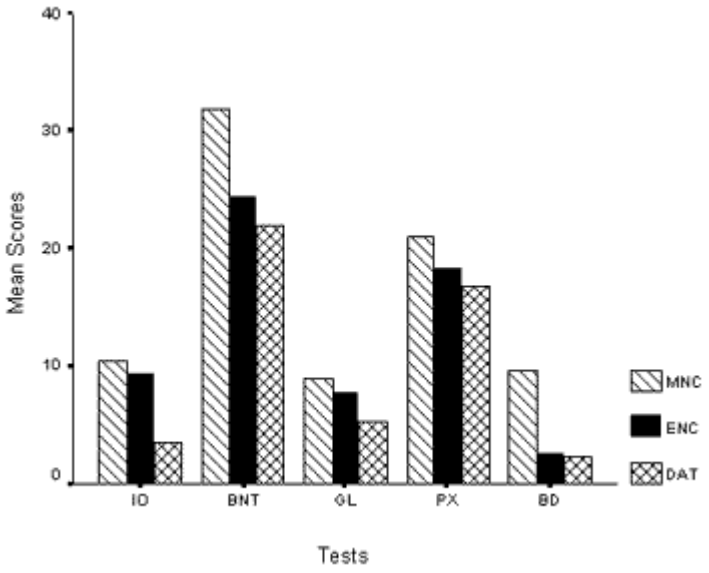
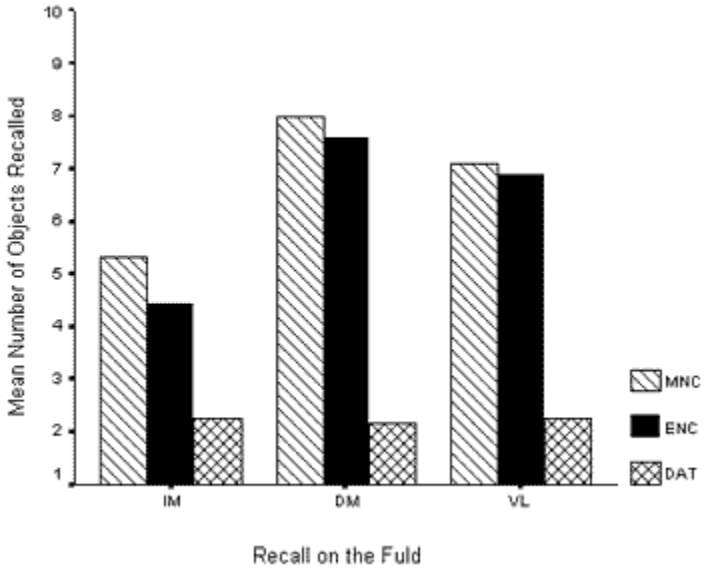


Figure 2. Mean scores of immediate (IM) and delayed recall (DM), and verbal learning (VL) on the Fuld Test for DAT, MNC, and ENC groups.



Discussion

Although various neuropsychological instruments have been recommended to assess dementia, there is no consensus among clinicians regarding the best tests for detecting dementia of the Alzheimer type in Down syndrome. This study was undertaken to evaluate the efficacy of a neuropscyhological test battery in distinguishing individuals with Down syndrome and dementia from similarly-aged and younger people with Down syndrome who showed no signs of dementia. Consistent with results from earlier studies (Brugg et al.,1994; Deveny et al., 1996; Oliver et al., 1998; Thase, Tigner, Smeltzer & Liss, 1984), we found age-related decline across all domains of cognitive functioning, as measured by the neuropsychological test battery. Congruent with published reports on aging and cognitive functioning in DS (e.g., Campbell-Taylor, 1993; Gibson, Groeneweg, Jerry & Harris, 1988), we found that scores on the BD and the BNT tests were most affected by aging. It is possible that lowered performance on these tests indicates very early stages of DAT, since constructional apraxia and anomia are known to be among the earliest signs of DAT in the general population. However, since our study shows

older people with and without dementia to have similar BNT and BD performance, these tests appear to have little diagnostic or practical value in the assessment and diagnosis of dementia in DS. It is possible, however, that these two groups may differ in their patterns of responding, although this needs to be tested in furutre research.

Findings of this study suggest that the most useful cognitive measures for differentiating older (40+) people with DS who have and do not have dementia are tests of information and orientation, verbal learning ability, and short and long-term verbal memory. Thus, the FULD and IO tests are the most valuable instruments for inclusion in diagnostic and cognitive baseline batteries for DS. It is reasonable to assume that these tests will be useful for detecting longitudinal changes that occur in the course of dementia. However, practice effects may confound the results of repeated assessment with the same instrument, particularly when the instrument has no, or only one, alternate version.

A limitation of the present study was that it did not include individuals functioning below the moderate level of cognitive disability. People functioning within the severe range of cognitive disability typically cannot be assessed using standardized neuropsychological instruments due to their limited comprehension and limited verbal abilties. One important area of future research is designing and evaluating instruments suitable for diagnosing clinical features of dementia in this population.

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