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The extent of working memory deficits associated with Williams syndrome: Exploration of verbal and spatial domains and executively controlled processes

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ABSTRACT

The present study investigated verbal and spatial working memory (WM) functioning in individuals with the neuro-developmental disorder Williams syndrome (WS) using WM component tasks. While there is strong evidence of WM impairments in WS, previous research has focused on short-term memory and has neglected assessment of executive components of WM. There is a particular lack of consensus concerning the profile of verbal WM functioning in WS. Here, WS participants were compared to typically developing participants matched for (1) verbal ability and (2) spatial ability ($N = 14$ in each of the 3 groups). Individuals with WS were impaired on verbal WM tasks, both those involving short-term maintenance of information and executive manipulation, in comparison to verbal-matched controls. Surprisingly, individuals with WS were not impaired on a spatial task assessing short-term maintenance of information in memory (remembering spatial locations) compared to spatial-matched controls. They were, however, impaired on a spatial executive WM task requiring the manipulation of spatial information in memory. The present study suggests that individuals with WS show WM impairments that extend to both verbal and spatial domains, although spatial deficits are selective to executive aspects of WM function.

1. Introduction

Williams syndrome (WS) is a neurodevelopmental disorder with confirmed genetic origin. The disorder is characterised by a sporadic deletion of approximately 25 genes on chromosome 7, specifically 7q11.23 (Ewart et al., 1993). The prevalence of WS is estimated to be between 1 in 7500 (Strømme, Bjørnstad, & Ramstad, 2002) and 1 in 20,000 live births (Morris & Mervis, 1999). The disorder has attracted research attention due to unique aspects of its associated behavioural, social and cognitive phenotypes. Cognitively, WS is associated with a distinct profile of relatively more proficient skills within the verbal domain compared to the severe impairments of visuo-spatial processing (e.g. Hoffman, Landau, & Pagani, 2003; Jarrold, Baddeley, & Hewes, 1999; Mervis, 1999; Vicari, Bellucci, & Carlesimo, 2003; Wang & Bellugi, 1994), although this distinct profile of relative preservation and impairment has been challenged (e.g. Brock, 2007; Porter & Coltheart, 2005, 2006). Importantly, performance within both domains falls short of that expected by chronological age due to overall functioning within the mild-moderately learning difficulties range (Searcy et al., 2004).

With regards to a profile of memory skills, accumulating evidence suggests impaired working memory (WM) function in individuals with WS (Jarrold, Cowan, Hewes, & Riby, 2004; Menghini, Addona, Costanzo, & Vicari, 2010; O'Hearn, Courtney, Street, & Landau, 2009; Rhodes, Riby, Park, Fraser, & Campbell, 2010). Importantly, in a recent study of short-term memory (STM), functioning was shown to be correlated with behavioural difficulties in children with WS (Rhodes et al., 2010). Surpisingly, despite the extensive focus on a verbal/spatial distinction previously noted in the literature and the distinction made between these components within popular models of WM (e.g. Baddeley, 1986; Baddeley & Hitch, 1974), no study to date has directly examined both verbal and spatial short-term memory (storage) and executive aspects of memory (storage + processing) in the same individuals who have the disorder. The present study set out to address this gap in the literature.

While various models of WM have been proposed (e.g. Baddeley, 1986; Cowan, 1995; Miyake, Friedman, Rettinger, Shah, & Hegarty, 2001; Shah & Miyake, 1996) the weight of evidence supports Baddeley's theoretical WM component model (Baddeley, 1986, 2006; Baddeley & Hitch, 1974). This model identifies verbal (phonological loop) and spatial (visuo-spatial sketchpad) components of WM involved in the storage of information and a 'central executive' wherein stored information is controlled and

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manipulated. The central executive is involved for conditions of high level processing, while the storage of information in memory is thought to involve domain-specific storage components; namely the phonological loop and visuo-spatial sketchpad. Furthermore, there is evidence that the visuo-spatial sketchpad comprises separate visual and spatial subcomponents: the visual cache and the inner scribe (Logie, 1995). A common mechanism is thought to underlie both verbal and spatial information processing (Baddeley & Logie, 1999) although some models (e.g. Cowan, 1999) argue that domain-general resources are responsible for processing and storage. Accumulating evidence supports the Baddeley model (e.g. Alloway, Gathercole, & Pickering, 2006; Bayliss, Jarrold, Gunn, & Baddeley, 2003). For example, Bayliss et al. (2003) reported that complex span performance was independently limited by individual differences in domain-general processing efficiency and domain-specific storage capacity. This finding questions a shared resource pool between processing and storage. Evidence from developmental studies suggest that this theoretical structure of WM, with a common processing mechanism but distinct storage systems, is evident from as young as 4 years of age in neuro-typical development (Alloway et al., 2006).

While limited research has assessed both STM (i.e. storage) *and* executive aspects of WM in WS, it is possible to gain an idea of WM profiles associated with the disorder across a number of published studies. Several recent studies have reported strategic/executive cognitive difficulties including the ability to hold and update spatial information and use strategies within WM (Menghini et al., 2010; Rhodes et al., 2010). Impaired executive function goes beyond WM to include components of inhibition, planning, and attention set-shifting (Menghini et al., 2010; Rhodes et al., 2010). In a recent study we reported that the profile of executive function impairments associated with WS is remarkably similar to that of ADHD (Rhodes, Riby, Matthews, & Coghill, 2011) a neurodevelopmental disorder known to be associated with impaired executive functioning (Rhodes, Coghill, & Matthews, 2004, 2005). To date, within the WS literature, reports of executive WM impairments are restricted to the non-verbal domain. An important aim of the current study was therefore to examine both verbal *and* spatial executive aspects of WM in a sample of individuals with WS.

Memory difficulties in WS appear to extend beyond executive aspects of processing to include short-term memory under particular task conditions. While individuals with WS appear relatively competent at holding visual information such as objects in memory (Vicari et al., 2003), they show significant difficulties when they have to link visual and spatial information; such as the location of that object within an array (Jarrold et al., 1999; Vicari, Bellucci, & Carlesimo, 2006). O'Hearn and colleagues (2009) and Rhodes et al. (2010) have reported that the length of delay between remembering and recalling information is also critical to short-term memory performance in this group (O'Hearn et al., 2009). In terms of the distinction between short-term and executive aspects of WM, these studies suggest that individuals with WS show particular difficulties on strategic / executive tasks or in short-term memory when multiple items have to be held in memory or held in memory over a delay interval beyond a few seconds duration. The studies outlined here reported short-term and executive WM impairments across a range of tasks with different stimuli and varying task requirements. The current study aimed to address this using short-term and executive WM tasks designed with identical task parameters with the sole distinction between them being the requirement to manipulate (in addition to maintain/store) information in WM.

Research that has examined verbal WM in WS samples is limited and all published studies, to our knowledge, specifically focus on short-term aspects of verbal WM functioning. These studies have produced highly inconsistent findings. While a number of studies report intact verbal short-term memory (Bellugi, Wang, & Jernigan, 1994; Crisco, Dobbs, & Mulhern, 1998; Majerus, Barisnikov, Vuillemin, Poncelet, & Van der Linden, 2003; Mervis, Morris, Bertrand, & Robinson, 1999; Nichols et al., 2004; Sampaio, Sousa, Fernandez, Henriques, & Goncalves, 2008; Vicari, Brizzolaram, Carlesimom, Pezzinim, & Volterram, 1996; Vicari, Carlesimo, Brizzolara, & Pezzini, 1996) others report impaired verbal short-term memory functioning (Barisnikov, Van der Linden, & Poncelet, 1996; Grant et al., 1997; Jarrold, Baddeley, Hewes, Leeke, & Phillips, 2004). Further inspection of these studies reveals that the majority reporting no significant impairment of verbal WM did not include a verbal ability matched comparison group. In contrast, the studies which report significant verbal short-term memory impairment all included some type of verbal matched comparison group. Therefore, despite their relative strengths in verbal functioning it seems likely that individuals with WS show verbal short-term memory impairments. Whether this extends to executive aspects of verbal WM is, however, currently unknown.

The current study set out to systematically examine verbal WM components in WS with direct comparison to a vocabulary matched comparison group of typically developing individuals. In addition an important question that is unanswered in the literature concerns whether the verbal/spatial dissociation that has been reported in the cognitive profile of the disorder, extends to memory functioning. The widely held view of impaired spatial functioning in the face of relatively preserved verbal functioning suggests that individuals would show greater impairment on spatial than verbal WM tasks. A detailed examination of verbal and spatial WM components (STM storage and central executive involvement) is clearly warranted and the current study set out to address this. An important aim of this study was to identify whether individuals with WS showed significant WM impairment over and above their spatial difficulties. We predicted that individuals with WS would show impaired WM performance on both spatial and verbal tasks, with a larger deficit on spatial tasks. We thus compared the performance of individuals with WS on verbal and spatial short-term and executive WM tasks with verbal and spatial ability matched control groups who were developing typically.

2. Method

2.1. Participants

Nineteen participants with WS were recruited via the William syndrome Foundation. In line with researchers who have reported performance on similar WM component tasks with clinical samples (e.g. Cannon et al., 2005; Kim, Glahn, Nuechterlein, & Cannon, 2004), only individuals who scored above chance on the verbal and spatial storage (i.e. STM) tasks were included in the final dataset ($N = 14$). All participants had previously been diagnosed phenotypically by clinicians and all had previously had their diagnosis confirmed with positive FISH testing. All participants were familiar with the research process. Participants with WS (i.e. $N = 14$) ranged between 11 years and 6 months of age and 29 years and 6 months of age (see Table 1 for mean and demographics).

In line with the verbal/spatial focus of the current study, participants with WS were matched to one typically developing individual on the basis of spatial ability (SM group) and one typically developing individual on the basis of verbal ability (VM group) (See Table 1). The matching measures were two of the most common implemented in learning difficulty research; namely the *Ravens Coloured Progressive Matrices* task (RCPM; Raven, Court, & Raven, 1990) and the *British Picture Vocabulary Scale* (BPVS II; Dunn, Dunn, Whetton, & Burley, 1997). Spatial ability was assessed using the RCPM, a child-version of the Ravens' Standard Progressive Matrices (Raven