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How executive functions are related to intelligence in Williams syndrome

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ABSTRACT

Williams syndrome is characterized by impairments in executive functions (EFs). However, it remains unknown how distinct types of EFs relate to intelligence in this syndrome. The present study analyzed performance on working memory, inhibiting and shifting, and its links to IQ in a sample of 17 individuals with WS, and compared them with a group of 17 typically developing individuals matched on chronological age and gender. In conclusion, our results suggest that working memory, inhibiting, and shifting relate differently to intelligence in WS as well as in typical development, with working memory being the EF most closely related to intelligence in both groups. Notably, the magnitude of the associations between the three EFs and IQ was substantially higher in the WS group than in the TD group, bringing further confirmation to the notion that frontal lobe impairments may produce a general compromise of several EFs.

1. Introduction

Williams syndrome (WS) is a rare neurodevelopmental disorder caused by a submicroscopic deletion on the long arm of chromosome 7q11.23 (Korenberg et al., 2000), with an approximate incidence of 1 in 7500 live births (Strømme, Bjømstad, & Ramstad, 2002). This syndrome is characterized by a distinctive pattern of physical (e.g. facial dysmorphia), medical (e.g. cardiovascular problems), socio-emotional (e.g. heightened empathy) and cognitive (e.g. moderate mental delay) features (Bellugi, Korenberg, & Klima, 2001; Mervis & Klein-Tasman, 2000; Sampaio et al., 2009). Individuals with WS have been of interest to researchers in the area of cognitive neuroscience because of their distinct cognitive profile of peaks and valleys, with relative verbal strengths contrasting with weaknesses in non-verbal domains (Atkinson et al., 2003; Bellugi, Lichtenberger, Jones, Lai, & St. George, 2000; Farran, Jarrold, & Gathercole, 2001; Mervis & Klein-Tasman, 2000; Semel & Rosner, 2003). Furthermore, individuals with WS show ample evidence of frontal lobe dysfunction (Menghini, Addona, Costanzo, & Vicari, 2010; Mobbs et al., 2007; Rhodes, Riby, Park, Fraser, & Campbell, 2010). In fact, these individuals show a higher prevalence of hyperactive and impulsive symptoms than expected, with attention-deficit/hyperactivity disorder (ADHD) as one of the most frequent co-morbid diagnoses (Dodd & Porter, 2009; Kennedy, Kaye, & Sadler, 2006).

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There has been increasing interest in studying the level of impairment in the executive functioning of individuals with WS. The term "executive function" (EF) refers to a range of processes such as working memory, inhibitory control and attentional shifting, which are believed to depend on frontal lobe activity and underlie goal-directed responses to novel and challenging situations (Friedman et al., 2006; Miyake et al., 2000). Porter, Coltheart, and Langdon (2007) found that a sample of 20 individuals with WS (aged 5–46 years) displayed significantly poorer response inhibition than expected based on their mental age and intellectual functioning. Rhodes et al. (2010) compared the performance of a sample of 19 individuals with WS (11–29 years of age) on a set of three EF tasks with two control groups: a chronological age and gender-matched group and a verbal ability and gender-matched group. They found that the WS group presented with impairments compared to typically developing groups in all three tasks, which involved attention set-shifting, spatial working memory and planning. Menghini et al. (2010) extended these results by providing evidence of deficits in both verbal and visuospatial EF tasks in WS participants compared with mental age-matched controls.

One important issue that has been neglected is how distinct types of executive functions relate to intelligence in WS. Because this syndrome is characterized by frontal lobe dysfunction, which is associated with executive impairments, it seems necessary to study the pattern of associations between different types of EF and intelligence. As no WS studies have been conducted on this subject, we review evidence from normative samples. Although measures of EF are moderately correlated in typical development, they are not redundant, which suggests that that they may have different associations with intelligence (Miyake et al., 2000). Indeed, over the course of the past two decades, research with normative samples has shown that inhibiting, working memory and shifting display distinct patterns of association with general cognitive ability. Regarding intelligence and inhibiting, Anderson and Spellman (1995) state that inhibition is a necessary mechanism in human cognition that relies on both excitatory and inhibitory processes and contributes to the stability of the neural networks. Despite scant empirical supporting evidence, the role of inhibition is accepted as being present in several cognitive skills such as selective attention, language comprehension and production, memory retrieval and analogical reasoning (Anderson & Spellman, 1995; Das, 2002; Michel & Anderson, 2009). In addition, Dempster (1991) and Dempster and Corkill (1999) reviewed the available evidence and concluded that inhibition seems to be an important factor in intelligence in non-clinical samples. More recently, Polderman et al. (2009) found an association between inhibitory control and intelligence in 9-, 12-, and 18-year-olds. Regarding working memory, a meta-analysis of 86 samples found evidence of moderate to strong correlations between working memory and intellectual ability (Ackerman, Beier, & Boyle, 2005). Indeed, working memory has been implicated in distinct mechanisms of human cognitive function (Jarrold & Towse, 2006). Some studies have failed to confirm a link between intelligence and shifting (Friedman et al., 2006; Rockstroh & Schweizer, 2001). However, Ardila, Pineda, and Rosselli (2000) reported correlations between the Wisconsin Card Sorting Test, a measure of shifting, and adult IQ on the WAIS. Similar results were found in a sample of children aged 3–6 years, whose performance on a dimensional-change card sort task (with high demands in terms of shifting) was strongly correlated with measures of intelligence (Hongwanishkul, Happaney, Lee, & Zelazo, 2005). Interestingly, only one study has analyzed the association between performance on tasks requiring all three types of EF and intelligence (Friedman et al., 2006). The authors found that updating working memory significantly correlated with intelligence in a very large sample of healthy adolescents aged 16–18 years, although the same results were not found with inhibiting and shifting.

To date, there have been no similar studies exploring the possible links between distinct types of EF and intelligence in WS.

The purpose of the present work was threefold. First, we compared performance on tasks assessing working memory, inhibiting and shifting between two groups: (a) a group of individuals with WS aged 10–29 years and (b) a group of typically developing individuals matched by chronological age and gender. Given the wealth of studies suggesting that frontal lobe impairments are a characteristic of this syndrome, and based on prior evidence, we expect individuals with WS to show significantly poorer performance than TD individuals on all three types of EF.

Second, we aimed to explore the pattern of associations between the three types of EF. We expect significant intercorrelations for both groups (for evidence for the TD group, see Friedman et al., 2006; Miyake et al., 2000), but these intercorrelations may be particularly evident in the clinical group. Because WS is characterized by frontal lobe impairments, we anticipate that all measures of EF may be affected and may vary together.

Finally, we aimed to explore the pattern of associations among the three types of EF (working memory, inhibiting and shifting) and intelligence in the two groups. In accordance with a recent report by Friedman et al. (2006), we posit that only working memory is associated with intelligence in the TD group. Due to frontal lobe impairments in the WS group, a more general compromise of EF may be present, which may be reflected in significant associations between EF and intelligence measures.

2. Materials and methods

2.1. Participants

Two groups of participants took part in this study. Seventeen participants with a diagnosis of WS (10 females and 7 males, with ages ranging from 9.6 to 29.3 years ($M = 20.6$, $SD = 5.9$)) with previously confirmed positive fluorescence in situ hybridization (FISH) to an elastin gene deletion in chromosome 7 (Ewart et al., 1993) were recruited at Fundación Pública Galega de Medicina Xenómica (Santiago de Compostela, Spain). The exclusion criteria were the presence of any sensorial or speech disorder, as well as comorbidity with severe psychopathology not associated with the syndrome. The control group