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Memory abilities in Williams syndrome: Dissociation or developmental delay hypothesis?

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Abstract

Williams syndrome (WS) is a neurodevelopmental genetic disorder often described as being characterized by a dissociative cognitive architecture, in which profound impairments of visuo-spatial cognition contrast with relative preservation of linguistic, face recognition and auditory short-memory abilities. This asymmetric and dissociative cognition has been also proposed to characterize WS memory ability, with sparing of auditory short-term memory and impairment of spatial and long-term memory abilities. In this study, we explored the possibility of a double memory dissociation in WS (short- versus long-term memory; verbal versus visual memory). Thus, verbal memory abilities were assessed using California Verbal Learning Test and Digit Span and Rey-Osterrieth Complex Figure and Corsi Blocks was used to assess visual–spatial memory abilities. Overall, WS subjects were found to present a generalized significant impairment in verbal and visuo-spatial components either in short- or long-term memory. In sum, data from this study brings support for a developmental delay hypothesis, rather than a double dissociation within memory systems in WS.

Keywords: Williams syndrome; Memory dissociation; Neurodevelopment

1. Introduction

Williams syndrome (WS) is a neurodevelopmental disorder, with a prevalence of 1 in 7500 (Stromme, Bjornstad, & Ramstad, 2002), characterized by a deletion on chromosome 7 q11.22-23 (Korenberg et al., 2000). Of further note, WS patients have an unusual phenotype, which includes a distinctive profile of physical, medical, neurocognitive and neuroanatomical characteristics. Physical characteristics include craniofacial and cardiac/pulmonary abnormalities, growth delay, hypercalcemia, hyperacusis and feeding difficulties (Metcalfe, 1999). The other main component of the WS phenotype is a neurodevelopment/cognitive profile characterized by mental retardation and an asymmetrical development, with weak and strong areas of performance. Specifically, severe impairment of visuo-spatial cognition has been described as coexisting with a relative preservation of face recognition, auditory short-term memory, language and narrative skills (Bellugi, Lichtenberger, Jones, Lai, & St. George, 2000; Gonçalves et al., 2004, 2005; Mervis et al., 2000).

This uneven cognitive profile of relative strengths and weaknesses also seems to be evident within each cognitive domain. For example, several studies found evidence for the existence of a good short-term verbal memory, even when compared with normal development controls (Bellugi, Wang, & Jernigan, 1994; Mervis, Morris, Bertrand, & Robinson, 1999; Nichols et al., 2004). However, several authors have been finding that this short-term verbal memory strength is not generalized across different memory sys-

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tems (Vicari, Bellucci, & Carlesimo, 2003; Vicari, Brizzolara, Carlesimo, Pezzini, & Volterra, 1996). For example, differences of ability in short- and long-term memory storage systems were found in a study by Vicari et al. (1996). The authors compared memory performance on Digit Span, Corsi Blocks, Rey-Osterrieth Complex Figure and a word list task in a WS group and a verbal mental age matched group. While a spatial span (Corsi Blocks) was found to be significantly reduced in the WS group, no significant differences were found in terms of verbal span. Additionally, no primacy effect in the serial position curve was found with the WS group. The authors interpreted their results as an evidence for the impairment of semantic long-term memory along with the preservation of short-term phonological memory. This claim was further supported by Nichols et al. (2004) using California Verbal Learning Test (CVLT).

More recently, Brock, Brown, and Boucher (2006) replicated the Vicari et al. (1996) study, introducing some procedural changes. Namely, the serial position curve was obtained in three different presentation conditions: different words, repeated words in different order and a repeated list. Using this strategy, both groups (WS and control) showed a similar pattern of recency, but no primacy effects in serial position curve. Additionally, participants were trained with a cumulative rehearsal strategy in order to determine the primacy effect extension. Again, no significant differences were found between groups in global performance (i.e., the rehearsal strategy was not associated with a better performance), but now, a primacy effect was observable in both groups. The authors suggested that there is not a selective long-term memory deficit in WS and that the absence of a primacy effect in WS may related with the inability in using rehearsal strategies.

A dissociation in working memory components (between phonological loop and visuo-spatial sketchpad) has been also proposed for characterizing cognitive profile of WS (Jarrold, Baddeley, & Hewes, 1999). These authors tested the hypothesis of the preservation of phonological loop coexisting with impaired visuo-spatial sketchpad in WS. Twenty-five children with Down syndrome (DS) were compared with 16 children and young adults with WS and 17 children with moderate learning disabilities, in short-term verbal memory (Digit Span) and short-term visuo-spatial memory (Corsi Blocks). The results revealed that WS had the lowest score in short-term visuo-spatial memory contrasting with a superior Digit Span performance. In a second study, the authors matched three groups in terms of nonverbal mental age (WS, learning disabilities and a normal development), comparing their performance in three short-term memory measures: one verbal (Digit Span) and two nonverbal (Corsi Span and Pattern Memory). Again, the results showed that WS group had significant lower scores in Corsi Blocks. However, in the Pattern Memory test, WS group was not impaired with respect to the control group. The authors concluded suggesting the existence of a specific impairment in visuo-spatial sketchpad and a relative sparing of the phonological loop in WS. However, more recently, Jarrold, Baddeley, and Phillips (2007) proposed that these visuo-spatial memory deficits may be secondary to visuo-spatial processing difficulties, a main feature of WS cognitive profile (Mervis et al., 2000). Indeed, WS patients exhibit difficulties in several aspects of visual–spatial cognition, including spatial representations (Farran & Jarrold, 2005; Hoffman, Landau, & Pagani, 2003), abilities that are necessary to perform some visual–spatial memory tasks. The preservation of WS performance in Pattern Memory Test was interpreted by Vicari and colleagues (2003) as suggesting that WS represents a natural case of dissociation within the two components of visuo-spatial sketchpad, in which visual perception is relatively preserved. These results were further investigated with respect to long-term memory system, showing that individuals with WS had impairments in learning visual–spatial material, although a typical performance in visual-object long-term memory (Vicari, Bellucci, & Carlesimo, 2005).

Overall, previous studies have been showing evidence for an uneven performance in WS individuals across different memory tests. The current study attempts to confirm the existence in WS of a double dissociation in memory systems between short- versus long-term memory as well as between verbal and visuo-spatial working memory components.

2. Method

2.1. Participants

A group of 14 individuals with WS (7 male and 7 female participants), with age range between 8 and 29 years ($M = 16.79$, $SD = 5.68$; mean Full Scale IQ = 49.14, $SD = 7.50$, range 40–61) was compared with a normal development group ($N = 14$, 5 male and 9 female) ranging in age from 8 to 29 years ($M = 17.93$, $SD = 6.10$; mean Full Scale IQ = 110.50, $SD = 10.48$, range 90–124). Given that the neurological profile of WS may change across development (Gagliardi, Martelli, Burt, & Borgatti, 2007), two subgroups were compared on the basis of chronological age for both WS and normal development participants: children ($N = 5$, age 8–14 years) and adult ($N = 9$, age 15–29 years) subgroups.

WS participants were recruited at Genetic Medical Institute (Portugal) and Genomic Foundation in Galicia (Spain). WS diagnoses were made by FISH confirmation of elastin gene deletion (Korenberg et al., 2000). Controls were typically developing individuals without evidence of psychiatric, neurological disorder or cognitive impairment. Each participant gave written informed consent for their participation in the study via consent forms, after a complete description of the study.

2.2. Instruments

To assess general cognitive functioning (Full Scale IQ), participants 8–16 years of age were administered the