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Executive functions in intellectual disabilities: A comparison between Williams syndrome and Down syndrome

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1. Introduction

Executive functions (EF) are a set of high cognitive abilities that control and regulate other functions and behaviors (Welsh, Pennington, & Groisserc, 1991). They encompass strategic planning, flexibility of thought and action (shifting), inhibition of inappropriate responses, generation of new responses (fluency) and concurrent remembering and processing (working memory) (Friedman et al., 2006; Pennington & Ozonoff, 1996). EF processes emerge in the first few years of life

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(e.g., Diamond, 1990) and continue to develop from childhood into adulthood (Hughes, Ensor, Wilson, & Graham, 2010; Huizinga, Dolan, & van der Molen, 2006; Lehto, Juujärvi, Kooistra, & Pulkkinen, 2003; Somerville & Casey, 2010). Due to their role in initiating and stopping actions, in monitoring and changing behavior and in planning future actions, EF are crucial for adaptive behavior. EF deficits have been described in developmental disorders, which are often characterized by low adaptive level. In particular, attention, inhibitory control, cognitive flexibility and working memory deficits are reported in individuals with attention and hyperactivity disorders (Abad-Mas et al., 2011; Corbett, Costantine, Hendren, Rocke, & Ozonoff, 2009; Sergeant, Geurts, & Oosterlaan, 2002); inhibition of responses (Stroop, Junior Hayling Test) and planning (Tower of London) impairments are described in children with autism (Hill, 2004; Kenworthy et al., 2005; Rinehart, Bradshaw, Moss, Brereton, & Tonge, 2001; Robinson, Goddard, Dritschel, Wisley, & Howlin, 2009); visual-spatial and auditory attention as well as shifting deficits are often found in dyslexic children (Altemeier, Abbott, & Berninger, 2008; Helland & Asbjørnsen, 2000; Menghini, Addona, Costanzo, & Vicari, 2010); planning and memory weakness are documented in fetal alcohol spectrum disorder (Green et al., 2009; Pei, Job, Kully Martens, & Rasmussen, 2011; Rasmussen, 2005). EF deficits have been also described in people with intellectual disability (ID) substained by different etiology. For example, both adults (Rowe, Lavender, & Turk, 2006) and adolescents (Lanfranchi, Jerman, Dal Pont, Alberti, & Vianello, 2010) with Down syndrome (DS) show impairment in set-shifting, conceptual shifting, sustained attention, planning, inhibition and working memory and deficits are age-related and seem to be associated with the onset of early dementia (Rowe et al., 2006). Moreover, performance in some EF have been found more impaired in people with DS than people with ID of unknown etiology and comparable mental-age (MA) (Lanfranchi et al., 2010; Rowe et al., 2006). However, other studies showed a preservation of planning, verbal and non verbal fluency, inhibition, spatial and verbal working memory in DS compared to MA-matched TD toddlers (Lanfranchi et al., 2010; Pennington, Moon, Edgin, Stedron, & Nadel, 2003; Vicari, Bellucci, & Carlesimo, 2000).

EF have been investigated also in other syndromic population such as Williams syndrome (WS). Namely, a number of studies documented deficits in inhibition (Atkinson et al., 2003; Atkinson, 2000; Menghini et al., 2010; Mobbs et al., 2007; Porter, Coltheart, & Langdon, 2007), planning (Menghini et al., 2010; Mobbs et al., 2007), and working memory (Menghini et al., 2010; Rhodes, Riby, Park, Fraser, & Cambell, 2010). Impairments in visual selective (Cornish, Scerif, & Karmiloff-Smith, 2007; Scerif, Cornish, Wilding, Driver, & Karmiloff-Smith, 2004) and visual and auditory sustained attention (Atkinson & Braddick, 2011; Menghini et al., 2010) as well as in attentional set-shifting (Atkinson, 2000; Rhodes et al., 2010) have been also reported. However, some aspects of EF as auditory selective attention, categorization, and shifting have been found preserved in WS when verbal material is processed (Atkinson & Braddick, 2011; Menghini et al., 2010; Tavano, Gagliardi, Martelli, & Borgatti, 2010).

All these findings support people with DS and WS are not fully impaired on EF. Moreover, specific EF profile in each syndrome may be supposed. Unfortunately, no definitive data are available so far.

This issue concerns the more general debate on etiological specificity hypotheses pertaining to the skill abilities of individuals with ID. The "syndrome specific hypothesis" (Conners, Moore, Loveall, & Merrill, 2011; Cornish et al., 2007) supports an asynchrony of cognitive and brain maturation for distinct etiological groups with ID. Conversely, the "syndrome independent theoretical perspective" (Zigler, 1969; Zigler & Balla, 1982) claims that a similar level of cognitive functioning is predicted by the same cognitive level. To address this issue, studies should directly compare groups with distinct etiology on specific neuropsychological abilities. To date, only few studies have jointly examined EF abilities in different genetic groups, and focused on few abilities each time: attention and inhibition (Brown et al., 2003; Cornish et al., 2007; Mervis et al., 2003), or shifting and working memory (Landry, Russo, Dawkins, Zelazo, & Burack, 2012).

The aim of the present study was to evaluate the etiological specificity hypotheses pertaining to EF abilities by comparing DS and WS individuals in different aspects of EF. We selected individuals from DS and WS populations because they are some of the most studied populations with ID, which have been often compared showing distinctive cognitive profiles (Vicari et al., 2004; Wang & Bellugi, 1993). Although EF is often considered a domain-general cognitive, distinction has been made between the more "cool," cognitive aspects of EF usually associated with lateral prefrontal cortex, and the relatively "hot," affective aspects of EF, usually associated with orbitofrontal cortex and other medial regions (Zelazo & Müller, 2002). Because we were interested in evaluating the role of low IQ in EF deficits we focused specifically on the so called "cool" EF.

2. Method

2.1. Participants

We evaluated EF abilities of 15 children, adolescents and adults with WS (F/M = 7/8), 15 with DS (F/M = 8/7) matched for MA to a group of 16 TD children (F/M = 8/8).

The individuals with WS exhibited a diagnosis established by FISH analysis and those with DS a diagnosis of free trisomy 21 documented by karyotyping. The participants with WS and DS were recruited at the Children's Hospital Bambino Gesù in Rome (Italy), at WS Association Marche and Umbria and at Verona DS Family Association. TD children were recruited in two primary schools. Inclusion criteria for all participants were the absence of neurosensory deficits, such as hypoacusia or serious visual impairment and epilepsy, and psychopathological disorders. All participants lived with their own families. Observations were carried out after informed consent has been obtained from all participants and their families. Demographic data of groups are reported in Table 1.

Table 1
Demographic characteristics of the groups.

	WS (n = 18)	DS (n = 15)	TD (n = 17)
CA–M (SD); range	17.6 (7.4); 10.7–34.9	14.5 (3.7); 8.6–21.2	7.4 (0.8); 6.1–8.4
MA–M (SD); range	6.7 (0.9); 5.10–7.8	6.2 (0.9); 4.8–8.7	6.9 (0.7); 5.-7.10
IQ–M (SD); range	53 (10.2); 36–71	53 (13.5); 36–83	94 (8.7); 85–119

CA, chronological age; MA: mental age; IQ, intelligence quotient; M, mean; SD, standard deviation.

2.2. Materials

2.2.1. Intelligence evaluation

The cognitive profile of all participants was assessed by the Leiter International Performance Scale – Revised, brief version (Leiter – R, Roid & Miller, 2002), with the exception of one participant with WS who performed the Stanford-Binet Intelligence Scale edited by Terman-Merrill (Bozzo & Mansueto-Zecca, 1993). The brief IQ test of the Leiter – R and the general composite score of the Stanford-Binet Intelligence Scale were considered as IQ measures. The corresponding MA was also computed.

2.2.2. Evaluation of the EF

Attention, memory, planning, categorization, shifting and inhibition domains were investigated. Selective attention was assessed by the BVN test (Bisiacchi, Cendron, Gugliotta, Tressoldi, & Vio, 2005) for the auditory domain and by the Sky Search (SKY; TEA-Ch, Manly et al., 2001; Manly, Robertson, Anderson, & Nimmo-Smith, 1999) for the visual domain. Auditory sustained attention was assessed by the Score task (SCO; TEA-Ch, Manly et al., 1999, 2001) while visual attention by the Bells Test (BELLS; Gauthier, Dehaut, & Joanette, 1989; Italian version: Biancardi & Stoppa, 1997).

Verbal short-term memory was evaluated by the forward version of the Digit Span test (F-DST, Orsini et al., 1987) and verbal working memory by the backward version of the Digit Span (B-DST) and by the Nonword Repetition task (NWR) (Vicari, 2007). Conversely, visual-spatial short-term memory was evaluated by the forward version of Corsi Block Test (F-CBT; Orsini et al., 1987) and visual-spatial working memory was evaluated by the backward version (B-CBT). Planning was measured by the Tower of London test (TOL; Shallice, 1982; SannioFancello, Vio, & Cianchetti, 2006). Verbal categorization was evaluated by the Category Fluency Test (CAT) (Mantyla, Carelli, & Forman, 2007) while visual categorization was evaluated by the Weigl Color Form Sort Test (WEIGL; Spinnler & Tognoni, 1987). Shifting in verbal modality was measured by the Alternate Category Fluency Task (CAT-A; Mantyla et al., 2007). Visual-spatial modality of shifting was investigated by the Trail Making Test (TMT) (Reitan, 1958) and Forma/Forma task (F/F) (Scarpa, 2006). Verbal response inhibition was measured by the Stroop task (STROOP; Stroop, 1935). Visual response inhibition was evaluated by the go-nogo task (G/NG; Van der Meere, Marzocchi, & De Meo, 2005). Depending of the task, both accuracy (correct responses) and speed (total time or reaction times-RTs) were scored. See Appendix A for detailed description of the tasks and measures adopted.

2.3. Procedure

Participants were evaluated individually in three testing sessions on three separate days. Each day the evaluation lasted approximately 1 h and a half with a pause of about an hour. The task order was randomized.

For each task, the experimenter verified that the participant understood the instructions. All of the tasks included a practice phase during which the experimenter illustrated the task instructions. People who did not show a proper task comprehension did not take part in the study.

2.4. Statistical analysis

Data were analyzed using the SPSS statistical software (SPSS version 13.0, for windows, SPSS Inc., Chicago, IL). A series of one-way ANOVAs on the neuropsychological measures of participants were carried out. For measures that presented heterogeneous variances, a Welch-ANOVA was employed. Row scores for each measure were considered. Post hoc analyses were performed by means of Tukey's test or Games–Howell post hoc test (following the Welch-ANOVA) to characterize the significant effects. Cohen's d (d) has been reported as effect size measure. Significant differences were considered for $p < .05$. To correct for multiple testing, p-values were adjusted according to the Benjamini and Hochberg procedure.

3. Results

3.1. Mental-age, chronological age and IQ comparison

WS, DS and TD participants did not differ for MA ($F(2,47) = 2.70$, $p = .08$), thus confirming the matching criterion. However, groups differed for CA ($F(2,47) = 19.91$, $p < .001$), in particular the two ID groups did not differ (WS vs DS $p = .15$) but were

older than TD people (WS vs TD $p < .001$; DS vs TD $p < .001$). The groups also differed for IQ ($F(2,47) = 77.08$, $p < .001$), again the two ID group did not differ (WS vs DS $p = .99$) but had lower IQ scores than TD participants (WS vs TD $p < .001$; DS vs TD $p < .001$) (see Table 1 for means and SDs).

3.2. Executive function assessment

3.2.1. Attention

On auditory selective attention task (BVN) no significant difference was found between groups in the number of correct responses (Welch's $F(2,28.72) = 1.87$, $p = .18$). Conversely, on auditory sustained attention (SCO) a significant effect emerged in the number of correct trials (Welch's $F(2,27.30) = 19.49$, $p < .001$): WS and DS groups did not differ but both groups scored lower than TD. On visual selective attention (SKY) a significant difference between groups was found on time-per-target (Welch's $F(2,25.70) = 11.41$, $p < .001$): while WS did not differ from DS, WS and DS individuals scored significantly lower than TD, with large effect sizes (see Table 2).

Results also showed a significant group effect ($F(2,47) = 6.24$, $p < .01$) on visual sustained attention (BELLS): DS individuals scored lower than WS individuals with a large effect size, but the two syndromic groups did not differ from TD.

3.2.2. Short-term and working memory

Significant differences between the three groups in the verbal short-term memory task (F-DST) were found ($F(2,42) = 12.14$, $p < .001$). In particular, DS group had lower score than WS and TD. No difference emerged between WS and TD. On verbal working memory (B-DST) results also showed difference between groups ($F(2,41) = 20.03$, $p < .001$). Post hoc analysis revealed that the two ID groups obtained lower score than TD and DS participants scored lower than WS. Moreover,

Table 2
Performance of the three groups in the executive functions tasks.

Measure	WS Mean (SD)	DS Mean (SD)	TD Mean (SD)	$p^{adjusted}$	Post hoc comparisons Tukey test or Games-Howell test (Cohen's *d*)			Post hoc summary
					WS vs DS	WS vs TD	DS vs TD	
Attention								
BVN[c]	33.8 (10.8)	31.8 (5.9)	35.4 (4.4)	.18	–	–	–	–
SCO[c]	5.3 (3.2)	3.6 (3.0)	8.6 (1.6)	<.001	.26 (0.6)	<.01 (1.3)	<.001 (2.2)	WS = DS < TD
SKY[c]	14.2 (6.6)	21.2 (12.8)	8.0 (3.7)	<.001	.16 (0.7)	<.01 (1.2)	<.01 (1.4)	WS = DS < TD
BELLS	103.6 (16.0)	81 (22.1)	96.8 (17.6)	<.01	<.01 (1.1)	.54 (0.4)	.05 (0.7)	WS = TD; DS = TD; DS < WS
Short-term and working memory								
F-DST	4.1 (1.2)	2.7 (1.0)	4.6 (0.8)	<.001	<.01 (2)	.40 (0)	<.001 (2.8)	DS < WS = TD
B-DST	2.4 (0.8)	1.3 (0.6)	3.4 (1.1)	<.001	<.01 (1)	<.01 (0.9)	<.001 (1.9)	DS < WS < TD
NWR[c]	34.2 (4.4)	22.1 (14.6)	32.4 (3.9)	<.05	<.05 (1.1)	.45 (0.4)	<.05 (0.9)	DS < WS = TD
F-CBT	2.7 (1.1)	3.2 (1.4)	4.9 (0.9)	<.001	.45 (0.4)	<.001 (2)	<.001 (1.4)	WS = DS < TD
B-CBT	1.8 (1.0)	1.7 (0.9)	3.8 (1.5)	<.001	.96 (0.1)	<.001 (2)	<.001 (1.7)	WS = DS < TD
Planning								
TOL								
Score[c]	16.4 (6.2)	25.4 (7.3)	25.7 (3.2)	<.001	<.01(1.3)	<.001 (1.8)	.98 (0)	WS < DS = TD
Time[a]	113.9 (53.5)	176.6 (60.4)	128.9 (33.8)	<.01	<.01 (1)	.65 (0.3)	<.05 (0.9)	DS < WS = TD
Categorization								
CAT	23.4(5.8)	18 (6.2)	21.7 (4.4)	<.05	<.05 (0.9)	.62 (0.3)	.16 (0.6)	WS = TD; DS = TD; DS < WS
WEIGL[c]	7.3 (3.7)	5.6 (2.8)	12.5 (2.1)	<.001	.29 (0.5)	<.001 (1.7)	<.001 (2.8)	WS = DS < TD
Shifting								
CAT-A	6.5 (2.6)	3.5 (2.4)	6.7 (2.3)	<.01	<.01 (1.2)	.97(0)	<.01 (1.4)	DS < WS = TD
TMT[a,c]	84.4 (59.6)	122.3 (71.2)	52.3 (32.4)	<.05	.32 (0.6)	. 13 (0.7)	<.05 (0.5)	WS = DS; WS = TD; DS < TD
F/F[a]	143.2 (54.1)	239.6 (139.3)	85 (50.7)	<.001	<.001 (0.9)	.12 (1.1)	<.001 (1.5)	DS < WS = TD
Inhibition								
STROOP								
Index[a]	1.4 (0.8)	2.6 (2.3)	0.7 (0.7)	<.01	<.05 (0.7)	.33 (0.9)	<.001 (1.2)	DS < WS = TD
G/NG								
RTs[b]	511.9(89.8)	494.2 (141.9)	577.6 (76.7)	.08	–	–	–	–
Correct responses[c]	89.9 (7.9)	85.5 (12.2)	93 (4.0)	.08	–	–	–	–

WS, Williams syndrome; DS, Down syndrome; TD, typically developed children; SD, standard deviation; BVN, Batteria per la Valutazione Neuropsicologica; SKY, sky search; SCO, Score!; BELLS, Bells Test; F-DST, Forward Digit Span Test; B-DST, Backward Digit Span Test; NWR, Nonword Repetition; F-CBT, Forward Corsi Block Test; B-CBT, Backward Corsi Block Test; TOL, Tower of London; CAT, Category Fluency; WEIGL, Weigl Color-Form Sort Test; CAT-A, Category fluency-alternated; TMT, Trail Making Test; F/F, Forma Forma; STROOP, Stroop task; G/NG, Go/NoGo task.

[a] Measurement in seconds.

[b] Measurement in milliseconds.

[c] Welch-ANOVA test and Games-Howell post hoc test were applied.

on NWR results documented significant difference between groups (Welch's $F(2,25.01) = 4.70$, $p < .05$), with participants with DS showing lower score than those with WS and TD, while WS and TD groups did not differ.

On visual-spatial short-term memory (F-CBT; $F(2,43) = 16.32$, $p < .001$) both DS and WS group obtained lower score than TD. Similarly, on visual-spatial working memory (B-CBT; $F(2,43) = 17.06$, $p < .001$) WS and DS groups scored lower than TD, with large effect sizes.

3.2.3. Planning

A difference between groups emerged on accuracy (Welch's $F(2,25.44) = 15.69$, $p < .001$), since WS group scored lower than DS and TD, but DS and TD groups did not differ. Difference between groups also emerged in execution time ($F(2,46) = 6.57$, $p < .01$) since DS spent more time than WS and TD. No difference emerged between WS and TD.

3.2.4. Categorization

On verbal categorization (CAT) a difference between groups emerged ($F(2,46) = 3.92$, $p < .05$), since DS group scored lower than WS, however both ID groups had comparable to TD group scores. On visual categorization (WEIGL; Welch's $F(2,29.47) = 34.07$, $p < .001$) the groups with ID did not differ and scored lower than TD.

3.2.5. Shifting

On verbal shifting (CAT-A; $F(2,45) = 7.57$, $p < .01$) DS participants generated lower number of pairs than WS and TD; however WS and TD performance did not differ.

Regarding visual-spatial shifting, a difference was found on execution time of TMT (Welch's $F(2,21.43) = 5.64$, $p < .05$), due to DS had longer time than TD. In the execution of F/F ($F(2,46) = 12.41$, $p < .001$) again DS spent more time than WS individuals and TD; no difference emerged between WS and TD group.

3.2.6. Response inhibition

As concerns verbal inhibition (STROOP), a difference between groups emerged ($F(2,46) = 7.27$, $p < .01$), since the index of interference for DS was higher than TD and WS, but WS did not differ from TD.

As concerns visual inhibition (G/NG) the two ID groups did not differ from controls both in accuracy (correct responses: Welch's $F(2,20.46) = 2.79$, $p = .08$) and in execution time (RTs: $F(2,41) = 2.77$, $p = .08$).

4. Discussion

The study investigated the EF profile of two different syndromic groups with ID of different etiology with an extensive battery tapping attention, working memory, planning, categorization, shifting and inhibition, on the verbal and the visual-spatial modality. The ID groups demonstrated a distinctive EF profile, with specific weakness and straights but with some common characteristics.

Both groups were impaired in auditory sustained attention (but not in the selective one), visual selective attention (but not in the sustained one), visual categorization (but not in the verbal one) and working memory.

However, a distinctive pattern of performance between the two syndromes emerged in other EF. While participants with DS were specifically poor in shifting and verbal aspects of memory, participants with WS were specifically poor in planning. Moreover participants with WS were stronger than DS in visual sustained attention, verbal categorization and verbal inhibition.

Visual selective attention deficits in both participants with WS and DS are in line with evidences from literature (Brown et al., 2003; Cornish, Sudhalter, & Turk, 2004; Cornish et al., 2007; Menghini et al., 2010; Munir, Cornish, & Wilding, 2000a, 2000b; Scerif et al., 2004; Wilding, Cornish, & Munir, 2002). In particular, our results agree with Cornish et al. (2007) and Munir et al. (2000a, 2000b) who found DS having lower performance than controls in visual selective attention, but not in visual sustained attention (Cornish et al., 2007), and with Brown et al. (2003), showing participants with DS exhibited lower performance on sustained visual attention than WS. Conversely, in younger children with WS and DS an opposite pattern of visual attention abilities has been described (Cornish et al., 2007) because DS toddlers showed a better performance than WS peers. Difference in age between Cornish et al.' and ours participants may explain the contrasting findings. Indeed, it could be that the pattern of visual attention abilities varies across age in the two syndromes as documented in other cognitive domains (Vicari et al., 2004).

The results on auditory attention delineated a similar profile between DS and WS group. While in individuals with WS preserved auditory selective attention and impaired sustained auditory attention are not surprising (Lincoln, Lai, & Jones, 2002; Menghini et al., 2010), in DS we expected deficits in both selective and sustained auditory attention, due to their acknowledged weakness in hearing and in verbal material processing. At the best of our knowledge, no studies have directly compared selective and sustained auditory attention in the same population of DS. According to our results, sustained auditory attention of individuals with DS has been found similar to individuals with ID of unspecific etiology (Trezise, Gray, & Sheppard, 2008). However, given the little available data, more investigations are needed to deeper define whether a syndrome specific profile could exists on auditory attention, looking at both selective and sustained abilities.

Results on short-term memory and WM revealed a general impairment for the DS group, while the WS group showed deficits only in visual-spatial memory. In particular, the preservation of verbal short-term memory and phonological WM in

our group with WS is in line with amount of data documenting higher performance on verbal than visual-spatial short-term memory in WS (Bellugi, Bihrle, Jerningan, Trauner, & Doherty, 1990; Rhodes et al., 2010; Vicari, Bellucci, & Carlesimo, 2001, 2006). However, both our ID groups showed deficits in WM tasks requiring high load (backward tasks), probably linked to a central executive deficit of WM. Deficits in the central executive component of WM in both individuals with WS and DS has been documented previously (Lanfranchi, Carretti, Spanò, & Cornoldi, 2009; Lanfranchi et al., 2010; Rhodes et al., 2010). Authors also found individuals with DS having more difficulties on backward tasks than persons with ID with different etiology (Lanfranchi et al., 2010; Vicari et al., 2001). Our results confirm such a difficult in DS compared to WS only for the verbal modality. We speculate that the degree of impairment in the central executive component of WM, as well as the specific deficits on the slave components of WM system, may vary between syndromes on the basis of the modality.

Concerning planning ability, WS participants were less accurate than DS and controls. Planning deficits have been yet described in WS (Arnold, Yule, & Martin, 1985; Bellugi et al., 1990; Menghini et al., 2010; Rhodes et al., 2010; Vicari et al., 2001) and may be explained by the presence of impulsive traits typically displayed by individuals with WS. DS group exhibited an opposite profile, confirming previous data in individuals with DS showing comparable accuracy but longer execution times in planning tasks compared to MA matched groups (Pennington et al., 2003; Rowe et al., 2006; Vicari et al., 2000). Notably, the only study reporting deficits in TOL in DS individuals (Lanfranchi et al., 2010) adopted a scoring procedure which is not directly comparable with the present study data.

As concerns categorization abilities, in the visual domain the two ID groups were commonly impaired while in the verbal domain they showed preserved abilities, although WS group was better than DS. Preserved verbal categorization abilities have been reported in literature in both syndromes (Jarrold, Baddeley, & Hewes, 1999; Vicari et al., 2004), although with exceptions (Landry et al., 2012; Lukács, Pleh, & Racsmany, 2004; Nash & Snowling, 2008; Volterra, Capirci, Pezzini, Sabbadini, & Vicari, 1996). As a limit of the study, the higher chronological age of the participants with ID compared to controls might have affected results particularly in this task, where the role of experience is more relevant. Indeed, given that semantic knowledge (needed to perform the task) increases with experience (Kail, 1990), the two ID groups might have been advantaged by their older age in performing the task. Visual categorization, instead, has been rarely investigated in groups with ID and the few available data are in line with our findings, reporting deficits in both WS (Menghini et al., 2010) and DS (Rowe et al., 2006) groups. Although further confirmations needs, deficits in categorization seems to be similar between syndromes, identifying a general weakness in the visual domain which is independent of the syndrome diagnosis.

Conversely, shifting abilities profile appeared to be dependent of the etiology of ID and preserved in WS but not in DS individuals. Only few studies have focused on shifting in individuals with WS and DS. On WS, preserved verbal but not visual-spatial shifting abilities have been found (Menghini et al., 2010). On DS, significantly lower performance than TD on verbal shifting or verbal mediated shifting tasks has been documented (Landry et al., 2012; Lanfranchi et al., 2010). According to literature, our results indicate shifting ability is particularly poor for persons with DS and poorer than in persons with WS.

Concerning inhibition, only DS group showed impairment and specifically in the verbal modality, which was lower even than WS. These results are in agreement with studies documenting a preservation of visual inhibition in DS group (Cornish et al., 2007; Pennington et al., 2003) and are in line with the study of Munir et al. (2000a, 2000b) documenting a verbal inhibition deficit but a less severe visual inhibition deficit in people with DS compared to controls and other groups with ID. Surprisingly, in WS group our results showed a general preservation of inhibition independently of the modality and did not confirm data reporting difficulties on visual inhibition in WS individuals (Atkinson et al., 2003; Mobbs et al., 2007). Perhaps, difference in the task and methodology may explain such contrasting findings. However inhibition abilities appear to be negatively influenced by DS weakness in the elaboration of verbal material, and then associated to the characteristic cognitive profile linked to the etiology of ID.

In summary, our results are in line with the "syndrome specific hypothesis" (Zigler, 1969; Zigler & Balla, 1982), documenting most EF in individuals with ID are dependent from the etiology of the syndrome and providing support for the etiological specificity of the cognitive and brain development.

The finding of non-homogeneous cognitive profiles in individuals with DS and WS is not new. Indeed, previous studies directly comparing individuals with DS and WS revealed syndrome-specific abilities in language (Mervis et al., 2000; Vicari et al., 2004), visual-spatial abilities (Wang & Bellugi, 1993; Vicari et al., 2004) and memory functioning (Brock, 2007; Vicari & Carlesimo, 2002; Vicari & Carlesimo, 2006; Vicari, Verucci, & Carlesimo, 2007).

Specifically, studies revealed visuo-spatial processing as relative strong in individuals with DS (Fidler, Hepburn, & Rogers, 2006; Jarrold et al., 1999; Klein & Mervis, 1999). Moreover, substantial evidence points to verbal processing difficulties in DS (Jarrold & Baddeley, 2001; Laws, 2002). Conversely most studies have described WS with relative strengths in linguistic processing and severe weakness in visuo-spatial processing (Bellugi, Mills, Jernigan, Hickok, & Galaburda, 1999; Mervis et al., 2000).

These syndrome specific cognitive profiles seem to influence the difficulties on some EF, which varied according to task modality. In particular, on short-term memory, categorization and inhibition, DS showed more difficulties with the verbal task modality than WS. However, task modality may not always be the most influent on EF, but difficulties in some EF domains may be general (such as working memory, visual categorization and components of attention) or relative to a specific syndromic group, such as shifting deficit in DS, or planning deficit in WS.

The EF profile exhibited by the two groups with ID is presumably the result of their characteristic cognitive and brain development. Recent neuroimaging studies (Atkinson & Braddick, 2011; Ball, Holland, Watson, & Huppert,

2010; Menghini et al., 2011) have attempted to document the presence of particular morphological cerebral characteristics to explain the distinct cognitive and behavioral profiles observed in persons with ID, especially of known genetic syndromes.

The pathophysiology of EF deficits is complex and includes abnormalities not limited to a single region (Hedden & Gabrieli, 2010). Indeed, EF have been linked to prefrontal cortex functioning, but activities in distributed neural systems, that include parietal and temporal cortices, also contribute to some EF. Studies on adults (Rowe et al., 2006) have suggested that abnormal development of the prefrontal cortex in individuals with DS (Ball et al., 2010; Brunamonti et al., 2011; Menghini et al., 2011; Rigoldi et al., 2009) may be linked to their EF deficits. Also in WS, deficits on EF tasks have been linked to frontal lobe dysfunctions, such as planning, working memory and attention set-shifting deficits (Rhodes et al., 2010). However, WS individuals differ from frontal-lobe patients, thus only frontal lobe dysfunctions cannot account for the whole pattern of EF peaks and valleys found in WS.

It is supposable that, structural abnormalities consistently reported in the fronto-parietal brain regions in WS (Atkinson et al., 2003; Hocking, Bradshaw, & Rinehart, 2008) and in the fronto-temporal regions in DS (Menghini et al., 2011; Smigielska-Kuzia et al., 2011) may account for the differential profile of EF between the syndromes. Studies should be developed to deeper investigate the relation between structural and functional brain abnormalities and EF in individuals with ID, by comparing a groups with ID of different etiology and with larger samples.

5. Conclusions

Evaluating distinctive EF abilities in ID may help clinical people to identify which skills should be targeted for early intervention. Given the inhomogeneous profile of EF skills in people with ID, it is essential that we gain an understanding of this profile evaluating both verbal and visual domain, to help people with ID to develop adaptive behaviors, autonomy and to live more independently in the community.

Appendix A

A.1. Attention

A.1.1. Auditory

Auditory attention was evaluated by two tests. A task from the *Batteria per la Valutazione Neuropsicologica* (Bisiacchi et al., 2005) was used as auditory selective attention measure (BVN). The examiner asked participants to listen to a series of 283 words and to respond by beating a hand on the table when the word *sun* was presented. The final score was the number of stimuli correctly identified (max = 45).

The Score task (SCO), a subtest taken from *Test of Everyday Attention for Children* (TEA-Ch, Manly et al., 1999, 2001), was used as a measure of sustained attention. The task required participants to count silently some tones and announce the total number of tones at the end of each trial. The number of tones ranged from 9 to 15, with a total of 10 trials in this subtest. The score was the number of correct trials (max = 10).

A.1.2. Visual

Visual attention was evaluated by two tests. The first one was the Sky Search (SKY), taken from *Test of Everyday Attention for Children* (TEA-Ch, Manly et al., 1999, 2001), and was used to evaluate visual selective attention. It was composed by a colorful A3 sheet in which there were distributed numerous pairs of aircraft (20 pairs). The children have to circle the pairs of identical aircraft as quickly as possible. The examiner registered the execution time (s) and the number of correct targets. To avoid the effect of the confused factor motor speed, on visual selection, the participant then completed a motor control version of the test. The sky search Attention Score was obtained by subtracting the motor control time-per-target ($F = D/E$; D = time, E = number of correct targets) from the more attentionally demanding sky search time-per-target ($C = A/B$; A = time, B = number of correct targets). This method of calculation ensured that the sky search attention score was free from the impact of motor speed.

The second test was the Bells Test (BELLS; Gauthier et al., 1989; Italian version: Biancardi & Stoppa, 1997) that consisted by four different sheets and was used to evaluate visual sustained attention. Each sheet contained 35 bells mixed with many other stimuli (animated and unanimated objects). The participants have to find and to check off only the bells in a fixed amount of time (2 min). The examiner recorded the total number of bells detected in four sheets and the number of bells identified in the first 30 s per sheet. The total number of bells was then considered as sustained attention score (max = 140).

A.2. Short-term and working memory

A.2.1. Verbal

The Digit Span test (DST, Orsini et al., 1987) was composed by two versions: forward version (F-DST) as a measure of short-term memory, and backward version (B-DST) as a measure of working memory. The examiner read aloud digits in one second intervals. The participants were asked to repeat the sequence in the same order (F-DST) and in the reverse order (B-DST). The length of the digit sequences was increased over trials. If participants correctly repeated three out of the five sequences presented

in the same list, a longer list of digits was proposed. If the participants failed on three sequences in the same list the test end. The score considered was the highest span achieved, plus the number of correctly repeated sequences (over the span) multiplied by 0.5 (F-DST: max = 10.5; B-DST: max = 7.5).

Working memory was also evaluated by Nonword Repetition task (NWR) taken from Promea memory battery (Vicari, 2007). It included 40 items (i.e., non-words). Covering her/his lips, the examiner read one non-word every 2 s and participants were asked to repeat it immediately. The score was the number of correct repetitions (max = 40).

A.2.2. Visual-spatial

We used the Corsi Block Test (CBT; Orsini et al., 1987) in the two versions: forward version (F-CBT), as a measure of short-term memory, and backward version as a measure of working memory (B-CBT). The experimenter pointed at a sequence of blocks which had to be reproduced by the participants in the same order (F-CBT) and in the reverse order (B-CBT). The length of the block sequences was increased over trials. If participants were able to repeat correctly three out of five sequences presented in the same list, a longer sequence of blocks was then proposed. If the participants failed on three sequences the test ended. The score considered was the highest span achieved, plus the number of correctly repeated sequences (over the span) multiplied by 0.5 (F-CBT: max = 10.5; B-CBT: max = 7.5).

A.3. Planning

Planning was measured by the Tower of London test (TOL; Shallice, 1982; Italian version: Sannio Fancello et al., 2006). Twelve different patterns were shown and three attempts were allowed at recreating each given pattern. Scoring was computed on the sum of the total points obtained (max = 36) and the total time spent to complete the patterns correctly.

A.4. Categorization

A.4.1. Verbal

The Category fluency test (CAT) was used as a measure of verbal fluency (Mantyla et al., 2007). Participants had to say as many words as possible from a semantic category (animals and fruits) in a given time (usually 60 s). The examiner recorded all words named by the participants and the number of valid responses given within time limits was calculated. Errors could be repetitions and identification of items that were not samples of the proposed category.

A.4.2. Visual

The Weigl Color – Form Sort Test (WEIGL; Spinnler & Tognoni, 1987) was used to assess the ability to classify 12 wooden objects on the basis of different categories: shape (circle, triangle or square), color (blue, red, green or yellow), a central symbol depicted on them (diamond, heart or flower), size (small, medium or large) and thickness (thin, medium or thick). The participant could categorize the objects in two modalities (active and passive) and received a score of 3 for each category found in the active modality (spontaneous categorization) and a score of 1 for each category they were able to detect in the passive mode (the examiner subdivided the test materials). Final score was calculated by summing the scores obtained in both modalities (max = 15).

A.5. Shifting

A.5.1. Verbal

To measure verbal shifting we used an alternate version of the Category fluency task (CAT-A; Mantyla et al., 2007). The participants had to generate pairs of words belonging to two categories (animals and fruits) and to shift between each opposite category. Total score was the number of correctly generated pairs within a time limit of 2 min.

A.5.2. Visual-spatial

The Trail Making Test (TMT) (Reitan, 1958) assesses the subject's attentional capacity and its ability to quickly switch from one stimulus to a numeric type alphabetically. The TMT consists of two subtests. In the first (test A) stimuli consist of a series of numbers from 1 to 25, circled in random order and printed on a sheet of A4, the number 1 corresponds to 'start, 25 at the end. In the second (test B) stimuli are formed both by numbers of letters, the number 1 is the beginning and 13 is the end, the letters range from A to N. Each of the two tests is preceded by a comprehension test in order to properly understand the subject to the rules of the task. In part A the subject were requested to merge with segments of circles drawn in pen numbers following the exact order beginning (No. 1) at the end (No. 8). The principle of the subtest B is the same, except that here the patient should switch to the numerical stimulus alphabetically, always following the 'order.

Another test used is Forma/Forma (F/F). The participants have to connect some shapes (circles, rectangles, squares, diamonds, etc.) according to an 'insideoutside' scheme. Each item consisted of two shapes, one inside the other, and the participants were asked to draw some lines between the items as quickly as possible, connecting all forms (from the inside form of an item to the outside same form of another item). The examiner recorded the total time spent (s) in completing the task.

A.6. Inhibition

A.6.1. Verbal

The Stroop task (STROOP; Stroop, 1935) was adopted to measure verbal response inhibition. Three conditions were considered: two neutral conditions and one incongruent condition. In the first neutral condition participants were required named the color of 30 circles printed in one of three colors: blue, green or red. In the second neutral condition participants had to read 30 color words printed in black. Finally, in the incongruent condition it was required to name 30 color words printed in a different color (e.g., 'blue' printed in red color). The execution time (s) and the error rate for each task were recorded. The index of interference was the difference relative to the baseline in execution time, computed as [(incongruent-control)/control], where the control condition was the second neutral condition (the list of color words printed in black). The index of interference was considered as score.

A.6.2. Visual

The G/NG version used in our study was presented through E-Prime software on a laptop computer screen. The task required the child to press a button as quickly as possible when blue, green and yellow circles appeared in the center of the screen but not when the red one was shown.

The response times (i.e., reaction times, RTs) and correct responses (the correct pressure of the space bar upon appearance of a red circle) were automatically recorded by the software.

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