

Executive function in adolescents with Down Syndrome

S. Lanfranchi,¹ O. Jerman,² E. Dal Pont,¹ A. Alberti¹ & R. Vianello¹

¹ Department of Developmental and Socialization, University of Padova, Padova, Italy

² Frostig Center, Pasadena, USA

Abstract

Background The present work is aimed at analysing executive function (EF) in adolescents with Down Syndrome (DS). So far, EF has been analysed mainly in adults with DS, showing a pattern of impairment. However, less is known about children and adolescents with this syndrome. Studying adolescents with DS might help us better understand whether performances on EF tasks of individuals with DS are determined by age or by Alzheimer disease, as some studies suggest, or whether their performances are directly related to DS cognitive profile.

Method A battery of EF tasks assessing set shifting, planning/problem-solving, working memory, inhibition/perseveration and fluency, as well as a tasks assessing sustained attention has been administered to a group of 15 adolescents with DS and 15 typically developing children matched for mental age. All EF tasks were selected from previous studies with individuals with intellectual disabilities or from developmental literature and are thought to be useful for the samples considered.

Results The present results revealed that the group of individuals with DS performed at a significantly lower level on tasks assessing set shifting, planning/

problem-solving, working memory and inhibition/perseveration, but not on the tasks assessing fluency. In addition, individuals with DS demonstrated a greater number of errors and less strategy use for the sustained attention task.

Conclusions The results suggest a broad impairment in EF in adolescents with DS, and are consistent with several similar studies conducted with adults with DS. We assume that EF deficit is a characteristic of DS.

Keywords Down Syndrome, executive function, working memory

Introduction

The concept of executive function (EF) refers to a set of interrelated abilities that are thought to be associated with the activity in the frontal parts of the brain, although it is also recognised that a number of different brain areas may be involved (Cummings 1993). EF abilities are described as higher-order control processes that subserve problem-solving, concept formation, task switching, inhibition, initiation of rapid and fluent responses, and planning (Stuss & Benson 1986; Shallice 1988; Rabbitt 1997). On the one hand, EF processes can be considered something unitary, as all of them can be attributed to the activity of frontal lobes; on the other hand, there is also some evidence for the

Correspondence: Phd Silvia Lanfranchi, Dipartimento di Psicologia dello Sviluppo e della Socializzazione, Via Venezia 8, 35131 Padova, Italy (e-mail: silvia.lanfranchi@unipd.it).

non-unitary nature of EF. Clinical observations, as well as studies of individual differences, found some dissociations in performance among the executive tasks (e.g. Miyake *et al.* 2000). Based on clinical observations, for example, some patients fail on the Wisconsin Card Sorting Task, but not on the Tower of Hanoi task (both considered to be executive tasks), whereas other individuals may show the opposite pattern, suggesting that EF may not be completely unitary (e.g. Shallice 1988; Godefroy *et al.* 1999). Moreover, studies on individual differences showed that, although EFs, such as shifting, updating and inhibition, are moderately correlated with one another, they are clearly separable (e.g. Miyake *et al.* 2000).

An important aspect of EF is working memory, which is the limited capacity memory system responsible for the temporary storage and processing of information while cognitive tasks are being carried out. One of the most prominent cognitive frameworks that has been associated with the study of working memory is Baddeley's (1986) multicomponent model. This model includes three components, two of which are specialised: one for the maintenance of speech-based, phonological information (the phonological loop), and the other for the visual and spatial information (the visuo-spatial sketchpad). In addition to these two 'slave' systems, the model also includes a central control structure called the central executive, which is considered to be responsible for the control and regulation of cognitive processes and is often linked to the functioning of the frontal lobes. Therefore, the activity of central executive can be considered as a link between working memory and inhibition, shifting, and planning (e.g. Miyake *et al.* 2000).

It has been demonstrated that EF is important early in development, and the first signs of its functioning are already observed in very young children (Welsh & Pennington 1988). However, the full mastery of EF may not be complete until adolescence or early adulthood, when the frontal areas of the brain achieve their maturation (Yakovlev & Lecours 1967; Tatcher 1991).

Deficits on EF tasks have been reported in a number of developmental disorders, such as attention deficit and hyperactive disorder, autism, Tourette Syndrome, conduct disorder, Turner's Syndrome, Klinefelter Syndrome and Fragile X

Syndrome (Pennington & Ozonoff 1996; Temple *et al.* 1996; Temple & Martin Sanfilippo 2003; Kirk *et al.* 2005). Recently, a number of studies have been carried out with the focus of exploring EF in individuals with Down Syndrome (DS). DS is caused by abnormalities of chromosome 21, in particular Trisomy 21, which is the most common karyotype accounting for 95% of cases. DS affects about 1 in 1000 live births (McGrowther & Marshall 1990). The great majority of people with DS have mild to severe levels of intellectual impairment and a wide range of associated physical, medical and cognitive deficits, including language impairment (e.g. Dykens *et al.* 2000; Silverman 2007, for a review). The severity of language impairment is highly variable, and different components of the language system are affected to a varying degree. In general, expressive language is affected to a greater extent when compared with receptive language and/or language comprehension (e.g. Miller 1992; Rondal 1996). At the same time, individuals with DS show relatively intact visuo-spatial abilities.

There are a number of studies that have examined memory system, structure and processes in individuals with DS (for a more comprehensive review, see Baddeley & Jarrold 2007, or Lanfranchi & Vianello 2006). Although less is known about EF in individuals with DS, some researchers propose the presence of deficits in EF area. Several research studies that included samples of adults with DS have been carried out. For example, Kittler *et al.* (2006) found that adults with DS make more verbal intrusion errors during verbal working memory tests than individuals with an unspecified intellectual disability (ID) matched for IQ and receptive vocabulary. These findings suggest deficient EF and inhibitory control in adults with DS. More recently, Kittler *et al.* (2008) showed that adults with DS (aged over 30) performed comparably to adults with Williams syndrome and individuals with ID of mixed etiologies (all three groups matched on IQ measure) on short-term memory tasks, but showed a greater decline in performance in the dual-task condition when compared with the other two groups. A dual-task condition involved short-term storage and a simultaneous processing component.

In a more comprehensive study, Rowe *et al.* (2006) reported that adults with DS, aged between 23 and 40, showed impairment on a number of EF

tasks, specifically on the tasks assessing set-shifting, sustained attention and planning, when compared with learning disabled controls, matched for age and verbal ability. Moreover, some studies suggest that performance on EF tasks might be age-related and might be associated with the presence of dementia in individuals with DS. For example, in a sample of 20 individuals with DS (age range 22–58 years), Nelson *et al.* (2005) found that their performance on an object reversal learning task (a task thought to be associated with frontal lobe functioning) was related to both age and the presence of dementia. Further, Das *et al.* (1995) compared a group of individuals with DS to a group of individuals without DS, matched for age and severity of learning disability, on a battery of tasks assessing cognitive processes, including planning and attention. Both groups were divided into ‘younger’ (40–49 years) and ‘older’ (50–62 years) subgroups. The authors found that, in the DS-older subgroup (but not in controls), participants showed poorer performance than in the DS-younger subgroup and suggested that impairments in planning and attention may occur in the early stages of dementia in this population. Finally, in a recent study, Ball *et al.* (2008) found that a group of adults (mean age 55 years) with DS and established dementia of Alzheimer’s type showed a consistent pattern of impaired performance on a battery of memory and EF tasks, when compared with adults (mean age 47) with DS without established dementia, even after the effect of age and LD severity was partialled out.

However, to examine the hypothesis whether age or dementia of Alzheimer’s type account for EF deficit in individuals with DS, a systematic study of children or adolescents with DS is needed. So far, only a few studies have been carried out on this population and the results are still not clear. On the one hand, Pennington *et al.* (2003) found that individuals with DS, aged between 11 and 19, years showed impairments on tasks associated with hippocampal functioning (tasks measuring verbal and visual long-term memory, as well as a measure of ecological memory) but not on tasks associated with prefrontal functioning (measures of planning, verbal and non-verbal fluency, inhibition, spatial and verbal working memory), when they were compared with a mental age (MA)-matched typically

developing (TD) group. On the other hand, Lanfranchi *et al.* (2004) and Lanfranchi *et al.* (2009b) found that children and adolescents with DS performed worse on both verbal and visuo-spatial dual tasks when compared with TD children matched for verbal MA. Thus, these results further point to a weakness in the executive system of individuals with DS. Moreover, Lanfranchi *et al.* (2009a) showed that individuals with DS are poorer than TD children of the same MA on working memory spatial-simultaneous tasks, but not on spatial-sequential tasks. The authors suggested that the simultaneous visuo-spatial working memory deficit of individuals with DS might be caused by difficulty to process more than one item at a time. Finally, Kogan *et al.* (2009) found impaired performances in individuals with DS in visuo-spatial working memory task, visual-perceptual and visual-spatial reversal learning tasks, while the performances in egocentric spatial learning and object discrimination tasks seem to be relatively preserved.

To summarize, the review of current published articles clearly points to an impairment of EF in adults with DS (e.g. Das *et al.* 1995; Ball *et al.* 2008). However, very little is examined with regard to adolescents and children with DS and their EF, and the few studies that have been conducted provide mixed results. It is clear that further research is necessary to explore EF in children with DS, as well as to determine the causes of EF impairments in this population: whether EF deficit is related to cognitive decline associated with either age and/or Alzheimer disease, or whether it is a characteristic of DS cognitive profile. This is the purpose of our study, and we included adolescents with DS, as adolescents do not show signs of cognitive decline or Alzheimer’s. Moreover, exploring a younger population, compared with the previous studies, would allow us to examine developmental trajectories of EF in DS. A growing number of studies (e.g. Karmiloff-Smith 1998, for a review) support the hypothesis that, in developmental disorders, the whole cognitive system might differ in processing ability when compared with a non-affected cognitive system and thus, in addition, the developmental trends of a given ability might also differ. We believe it is very important to explore the wide range of processes falling under the umbrella of EF in individuals with DS in order to better under-

stand the areas of strength and weakness within EF in the DS cognitive profile.

Therefore, the main purpose of the present study is to analyse the EF profile in adolescents with DS and compare them to an MA-matched TD group in order to identify strengths and weaknesses in EF with respect to MA. As EF is not a unitary function and there is a possibility that developmental neuro-pathology may differentially affect different aspects of EF, we chose a battery of tasks that allowed us to explore the areas of EF as listed by Pennington & Ozonoff (1996): set shifting; planning/problem-solving; working memory; inhibition/perseveration; and fluency. In addition, as suggested by Rowe *et al.* (2006), we examined sustained attention which is believed to overlap with EF and to be served by the prefrontal cortex (Posner & Petersen 1990; Mirsky *et al.* 1991; Manly & Robertson 1997).

Method

Participants

Participants were 15 adolescents with DS (eight males and seven females) with a mean chronological age (CA) of 15 years and 2 months ($SD = 2.2$ years; age range 11 years to 18 years and 5 months) and a mean MA of 5 years and 9 months ($SD = 8$ months; age range 4 years and 6 months to 6 years and 10 months). All our participants were still attending school and all of them were included in regular schools. Participants were contacted through several associations for people with DS in Northern Italy. Selection criteria were age and the absence of behavioural and hearing problems.

The control group was comprised of 15 TD children (eight males and seven females) with a mean chronological age of 5 years and 9 months ($SD = 8$ months), ranging between 4 years and 6 months to 6 years and 10 months. These children were contacted in a preschool and a regular primary school.

Both groups were individually matched on MA within 3 months, as assessed by the Logical Operations test (Vianello & Marin 1997), an intelligence test that provides an MA measurement in terms of logical thinking development. This test seems particularly appropriate for children with ID. It is inspired by the Piagetian theory of operational thinking and includes 18 tasks that assess the fol-

lowing areas of logical thinking: seriation, numeration and classification. In comparison with the Wechsler Intelligence Scales (the correlation of the Wechsler Intelligence Scale for Children with the Logical Operations test is 0.68), the Logical Operations test is less affected by cultural and verbal components. In comparison with the Columbia Mental Maturity Scale ($r = 0.78$), the Logical Operations test is less affected by visuo-spatial components. This test therefore seems particularly appropriate for matching children with DS with TD children on a central intelligence component, while limiting the influence of culture, linguistic ability (a weak ability in DS) (for a review, see Vianello & Marin 1997). Of course, we should keep in mind that matching groups on logical thinking does not eliminate the group difference in their verbal skills (our DS group might have lower verbal abilities than the control group). However, apart from verbal fluency, the utilised tasks contained only minimal verbal demands, such as repeating simple and familiar words. Therefore, we believe that any differences between the two groups on their verbal skills, did not affect the participants' performances on any of the EF tasks.

Tasks

Executive function was assessed by a battery of measures selected from previous studies with individuals with ID or from developmental literature. The description of the administered tasks follows below.

Working memory

Verbal and visuo-spatial dual tasks (Lanfranchi *et al.* 2004)

In the *verbal dual task*, the child was presented orally with a list of two to five words and was asked to remember the first word on the list and to tap on the table when the word *ball* was presented. The word *ball* was presented once in every list. Its position in the list varied across trials. The score of 1 was given for every trial performed correctly, with the child both remembering the first word of the list and performing the tapping task. Otherwise, the score of 0 was given. In each task, the minimum score was 0 and the maximum score was 8.

In the *visuo-spatial dual* task, the child was asked to remember the frog's starting position on a pathway on a 4×4 chessboard (where one of the sixteen cells was coloured in red). The child also had to tap on the table when the frog jumped onto the red square. The frog jumped once in every trial onto the red square. The position of the step onto the red square varied across trials. The task had four different levels of difficulty, according to the number of steps in the pathway: two, three, four and five steps, respectively. The score of 1 was given for every trial performed correctly, with the child both remembering the first position of the pathway and performing the tapping task. Otherwise, the score of 0 was given. In each task the minimum score was 0 and the maximum score was 8.

Inhibition

Stroop Type Task – Day/Night Version
(Gerstadt *et al.* 1994)

This task assessed the ability to inhibit a predominant response. In this task, participants were asked to say 'night' when they saw a picture of the sun drawn on a white card, and 'day' when they saw a picture of moon and stars drawn on a black card. After being given a brief practice trial, participants were then shown a sequence of 16 cards arranged in a set order. Participants were also given a control version of the task, in which they were taught to associate two separate abstract drawings with the words 'night' and 'day' and then again after a short practice given a trial of 16 cards in an arranged order. According to Gerstadt *et al.* (1994), the following indices were registered: number of correct responses for experimental and control condition, number of correct responses in the first four trials for experimental and control condition, and number of correct responses in the last four trials for experimental and control condition. The minimum score was 0 and the maximum was 16, for both experimental and control conditions.

Set shifting

*Rule Shift Card Test – (Wilson *et al.* 1996)*

This test assessed the ability to respond correctly to a rule and then to shift from that rule to another

one. In the first part of the task, participants were asked to say 'yes' when they saw a red card, and 'no' when they saw a black card. In the second part of the test, participants were asked to respond 'yes' if the card that had just been turned over was the same colour as the previously turned card, and 'no' if it was a different colour. The challenge in the second part of the task was therefore to concentrate on applying the new rule. The number of correct responses in both tasks was recorded. The minimum score was 0, and the maximum was 21 for both the first and the second parts.

Conceptual shifting

Modified Card Sorting Test (Nelson 1976)

The Modified Card Sorting Test (MCST) was proposed by Nelson (1976) as a reduced version of the Wisconsin Card Sorting Test (Berg 1948). In the MCST, four stimulus cards are placed in front of the child: the first card portrays a single red triangle, the second card shows two green stars, the third one, three yellow crosses and the fourth one, four blue circles. Participants are required to match each card according to colour, shape or number with every consecutive card of a set of 48 response cards, portraying all possible combinations of colour (red, green, yellow, blue), shape (triangles, stars, crosses, circles) or number (one to four). Participants are informed if their choice is correct or not, without, however, making suggestions regarding the sorting criterion. After the participant had completed six consecutive correct sorts (completion of a 'category'), he or she was explicitly informed that the sorting criterion had changed, with the phrase: 'The rules have now changed. I want you to find another rule'. The procedure continued until the participant completed the three categories twice, or until all 48 response cards were used.

The following parameters were recorded, according to Cianchetti *et al.* (2007) and Temple *et al.* (1996):

- 1 Categories completed. This criterion corresponds to the number of sequences of six consecutive correct matches the participant completes during the test (minimum score 0, maximum score 6).
- 2 Percentage of responses that were correct.
- 3 Perseverative errors: according to this criterion,

an error is perseverative when the participant persists in the same category as that of the previously incorrect response. We also included in this criterion errors made when the participant did not change category after being told that the rules had changed.

4 Non-perseverative errors: all errors not classified as 'perseverative errors'.

Planning

Tower of London

The Tower of London (Shallice 1982) assesses problem-solving and spatial planning. Instructions for administration were taken from Sannio Fancello *et al.* (2006). Test material consisted of a board, 13 cm × 5 cm × 1.5 cm, with three pegs of increasing height (9.5 cm, 6.5 cm and 3.5 cm) and three balls red, blue and green. The balls were 3 cm in diameter with 1.3 cm diameter central hole. A5 (paper size) coloured photographs of the target configurations were provided. The child was asked to match the target configuration, shown on a photograph, from a standard start configuration by moving the balls in accordance with set rules. The child had three attempts to solve each problem. Problems increased in difficulty from those which could be performed in a minimum of three moves to problems that could only be performed in a minimum of seven moves. Each level of move had three different problems, giving a total of 12 problems, altogether. Participants were given a practice two-move problem, in order to help them understand what they had to do. The following scores were recorded: the number of problems solved correctly (min 0, max 12), the number of problems solved correctly in the minimum number of moves (min 0, max 12) and the number of problems solved correctly at the first attempt (min 0, max 12). The first problem was a training task, following which 12 problems of increasing complexity were presented.

Fluency

Verbal fluency

Verbal fluency, in which participants produce as many words belonging to the same category (e.g. same initial letter or same semantic category) as

they can within a fixed period of time, is an established test of EF (e.g. Pennington & Ozonoff 1996). Two variations of this task were given to participants. The first was the FAS task, a traditional verbal fluency test (Newcombe 1969). Participants were asked to produce as many words as they could that started with the letters F, A and S within a minute. Participants were given a score for the number of words produced for each letter and, from that, a total score was derived. The number of errors and number of words that were repeated were also recorded. To assess semantic fluency, participants were asked to name, within a minute, as many items as possible from three semantic categories: animals, objects and occupations (Temple *et al.* 1996). Participants were given a score for the number of words generated for each of the three categories. From that, a total score was derived. The number of errors and number of repeated words was also recorded.

Sustained attention

Self-ordered Pointing Test – Temple *et al.* (1996)

In this task, which is an adaptation of a self-ordered pointing task devised by Petrides & Milner (1982), participants had to remember the sequence in which they pointed to a set of visually displayed stimuli by using their own strategies. Considering the age, both mental and chronological, of the individuals in the two groups, we decided to use only one of two versions of the task proposed by Temple *et al.* (1996): using the drawings of everyday objects. The task is divided in four sections. The first part consists of a set of six pages, on which an array of six different drawings is printed. The actual drawings are the same on each page, but they appear in different spatial positions. The child is asked to point to one drawing at a time, and to point to each drawing only once. Therefore, each time a page is turned, the child is required to remember which drawings have already been pointed to, and to point to a novel stimulus. Participants are open to determine their own order of responding. The other sections of the task are the same, except that they contain 8, 10 and 12 stimuli, respectively, with the corresponding number of pages. Scores were based on the number of pages correctly completed (min

0, max 36), the number of times the same item was pointed to in a set (item preservation) and the number of repeated points made to the same spatial position (position preservation). Overall, total scores were obtained, as well as separate scores for each set of stimuli.

Although this task is often used to assess sustained attention (see, for example, Temple *et al.* 1996), we should note that it also involves a memory component as a child has to hold in mind all the pictures that s/he has already pointed to in order to point to all the others.

Procedure

All the tasks were administered individually in three sessions separated by approximately 1 week, with each session lasting approximately 30 min. During the first session, participants completed a Logical Operation test that allowed us to closely match groups. During the two subsequent sessions, the EF tasks were administered. The order of presentation of the tasks was counterbalanced across participants to avoid testing effects. All participants were tested in a quiet room during or after school hours. Parental consent forms were obtained prior to testing.

Results

A preliminary exploration of the data revealed no outliers. Table 1 shows the mean score differences on all EF tasks (independent *t*-tests with, when needed, correction for non-homogeneous variances) between the group of adolescents with DS and the comparison group. As presented in Table 1, the group of adolescents with DS performed significantly worse than TD children for both *Verbal* and *Visuo-spatial Dual* tasks. On the *Inhibition* tasks, the individuals with DS had inferior scores in the experimental condition for the total number of correct answers ($t = -2.31, P = 0.028$) and the number of correct responses in the first four answers ($t = -2.43, P = -0.022$), but not for the number of correct responses for the last four answers of the task ($P > 0.05$). No differences between the two groups were observed in the control condition.

For the Rule Shift Card Test that assessed set shifting, no differences were found in scores for

Task 1 (indication that both groups understood the basic task), while individuals with DS performed worse on Task 2 ($t = -2.36, P = 0.025$), showing greater difficulties in shifting task rule. Significant group differences were found for the MCST, with the DS group completing fewer number of categories ($t = -4.66, P < 0.001$) and making a higher number of non-perseverative errors ($t = 2.54, P = 0.017$) when compared with the TD group. Moreover, the DS group's performance was significantly lower than that of the TD group for the Tower of London task. No differences were found between the two groups for the Verbal Fluency task ($P_s > 0.05$).

With regard to performance on self-ordered pointing tasks, the adolescents with DS completed a number of pages comparable to the TD-matched controls, but made a greater number of errors.

To summarize, adolescents with DS performed worse than TD children for all EF tasks, with the exception of verbal fluency. All these group differences are important as the effect sizes range from 0.72 to 2.26. However, adolescents with DS showed the greatest impairments (effect sizes greater than 1) for the following tasks: verbal dual task, modified card sorting task and Tower of London task.

Finally, a series of ANCOVAs, one for every variable of interest, was carried out with the effect of age partialled out. However, the effect of group did not change for any variable. This result might be better understood by examining the pattern of correlation between age and the EF variables analysed (rank-order correlations). None of the correlations in the DS group were significant, while in the control group age was significantly related to some variables [verbal working memory (0.58) and Tower of London (0.57)].

Discussion

The present work was aimed at analysing EF in adolescents with DS. On all EF tasks, except for verbal fluency, the group of adolescents with DS scored at a lower level when compared with the control group of TD children matched for MA. Specifically, based on their MA, the adolescents with DS showed impairments in their working memory system, planning, inhibition, set shifting

Table 1 Descriptive statistic and results of the comparison between the groups (Student's *t*) for EF tasks

Tasks	DS Mean (SD)	TD Mean (SD)	<i>t</i>	<i>P</i>	Cohen's <i>d</i>
Working memory					
Verbal dual task	2.87 (2.72)	5.00 (1.07)	-2.82	0.01	1.03
Visuo-spatial dual task	2.87 (2.20)	4.73 (1.67)	-2.62	0.01	0.95
Inhibition					
Stroop night and day: exp. condition					
Number of correct responses	13.87 (2.9)	15.67 (0.82)	-2.31	0.03	0.84
Number of correct responses first four items	3.53 (0.74)	4.00 (0)	-2.43	0.03	0.90
Number of correct responses last four items	3.27 (1.1)	3.80 (0.41)	-1.76	0.10	0.63
Stroop night and day: control condition					
Number of correct responses	14.93 (2.05)	15.40 (1.3)	-0.74	0.46	0.27
Number of correct responses first four items	4.00 (0)	3.87 (0.35)	1.47	0.16	0.52
Number of correct responses last four items	3.60 (0.83)	3.67 (1.05)	-0.19	0.85	0.07
Set shifting					
Rule Shift Card Test					
Number of correct responses task 1	19.00 (1.96)	19.87 (1.46)	-1.37	0.18	0.50
Number of correct responses task 2	12.80 (2.43)	15.40 (3.5)	-2.36	0.02	0.86
Conceptual shifting					
Modified Card Sorting Test					
Number of categories completed	2.0 (0.85)	3.87 (1.3)	-4.66	<0.001	1.7
Number of perseverative errors	12.07 (10.43)	14.07 (10.79)	-0.52	0.61	0.18
Number of non-perseverative errors	11.07 (7.88)	5 (4.88)	2.54	0.02	0.93
Planning					
Tower of London					
Number of problems solved correctly	3.8 (2.68)	9.67 (2.5)	-6.21	<0.001	2.26
Number of problems correctly solved at the first attempt	2.67 (1.99)	5.67 (3.66)	-2.79	0.01	1.02
Fluency					
Verbal fluency FAS					
F-letter	3.73 (2.19)	3.27 (1.98)	0.61	0.55	0.22
A-letter	2.47 (1.55)	2.60 (1.92)	-0.21	0.84	0.07
S-letter	2.93 (1.91)	2.13 (1.46)	1.29	0.21	0.47
Total FAS	9.13 (5.36)	8.00 (4.47)	0.63	0.53	0.22
Repeated words	0.07 (0.26)	0.47 (0.74)	1.97	0.06	0.72
Errors	0	1.73 (3.73)	1.8	0.09	0.65
Verbal fluency categories					
Animals	7.67 (3.44)	6.47 (4.29)	0.85	0.40	0.30
Objects	8.87 (5.74)	4.67 (3.35)	2.45	0.02	0.89
Occupations	3.07 (2.52)	2.07 (1.22)	1.38	0.18	0.51
Total categories	19.60 (10.19)	13.20 (7.1)	2.00	0.06	0.73
Repeated words	0.27 (0.59)	0.27 (0.46)	0	1	0
Errors	0.67 (1.45)	1.13 (1.46)	-0.88	0.39	0.32
Sustained attention					
Self-ordered Pointing Test					
Total number of pages correctly completed	25.67 (4.12)	22.20 (8.50)	1.42	0.17	0.52
Total number of errors	10.2 (4.23)	6.67 (5.15)	2.05	0.05	0.75
Total item preservations	5.00 (3.67)	3.60 (2.72)	1.19	0.24	0.43
Total position preservations	1.20 (1.21)	5.40 (3.2)	-4.78	<0.001	1.74

DS, Down Syndrome; EF, executive function; FAS, TD, typically developing.

and conceptual shifting skills, but not in their verbal fluency abilities. This might be an aspect of EF that is relatively preserved in DS. However, it is a quite surprising result as individuals with DS have weak verbal abilities. Therefore, further research is required. For the sustained attention task, the adolescents with DS completed the same number of pages than TD children, but made a greater number of errors. This is probably due to the fact that TD children make errors while performing the first and the second sets, which are the shortest sets, but then they spontaneously find a strategy to apply to the task at hand, such as, for example, focusing on a particular spatial position. This is confirmed by the fact that TD children made significantly more position perseverations (pointing always to the items in the same spatial position within the matrix), that can be considered a strategy in solving this task. On the other hand, the children with DS failed to come up with any strategy spontaneously and continued making mistakes when the sets of items became longer.

The present study provides a picture of a broad impairment of EF in adolescents with DS. The areas of major concern are those related to verbal working memory, conceptual shifting and planning.

The results of this study tend to be in line with the results of previous studies with adults with DS in confirming the evidence for executive dysfunction in DS (e.g. Rowe *et al.* 2006, Kittler *et al.* 2008). Our results further support the hypothesis that an impairment of EF, relative to intellectual ability, is a feature of DS. Our results are somewhat different from Pennington *et al.*'s (2003) study, the only study, to our knowledge, on EF conducted with adolescents with DS that did not find an EF deficit in adolescents with DS when compared with controls. However, these discrepancies could result from methodological differences between the two studies. First, they could be related to the different tasks that were administered (except for the verbal fluency task, the only task where no group differences were found in both studies). As such, these results suggest that not all EF processes are impaired, and provide encouragement to carry out further studies to better understand which tasks do not involve impaired performance. Second, the groups in the two studies differed in their MA. In the present study, the groups had an MA of 5 years

and 9 months, while the sample in Pennington *et al.*'s study (2003) had an MA of 4.92 years. As suggested by Garon *et al.* (2008), the elementary forms of the core EF components are present early during the preschool period, but several improvements occur between 3 and 6 years because of the development of attention and the integration of EF components. This means that even 1 year of difference in age in TD children might have a direct effect on the children's performance and, consequently, on the study results.

Clearly, more research is needed that would involve younger samples, as well as longitudinal analyses to clarify whether EF impairment is, in fact, a characteristic of the DS cognitive profile.

Our study also confirms that the EF tasks employed could be included in baseline batteries for individuals with ID [as also suggested by Rowe *et al.* (2006) and Ball *et al.* (2008)], although longitudinal data are further needed to validate their sensitivity and specificity. Overall, the tests used in this study were shown to be suitable for administration to a population with moderate to mild ID, as the participants were able to understand and perform the tasks with a low number of floor and ceiling effects.

Unlike other genetic disorders, such as Turner Syndrome and Klinefelter Syndrome, that showed a specific pattern of impaired and preserved executive skills (e.g. Temple *et al.* 1996; Temple & Martin Sanfilippo 2003), we found a generalised impairment of executive skills in adolescents with DS. However, for other genetic disorders, such as Fragile X Syndrome, a similar pattern of generalised impairment of EF has been found (e.g. Hooper *et al.* 2008). Hooper *et al.* (2008) and Kirk *et al.* (2005) found that, for individuals with Fragile X, low IQ or MA status significantly impacted performance on EF tasks. Therefore, we could hypothesise that both genetic status and MA contribute to EF limitations in genetic syndromes. In genetic syndromes with no ID, we found a selective pattern of impairments of EF, while in disorders with mild to severe ID, it is more likely to find a generalised pattern of impairment of EF.

This hypothesis is also consistent with previous studies with a non-clinical population that found EF to be related to performance on tasks closely associated with intelligence (Carpenter *et al.* 1990;

Salthouse *et al.* 1998; Engle *et al.* 1999; Miyake *et al.* 2001). These studies speculate that EF and intelligence share a common genetic variance (Luciano *et al.* 2001). However, Friedman *et al.* (2006) also point to the fact that not all EF are related to intelligence. It is possible therefore that in DS and other genetic disorders that involve ID, low performance on some EF tasks would be constrained by genetic factors, while on other EF tasks it would be constrained by an individual's overall low level of intelligence. Of course, more work on developmental disorders for genetic causes is needed in order to better understand EF and to further test these important theoretical assumptions.

Acknowledgements

We would like to thank all children who participated in this study and their families. Moreover, we would like to thank Susie Hartley for her editorial support.

References

- Baddeley A. D. (1986) *Working Memory*. Clarendon Press, Oxford.
- Baddeley A. D. & Jarrold C. (2007) Working memory and Down syndrome. *Journal of Intellectual Disability Research* **51**, 925–31.
- Ball S. L., Holland A. J., Treppner P., Watson P. C. & Huppert F. A. (2008) Executive dysfunction and its association with personality and behaviour changes in the development of Alzheimer's disease in adults with Down syndrome and mild to moderate learning disabilities. *British Journal of Clinical Psychology* **47**, 1–29.
- Berg E. A. (1948) A simple objective technique for measuring flexibility in thinking. *Journal of General Psychology* **39**, 15–22.
- Carpenter P. A., Just M. A. & Schell P. (1990) What one intelligence test measures: a theoretical account of the processing in the Raven Progressive Matrices test. *Psychological Review* **97**, 404–31.
- Cianchetti C., Corona S., Foscoliano M., Contu D. & Sannio-Fancello G. (2007) Modified Wisconsin Card Sorting Test (MCST, MWCST): normative data in children 4–13 years old, according to classical and new types of scoring. *The Clinical Neuropsychologist* **21**, 456–78.
- Cummings J. (1993) Frontal-sub cortical circuits and human behaviour. *Archives of Neurology* **50**, 873–80.
- Das J. P., Divis B., Alexander J., Parrila R. K. & Naglieri J. A. (1995) Cognitive decline due to aging among persons with Down syndrome. *Research in Developmental Disabilities* **16**, 461–78.
- Dykens E. M., Hodapp R. M. & Finucane B. M. (2000) *Genetics and Mental Retardation Syndromes*. Paulh Brookes, New York.
- Engle R. M., Tuhloski S. W., Laughlin J. E. & Conway A. R. A. (1999) Working memory, short-term memory and general fluid intelligence: a latent variable approach. *Journal of Experimental Psychology: General* **128**, 309–31.
- Friedman N. P., Miyake A., Corley R. P., Young S. E., DeFries J. C. & Hewitt J. K. (2006) Not all executive functions are related to intelligence. *Psychological Science* **17**, 172–9.
- Garon N., Bryson S. E. & Smith I. M. (2008) Executive function in preschoolers: a review using an integrative framework. *Psychological Bulletin* **134**, 31–60.
- Gerstadt C., Hong Y. J. & Diamond A. (1994) The relationship between cognition and action: performance of children 3 1/2–7 years old on a Stroop-like day-night test. *Cognition* **5**, 129–53.
- Godefroy O., Cabaret M., Petit-Chenal V., Pruvo J.-P. & Rousseaux M. (1999) Control functions of the frontal lobes: modularity of the central-supervisory system? *Cortex* **35**, 1–20.
- Hooper S. R., Hatton D., Sideris J., Sullivan K., Hammer J., Schaaf J. *et al.* (2008) Executive functions in young males with fragile X syndrome in comparison to mental age-matched controls: baseline findings from a longitudinal study. *Neuropsychology* **22**, 36–47.
- Karmiloff-Smith A. (1998) Development itself is the key to understanding developmental disorders. *Trends in Cognitive Sciences* **2**, 389–98.
- Kirk J. W., Mazzocco M. M. & Kover S. T. (2005) Assessing executive dysfunction in girls with fragile-or Turner syndrome using the Contingency Naming Test (CNT). *Developmental Neuropsychology* **28**, 755–77.
- Kittler P., Krinsky-McHale S. J. & Devenny D. A. (2006) Verbal intrusions precede memory decline in adults with Down syndrome. *Journal of Intellectual Disability Research* **50**, 1–10.
- Kittler P. M., Krinsky-McHale S. J. & Devenny D. A. (2008) Dual-Task processing as a measure of executive function: a comparison between adults with Williams and Down syndrome. *American Journal on Mental Retardation* **113**, 117–32.
- Kogan C. S., Boutet I., Cornish K., Graham G. E., Berry-Kravis E., Drouin A. & Milgram N. W. (2009) A comparative neuropsychological test battery differentiates cognitive signatures of Fragile-and Down syndrome. *Journal of Intellectual Disability Research* **53**, 125–42.

- Lanfranchi S. & Vianello R. (2006) Memory profiles in individuals with mental retardation due to genetic causes. In: *Focus on Birth Defects Research*, 53–80 (ed. J. V. Engels), pp. 53–80. Nova Science Publishers, New York.
- Lanfranchi S., Cornoldi C. & Vianello R. (2004) Verbal and visuo-spatial working memory deficits in children with Down syndrome. *American Journal on Mental Retardation* **109**, 456–66.
- Lanfranchi S., Carretti B., Spanò G. & Cornoldi C. (2009a) A specific deficit in visuo-spatial simultaneous working memory in Down syndrome. *Journal of Intellectual Disability Research* **53**, 474–83.
- Lanfranchi S., Jerman O. & Vianello R. (2009b) Working memory and cognitive skills in individuals with Down syndrome. *Child Neuropsychology* **15**, 397–416.
- Luciano M., Wright M. J., Smith G. A., Geffen G. M., Geffen L. B. & Martin N. G. (2001) Genetic covariance among measures of information processing speed, working memory, and IQ. *Behavior Genetics* **31**, 581–92.
- McGrowther C. W. & Marshall B. (1990) Recent trends in incidence, morbidity, and survival in Down's syndrome. *Journal of Mental Deficiency Research* **34**, 49–57.
- Manly T. & Robertson I. H. (1997) Sustained attention and the frontal lobes. In: *Methodology of Frontal and Executive Function* (ed. P. Rabbitt), pp. 135–50. Psychology Press, Hove.
- Miller J. (1992) Development of speech and language in children with Down syndrome. In: *Down Syndrome* (eds I. T. Lott & E. E. McCoy), pp. 39–50. Wiley-Liss, New York.
- Mirsky A. F., Anthony B. J., Duncan C. C., Ahearn M. B. & Kellam S. G. (1991) Analysis of the elements of attention: a neuropsychological approach. *Neuropsychology Review* **2**, 109–45.
- Miyake A., Friedman N. P., Emerson M. J., Witzki A. H. & Howerter A. (2000) The unity and diversity of executive functions and their contributions to complex 'frontal lobe' tasks: a latent variable analysis. *Cognitive Psychology* **41**, 49–100.
- Miyake A., Friedman N. P., Rettinger D. A., Shah P. & Hegarty M. (2001) How are visuospatial working memory, executive functioning, and spatial abilities related? A latent variable analysis. *Journal of Experimental Psychology: General* **130**, 621–40.
- Nelson H. E. (1976) A modified card sorting test sensitive to frontal lobe defects. *Cortex* **12**, 313–24.
- Nelson L., Johnson J. K., Freedman M., Lott I., Groot J., Chang M. *et al.* (2005) Learning and memory as a function of age in Down syndrome: a study using animal-based tasks. *Progress in Neuropsychopharmacology and Biological Psychiatry* **29**, 443–53.
- Newcombe F. (1969) *Missile Wounds of the Brain: A Study of Psychological Deficits*. Oxford University Press, Oxford.
- Pennington B. F. & Ozonoff S. (1996) Executive functions and developmental psychopathology. *Journal of Child Psychology and Psychiatry* **37**, 51–87.
- Pennington B. F., Moon J., Edgin J., Stedron J. & Nadel L. (2003) The neuropsychology of Down syndrome: evidence for hippocampal dysfunction. *Child Development* **74**, 75–93.
- Petrides M. & Milner B. (1982) Deficits in subject-ordered tasks after frontal and temporal lobe lesions in man. *Neuropsychologia* **20**, 249–62.
- Posner M. I. & Petersen S. E. (1990) The attention system of the human brain. *Annual Review of Neuroscience* **13**, 25–42.
- Rabbitt O. (1997) Introduction: methodologies and models in the study of executive functions. In: *Methodology of Frontal and Executive Function* (ed. P. Rabbitt), pp. 1–38. Psychology press, Hove.
- Rondal J. A. (1996) Oral language in Down's syndrome. In: *Down's Syndrome: Psychological, Psychobiological and Socio-Educational Perspectives* (eds J. Rondal, J. Perera, L. Nadel & A. Comblain), pp. 99–117. Whurr Publisher, London.
- Rowe J., Lavender A. & Turk V. (2006) Cognitive executive function in Down's syndrome. *British Journal of Clinical Psychology* **45**, 5–17.
- Salthouse T. A., Fristoe N., McGuthry K. E. & Hambrick D. Z. (1998) Relation of task switching to speed, age, and fluid intelligence. *Psychology and Aging* **13**, 445–61.
- Sannio Fancello G., Vio C. & Cianchetti C. (2006) *TOL – Torre di Londra Testi di valutazione delle funzioni esecutive (pianificazione e problem solving)*. Edizioni Erickson, Trento.
- Shallice T. (1982) Specific impairments of planning. *Philosophical Transactions of the Royal Society of London B* **298**, 199–209.
- Shallice T. (1988) *From Neuropsychology to Mental Structure*. Cambridge University Press, Cambridge.
- Silverman W. (2007) Down syndrome: cognitive phenotype. *Mental Retardation and Developmental Disabilities Research Reviews* **13**, 228–36.
- Stuss D. T. & Benson D. F. (1986) *The Frontal Lobes*. Raven Press, New York.
- Tatcher R. W. (1991) Maturation of the human frontal lobes: physiological evidence for staging. *Developmental Neuropsychology* **7**, 397–426.
- Temple C., Carney R. & Mullarkey S. (1996) Frontal lobe function and executive skills in children with Turner's syndrome. *Developmental Neuropsychology* **12**, 343–69.
- Temple C. M. & Martin Sanfilippo P. (2003) Executive skills in Klinefelter's syndrome. *Neuropsychologia* **41**, 1547–59.
- Vianello R. & Marin M. L. (1997) *OLC: Operazioni Logiche e Conservazione. Progetto MS*. Junior, Bergamo.

Welsh M. C. & Pennington B. F. (1988) Assessing frontal lobe functioning in children: views from developmental psychology. *Developmental Neuropsychology* **4**, 199–230.

Wilson B., Alderman N., Burgess P., Emslie H. & Evans J. (1996), *Behavioural Assessment of the Dysexecutive Syndrome (BADS)*. Thames Valley Test Company, Suffolk.

Yakovlev P. I. & Lecours A. R. (1967) The myelogenetic cycles of regional maturation of the brain. In: *Regional Development of the Brain in Early Life* (ed. A. Minkowski), pp. 3–70. Blackwell Scientific Publishers, Oxford.

Accepted 8 February 2010