Ageing in adults with Down's syndrome in institutionally based and community-based residences

J. M. Roeden¹ & F. G. Zitman²

¹ Sterrenberg, Institute for Mentally Retarded People, Huis ter Heide
² Department of Psychiatry, University of Nijmegen, Nijmegen, the Netherlands

Abstract

Seventy-one subjects with Down's syndrome (DS), between the ages of 29 and 68 years, and 46 matched controls (without DS) were examined for intelligence, memory (short- and long-term memory, and spatial and temporal orientation), communication (receptive, expressive and written language) and daily living skills (personal, domestic and community daily living skills). All subjects were screened on hearing and visual functions, thyroid functions, depression and dementia. DS-subjects and controls were matched on chronological age, mental age, living conditions and male/female ratio. Comparisons were made between five subgroups (1) non-demented institutionalized subjects with DS (DSi-group; n=35); (2) non-demented institutionalized controls without DS (Ci-group; n=22); (3) demented institutionalized subjects with DS (n=10); (4) non-demented subjects with DS living in group homes (DSg; n=26); and (5) non-demented controls without DS living in group homes (Cg; n=24). Institutionalized and non-institutionalized subjects, as well as demented and non-demented subjects differed significantly on all functions measured. Multiple regression analysis was performed to examine the influence of age and sensory deficits on adaptive and cognitive functioning. In DSg subjects, significant associations were found between age and mental age, and between age and performances on written language. In (non-demented) DSi subjects, significant relations were found between age and memory functions observed in daily circumstances. Moreover, in the (non-demented) DSi elderly, visual impairment was significantly related to depressed performance on daily living skills. No age effects were seen in control subjects. Infirmities of old age like dementia and sensory deficits were far more common in people with DS than in controls. Psychiatric and diagnostic aspects of clinical depression and dementia were emphasized in particular.

Introduction

A review of all of the neuropathologic studies of Alzheimer disease (AD) in Down's syndrome (DS) (Mann 1988a) has indicated that nearly 100% of DS cases that come to autopsy after the age of 40 years show senile plaques and neurofibrillary tangles. The form and distribution of senile plaques and tangles qualitatively and quantitatively meet the criteria recommended for a positive diagnosis of AD (Mann 1988b). In spite of the Alzheimer-type neuropathological changes, the major clinical sign of dementia of AD, progressive dementia, is not commonly observed in most mature and elderly people with DS. The available literature suggests...
that between 15 and 30% of people with DS develop dementia (Wisniewski & Rabe 1986), a percentage which is far below the near 100% presence of AD neuropathology in people with DS over 40.

Because of the high risk of developing AD, the process of ageing in DS is of special interest. This subject has been addressed in a number of studies. Comparing patients with DS below and above age 35 in a cross-sectional study, Wisniewski et al. (1978) found a higher prevalence of recent memory loss, impairment of short-term visual retention and neurological symptoms in older subjects. Because a control group is lacking, it was unclear whether these findings are specific for patients with DS.

Several research groups did cross-sectional studies with control groups. Thase et al. (1982, 1984) found lower scores for orientation, digit span, visual memory, object naming and general knowledge in the DS group. Zigman et al. (1987) found a decline in adaptive competence with increasing age. In both studies, the most evident impairment was found in DS subjects above age 50. A third research group (Silverstein et al. 1986, 1988) found a drop in the scores on the Motor Development Factor on the Client Development Evaluation Report specifically in DS subjects. In all three studies, demented and non-demented DS-subjects were grouped together. Therefore, the results of these studies may reflect the greater number of demented DS-subjects in older age groups. Haxby (1989) studied demented and non-demented DS subjects separately. The latter group showed an age-related decrease in the ability to form new long-term memories and in visuospatial construction. In a later paper, Haxby & Schapiro (1992) also demonstrated the decline in the non-demented group in a longitudinal study. However, in both studies, a control group is lacking. Besides, it is not clear whether Haxby & Schapiro (1992) tried to differentiate between dementia and depression. In a series of prospective studies, Dalton & Crapper (1984) found a higher incidence and prevalence of memory deterioration in adults with DS over the age of 40 years than in age-matched mentally retarded controls. In their studies, Dalton & Crapper (1984) only made use of selected research populations, excluding persons with gross sensory deficits, for instance.

All the studies discussed here did not take into account several possibly confounding factors. Many older DS subjects develop substantial visual and hearing defects which negatively influence the performance on cognitive and other tests. This factor was not analysed in any of the studies.

The studies mentioned above have selected subjects from institutionalized populations or from a mixture of populations from institutionally based and community-based residential settings. Generalizing findings from these studies to all people with DS, regardless of their level of mental retardation and/or their living conditions, is questionable.

In an attempt to overcome most of these shortcomings, the present authors have carried out a cross-sectional study in which five groups are compared: non-demented DS subjects living in group homes, non-demented DS subjects who are institutionalized, and two control groups matched for age, mental age and male/female ratio with the first and second DS-subjects group, respectively; the fifth group consisted of demented DS-subjects. The groups were compared on intelligence, memory, communication and daily living skills. Data on visual and auditory handicaps were gathered from all the subjects. The subjects were also screened on depression.

**Materials and methods**

**Subjects**

The subjects were 71 adults with karyotypically verified Down's syndrome between the ages of 29 and 68 years. Forty-five subjects of the DS population are residents of an institute for mentally retarded adults (DSi group) and 26 subjects are inhabitants of group homes (DSg group). Dementia was judged to be present in 10 DSi subjects. Dementia was not present in DSg subjects.

Controls were mentally retarded subjects without DS. There were two control groups. One control group (Ci; n=22) had IQ, age and living conditions similar to the non-demented DSi group. The other control group (Cg; n=24) had IQ, age and living conditions similar to the DSg group. Since it is hard to find sufficient numbers of Ci-subjects with (presenile) dementia, no control group was formed for the demented DSi subjects. Permission for this
study was obtained from the family and an institutional ethical commission.

Instruments

Tests, interviews and behaviour rating scales were administered to assess cognitive and adaptive functioning. A standardized clinical interview was used to diagnose dementia and depression.

Mental age

Mental age was assessed using the Vineland Adaptive Behaviour Scales (VABS, interview version; Sparrow et al. 1984). The interview version of this instrument is a semi-structured informant-based questionnaire. Respondents are care-givers who are familiar with the adaptive behaviour of the individual being evaluated. Mental age based on the adaptive level of functioning can be seen as the best estimate of overall intelligence in severely and profoundly mentally retarded people (Maloney & Ward 1979). Raw scores on six subscales (receptive, expressive and written language, and personal, domestic and community daily living skills) were transposed into mental ages. The median of the subscale mental ages was considered to be an index of mental age.

When there were sufficient conditions for testing (no severe visual loss, mental ages over 2.5 years and sufficient cooperation), the Snijders-Oomen Nonverbal Intelligence Test; 2.5–7 years (SON; Snijders & Snijders-Oomen 1975) was also used. When possible, it is better to use mental age scores based on conventional tests than mental age scores based on the adaptive level of functioning. An extra advantage of the SON-test is its suitability for deaf or hearing-impaired people and for people with speech impediments. The test consists of five subtests (sorting, mosaic, combination, memory and copying), each one providing a separate mental age. The median of the subscale mental ages was considered to be an overall index of mental age.

Memory

Memory functions were determined using the Dementia Questionnaire for Mentally Retarded People (DMR; Evenhuis et al. 1990). The questionnaire is a standardized interview to be completed by caregivers. Scores of the first three subscales, short- and long-term memory and orientation, as well as the Sum of Cognitive Scores (SCS: sum of the first three subscale raw scores) were recorded. Higher scores correspond to more severe deterioration.

Communication

Communication levels were determined using the Vineland Adaptive Behaviour Scales (VABS). Separate scores on three single subscales (receptive language: ‘what the individual understands’; expressive language: ‘what the individual says’; and written language: ‘what the individual reads and writes’) were recorded. The total score of three subscales was used as an overall index of communication.

Daily living skills

Adaptive levels for daily living skills were measured using the Vineland Adaptive Behaviour Scales (VABS). Separate scores were recorded on three single subscales: personal living skills (‘how the individual eats, dresses and practices personal hygiene’); domestic living skills (‘what household tasks the individual performs’); and community living skills (‘how the individual uses time, money, telephone and job skills’). Total score of three subscales was used as an overall index of daily living skills.

Diagnosis of depression and dementia

Specific standardized diagnostic methods for depression and dementia in mentally retarded people in the Netherlands are not available. Therefore, a standardized informant-based clinical interview was developed. At the moment, this interview is still in a phase of evaluation. The interview was administered by a psychologist who is experienced in interviewing techniques and in the interpretation of mental disorders. Respondents were care-givers who were familiar with the behaviour of the individual being evaluated.
The questions in the first part of the interview reflect the diagnostic criteria for ‘major depression for use with mentally retarded people’ [modified DSM-III criteria (APA, 1980) by Sovner (1986)]:

(1) a disturbance of mood characterized by sadness, withdrawal or agitation.

(2) any four of the following nine symptoms: (a) change in sleep; (b) change in appetite and/or weight; (c) onset of or increase in severity of self-injurious behaviour; (d) apathy; (e) psychomotor retardation; (f) loss of activity of daily living skills; (g) catatonic stupor and/or rigidity; (h) spontaneous crying; and (i) fearfulness. The diagnosis ‘minor depression’ is made if one of the symptoms is either sadness, withdrawal or agitation, and if at least two of the nine symptoms in (2) are met.

Sovner (1986) reasoned that the original DSM-III criteria have to be modified because the confluence of intellectual distortion, psychosocial masking, cognitive disintegration and baseline exaggeration makes it difficult to use the operational criteria in their current form.

The questions in the second part of the interview reflect the diagnostic criteria for ‘dementia for use among mentally retarded adults’ [modified DSM-III-R criteria by Evenhuis (1992)]:

(1) demonstrable evidence of decline of original level of short- and long-term memory (observed in daily circumstances).

(2) at least one of the following (observed in daily circumstances): 1. disturbance of original level of spatial and temporal orientation, 2. aphasia, 3. apraxia, 4. personality change.

(3) the disturbance in A and B significantly interferes with work or usual social activities or relationships with others.

(4) not occurring exclusively during the course of delirium.

A diagnosis of dementia was made when a permanent and progressive decline of cognitive and social functioning over several years of observation was observed, according to the modified DSM-III-R criteria. Previous and current levels of functioning were available for all subjects. In this study, mean time elapsed since the last intellectual, mnestic and adaptive assessment was 3 years and 8 months.

Medical examination

Each person received pure tone, speech or brainstem electric response audiometry, or was assessed by the Stycar Hearing Test (Sheridan 1976a). The latter test was used when no specific audiological assessments were available. The test assesses the ability to hear under everyday life circumstances. Procedures varied depending upon the subjects level of functioning, cooperation and concentration. A hearing handicap in this paper is defined as an insufficiently corrected hearing loss of over 30 dB.

Visual impairments were derived from medical chart, using ophthalmological and orthoptist information. When no specialist information was available, the Stycar Vision Test (Sheridan 1976b) was used to assess visual acuity. A visual handicap in this paper is defined as an insufficiently corrected visual acuity bilaterally less than 0.1. All the subjects in this study diagnosed as hypothyroid received thyroid hormone therapy.

Statistical procedures

The Scheffé multiple comparison procedure was used for determining the differences between population means of DSg subjects, DSi subjects without dementia and DSi subjects with dementia, and between Cg subjects and Ci subjects. The Scheffe method is conservative for pairwise comparisons of means. It requires larger differences between means for significance than most of the other methods.

Multiple regression analysis was performed to give insight into the association between age, auditory handicaps and visual handicaps on one side (the independent variables), and intellectual, mnestic, adaptive and communicative functioning on the other (the dependant variables). It is hypothesized that age and the infirmities of old age (auditory and visual handicaps) will have significant impact on the level of functioning of the subjects being investigated. Critical values of \( P < 0.05 \) were used for
Table 1 Prevalence of hearing and visual loss in Down's syndrome subjects and control subjects*

<table>
<thead>
<tr>
<th></th>
<th>DSi</th>
<th>CI</th>
<th>DSg</th>
<th>Cg</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Hearing loss</strong></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>n</td>
<td>30</td>
<td>22</td>
<td>26</td>
<td>24</td>
</tr>
<tr>
<td>%</td>
<td>36</td>
<td>9</td>
<td>19</td>
<td>8</td>
</tr>
<tr>
<td><strong>Visual loss</strong></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>n</td>
<td>29</td>
<td>22</td>
<td>26</td>
<td>24</td>
</tr>
<tr>
<td>%</td>
<td>23</td>
<td>0</td>
<td>11</td>
<td>0</td>
</tr>
</tbody>
</table>

*DSi, subjects with Down's syndrome living in an institute; DSg, subjects with Down's syndrome living in group homes; CI, controls living in an institute; Cg, controls living in group homes; n, number of subjects investigated; %, prevalence of hearing or visual loss.

comparisons utilizing the Scheffé method or multiple regression analysis.

Results

The matching procedure

All subjects of the group homes were tested on the SON-test. In only 47% of the institutionalized subjects, the conditions were good enough to administer a complete SON-test. Since scores on the VABS were available for all institutionalized subjects the medians of the subscale mental ages were considered as the best estimate for mental functioning in these subjects.

Statistical comparisons of means of mental age chronological age and male/female ratio of non-demented DSi and CI subjects on the one hand and of DSg and Cg subjects on the other hand indicate that the matching procedure had been successful. The mean mental age of DSi and CI subjects was 36.4 (±11.2) and 34.2 (±11.6) months respectively, the mean chronological age of DSi and CI was 53.8 (±7.6) and 51.0 (±8.4) years, respectively and the male/female ratio in the DSi subgroup was 14/21 compared to 9/13 in the CI subgroup. In group homes, the mean mental age of DSg and Cg subjects was 61.6 (±12.2) and 61.6 (±15.4) months, respectively, the mean chronological age was 40.5 (±8.3) and 42.7 (±7.6) years, respectively, and the male/female ratio in the DSg subgroup was 12/14 compared to 12/12 in the Cg subgroup.

Medical examination

Hearing and visual functions could not be assessed reliably in 14 and 17% of the non-demented DSi subjects, respectively. In the remainder of the research population, the prevalence of insufficiently corrected hearing loss (loss bilaterally of over 30 dB) and insufficiently corrected visual loss (visual acuity bilaterally <0.1) was relatively high in the DS subgroups compared to the control subgroups (Table 1).

Since a large majority of DS subjects with a hearing or visual handicap were over 50 years old (79 and 82% respectively), these handicaps can be considered as infirmities of old age. Twelve people (17%) with DS had hypothyroidism. All subjects with hypothyroidism were treated and determined to be euthyroid by blood tests.

Differences between groups

The evaluation of scores on mental age, memory, communication and daily living skills by means of multiple comparison analysis revealed significant differences between institutionally and community-based residents on the one hand, and between demented and non-demented institutionalized subjects on the other. The results of the comparisons are summarized in Table 2.

Significant differences on all measures demonstrate that institutionally- and community-based residents and demented and non-demented subjects have to be considered as separate subgroups with divergent levels of functioning. The objective for further analysis is to consider to what degree ageing, hearing or visual deficiencies influence cognitive and adaptive processes in the subgroups (DSg, DSi-non-demented, DSi-demented, Cg and CI).

Regression analysis

Separate regression analysis for DS subjects and controls supported the hypothesis that the effects of age
Table 2: Mean scores on mental age, memory, communication and daily living skills for people with Down's syndrome and controls.

<table>
<thead>
<tr>
<th>Scores</th>
<th>DSg (n=26)</th>
<th>DSI (n=35) (non-demented)</th>
<th>DSI (n=10) (demented)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Mental age¹</td>
<td>61.6±12.3³</td>
<td>36.4±11.2**</td>
<td>11.8±9.1</td>
</tr>
<tr>
<td>Memory²</td>
<td>9.5±6.9*</td>
<td>20.2±11.1**</td>
<td>39.7±3.7</td>
</tr>
<tr>
<td>Communication²</td>
<td>154.8±46.8*</td>
<td>109.8±42.6**</td>
<td>33.1±29.1</td>
</tr>
<tr>
<td>Daily living skills²</td>
<td>224.2±19.8*</td>
<td>137.5±49.9**</td>
<td>32.0±34.5</td>
</tr>
<tr>
<td>Cg (n=24)</td>
<td></td>
<td>Cl (n=22)</td>
<td></td>
</tr>
<tr>
<td>Mental age¹</td>
<td>61.6±15.4***</td>
<td>34.2±11.6</td>
<td></td>
</tr>
<tr>
<td>Memory²</td>
<td>8.9±7.7***</td>
<td>22.4±10.6</td>
<td></td>
</tr>
<tr>
<td>Communication²</td>
<td>159.0±54.8***</td>
<td>104.2±38.9</td>
<td></td>
</tr>
<tr>
<td>Daily living skills²</td>
<td>243.6±48.6***</td>
<td>117.8±55.7</td>
<td></td>
</tr>
</tbody>
</table>

DSi, DSg, Cl, Cg: for abbreviations, see Table 1.
¹Mental age in months. ²Scores are raw scores. ³Values are means±standard deviations.
*Differs significantly (P<0.05) from DSi non-demented subjects.
**Differs significantly (P<0.05) from DSi demented subjects.
***Differs significantly (P<0.05) from Cl non-demented subjects.

Table 3: Influences on intellectual, mnestic and adaptive functioning based on results of multiple regression analysis for people with Down's syndrome living in group homes.

<table>
<thead>
<tr>
<th></th>
<th>Mental age</th>
<th>Memory</th>
<th>Communication</th>
<th>Daily living skills</th>
</tr>
</thead>
<tbody>
<tr>
<td>β</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Age</td>
<td>-0.55</td>
<td>0.01*</td>
<td>-0.33</td>
<td>-0.37</td>
</tr>
<tr>
<td>Visual handicap (±)</td>
<td>-0.16</td>
<td>0.37</td>
<td>-0.14</td>
<td>-0.10</td>
</tr>
<tr>
<td>Auditory handicap (±)</td>
<td>-0.14</td>
<td>0.98</td>
<td>-0.13</td>
<td>0.04</td>
</tr>
</tbody>
</table>

The column labelled β contains standardized regression coefficients; the column labelled P contains the observed significant level. *Significant.

and age-related infirmities differ significantly for DS and non-DS groups. Tests of regression analysis were significant only for the DS groups. The results of regression analysis for DS-groups are summarized in Tables 3 and 4.

In DSg subjects, a significant relation existed between advanced age and lower mental age score. Analysis of subtest scores revealed that the association was caused by the performance on three specific subtests: copying (β=-0.36; P<0.05), sorting (β=-0.45; P<0.05) and memory (β=-0.33; P<0.05). In the first test, a geometric design had to be copied in the second test, representations had to be categorized logically, and in the third test, hidden objects had to be found.

In DSg subjects, no significant relations were found between age visual and auditory handicaps on the one hand, and memory communication and daily living skills on the other. The only notable exception was a significant relation between age and depressed performance on the written subscale of the VABS communication scale (β=-0.47; P<0.05).

In Cg subjects, no significant relations were found.

In DSi (non-demented) subjects, a significant relation was noted between memory disturbances and advanced age. Analysis of subscales demonstrated significant correlations between age and short-term memory (β=0.41; P<0.05) and orientation (β=0.54; P<0.05). The correlation coefficients are positive because higher memory scores correspond to more severe deterioration.

In addition, a significant relation existed between visual loss and performance on daily living skills. On the subscale level DSi people with a visual handicap

Table 4 Influences on intellectual, mnestic and adaptive functioning based on results of multiple regression analysis for institutionalized non-demented people with Down's syndrome

<table>
<thead>
<tr>
<th></th>
<th>Mental age</th>
<th></th>
<th>Memory</th>
<th></th>
<th>Communication</th>
<th></th>
<th>Daily living skills</th>
<th></th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>$\beta$</td>
<td>$P$</td>
<td>$\beta$</td>
<td>$P$</td>
<td>$\beta$</td>
<td>$P$</td>
<td>$\beta$</td>
<td>$P$</td>
</tr>
<tr>
<td>Age</td>
<td>-0.32</td>
<td>0.12</td>
<td>0.48</td>
<td>0.02*</td>
<td>0.02</td>
<td>0.94</td>
<td>-0.20</td>
<td>0.35</td>
</tr>
<tr>
<td>Visual handicap (±)</td>
<td>0.05</td>
<td>0.81</td>
<td>0.12</td>
<td>0.10</td>
<td>0.30</td>
<td>0.20</td>
<td>-0.73</td>
<td>0.00*</td>
</tr>
<tr>
<td>Auditory handicap (±)</td>
<td>-0.29</td>
<td>0.17</td>
<td>-0.03</td>
<td>0.88</td>
<td>0.12</td>
<td>0.63</td>
<td>-0.32</td>
<td>0.09</td>
</tr>
</tbody>
</table>

The column labelled $\beta$ contains standardized regression coefficients; the column labelled $P$ contains the observed significant level. *Significant.

had lower personal skills ($\beta=-0.38; P<0.05$) domestic skills ($\beta=-0.23; P<0.05$) and community skills ($\beta=-0.24; P<0.05$).

In Ci subjects, no significant relations were found.

The analysis of scores of demented DSi persons revealed no significant correlations between age and scores on mental age, memory, communication or daily living skills. No reliable data on visual and auditory deficits were available for most of the demented patients so no multiple regression analysis was performed.

Dementia: clinical symptoms

Based on care-giver report test report and medical chart, 10 individuals met the criteria for a diagnosis of dementia. All clinically demented subjects had apparent declines in intellectual functioning (decline of mental age $>6$ months) mnestic functioning (increase of SCS-scores $>20$ points) and adaptive functioning (decline of VABS daily living skills-scores $>20$ points). All demented individuals were institutionalized residents with DS.

Some clinical symptoms and features of demented subjects were: epileptic seizures (eight out of 10 patients) bedridden or wheelchair-bound (seven out of 10 patients), and incontinence (six out of 10 patients). All demented subjects were over 50 years old. (The data reported reflect the most recent observation.)

Notable changes in behaviour were seen in eight out of 10 demented individuals at the onset of the deterioration process. In three cases, these changes met the criteria of a 'major depressive episode'. In two cases, the changes met the criteria of a 'minor depressive episode'. A period characterized by 'acting-out' behaviour (agression, restlessness, irritation and destruction) and symptoms of fear was seen in the remaining three individuals.

Depression: clinical symptoms

Two institutionalized non-demented individuals with DS and two inhabitants of group homes met the criteria of depression. Two individuals suffered a major depressive episode and two a minor depressive episode. Only one person without DS suffered an episode of minor depression. In demented subjects, episodes of depression were diagnosed in five cases (see previous section). Case studies revealed that behavioural components of the illness, like disturbance of sleep or appetite, social withdrawal and loss of interest in normal activities, psychomotor retardation, apathy, weeping or crying, and loss of daily living skills were common (each symptom in over half of the cases). Probably because of the reduced cognitive abilities as well as the reduced ability to verbally express emotions, complaints of depressive feelings, hopelessness, pessimism and guilt were not seen. Atypical features like attention seeking, aggressive acting-out behaviour, negativism and somatic complaints occurred frequently.

Discussion

The most important findings in the present study are that, in DSg subjects, significant associations existed between age and mental age and between age and performances on written language. In DSi subjects, significant relations were found between age and memory and orientation observed in daily
circumstances. Since no age-effects were seen in control subjects, the relatively poor performance of the older DS subjects can most likely be attributed to a syndrome-related process. These results resemble those of earlier studies (Thase et al. 1982, 1984; Silverstein et al. 1986, 1988; Zigman et al. 1987) which have various methodological shortcomings. In the study presented here, many of the these shortcomings were overcome by differentiating between institutionalized and non-institutionalized subjects, by including control groups, by studying demented subjects as a separate group, and by taking visual and hearing impairments into account. Besides, the present authors studied a broader range of aspects of intelligence, memory, communication and daily living skills. So, it can be concluded that the present study makes the data in the effects of ageing in DS more solid.

This study demonstrates that not only AD contributes to cognitive and adaptive decline in DS. Sensory deficits, which were far more common in DS subjects compared to controls, also contribute to the decline. The study also demonstrates that, in order to study the influence of ageing, different parameters must be used for institutionalized and for non-institutionalized subjects. For instance, in DSI residents, reduced performance was for the greater part easily observable in daily living activities like eating, washing, knowing names and recognizing people. In DSg subjects, the age-effects were more subtle. They did not have appreciable impact on the ability to meet the demands of everyday life. For some mental functions, only specific cognitive tests (some of the SON-subtests) and specific writing tasks (VABS) revealed age-related dysfunctions.

Notwithstanding the methodological advantages mentioned above, the present study is limited by the use of a cross-sectional design. While this design provides an accurate assessment of current functioning, cross-sectional age differences may not reflect reductions in mental ability. Sample selection bias, survivor effects and cohort effects can influence cross-sectional differences. For example, improvements in education of people with DS may explain why the younger adults do better on tests and ratings. Longitudinal studies are preferred, particularly since deterioration from a previous higher level of functioning is required for clinical diagnosis of dementia.

Moreover, like similar studies, this study was hampered by the paucity of appropriate diagnostic tools. Although the instruments chosen for this study were promising as valuable indicators of age-related dysfunction, they were not fully satisfactory for the hard-to-test DS population. For instance, the SON-test showed pronounced ‘floor effects’ which makes this test useless for severely handicapped individuals and those who are in the late stages of dementia. Other instruments employed (DMR and the clinical interview for depression and dementia) are still in a phase of evaluation, so at the moment, their psychometric properties are unknown.

Some clinical findings in this study are of diagnostic importance. Generalized tonic-clonic epileptic seizures developed in eight out of 10 DS individuals who had dementia. This finding is similar to prevalences of epilepsy found in other clinical studies: 84% (Lai & Williams 1989) and 78% (Evenhuis 1990). Therefore, the onset of epileptic seizures in adults with DS is strongly suggestive of dementia.

Clinical depression was diagnosed in four non-demented people with DS. The differential diagnosis of dementia must include depression. Depressed adults with DS may have clinical symptoms which can be mistaken for dementia, but which respond to pharmacological treatment. In addition, depression and other emotional symptoms occurred in the early stages of dementia in eight out of 10 people with DS. Although it remains unclear at this point to what extent affective disorder is associated with early dementia, the frequent occurrence of depressive symptoms during the onset of dementia is notable. The present authors suggest that, in clinical practice, the differential diagnosis is made as a result of the clinical history, time factor and the response to antidepressants.

References


Received 22 October 1993; revised 29 July 1994