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Attentional disengagement in adults with Williams syndrome

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ABSTRACT

Williams syndrome (WS) is a neurodevelopmental disorder characterized by a distinctive behavioral and cognitive profile, including widespread problems with attention. However, the specific nature of their attentional difficulties, such as inappropriate attentional allocation and/or poor attentional disengagement abilities, has yet to be elucidated. Furthermore, it is unknown if there is an underlying difficulty with the temporal dynamics of attention in WS or if their attentional difficulties are task-dependent, because previous studies have examined attention in established areas of deficit and atypicality (specifically, visuospatial and face processing). In this study, we examined attentional processing in 14 adults with WS (20–59 years) and 17 typically developing controls (19–39 years) using an attentional blink (AB) paradigm. The AB is the decreased ability to detect a second target when it is presented in close proximity to an initial target. Overall, adults with WS had an AB that was prolonged in duration, but no different in magnitude, compared with typically developing control participants. AB performance was not explained by IQ, working memory, or processing speed in either group. Thus, results suggest that the attention problems in WS are primarily due to general attentional disengagement difficulties rather than inappropriate attentional allocation.

1. Introduction

Williams syndrome (WS) is a neurodevelopmental disorder caused by a microdeletion of ∼28 genes on chromosome 7q11.23 (Ewart et al., 1993). Occurring in 1 in 7500 births, WS is associated with cardiovascular, endocrine and neurological problems, as well as a distinctive cognitive–linguistic profile (see Martens, Wilson, and Reutens (2008) for a review). Superimposed upon the intellectual disability usually occurring in the syndrome, individuals with WS show a significant relative weakness in visual–spatial processing but relative strengths in expressive language and facial recognition (although face processing may be atypical, Mills et al., 2000).

Considerable research has focused on cognition, language, and face processing in WS (Martens et al., 2008), yet research is needed to elucidate other areas of the WS phenotype (Dykens, 2003a, 2003b). One such area of particular relevance to WS is attention, as individuals with WS of all ages show difficulties with attentional processes. Distractibility or a short attention span has been endorsed by up to 95% of parents of children with WS on standardized checklists (Einfeld, Tonge, & Florio, 1997; Tomc, Williamson, & Pauli, 1990). In a recent study using the Conners Parent Rating Questionnaire, a measure of inattention, hyperactivity, and oppositional behaviors, 11 of 11 children were rated as "abnormal" on the ADHD index (Rhodes et al., 2010). Furthermore, Leyfer et al. (2006) found that 65% of 119 children with WS met DSM-IV criteria for Attention Deficit Hyperactivity Disorder (ADHD). Problems with attention appear to persist into adulthood. In semi-structured interviews with caregivers of adults with WS aged 19–55 years, up to 85% reported problems with distractibility (Elison, Stinton, & Howlin, 2010).

Beyond symptoms of ADHD, several studies point to difficulties with attentional disengagement in WS. Despite varying methodologies, these studies find that infants, children and adults with WS have difficulties disengaging attention, be it from initial fixation points or other static stimuli (Cornish, Scerif, & Karmiloff-Smith, 2007; Montfoort et al., 2007), incongruent information in a hierarchical stimulus (Farran, Jarrold, & Gathercole, 2003), human faces (Laing et al., 2002; Mervis et al., 2003; Riby & Hancock, 2009), or switching between auditory and visual modalities (Lincoln, Lai, & Jones, 2002). Individuals with WS also appear to have slower processing speed (Sampaio et al., 2009) and may experience difficulties choosing a target amongst distractors (Scerif et al., 2004). However, since the temporal dynamics of attention have yet to be assessed in WS, it is unknown if these results are task-dependent.

One way to address this question is to use a task that measures a number of discrete attentional domains without tapping

Abbreviations: WS, Williams syndrome; TD, typically developing; AB, attentional blink; RSVP, rapid serial visual presentation; DS, digit span; CN, cancellation.

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processes that are known to be aberrant in WS. The attentional blink (AB) paradigm does so, as it enables exploration of attentional processing in the temporal domain, and involves sustained attention, selective attention to targets, shifting attention between multiple targets, working memory, and response selection (Raymond, Shapiro, & Arnell, 1992). In typical AB paradigms, participants view rapid serial visual presentations (RSVP), in which a series of stimuli appear briefly (~100 ms or less) at the same location. Participants are asked to detect specific target stimuli in the visual stream. The AB refers to the decreased accuracy in reporting a second target that appears in close temporal proximity to a first target, which has been correctly identified. The AB, which generally occurs when the second target lags 200–400 ms behind the first target, is quite robust, occurring despite experimental manipulations regarding recognition vs. identification of targets, luminosity, presentation speed, and target color and type (for a review, see Dux & Marois, 2009).

The AB can be explained by a two-stage model of attention, in which stimuli are first rapidly identified (but susceptive to decay), and then consolidated in working memory (Chun & Potter, 1995). Only targets that reach the second stage undergo the necessary encoding and response selection that make them available for report. The common capacity-limited attentional resource model (Dux & Marois, 2009) proposes that stimulus encoding and response selection processes are attentionally demanding, and therefore limit the available attentional resources for handling subsequent stimuli. When two targets appear in close temporal proximity, they compete with each other to enter the second stage of processing, with the first target typically winning this competition, and the second target decaying before being encoded. This decay may be due to limited resources that prevent the second target from being encoded in stage 2, or to the simultaneous suppression of the second target (which should otherwise be enhanced because of its role as a target among distractors). Either way, the AB is due to the limits of the attentional system in processing successive stimuli presented at rapid speeds (Dux & Marois, 2009).

Beyond processing speed of individual targets, the AB paradigm explores both the attentional allocation to different targets and the attentional disengagement between targets. Inappropriate attentional allocation occurs when an individual focuses on one target at the expense of a second target, and may be deliberate or due to inappropriate attentional capture by the first stimulus. Attentional allocation can be quantified in the AB via the magnitude of the AB (i.e., decrement in accuracy for the second target, compared to first target accuracy). Greater AB magnitude would indicate inappropriate attentional allocation. Attentional disengagement refers to the amount of time an individual requires to disengage from a first target and engage with a second target and thus reflects the processing time needed between recognizing successive targets. Attentional disengagement time is quantified via the duration of the AB (i.e., the length of the temporal window with poorer accuracy for the second target). Individuals with greater attentional disengagement difficulties have an AB that is longer in duration. Thus, the AB task can reveal whether inappropriate attentional allocation and/or attentional disengagement difficulties contribute to the anomalous performance of people with WS on previously studied spatial or social attention tasks (e.g., Cornish et al., 2007; Riby & Hancock, 2009). If there is indeed an underlying difficulty with temporal attentional dynamics in WS, it may suggest that basic attentional training could improve their performance in these other domains.

The current study used an AB paradigm in a sample of adults with WS and typical age- and gender-matched controls. The first aim of the study was to identify to what extent anomalous attention processing in WS is associated with difficulties allocating attention versus disengaging attention. Additionally, although the AB has been studied in other developmental disabilities, including children with dyslexia (Facoetti et al., 2008), specific language impairment (Lum, Conti-Ramsden, & Lindell, 2007), high functioning autism (Rinehart et al., 2010), and ADHD (Mason, Humphreys, & Kent, 2005), to our knowledge an AB task has not been administered to a group that also has intellectual disabilities. As such, the second aim of the study was to identify neuropsychological correlates of the AB. We specifically examined the role of IQ, processing speed, and working memory in the AB in both the WS and control groups.

2. Methods

2.1. Participants

Fourteen adults with genetically-confirmed WS and 17 typically developing (TD) control participants completed the AB paradigm and neuropsychological testing. Individuals with WS were recruited from a residential summer camp, while TD participants were recruited from the community. There were no significant gender or age differences between the WS and TD groups; Table 1 summarizes demographic data and neuropsychological test scores across groups. Four additional WS participants were tested but were excluded from subsequent analysis because of their chance level performance. All participants with WS were reported to have normal or corrected-to-normal vision by their parents. All TD participants reported having normal or corrected-to-normal vision.

The study was approved by the Institutional Review Board (IRB) of the university and was conducted in accordance with the standards of the IRB and the Helsinki Declaration of 1975. Written informed consent was obtained from control participants and parents of participants with WS. Individuals with WS provided written assent.

2.2. Neuropsychological measures

Participants were administered the Kaufman Brief Intelligence Test, 2nd edition (KBIT-2; Kaufman & Kaufman, 2004), that yields verbal, non-verbal, and full scale IQ scores. Additionally, participants completed the Digit Span (DS) and Cancellation (CN) subtests from the Wechsler Adult Intelligence Scale, 4th edition (WAIS-IV) as measures of working memory and processing speed, respectively (Wechsler, Coalson, & Raiford, 2008). The WAIS-IV DS task includes forwards, backwards, and sequencing subscales, which are summed to form a total score. As expected, the TD group scored higher than the WS group on all measures (p's < 0.001). Consistent with previous research, WS participants had relative strengths in verbal vs. non-verbal IQ; such differences were not found in the TD group.

2.3. AB stimuli and procedure

AB stimuli were presented via RSVP, in which a series of letters flashed on a computer screen. Letters were presented in boldface

Table 1
Demographic information and neuropsychological measures.

	WS	TD	p-Value
Sex (% male)	64.3	52.9	0.524
Age (years)	28.36 ± 10.62	26.24 ± 5.71	0.483
Composite IQ (mean ± sd)	77.64 ± 16.31	109.19 ± 10.63	<0.001
Verbal IQ	83.00 ± 14.21	108.88 ± 10.97	<0.001
Non-verbal IQ	77.07 ± 16.38	106.56 ± 10.63	<0.001
Digit span scaled score	6.00 ± 2.77	11.19 ± 2.43	<0.001
Cancellation scaled score	3.50 ± 2.03	9.06 ± 2.91	<0.001