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Attentional lapse and inhibition control in adults with Williams Syndrome

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ABSTRACT

Research exploring cognitive processing associated with Williams Syndrome (WS) has suggested that executive functioning deficits exist across the developmental spectrum. Such executive functions include problem solving, planning, dividing attention and inhibiting responses. Within a framework of executive functions, the aim of the current study was to explore attentional lapse and inhibition skills in older adults with WS ($n = 20$; aged 36–61 yr) and consider the implications of deficits within this group. Participants with WS were compared to typical adults of the same chronological age and typical older adults (aged 65+ yr) to consider attentional changes seen in the ageing process. The study employed a sustained attention to response task known to assess inhibition and attentional lapse but which had not previously been used with this population. Compared to both groups of typical matches, the results indicated atypicalities of attention and inhibition in adults with WS. Specifically, compared to the typical matches, adults with WS failed to withhold a response (showing inhibition deficits), had problems re-engaging attentional control processes after making an error and showed a generalised deficit of concentration and task engagement. We conclude that further attention should be paid to the cognitive capacity of older individuals with WS in order to consider the everyday challenges faced by this group and to provide adequate intervention and support for daily living.

1. Introduction

Williams Syndrome (WS) is a neurodevelopmental disorder with a prevalence of 1:20,000 (Wang et al., 1997; but see also 1:7500 Strømme, Bjørnstad, & Ramstad, 2002) that is caused by a micro-deletion of approximately 28 genes on chromosome 7 (Osborne & Mervis, 2007). Individuals with the disorder tend to function within the mild-moderate range of intellectual difficulty (Searcy et al., 2004) and exhibit a cognitive profile of relative proficiency within the verbal compared to the non-verbal domain (Bellugi, Lichtenberger, Mills, Galaburda, & Korenberg, 1999). The cognitive profile of the disorder has attracted the attention of cognitive scientists for the last two decades due to this juxtaposition of relatively better verbal than non-verbal skill, but it is critical to emphasise that heterogeneity of cognitive function occurs (Porter & Coltheart, 2005) and the relative difference between verbal and spatial skill co-exists against a background of mild-moderate intellectual difficulty. Within the cognitive profile, research has recently highlighted the importance of exploring the area of executive

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functioning (e.g. Rhodes, Riby, Park, Fraser, & Campbell, 2010) since the successful engagement of such processing mechanisms is closely related to everyday cognitive ability. Executive function (EF) is an umbrella term that encompasses a range of higher order cognitive processes that control and regulate functions such as working memory, problem solving, planning, divided attention and inhibition and which are predominantly controlled by frontal brain regions (Alvarez & Emory, 2006). Here, we focus on response inhibition and lapses of attention as these are executive skills with clear implications for understanding wider deficits related to facets of the WS phenotype (e.g. the inability to inhibit inappropriate social approach behaviour, Little et al., 2013).

In research exploring executive functioning in WS, there is no consensus regarding the precise components of executive ability that are more or less impaired. However, in a recent paper in the area Costanzo et al. (2013) examined a variety of executive function tasks in children, and younger and older adults with WS (range 11–35 yr olds) compared to Down Syndrome (DS) and mental-age matched typical controls. Planning ability was particularly compromised in the WS group, with mixed finding found in categorization and inhibition, particularly with regards the modality of the tests employed (i.e. visual vs. auditory tasks yielding inconsistent results; see Osório et al., 2012 who also employed a battery of executive function tasks and again report inconclusive findings).

Somewhat more informative, research has suggested that some individuals with WS share executive function characteristics with individuals who have Attention Deficit Hyperactivity Disorder (ADHD; Rhodes et al., 2010). Comorbid ADHD is relatively more common in WS (64%; Leyfer, Woodruff-Borden, Klein-Tasman, Fricke, & Mervis, 2006) than it is in other disorders such as DS (6–8%; Dykens, 2007). Important here is the fact that ADHD is a neurodevelopmental disorder characterised by impaired attention, hyperactivity, impulsivity and disinhibition (Nigg, 2001; Rhodes, Riby, Matthews, & Coghill, 2011) and which is linked to executive-frontal lobe deficits within the brain (Willcutt, Doyle, Nigg, Faraone, & Pennington, 2005). Focussing specifically on inhibition, possible primary, and at least secondary, causes of the behavioural deficits observed in ADHD can be explained by disinhibitory deficits (Nigg, 2001). Recent fMRI work concurs that the executive impairment observed in WS mirrors the patterns seen in ADHD. In that study, Mobbs et al. (2006) employed fMRI while participants with WS (aged 15–48 yr) performed a *Go/No Go* measure of sustained attention and inhibition. The authors concluded that observed dis-engagement of the frontal-striatal networks of the brain contributed to the complex pattern of social and behavioural deficits associated with WS (see Hocking et al., 2013 who examined dual tasking and inhibition in the motor domain). In summary, work that has administered batteries of executive function tasks have been inconclusive while those that have specifically examined inhibition are promising in pinpointing the precise executive cognitive processes impaired in WS.

We have noted that EF has been linked to other facets of the WS phenotype. Cognitive aspects of inhibition can be linked to a social phenotype characterised by a tendency to indiscriminately approach both familiar and unfamiliar people (Jones et al., 2000). Using Cluster Analysis to explore heterogeneity of social approach within WS, Little et al. (2013) noted that the participants who showed most indiscriminate and atypically heightened approach ratings to unfamiliar faces were also those individuals who struggled with the Sun-Moon inhibition task (as opposed to relating to emotion processing ability or intellectual capability; Little et al., 2013). The authors proposed that the finding provided preliminary support for a frontal lobe hypothesis of atypical social behaviour within the disorder. The study emphasised the necessity to explore inhibition abilities in individuals with WS due to their link to other facets of the disorder. For example, identifying the role of inhibition in abnormal social approach may mean that intervention can target this skill within a broad intervention approach that considers the cognitive and behavioural needs of the individual.

The first aim of the current study was to investigate inhibitory processing in adults with WS (aged 35+ yr), as to date there is limited research that focusses on these behaviours in an older adult cohort. It is not unreasonable to predict particular inhibition deficits in an older WS sample given (1) typically developing older adults suffer from executive deficits (see frontal ageing hypothesis; Greenwood, 2000; inhibition deficit hypothesis; Hasher & Zacks, 1988) and (2) older adults with WS have been argued to suffer from "mild accelerated ageing" (Krinsky-McHale, Kittler, Brown, Jenkins, & Devenny, 2005; p. 483). For these reasons we also incorporated an elderly typically developing comparison group to help in the data interpretation. The second aim was to employ a task that would enable a comprehensive examination of lapses of attention and inhibition which had previously been demonstrated to be related to real world activities in other populations, including individuals with a neurodevelopmental disorder (e.g. ADHD as well as traumatic brain injury, TBI; see Smilek, Carriere, & Cheyne, 2010 for discussion). The paradigm used was the Sustained Attention to Response Task (SART; Robertson, Manly, Andrade, Baddeley, & Yiend, 1997), a vigilance task which required the participant to respond to a frequent non-target stimulus and withhold a response to an infrequent target stimulus. There were three main metrics derived from the task. First, false alarm (FA) commission errors where participants failed to inhibit a response to non-target infrequent stimuli were used as a measure automaticity and inhibition. Secondly, and arguably the most sensitive measure, pre- and post-error reaction times after a commission error to reflect error monitoring was utilised. Finally, as a general measure of task engagement, differences in the variability of reaction time during the task were gathered as a further measure of attentional lapse (see Dockree et al., 2004; Smallwood, Riby, Heim, & Davies, 2006). Here we aim to elucidate how inhibitory deficits observed in older adults with WS during the SART compare with typically developing individuals matched for chronological age (CA) and gender, and with a group of typically developing adults aged 65 yr and over (65 yr). It was hypothesised that (1) the WS group would report greater deficits in failing to withhold a response compared with the CA and more similar to the over 65 yr groups with known difficulties in inhibitory control (Greenwood, 2000), (2) there would be no difference in the WS group's RT before and after a failure to withhold a response, similar to other populations with known deficits in error monitoring and executive control