

THE ATTENTIONAL BLINK PARADIGM IN WILLIAMS SYNDROME

By

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CHAPTER I

INTRODUCTION

Williams syndrome (WS) is a neurodevelopmental disorder caused by a microdeletion of ~28 genes on chromosome 7q11.23 (Ewart et al., 1993). It is believed to be underdiagnosed, with newer prevalence estimates suggesting it may occur in 1 in 7,500 births, accounting for as many as 6% of individuals with genetic etiologies of intellectual deficit (Stromme, Bjomstad, & Ramstad, 2002). WS is associated with cardiovascular, endocrine and neurological problems, as well as a distinctive cognitive-linguistic profile (see Bellugi, Lichtenberger, Jones, Lai, & St. George, 2000 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 (though face processing may be atypical (Mills et al., 2000)).

As interest and research in WS has grown over the past three decades (Dykens & Hodapp, 2001), there is increasing awareness of the need to expand research into other domains of the WS profile and to tie together research in cognitive and non-cognitive domains (Dykens, 2003). One area of particular relevance to WS is attention. Research on individuals with WS of all ages suggests difficulties with attention and anomalous attentional processing. Indeed, nearly 65% of children with WS also meet DSM-IV criteria for an Attention Deficit Hyperactivity Disorder (ADHD) diagnosis (Leyfer, Woodruff-Borden, Klein-Tasman, Fricke, & Mervis, 2006). And many individuals who

may not meet criteria for ADHD appear to also struggle with problems with attention. In studies using parental checklists of behavior or temperament, distractibility is frequently endorsed by caregivers (Dilts, Morris, & Leonard, 1990; Tomc, Williamson, & Pauli, 1990), with short attention endorsed by 95% of caregivers in one study (Einfeld, Tonge, & Florio, 1997). These issues with attention appear to persist into adulthood. In recent studies based on semi-structured interviews with caregivers of adults with WS (19-55 years), between 45-85% reported problems with distractibility (Elison, Stinton, & Howlin, 2010; Stinton, Elison, & Howlin, 2010).

While most previous research has focused on the weaknesses in visual-spatial perceptual processing or atypical facial processing in WS in general rather than the role of attentional factors, a handful of these studies do provide a window into atypical attentional processes in WS through both observational (Mervis et al., 2003) and experimental paradigms (Cornish, Scerif, & Karmiloff-Smith, 2007; Farran, Jarrold, & Gathercole 2004; Laing et al., 2002; Lincoln, Lai, & Jones, 2002; Montfort, Frens, Hooge, Lagers-van Haselan, & van der Geest, 2007; Riby & Hancock, 2009; Scerif, Cornish, Wilding et al., 2004). Despite varying methodologies and populations ranging from infants to adults, all studies suggest difficulty with attentional disengagement in WS, be it from initial fixation points or other static stimuli (Cornish et al., 2007; Montfort et al., 2007), incongruent information in a hierarchical stimulus (Farran et al., 2004), or human faces (Laing et al., 2002; Mervis et al., 2003; Riby & Hancock, 2009). One pilot study of three adults with WS also found that they had difficulty disengaging their attention from visual to auditory modalities, and vice versa, especially at latencies below 2500 ms (Lincoln et al., 2002). However, since the temporal dynamics of attention have

yet to be explored in WS, it is unknown if these results are due to an underlying difficulty with attentional disengagement or if they are specific to tasks that tap areas with known atypicalities in WS. Additionally, without knowledge of the temporal attentional dynamics in WS, it is unclear if the pattern of performance is truly due to difficulty with attentional disengagement or if other factors, such as inappropriate attentional allocation to different stimuli, are involved.

The attentional blink (AB) is a well-studied phenomenon that enables exploration of attentional processing in the temporal domain (Broadbent & Broadbent, 1987; 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, and then participants report specific target stimuli from 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 is reported. The AB, which generally occurs when the second target lags 200-400 ms behind the first target, appears to be quite robust, occurring despite numerous experimental manipulations regarding recognition vs. identification of targets, luminosity, presentation speed, and target color and type (for reviews, see Dux & Marois, 2009 or Shapiro, Arnell, & Raymond, 1997).

The AB arises due to a two-stage model of attention, in which stimuli are first rapidly identified but susceptible to decay, and second are 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 encoding and response selection in the second stage is attentionally demanding, which limits the available attentional resources for handling subsequent stimuli. When two

targets appear in close temporal proximity to each other, they compete with each other to enter the second stage of processing, with the first target typically winning this competition (e.g., Marois & Dux, 2009; Potter, Staub, Rado, & O'Connor, 2002) and the second target decaying before being encoded. Thus, the AB results due to the limits of the attentional system in processing successive stimuli at rapid speeds.

There are many theories and frameworks for what specific processes cause the attentional bottleneck, such as limitations on attentional selection, working memory encoding, response selection, and distractor inhibition (see Dux & Marois, 2009, for a review). However, Dux & Marois (2009) also suggest that a common capacity-limited attentional resource may underlie all of these processes, leading to the AB. For example, this model proposes that while the first target is being processed in stage two, limited resources prevent the second target from entering stage two and being encoded, and also suppress the attentional representation of the second target (which should otherwise be enhanced because of its role as a target among distractors). Individuals with WS appear to have slower processing speed (Sampaio et al., 2009), and difficulty with both disengaging from stimuli (Cornish et al., 2007; Riby et al., 2009) and choosing a target amongst distractors (Farran et al., 2004; Scerif et al., 2004). The AB provides an opportunity to explore the limits of central attentional resources in WS in the temporal domain, without introducing confounds due to known difficulties with spatial processing or atypical face processing, as has been done in the previously mentioned studies in WS.

The AB paradigm is a useful tool in exploring attentional capacities because beyond allowing for examination of processing speed of individual targets (i.e., can an individual recognize a single target in an RSVP at a given speed), it explores both the

attentional allocation between different targets (i.e., focusing only on the first target at the expense of the second target), and the processing time between recognizing successive targets (i.e., the time an individual needs to disengage from the first target and engage the second target) in the temporal domain. Attentional allocation, which may be deliberate or due to inappropriate attentional capture by the first stimulus, can be quantified in the AB via the magnitude of the AB (i.e., poorer accuracy for the second target during the AB period due to over-allocation to the first target), while attentional disengagement time is quantified via the duration of the AB (i.e., poorer accuracy for the second target for an extended window following the first target). Therefore, the AB task provides an opportunity to isolate various aspects of attentional processing in WS, which may help elucidate whether inappropriate attentional allocation and/or an inability to disengage attention plays a role in their anomalous performance when factors such as spatial (Cornish et al., 2007; Montfort et al., 2007) or social (Laing et al., 2002; Mervis et al., 2003; Riby & Hancock, 2009) information are additionally involved in a task. These attentional anomalies can be detrimental for navigating the environment and learning about the world due to a decreased ability to monitor one's surroundings and link together related stimuli that are temporally or spatially distinct (Mervis et al., 2003).

The AB paradigm has been studied extensively in typical populations (Dux & Marois, 2009; Shapiro, Arnell, & Raymond, 1997), and in recent years, has been explored in atypical populations with aberrant attentional processes. An increase in AB magnitude or duration has been demonstrated in numerous clinical populations, including impulsive adolescents (Li, Chen, Lin, & Yang, 2005), children with dyslexia (Facoetti, Ruffino, Peru, Paganoni, & Chelazzi, 2008), Specific Language Impairment (Lum, Conti-

Ramsden, & Lindell, 2007), and high functioning autism (Amirault et al., 2009; Rinehart, Tonge, Brereton, & Bradshaw, 2009). An aberrant AB is also found in adults with dyslexia (Hari, Valta, Uutela, 1999), focal cerebral lesions (Rizzo, Akutsu, & Dawson, 2001; Shapiro, Hillstrom, & Husain, 2002), schizophrenia (Li et al., 2002; Wynn, Breitmeyer, Nuechterlein, & Green, 2007), and depression (Rokke, Arnell, Koch, & Andrews, 2002). Additionally, several studies have reported an aberrant AB in children with ADHD (Li, Lin, Chang, & Hung, 2004; Mason, Humphreys, & Kent, 2005) and adults with ADHD (Hollingsworth, McAuliffe, & Knowlton, 2001). However, the AB has yet to be explored in individuals with WS despite the high prevalence of ADHD and the anomalous attentional processing in the population. Additionally, to our knowledge, the AB has not been examined in populations with intellectual disabilities, so it is unknown how this might moderate the AB. This is a fundamental question because many populations with aberrant AB, such as ADHD and autism, also have lower IQ or intellectual deficit (Fombonne, 2003; Frazier, Demaree, & Youngstrom, 2004).

The aim of the current study was to assess the temporal dynamics of attention in WS via an AB paradigm in order to shed light on the nature of the proposed anomalous attention processing (attentional allocation versus attentional disengagement) in the syndrome, which may underlay their attentional difficulties when spatial or facial processing is additionally involved. As this is the first time an AB task has been administered to a group with intellectual disability, we also explored neuropsychological correlates of the AB to disentangle the role of intellectual abilities, processing speed, and working memory in the AB.

CHAPTER II

METHODS

Participants

Eighteen individuals with WS and 17 typically developing (TD) control participants completed the AB paradigm. Individuals with WS were recruited from the Vanderbilt Kennedy Center Williams Syndrome Music Camp while TD participants were recruited from the community. All individuals with WS had genetic testing confirming WS diagnosis. Demographic information, along with neuropsychological test scores (see *Neuropsychological Measures* below) can be found in Table 1. There were no gender or age differences between the WS and TD groups (p 's = NS).

Table 1: Demographic Information and Neuropsychological Measures

	WS	TD	p-value
Sex (% male)	61.1	52.9	0.625
Age (years)	28.11±9.80	26.24±5.71	0.497
Composite IQ (mean±sd)	71.50±18.55	109.19±10.63	<0.001
Verbal IQ	78.50±15.19	108.88±10.97	<0.001
Non-verbal IQ	70.06±19.75	106.56±10.63	<0.001
Digit Span Scaled Score	5.17±3.01	11.19±2.43	<0.001
Cancellation Scaled Score	3.17±1.98	9.06±2.91	<0.001

Neuropsychological Measures

Participants were administered the KBIT-2 (Kaufman & Kaufman, 2004), a brief IQ battery that reports a verbal, non-verbal, and full scale IQ composite score.

Additionally, participants completed the Digit Span (DS) and Cancellation (CN) subtests from the WAIS-4 as measures of working memory (WM) and processing speed (PS), respectively (Wechsler, Coalson, & Raiford, 2008). Neuropsychological measures were collected on all 18 WS participants and 16 of the 17 TD participants. Mean scores on these measures for the WS and TD groups can be found in Table 1. The TD group scored higher than the WS group on all measure ($p's < 0.001$). Consistent with other WS samples, WS participants had relative strengths in verbal IQ vs. non-verbal IQ. There was no difference between verbal and non-verbal IQ for the TD group ($p = .450$).

AB Stimuli and Procedure

Participants completed a rapid serial visual presentation (RSVP) task in which a series of letters flashed on a computer screen. Letter stimuli were presented in boldface Courier New font type, size 76, and were viewed from a distance of approximately 42 inches. Following a fixation asterisk, black letters were presented on a grey background for 107 ms each with a 13 ms interstimulus interval between letters (stimulus onset asynchrony of 120 ms). Each RSVP trial was randomly generated to contain 9 to 15 letters. Participants' task was to identify two targets embedded in the RSVP. The first target (T1) was a white letter (either B, G, or S), which appeared in serial positions 4-7 in the stream. The second target (T2) was a black X, which always appeared in position 4 or later, and was never the last letter presented. Distractor letters could be any letter of the alphabet except for B, D, G, K, S, X or Y.

Participants completed a total of 162 randomly ordered trials in one block, which lasted for approximately 18-20 minutes. In one-sixth of the trials, only T1 appeared; in

one-sixth of the trials, only T2 appeared; and in two-thirds of the trials, both T1 and T2 appeared. In these dual target trials, T2 appeared 1, 2, 3, or 7 positions after T1, with equal probability of T2 appearing at any lag position (see Figure 1). Following the last letter in the stream, participants were asked first “What was the white letter?” (response box options for B, G, S, or NO (if T1 was not seen)) and then “Was there a black X?” (responses for YES and NO). Participants recorded their own responses using a response box.

Prior to the full experiment, participants completed a practice block of 12 trials (three T1 only trials, three T2 only trials, three dual trials with T2 at lag 7). Practice trials were presented at the same speed as the full experiment. For both WS and TD participants, a research assistant sat in the testing room throughout the experiment to provide instructions for the task and answer any questions.

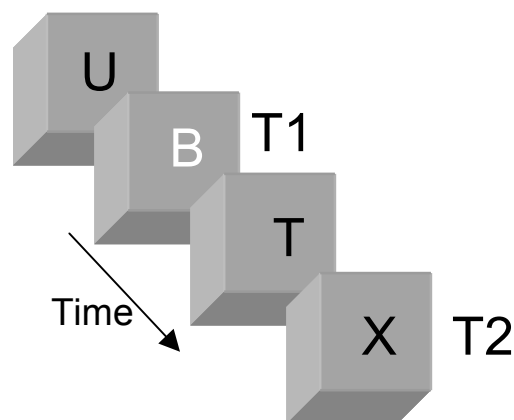


Figure 1. AB Paradigm

Statistical Analysis

Consistent with previous AB studies (Raymond et al., 1995; Rinehart et al., 2009; Rokke et al., 2002), target identification scores were converted to a criterion-free measure of discriminability, a' , for the dependent variable. A' is calculated as $(\text{hit} + \text{correct rejection}) / (\text{hit} + \text{correct rejection} + \text{miss} + \text{false alarm})$ and is more sensitive to true probe detection than using only number of hits. It ranges in value from 0 to 1 with values above 0.5 indicating positive discrimination.

T2 detection in the single target condition was compared against chance ($=0.5$). Only individuals who were detecting T2 at greater than chance levels were included in the analysis. For the single target conditions (T1 only or T2 only), mean a' scores were compared between the WS and TD groups using independent samples t-tests. For the dual target conditions, two 2 (group (WS vs. TD)) x 4 (lag (positions 1, 2, 3, 7)) repeated measures ANOVA were performed on T1 and T2 a' scores.

AB magnitude was calculated as the average of T2 detection given T1 detection at lag 2 and lag 3 according to the formula developed by Martens et al. (2006): $\{[(\text{T1 detection at Lag 2} - \text{T2} / \text{T1 detection at Lag 2}) / \text{T1 detection at Lag 2}] + [(\text{T1 detection at Lag 3} - \text{T2} / \text{T1 detection at Lag 3}) / \text{T1 detection at Lag 3}] / 2\} \times 100$. AB magnitude was compared between groups using Student's t-test.

As has been done previously in clinical populations (Amirault et al., 2009; Shapiro et al., 2002), AB duration was examined by comparing T2 performance in the single target condition with T2 performance at each lag in the dual target condition separately for WS and TD participants. The single target condition can be considered a test of the maximal ability of an individual to detect a target embedded in an RSVP

because there is no previous target present to distract the participant. Matched-pairs t-tests were used to compare the T2 a' scores between conditions, with significance set at $\alpha=0.0125$ ($0.05/4$) using the Bonferroni correction for multiple comparisons.

To examine associations between task performance and cognitive abilities, correlations among performance in the single target condition, IQ, working memory (DS task), and processing speed (CN task) were computed separately for the WS and TD groups. Additionally, associations between the neuropsychological tests and AB magnitude and duration were examined within each group.

CHAPTER III

RESULTS

Single Target Condition

T2 detection in WS was significantly above chance ($t=4.092$, $p=0.001$), indicating that WS participants were positively discriminating T2 in the RSVP. However, four WS participants did not detect T2 at a greater than chance level and were excluded from subsequent analysis. All TD participants detected T2 above chance.

TD participants were significantly better than WS participants at detecting T1 in the T1 only condition (0.98 ± 0.03 vs. 0.91 ± 0.06 , $t=3.991$, $p=.001$). Though TD participants had higher T2 detection rates than WS participants in the T2 only condition, it failed to reach significance (0.84 ± 0.08 vs. 0.76 ± 0.13 , $t=1.886$, $p=0.073$) (see Figure 2).

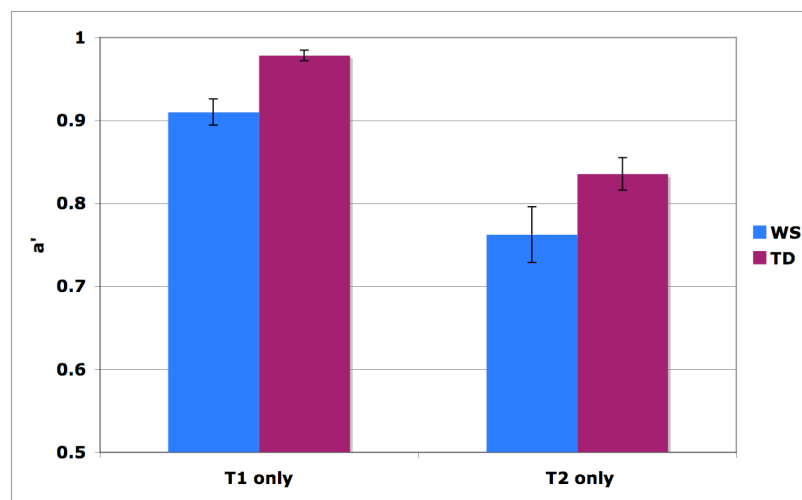


Figure 2. Average Target Detection in the Single Target Condition for WS and TD groups. Error bars represent ± 1 st. error.

Dual Target Condition

T1 a'. As depicted in Figure 3, *T1 a'* was higher in TD participants than in WS participants, with no effect of lag position. A 2 (group) x 4 (lag position) ANOVA revealed a main effect of group ($F_{1,29}=18.483$, $p<0.001$) but no effect of lag position ($F_{3,27}=1.853$, $p=0.161$) or group x lag interaction ($F_{3,27}=0.327$, $p=0.806$). Across all lags, TD participants had an average *T1 a'* of 0.98 ± 0.025 while WS participants had an average *T1 a'* of 0.91 ± 0.05 .

T2 a'. In measuring the AB, only trials in which T1 is correctly identified are included for determining T2 accuracy. As depicted in Figure 3, TD participants showed a drop in T2 detection at lag 2, but performance improved at lag 3 and again at lag 7, indicating the canonical AB. WS participants had poorer performance at each lag than did TD participants, with lower performance at both lags 2 and 3, and improving at lag 7. A 2 (group) x 4 (lag position) ANOVA revealed a main effect of group ($F_{1,29}=5.043$, $p=0.033$), main effect of lag position ($F_{3,27}=10.437$, $p<0.001$), but no group x lag interaction ($F_{3,27}=1.215$, $p=0.323$). Therefore, individuals with WS appear to have overall lower detection abilities than TD participants, but still show an AB.

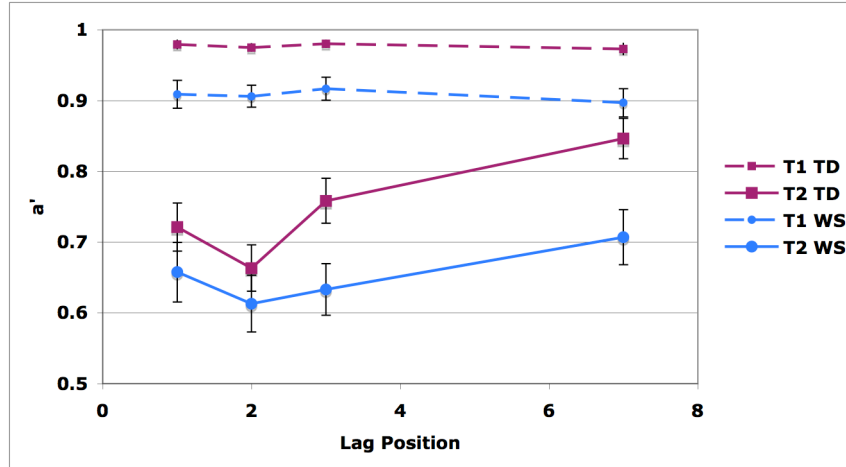


Figure 3. Average Target Detection Across Lag Positions in Dual Target Condition for WS and TD groups. Error bars represent ± 1 st. error.

AB Duration. We compared T2 a' at each lag position with T2 a' in the single target condition within each group in order to determine the duration of the AB. T2 a' in the single target condition can be viewed as the maximal ability of the individual to detect an embedded target in an RSVP. For TD participants, a' at lags 1 and 2 were significantly lower than in the single target condition ($p's \leq .003$) (Figure 4). By lag 3, however, T2 detection was not significantly different from detection in the single target condition. In contrast with TD individuals, in the WS group, T2 a' at lags 1, 2 and 3 was impaired compared with T2 a' in the single target condition ($p's \leq .001$) (Figure 4). It was not until lag 7 that T2 a' in the dual target condition reached the same level as in the single target condition. Recovery from lag 2 to lag 3 was compared between WS and TD individuals via difference scores. There was greater improvement in T2 performance from lag 2 to lag 3 in the TD group (improvement of $9.5\% \pm 13.1\%$ in TD vs. $2.0\% \pm 6.6\%$

in WS, $p=.05$). Thus, the AB appears to be prolonged in WS compared with TD individuals.

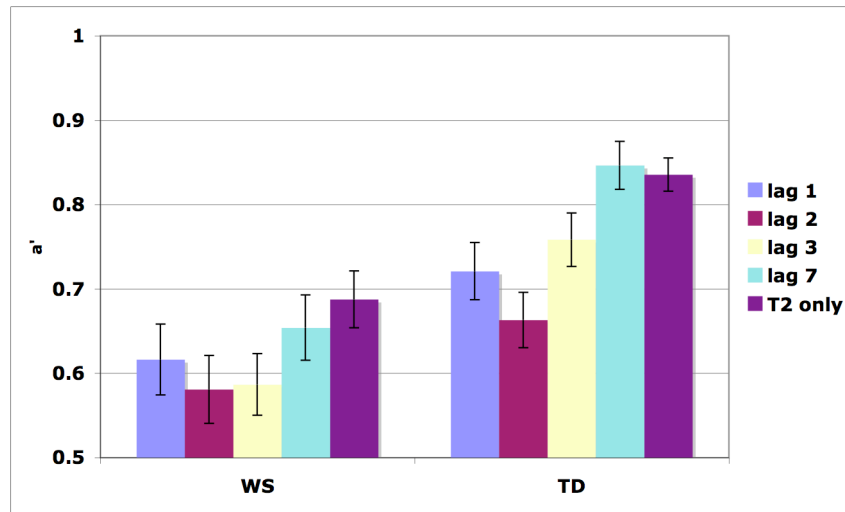


Figure 4. Average T2 detection in the single and dual target conditions for the WS and TD groups. Error bars represent ± 1 st. error.

AB Magnitude. We also compared the magnitude of the AB between the WS and TD groups. The magnitude of the AB is the average decrease in T2 performance relative to T1 performance at lags 2 and 3 (Martens et al., 2006). AB magnitude was not significantly different between the WS and TD groups ($31.76\% \pm 13.3\%$ vs. $27.38\% \pm 11.29\%$, $p=0.329$).

Correlates of AB Performance. Since this is the first time AB has been studied in a low IQ population, and given the overall lower rates of T2 detection in the WS group, we wanted to further examine correlates of task performance in the WS group.

Interestingly, average T2 a' across lag position did not significantly correlate with T2 a'

in the single target condition for the TD group ($r=0.371$, $p=0.143$), but it did for the WS group ($r=0.811$, $p\leq 0.001$). This suggests that for WS participants, but not TD participants, general perceptual or task-related difficulties may contribute to their overall lower performance in the dual target conditions. Indeed, among the WS participants, there was a trend for greater T2 a' in the single target condition to be associated with higher IQ ($r=0.475$, $p=0.086$), but not higher CN scores ($r=0.142$, $p=0.627$) or DS scores ($r=0.225$, $p=0.440$). However, IQ, CN, and DS scores were not associated with single target T2 a' in the TD group (see Table 2). IQ, CN, and DS scores did not correlate with T1 performance in the single or dual target conditions for the WS or TD groups.

None of the neuropsychological measures correlated with either AB magnitude or AB duration (quantified as improvement at lag 3 relative to lag 2) in the WS or TD groups (see Table 2).

Table 2. Neuropsychological correlates of AB Performance in WS (bold) and TD (italics)

	IQ	CN	DS	T1only	T2only	T1dual	T2dual	ABmag	ABdur
IQ		<i>.545*</i>	<i>.230</i>	<i>.341</i>	<i>.320</i>	<i>.223</i>	<i>.198</i>	<i>-.201</i>	<i>.180</i>
CN	.452		<i>.470</i>	<i>-.076</i>	<i>-.052</i>	<i>-.113</i>	<i>-.087</i>	<i>.040</i>	<i>.107</i>
DS	.714*	.643*		<i>.129</i>	<i>.384</i>	<i>.069</i>	<i>.440</i>	<i>-.389</i>	<i>-.117</i>
T1only	.112	.046	.002		<i>-.037</i>	<i>.869**</i>	<i>.068</i>	<i>-.016</i>	<i>.355</i>
T2only	.475	.142	.225	.424		<i>-.037</i>	<i>.371</i>	<i>-.376</i>	<i>-.226</i>
T1dual	.108	-.115	.066	.813**	.430		<i>.256</i>	<i>-.268</i>	<i>.283</i>
T2dual	.493	.066	.390	.329	.811**	.480		<i>-.952**</i>	<i>.003</i>
ABmag	-.402	-.281	-.429	-.028	-.646*	-.177	-.918**		<i>.066</i>
ABdur	-.100	-.209	-.459	-.357	-.172	-.490	-.200	.125	

Note. WS: bold-faced values below the diagonal; TD: italicized values above the diagonal.

*p<0.05; **p<0.001

IQ = Composite IQ (K-BIT-2)

CN = WAIS-4 Cancellation Scaled Score

DS = WAIS-4 Digit Span Scaled Score

T1only = T1 a' in single target condition

T2only = T2 a' in single target condition

T1dual = average T1 a' across lags in dual target condition

T2dual = average T2 a' across lags in dual target condition

ABmag = AB magnitude

ABdur = AB duration (Improvement in T2 a' from lag 2 to lag 3)

CHAPTER IV

DISCUSSION

To our knowledge, this is the first study to address the temporal dynamics of attention via the AB paradigm in Williams syndrome. Moreover, it appears to be the first study to address this question in individuals with intellectual disabilities in general.

Although their performance was poorer than that of TD individuals in the single target conditions, as a group, WS individuals were able to reliably complete the AB paradigm.

Key findings of the study are that individuals with WS have poorer target detection abilities than chronological age-matched TD individuals as indicated by both their T1 and T2 performance in the single and dual target conditions. Though they have an AB that is similar in magnitude between TD and WS individuals, the WS individuals exhibited a prolonged AB.

Although reasons for the prolonged AB in WS are uncertain, theoretical models of the AB shed some light on the WS findings. Many informal and formal theoretical accounts of the AB have been proposed, and there is much overlap among the different models (Dux & Marois, 2009). It is believed the AB arises due to a two-stage model of attention (Chun & Potter, 1995). In the first stage, stimuli are recognized but susceptible to rapid decay. In the second stage, stimuli are consolidated in working memory and thus become available for report. The common capacity-limited attentional resource model (Dux & Marois, 2009) proposes that a high level central resource underlies the attentional and working memory demands of the AB paradigm. When more attention is deployed for

T1 processing, less attentional resources are available for the processing and encoding of T2. The resulting AB reveals the limits of attention in the temporal domain.

Support for the common capacity-limited attentional resource model comes from both behavioral and psychophysiological studies. By manipulating participants' focus to decrease attention to T1 (though still identifying T1), researchers have induced a corresponding decrease in the magnitude of the AB, suggesting that overinvesting resources in T1 leads to poorer T2 performance (Arend, Johnston, & Shapiro, 2006; Olivers & Nieuwenhuis, 2005; Olivers & Nieuwenhuis, 2006). Using magnetoencephalography (MEG), one study revealed a tradeoff in target-related activation during an AB task (Shapiro, Schmitz, Martens, Hommel, & Schnitzler, 2006). Specifically, the amount of attentional resources devoted to T1 as indexed by T1-related activation, was positively related to the magnitude of the AB; greater T1 activation corresponded to less T2 activation, and participants were less likely to report seeing T2.

In the current study, WS participants had more difficulty identifying any target than did TD participants, and in particular, were poorer at identifying T1 in the single target condition. This may be due to a more limited central processing resource in WS making it more difficult to attend to and extract the relevant features of the stimuli. However, despite poorer T1 performance in the WS group, it is important to note that their performance was still quite high and well above chance for identifying T1 in both the single and dual target conditions. Additionally, the WS group did not significantly differ from the TD group at identifying T2 in the single target condition. Therefore, it appears that individuals with WS were still reliably able to pick out the targets in the RSVP even at the fast speed of presentation.

There are additional reasons to believe that central processing resources may underlie the target identification performance in WS rather than other explanations. First, all individuals in the current study were able to read letter stimuli, which is typical for individuals with WS (Laing, 2002). Second, target identification did not correlate with any of the neuropsychological measures in either the WS or TD groups, suggesting that poorer target performance in WS was not due specifically to lower IQ, processing speed, or working memory abilities. Finally, it appears that both the WS and TD groups appear to be processing the T1 stimuli in the same way because in both groups, T1 detection was highly correlated between the single and dual target conditions, and did not differ between the conditions for each individual. Therefore, though the dual target condition added a second target that needed to be attended to, encoded, and reported, these additional demands did not effect T1 processing and report in either the TD or WS group. Thus, it seems most likely that a central processing resource, such as extracting relevant features of the stimuli, is involved in the poorer target detection in WS.

Interestingly, the magnitude of the AB was no different in WS and TD participants, suggesting that WS participants remembered to look for both targets and that they were not simply overinvesting in T1 compared to the TD group. Despite their more limited resources (as indexed by their lower target detection levels for both T1 and T2 in the dual target condition), the individuals with WS do not appear to be only allocating attention to T1 (either deliberately or due to automatic attentional capture) at the expense of attending to T2.

Rather, the most striking component of the AB dynamics was the prolonged AB in the WS group, suggesting difficulty with disengaging attention from T1 and re-

allocating attentional resources to T2. While the slower processing speed in WS vs. TD individuals, as indicated by their lower scores on the CN task, may be involved in the longer AB in WS, it is unlikely that this explains the longer AB by itself for several reasons. First, processing speed was not related to T1 or T2 detection abilities in either the single or dual target conditions in both the WS and TD groups. Additionally, processing speed was not related to the magnitude or duration of the AB. If processing speed was the underlying factor leading to a prolonged AB, we would expect individuals with slower processing speed to have a longer blink and show less improvement in T2 performance from lag 2 to lag 3. As this was not supported by our data, it is unlikely that the slower processing speed in WS is solely responsible for the prolonged AB.

Additional reasoning about the nature of the tasks also supports the lack of association between processing speed and AB parameters in the current study. Processing speed has not traditionally been explored as a correlate of AB magnitude or duration because the AB assesses the temporal difference *between* recognizing two different targets rather than the time needed for recognition of a single target (Hari & Renvall, 2001). In addition, the measure of processing speed used in the current study, the Cancellation task, consists of two different shapes and involves a spatial display, which is notoriously difficult for individuals with WS. Therefore, the lack of association between the Cancellation task and the AB may be further evidence of a dissociation between processing abilities in the spatial and temporal domains in WS.

It also seems unlikely that the prolonged AB can be explained solely by the lower IQ in the WS group. Though there was a trend correlation between IQ and T2 detection in the single and dual target conditions in the WS group, the correlations were modest

and failed to reach significance. Additionally, T2 detection in the single target condition did not significantly differ between the TD and WS groups despite their IQ difference. It is possible that a minimum level of intellectual ability is needed to complete the AB task because, for example, participants must remember rules about the two different targets and respond to different questions about them. Indeed, the four WS participants who were excluded from analysis due to chance T2 detection in the single target condition had lower IQ and lower DS scores than did the included WS participants ($p's \leq .025$), but performed no differently on the CN task. Three of these four participants had extremely high false alarm rates ($>85\%$), suggesting an indiscriminant response style, perhaps due to difficulty understanding or remembering the directions, or executing their intended response. Therefore, a minimum level of intellectual ability may be a prerequisite for completing a dual target task. Nevertheless, IQ was still quite variable among included individuals, ranging from 51-102 in the WS group.

Importantly, among individuals who reliably completed the task, IQ did not correlate with AB magnitude or duration in either the WS or TD groups. These findings are consistent with other studies of healthy adults (Colzato, Spapé, Pannebakker, & Hommel, 2007), and in groups with developmental disabilities. Specifically, children with ADHD and SLI had a prolonged AB compared with TD children, results that could also not be explained by IQ (Li et al., 2004; Lum et al., 2007). Thus, it is unlikely that underlying intellectual factors are solely responsible for the prolonged AB in WS.

Of all the neuropsychological measures, it is most surprising that Digit Span, as a measure of working memory, did not correlate with the magnitude or duration of the AB. Electrophysiological studies reveal that working memory is indeed involved in the AB

(Kranzioch, Debener, & Engel, 2003; Vogel & Luck, 2002) and measures of working memory in TD individuals have predicted AB magnitude (Arnell, Stokes, MacLean, & Gicante, 2010; Colzato et al., 2007) and AB duration (Gillard-Crewther et al., 2007). However, the lack of association may be due to differences between the Digit Span and AB tasks. First, and most important, Digit Span is an aurally presented task while the AB is purely visual. As auditory working memory is a relative strength in WS (Jarrold, Baddeley, & Hewes, 1999), it is likely tapping a different domain than does the AB paradigm. Additionally, there are separate components of working memory (Baddeley, 1996), and recent research suggests it is the executive component aspect of working memory that relates to the AB, not storage capacity (which Digit Span measures) (Arnell et al., 2010). Therefore, it remains to be seen if a visual working memory measure, especially one that relates to the executive control system, would better predict AB performance in WS.

The AB performance found here in WS is remarkably similar to a study of adolescents with SLI (Lum et al., 2007). As would be expected, the adolescents with SLI had lower language scores than did TD peers. However, the SLI group was split between individuals with non-verbal IQ in the “low normal” range (85-96) and individuals with non-verbal IQ in the “high normal” range (97-115). All TD peers had non-verbal IQ in the “high normal” range, as well. When examining T1 performance in the dual target condition, individuals in the SLI “low normal” group performed less accurately than the SLI “high normal” or TD groups, who did not differ from each other. Similar to the SLI “low normal” group, our WS participants have both lower verbal and non-verbal IQ than the TD group, and were less accurate at detecting T1. (It is important to note that T1

detection was much more variable, and overall poorer, in the SLI study than in the current WS study, probably due to stimulus characteristics (T1 in the SLI study was not a different color from the distractors and so would not have a “pop out” effect). This may be why there was differential performance for T1 based on non-verbal IQ in the SLI study but not in the current study.) In terms of T2 processing, however, the SLI “low normal” and “high normal” groups did not differ from each other, and both showed an AB that was greater in duration than their TD peers. Therefore, regardless if there was an underlying difficulty with processing a given rapidly presented stimulus, the behavioral effects for processing a second stimulus were the same. The authors propose that a sluggish attention system (SAS), rather than simply slower processing speed, may be responsible for this pattern of performance in the AB. Individuals with SAS require more time to engage and disengage attention, and SAS has been previously linked to difficulties with rapid processing in persons with dyslexia (Hari & Renvall, 2001), who also show a prolonged AB (Facoetti et al., 2008; Hari et al., 1999). It is possible SAS also occurs in WS, given their comparable AB performance and their difficulties with attention disengagement.

Difficulty with attention disengagement and appropriate reallocation of resources as a potential explanation of the prolonged AB in WS fits well with other observations of attention in WS. Though most previous studies have focused on toddlers and children with WS and have contained a spatial or social factor, they reveal difficulty with disengaging attention from faces (Mervis et al., 2003; Riby & Hancock, 2009), fixation cues (Cornish et al., 2007), and items within a visual search display (Montfort et al., 2007). In the one previous pilot study of temporal attention in three adults with WS,

individuals had difficulty disengaging attention from visual to auditory modalities and vice versa (Lincoln et al., 2002). Thus, difficulty with attention disengagement appears to be a core component of the WS attentional profile, extending to temporal, spatial, cross-modal, and social domains.

Comparison of neuroimaging studies in WS with research aimed at identifying the neural underpinnings of the AB phenomenon may further elucidate our understanding of the prolonged AB in WS. Recent work suggests that the AB relies on a highly distributed parietofrontal network responsible for target selection (Gross et al., 2004; Hommel et al., 2006; Kranzloch, Debener, Schwarzbach, Goebel, & Engel, 2005; Marois et al., 2000; Marois & Ivanoff, 2005). In particular, the intraparietal sulcus (IPS) and inferior parietal lobe (IPL) appear to work in concert to contribute to the AB (Cooper, Humphreys, Hulleman, Praamstra, & Georgeson, 2004; Kihara et al., 2007; Kihara et al., 2010). Correspondingly, neuroimaging studies in children and adults with WS consistently reveal reduced gray matter density in parietal regions, including reduced sulcal depth and reduced gray matter in the IPS (Boddaert et al., 2005; Chiang et al., 2007; Eckert et al., 2006; Eckert et al., 2005; Kippenham et al., 2005; Meyer-Lindenberg et al., 2004; Reiss et al., 2004; Reiss et al., 2000; Van Essen et al., 2006).

Previous functional brain imaging studies in WS have focused on neural correlates of impaired dorsal stream functioning (the “where” pathway via occipital-parietal lobes) compared with intact ventral stream functioning (the “what” pathway via occipital-temporal lobes) (Meyer-Lindenberg et al., 2004; Sarpal et al., 2008; Ungerleider & Mishkin, 1982). The IPS appears to play a key role in this dysfunction. Research suggests reduced connectivity between the IPS and later dorsal stream regions involved

in spatial tasks (Meyer-Lindenberg et al., 2004), as well as between the IPS and areas that are involved in both dorsal and ventral stream processing (parahippocampal place area (PPA)) (Sarpal et al., 2008) in WS. Thus, there appears to be a structural-functional connection between the reduced gray matter and decreased sulcal depth of the IPS in WS (Kippenham et al., 2005; Van Essen et al., 2006) and the dorsal stream hypoactivation (Meyer-Lindenberg et al., 2004).

Therefore, it seems plausible that the abnormal IPS is also involved in the prolonged AB seen in WS. Kihara et al. (2010) suggest that the IPS and IPL may work cooperatively to disengage attention from T1 and engage attention with T2. While the IPL in WS appears to be preserved in volume (in contrast with the reduced SPL) (Eckert et al., 2005), less is known about the functionality or connectivity of the IPL.

Interestingly, in a study of TD older adults, those with lesions to the IPL showed a prolonged AB while those with lesions to the SPL had an AB equal in duration to individuals without lesions (Shapiro et al., 2002). Therefore, exploration of the neural correlates of non-spatial attention in WS is an area for future research to better understand if and how dysfunction extends beyond the IPS and how the IPL, in particular, may be involved.

In conclusion, this study is the first to examine altered temporal dynamics of attention in WS via the AB paradigm. It reveals that difficulties with attentional processing in WS extend beyond the spatial and social domains and demonstrates a need to extend research in WS to other salient aspects of their phenotype. Individuals with WS had a harder time detecting targets in a RSVP than did TD individuals. They had an AB that was equal in magnitude but prolonged in duration compared with a TD group. These

results are in concordance with the small body of work pointing to problems with attentional disengagement in WS (Cornish et al., 2007; Lincoln et al., 2002; Mervis et al., 2003; Montfort et al., 2007; Riby & Hancock, 2009), and suggest that an underlying difficulty with attentional disengagement may be responsible for the results of studies exploring spatial and social attentional processing in WS. Future research could extend this work by including neuroimaging to further elucidate the neural underpinnings of the attentional system in WS and including other measures of working memory to better understand the cognitive correlates of their AB performance. Given the high rates of anxiety in WS (Dykens, 2003), it would also be interesting to explore how emotional or anxiety-inducing stimuli might increase (when presented as T1) or diminish (when presented as T2) the magnitude of the AB in WS, an approach used in TD individuals (e.g., Anderson, 2005; Maratos, Mogg, & Bradley, 2008; Most, Chun, Widders, & Zald, 2005; Smith, Most, Newsome, & Zald, 2006; Stein, Zwickel, Ritter, Kitzmantel, & Schneider, 2009). Finally, this study appears to be the first to successfully report on the AB in a low IQ group. As many other clinical populations are increasingly being examined with the AB paradigm, future studies may not necessarily need to be limited to higher functioning individuals, but can likely include those along a broad spectrum of cognitive abilities.

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