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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 difference 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.

Keywords

Williams syndrome; attentional blink; attentional disengagement; 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 7,500 births, WS is associated with cardiovascular, endocrine and neurological problems, as well as a distinctive cognitive-linguistic profile (see Martens et al., 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).

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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, 2003). 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 et al., 1997; Tomc et al., 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 to 55 years, up to 85% reported problems with distractibility (Elison et al., 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 et al., 2007; Montfoort et al., 2007), incongruent information in a hierarchical stimulus (Farran et al., 2003), human faces (Laing et al., 2002; Mervis et al., 2003; Riby & Hancock, 2009), or switching between auditory and visual modalities (Lincoln et al., 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 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 et al., 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 susceptible 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 et al., 2007), high functioning autism (Rinehart et al., 2010), and ADHD (Mason et al., 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 et al., 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 Courier New font, size 76, and were viewed from a distance of approximately 42 inches (about 1.43° of visual angle). Following a fixation asterisk, black letters were presented on a grey background for 107 ms each with a 13 ms interstimulus interval between letters (see Figure 1). Each RSVP trial was randomly generated to contain 9 to 15 letters. Participants identified 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 serial 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 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 responses using a handheld 4-button response box.

Prior to the experiment, participants completed a practice block of 12 trials (three T1 only trials, three T2 only trials, six 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 to provide instructions and answer questions.

2.4. Statistical Analysis

Consistent with previous AB studies (e.g., Rinehart et al., 2010), 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 detected T2 at greater than chance levels were included in analyses. 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, Group (2: WS vs. TD) $\times$ Lag position (4: 1, 2, 3, 7) repeated measures ANOVAs were performed on T1 and T2 a' scores.

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. Therefore, AB duration was examined by comparing T2 performance in the single target condition with T2 performance at each lag in the dual target condition using matched-pairs t-tests. Significance was set at $\alpha=0.0125$ ($0.05/4$) using the Bonferroni correction for multiple comparisons. The AB duration was considered to be the latest lag position at which detection still differed from the single target condition.

The magnitude of the AB is the percent decrease in T2 performance relative to T1 performance at lags 2 and 3 (which is when the canonical AB occurs; Martens et al., 2009). Only trials on which T1 is correctly identified are included to ensure that the participant is trying to attend to both targets and not disregarding T1 while attending for T2. AB magnitude was calculated according to the formula developed by Martens et al. (2006): $\{[(T1 \text{ detection at Lag } 2 - T2/T1 \text{ detection at Lag } 2)/T1 \text{ detection at Lag } 2] + [(T1 \text{ detection at Lag } 3 - T2/T1 \text{ detection at Lag } 3)/T1 \text{ detection at Lag } 3]/2\} \times 100$. AB magnitude was compared between groups using Student's t-test.

To examine associations between task performance and cognitive abilities, correlations were computed among AB parameters (a' scores in the single and dual target conditions, AB duration, and AB magnitude) with IQ, DS, and CN scores. Differences in the magnitude of the correlations between the WS and TD groups were examined using Fisher's z transformation.

3. Results

3.1. Single Target Condition

Both groups identified T1 and T2 at a greater than chance level ($p's < .001$). 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 , $t_{29} = 3.991$, $p = 0.001$). Though the TD group had higher T2 detection rates than WS in the T2 only condition, this comparison failed to reach significance (0.84 ± 0.08 vs. 0.76 ± 0.13 , $t_{29} = 1.886$, $p = 0.073$) (see Figure 2).

3.2. Dual Target Condition

3.2.1. T1 a' —A Group (2) $\times$ Lag Position (4) ANOVA revealed a main effect of group ($F_{1,29} = 18.483$, $p < 0.001$) but no effects for lag position or Group $\times$ Lag interaction. As depicted in Figure 2, T1 a' was higher in TD versus WS participants. 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 .

3.2.2. T2 a' —In measuring the AB, only trials in which T1 is correctly identified were included for determining T2 accuracy. A Group (2) $\times$ Lag Position (4) ANOVA revealed main effects of Group ($F_{1,29} = 5.043$, $p = 0.033$), and Lag Position ($F_{3,27} = 10.437$, $p < 0.001$), but no interaction. As depicted in Figure 2, TD participants showed a drop in T2 detection at lag 2, but performance improved at lag 3 and again at lag 7. WS participants had poorer performance at each lag than did TD participants, with lower performance at lags 2 and 3, and improvement at lag 7. Individuals with WS appear to have lower target detection performance than TD participants, but still show an AB.

3.2.3. AB Duration—To quantify the duration of the AB, we compared T2 a' at each lag position with T2 a' in the single target condition using matched-pairs t-tests. For TD participants, a' at lags 1 and 2 were significantly lower than in the single target condition ($p's < .003$), but there was no longer a difference by lag 3 (see Figure 2). In contrast, in the

WS group, T2 a' at lags 1, 2 and 3 was impaired compared with T2 a' in the single target condition ($p's < .001$). 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 (TD $9.50\% \pm 13.13\%$ vs. WS $1.99\% \pm 6.58\%$, $t_{24,455} = 2.066$, $p = .05$). Thus, compared to TD controls, the AB appears to be prolonged in WS.

3.2.4. AB Magnitude—The magnitude of the AB is the average of the 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 ($31.76\% \pm 13.29\%$ vs. $27.38\% \pm 11.30\%$, $t_{29} = -0.993$, $p = 0.329$).

3.2.5. Correlates of AB Performance—Since this is the first time AB has been studied in a group with intellectual disabilities, correlations were conducted between task performance measures and also with IQ, CN, and DS. In both the TD and WS groups, T1 a' in the single target condition significantly correlated with average T1 a' across lag positions ($r's > .810$, $p's < .001$). 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 ($r = 0.811$, $p < 0.001$; Fisher's $Z = -1.838$, one-tailed $p = 0.03$). No significant correlations were found between the neuropsychological measures and AB performance parameters in either group.

4. Discussion

To our knowledge, this is the first study to address the temporal dynamics of attention via the AB paradigm in WS, or in individuals with intellectual disabilities in general. Participants with WS reliably completed the AB paradigm, but demonstrated 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. The magnitude of the AB, which quantifies attentional allocation, was similar between groups. However, WS participants exhibited a prolonged AB, suggesting a specific problem with attentional disengagement. AB performance was not correlated with neuropsychological measures in either group. Below we review the performance of the WS group on these different AB parameters within the framework of the common capacity-limited attentional resource model and discuss implications for the WS phenotype.

The WS group had more difficulty identifying any target than did TD participants, and in particular, were poorer at identifying T1 in the single target condition. Although T1 differed in color from the distractor letters, identifying T1 was a complex task. It required discerning features that could be any of three target letters, remembering the letter and delaying the response until the end of the RSVP, and then choosing the correct button on the response box. Limitations in central processing resources may make it more difficult for WS individuals to attend to and extract the relevant features of the stimuli, and store the target in working memory until prompted to respond. Nevertheless, it appears that both the WS and TD groups processed 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. Although the dual target condition involved additional demands, they did not affect T1 processing and reporting in either the TD or WS group. Thus, the individuals with WS were able to successfully detect white letters (T1) amongst black distractors.

We can also rule out other possible explanations for the poorer target performances in WS. First, all individuals could read letter stimuli, and despite their poorer T1 responses, their performance was still quite high and well above chance. Additionally, the WS group did not significantly differ from the TD group at identifying T2 in the single target condition, i.e., they were able to pick out the targets in the RSVP even at the fast speed of presentation. Though T2 was difficult to detect because it did not differ in color from the distractor letters, detecting T2 required extracting features of only a single possible target letter and choosing between only two response options. T2 detection in the single target condition may therefore have required fewer attentional resources than T1 detection.

The magnitude of the AB was not different between WS and TD participants, suggesting that WS participants remembered to look for both targets and that they were not simply overinvesting in T1 detection compared to the TD group. Therefore, it appears that they were not expending all of their limited resources on T1 at the expense of T2.

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. As suggested by the common capacity-limited model, if the attentional resources are deployed to select T1 (extracting features, storing in working memory, inhibiting distractors, etc.), then they are not available to select T2. It appears to take people with WS a longer time to disengage and then re-engage these attentional resources in order to select the second target.

To address the second study aim, we examined whether any of several neuropsychological measures were related to the dynamics of the AB in WS. While the slower processing speed on the CN task in WS vs. TD individuals may be contributing to the longer AB in WS, it is unlikely. First, processing speed was not related to T1 or T2 detection abilities in either the single or dual target conditions in either group. Additionally, processing speed was not related to the magnitude or duration of the AB, likely 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).

It also seems unlikely that the prolonged AB in WS can be explained solely by the lower IQ of this group. IQ was not correlated with AB performance, and T2 detection in the single target condition did not differ between the TD and WS groups despite their IQ difference. Even so, a certain minimum level of intellectual ability is likely needed to complete the AB task because participants must remember rules about two different targets and respond to questions about them. Indeed, the four WS participants who were not included in the analysis had lower IQ scores than their counterparts. 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. Nevertheless, IQ was still quite variable among included individuals with WS, ranging from 51-102, and with a mean (77.64) in the mild range of disability.

Beyond target detection, IQ did not correlate with AB magnitude or duration in either the WS or TD groups. These findings add to a corpus of studies revealing no relationship between IQ and AB parameters in individuals with developmental disabilities. Specifically, children with ADHD and Specific Language Impairment (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;). It is thus 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 (e.g., Vogel & Luck, 2002) and measures of working memory in TD individuals have predicted AB magnitude (e.g., Arnell et al., 2010) and AB duration (Gillard-Crewther et al., 2007). However, working memory contains separate components (Baddeley, 1996), and it was recently demonstrated that it is the executive component aspect of working memory that relates to the AB, not storage capacity (Arnell et al., 2010), which is measured by Digit Span. Additionally, Digit Span is an aurally presented task while the AB is purely visual. Though previous research has found associations between auditory working memory and the visual AB task in TD individuals (Gillard-Crewther et al., 2007), further suggesting that central attentional limitations give rise to the AB rather than purely visual processes, auditory working memory may be a relative strength in WS (Jarrold et al., 1999, though see Sampaio et al., 2008 for opposing findings), and so dissociations between modalities and the AB task may exist in WS though not in TD individuals.

The AB findings in WS are remarkably similar to a study of adolescents with (SLI) (Lum et al., 2007). Though non-verbal IQ impacted T1 accuracy in the dual target condition among SLI participants, all SLI participants, regardless of non-verbal IQ, had an AB duration that was longer than their TD peers. The authors propose that a sluggish attention system (SAS), rather than simply slower processing speed, may be responsible for these findings. Individuals with SAS require more time to engage and disengage attention, and SAS has been 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). Given their comparable AB performance and difficulties with attention disengagement, it is possible that people with WS also have a SAS, thus requiring more time to disengage and re-deploy limited attentional resources.

Neuroimaging studies of the AB may help explain the prolonged AB in WS. The AB relies on a highly distributed parietofrontal network (Kranzloch et al., 2005; Marois et al., 2000). The intraparietal lobe (IPL) is hypothesized to send a signal to the intraparietal sulcus (IPS) telling it to disengage from its current attentional set (Corbetta et al., 2008; Kihara et al., 2011). Structural and functional abnormalities of the IPS in WS, including reduced sulcal depth, reduced gray matter, and reduced connectivity between the IPS and other processing regions (Meyer-Lindenberg et al., 2004; Sarpal et al., 2008), may disrupt this coordination between the IPS and IPL, leading to the prolonged AB in WS.

An underlying difficulty with attentional disengagement in WS has several important clinical implications. It is plausible that basic attentional training to improve attentional disengagement could lead to gains in other areas of deficit. For example, attentional training could be included in social skills training to improve face gaze behavior, which is often prolonged and inappropriate in WS (e.g., Mervis et al., 2003). Attentional training could also extend beyond social appropriateness. Laing et al. (2002) reported impaired triadic interactions in toddlers with WS and suggested that this could contribute to the language delay in the syndrome because they miss opportunities for referential learning. Thus, toddlers with WS may need to be given more time to disengage their attention (e.g., from another person, from an initial object of fascination) and engage with the object being referenced. They may need to be explicitly encouraged to disengage their attention and rewarded for doing so.

Attentional difficulties may also contribute to other problematic behaviors. Indeed, Rhodes et al. (2010) reported associations between difficulties with attention switching and emotional and conduct problems in individuals with WS. Thus, attentional training may be important for these areas, as well. For example, individuals with WS frequently meet criteria for specific phobia and generalized anxiety (Dykens, 2003). Children and adults with

anxiety frequently manifest attention biases to threatening objects. Attention modification training programs (such as cuing) can be successfully used to improve attentional disengagement from the threatening objects and decrease anxiety (e.g., Amir et al., 2009, Bar-Haim et al., 2011). This may suggest an avenue for intervention in individuals with WS who have comorbid problems with anxiety, by focusing on improving their attention disengagement abilities.

Strengths of the current study are its distinction between attentional allocation versus attentional disengagement, and its isolation of the temporal dynamics of attention so that results were not confounded by spatial or social factors, which have been the focus of previous studies (e.g., Montfoort et al., 2007; Riby & Hancock, 2009). Additionally, it included a lower functioning population than has previously been examined with the AB task, demonstrating the feasibility of including individuals with lower IQ in AB paradigms.

However, the study had several weaknesses that should be addressed in follow-up work. Future studies should include more individuals, though it should be recognized that the current sample size and findings are comparable with those of previous WS studies (e.g., Riby & Hancock, 2009, Sampaio et al., 2008). The very recent findings isolating the executive component of working memory rather than storage capacity point to the need to include visual working memory measures that target the executive component.

Finally, we elected to include a chronological-age matched comparison group because of the large age range of our WS participants and evidence for effects of age on AB parameters, particularly AB duration, from childhood through older adulthood (Garrad-Cole et al., 2011; Maciokas & Crognale, 2003). Thus, the AB task has traditionally been assessed in disability groups by comparing their performance with chronological-age matched typical controls (e.g., dyslexia (Hari et al., 1999); SLI (Lum et al., 2007)). As discussed by Sampaio et al. (2008), when sub-processes within a domain have different developmental trajectories, such as occurs within the realm of attention (e.g., Garrad-Cole et al., 2011; Gupta & Kar, 2009; Scerif & Steele, 2011), data interpretation becomes confounded when choosing a control group matched in ability to only one cognitive process (e.g., verbal IQ vs. non-verbal IQ vs. working memory vs. sustained attention, etc.). A chronological-age matched comparison group more clearly compares functioning between WS and typical development (Sampaio et al., 2008). Nevertheless, inclusion of a mental-age matched comparison group could help address concerns about the role of general cognitive abilities in the AB.

Future studies could also use the AB framework to address specific aspects of the WS phenotype. Given the high rates of anxiety (Dykens, 2003) and hypersociability (Mervis et al., 2003) in WS, emotional social 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 to examine how emotionally arousing stimuli alter attentional dynamics (e.g., Most et al., 2005). For example, examination of how socially relevant targets such as faces alter AB parameters compared with non-social targets could reveal if attentional disengagement difficulties are more extreme for faces, as has been recently suggested (Riby et al., 2010). Due to dissociations between visual and auditory abilities in WS (e.g., Jarrold et al., 1999), future studies could also compare the dynamics of the AB in the auditory versus visual domain to test the domain specificity of these findings.

5. Conclusions

This study is the first to examine altered temporal dynamics of attention in WS via the AB paradigm. Compared to TD peers, individuals with WS had a harder time detecting targets in a RSVP, and they had an AB that was equal in magnitude but prolonged in duration.

Attentional disengagement difficulties, as revealed by the prolonged AB, are a general finding common to many disability groups. However, the results presented here suggest that attentional disengagement is an underlying difficulty in WS, and is not specific to areas of atypicality (such as the spatial and social domains), which have been the focus of previous studies (Cornish et al., 2007; Lincoln et al., 2002; Mervis et al., 2003; Montfoort et al., 2007; Riby & Hancock, 2009). Future research is needed to identify the neural underpinnings of the attentional system in WS, which should also include larger samples, a mental age-matched comparison group, and visual measures of working memory. Finally, this study is the first to successfully use an AB paradigm in a group with intellectual disabilities. As many other clinical groups are being examined with the AB paradigm, future studies can likely include those along a broad spectrum of cognitive abilities.

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Abbreviations

WS	Williams syndrome
TD	typically developing

AB	attentional blink
RSVP	rapid serial visual presentation
DS	digit span
CN	cancellation

Highlights

- Attentional problems are frequently reported in WS.
- The AB in WS is prolonged in duration, but not magnitude, compared to TD peers.
- The AB did not relate to IQ, working memory, or processing speed abilities.
- Results suggest an underlying difficulty with attentional disengagement in WS.
- The AB paradigm can be used in individuals with intellectual disability syndromes.

Fig. 1. AB Paradigm

Fig. 2.

Average Target Detection (a') in Single Target and Dual Target Conditions for WS and TD groups. Error bars represent ± 1 st. error. The WS group performed significantly more poorly than the TD group in the T1 only ($t_{29}=3.991$, $p=0.001$) but not the T2 only condition ($t_{29}=1.886$, $p=0.073$). In the dual target condition, the WS group performed more poorly than the TD group at detecting T1 ($F_{1,29}=18.483$, $p<0.001$) and T2 ($F_{1,29}=5.043$, $p=0.033$). In the dual target condition, there was also a main effect of Lag Position for T2 detection ($F_{3,27}=10.437$, $p<0.001$). No Group X Lag interactions were noted for either T1 or T2 in the dual target condition.

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