Research in Developmental Disabilities 34 (2013) 1770–1780

Contents lists available at SciVerse ScienceDirect

Research in Developmental Disabilities

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

Floriana Costanzo [a,1], Cristiana Varuzza [a,b,1], Deny Menghini [a], Francesca Addona [a], Tiziana Gianesini [c], Stefano Vicari [a,*]

[a] Department of Neuroscience, Bambino Gesù Children's Hospital, Piazza Sant'Onofrio 4, I-00165 Rome, Italy
[b] LUMSA University, Via della Traspontina 21, I-00193 Rome, Italy
[c] AGBD, Down's Syndrome Association, Physical Medicine and Rehabilitation Clinic, via Valpantena 116/A, I-37142 Marzana, Verona, Italy

ARTICLE INFO

Article history:
Received 28 November 2012
Received in revised form 28 January 2013
Accepted 28 January 2013
Available online 15 March 2013

Keywords:
Genetic syndromes
Executive functions
Intellectual disability
Neuropsychological profile

ABSTRACT

Executive functions are a set of high cognitive abilities that control and regulate other functions and behaviors and are crucial for successful adaptation. Deficits in executive functions are frequently described in developmental disorders, which are characterized by disadaptive behavior. However, executive functions are not widely examined in individuals with intellectual disability. The present study is aimed at evaluating the etiological specificity hypotheses pertaining to executive functions by comparing individuals with intellectual disability of different etiology, as Williams syndrome and Down syndrome, on different aspects of executive functions. To this aim a battery evaluating attention, short-term and working memory, planning, categorization, shifting and inhibition, was administered to 15 children, adolescents and adults with Williams syndrome, to 15 children, adolescents and adults with Down syndrome and to 16 mental-age-matched typically developing children. The two groups with intellectual disability showed impairment in a set of executive functions, as auditory sustained attention, visual selective attention, visual categorization and working memory, and preserved visual sustained attention, auditory selective attention and visual inhibition. However, a distinctive profile has been found between the two syndromic groups on other executive functions. While participants with Down syndrome were poor in shifting and verbal aspects of memory and inhibition, those with Williams syndrome were poor in planning. The specific weakness and straights on executive functions may support the etiological specificity hypothesis accounting for distinctive cognitive development syndrome-specific.

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

* Corresponding author. Tel.: +39 06 68592475.
E-mail addresses: stefano.vicari@opbg.net, stefanovicari@hotmail.it (S. Vicari).
[1] These authors contributed equally to this work.

0891-4222/$ – see front matter © 2013 Elsevier Ltd. All rights reserved.
http://dx.doi.org/10.1016/j.ridd.2013.01.024

(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.