

Attention Bias Reduction in Individuals with Williams Syndrome

Research Thesis

Presented in partial fulfillment of the requirements for graduation *with research distinction* in  
Psychology in the undergraduate colleges of The Ohio State University

by

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April 2014

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### Abstract

Individuals with Williams syndrome (WS) have high levels of anxiety. In attention tracking tasks, individuals with WS and typically developing individuals with high levels of anxiety allocate greater attention to threatening images than typical individuals. In individuals with high levels of anxiety, Attention Bias Reduction (ABR) tasks reduce attention bias and anxiety symptoms. The purpose of this project is to investigate the use of ABR to reduce attention bias toward pictures of lightning in individuals with WS. This research includes two studies. Study One is an internet based survey in which individuals with WS rate nonthreatening (nature scenes) and threatening (lightning) images. Study One demonstrates that individuals with WS rate pictures of lightning as significantly more upsetting (mean=4.92) than other nature scenes (mean=1.44,  $t(19)=19.80$ ,  $p<.001$ ). The results support lightning images as a stimulus for Study Two. Study Two utilizes reaction time and eye-tracking methodology during an ABR task, with each stimulus presentation consisting of one threatening image (lightning) and one nonthreatening image, followed by a probe. During pre and post-testing, the probe is placed randomly, but during ABR training the probe always follows the nonthreatening image. Individuals with WS show a faster reaction time to lightning images, even after ABR training, but this attention bias is not confirmed by the eye-tracking data. Future studies should consider reducing stimulus presentation time and increasing ABR training sessions to better evaluate the effectiveness of ABR training to reduce anxiety in individuals with WS.

### Attention Bias Reduction in Williams Syndrome

Williams syndrome (WS) is a complex disorder caused by genetic deletion on the seventh chromosome (Ewart et al., 1993), resulting in a variety of atypical physical, cognitive, and behavioral features. Individuals with WS present with atypical facial characteristics (e.g. broad forehead, full lips), medical abnormalities (most commonly a narrowing of the aorta), and intellectual impairment (Mervis, Robinson, Bertrand, Morris, Klein-Tasman, & Armstrong, 2000; Morris & Mervis, 2000). Children with WS display a hypersocial personality and score higher on sociability measures than typically developing (TD) individuals (Doyle, Bellugi, Korenberg, & Graham, 2004).

Adults with WS have a high level of psychopathology, the most common being anxiety and phobias (Stinton, Elison, & Howlin, 2010). Children and adolescents with WS also experience high levels of persistent anxiety and specific phobias (Woodruff-Borden, Kistler, Henderson, Crawford, & Mervis, 2010). Dykens (2003) found that individuals with WS had more fears than other groups with intellectual impairment and a greater range of fears, as well as high levels of generalized and anticipatory anxiety, persistent fears, and fear avoidance behaviors. Questionnaires given to parents of individuals with WS indicated that their children showed elevated levels of fear in every section of the Fear Survey Schedule for Children-Revised (FSSC-R; Ollendick, 1983) compared to controls with intellectual delay. Individuals with WS were asked about their fears, and the most commonly listed were thunderstorms, loud sounds, and death or dead people (Dykens, 2003). To treat anxiety in individuals with WS the use of medication is often employed. Individuals with intellectual delay may respond differently to psychotropic medication for mental health than TD controls and may require altered doses (Handen & Gilchrist, 2006). Matson and Neal (2009) studied the medication treatment of mental

disorders in individuals with intellectual disabilities, and found that information on treatment efficacy was lacking, and methods used by researchers made the available data unreliable.

An in-depth study of specific medication use in individuals with WS was carried out in 2012 (Martens et al., 2012). The study was comprised of a survey completed by parents in regard to their children (including adult children) with WS. The responses of the 513 parents indicated that 24% of the participants' children with WS had been prescribed Serotonin Selective Reuptake Inhibitors (SSRIs), the majority of these for anxiety. An additional 12% had been prescribed another type of antidepressant or medication for anxiety. Reduced anxiety was noted in 81% of those taking an SSRI, while only 64% of those receiving other medications noted benefits. Despite the effectiveness of reducing anxiety, rates of side effects (34%) were reported in SSRI users, such as appearing zoned, sleep disorders, tics, and increased anxiety.

Published studies examining the use of non-medication interventions to treat mental health disorders in individuals with WS are infrequent. In one case study, a young man with WS who had a tendency to show socially inappropriate behavior participated in cognitive-behavioral therapy to address his symptoms (Klein-Tasman & Albano, 2007). A follow-up interview with his mother determined that some interventions, such as role playing situations with females, helped him become more aware of his socially inappropriate behavior. In contrast, other interventions, such as using videos to elicit arousal, may have worsened his symptoms.

Another case study using cognitive-behavioral intervention for anxiety and behavioral/emotional issues in two children with WS determined that the impact of therapy on anxiety was unclear (Phillips & Klein-Tasman, 2009). The first child had difficulty recognizing anxious thoughts and distinguishing different emotions, which made cognitive restructuring too difficult. As a result, other strategies were employed, such as teaching the use of self-statements

in response to teasing (e.g. “Who cares what they think?”). Although the intervention resulted in a slight reduction in outbursts and increased independent behavior, behavioral outbursts still remained a concern. In addition, anxiety levels remained the same or worsened according to the mental health assessments utilized. The second participant also experienced challenges with cognitive-restructuring and more concrete strategies, such as her mother limiting the number of times she could ask a specific question, were utilized. The second child in the study showed an improvement in anxiety and worry within the first four therapy sessions of therapy, with no significant improvements after that point. A follow-up with anxiety instruments did not indicate a significant change after therapy. Therefore, the effectiveness of cognitive-behavioral intervention in individuals with WS is questionable.

A 2000 study noted problems in interventions for adults with an intellectual disability and determined that these individuals may first need to be trained in therapy skills for therapy intervention to be effective (Dagnam, Chadwick, & Proudlove, 2000). Sturmey (2005) highlights the lack of empirical studies of psychosocial interventions (e.g., psychotherapy, group therapy) in individuals with intellectual disabilities, while still supporting these approaches. One critical response to Sturmey highlighted the lack of evidence for using these interventions without evidence of their efficacy, and a need for more research (Beail, 2005).

Research in TD individuals who have anxiety indicates that anxious individuals tend to show increased attention to stimuli that appear threatening. This ‘attention bias’ is often measured using a dot probe task, where two stimuli are presented on a screen, then they disappear and a probe follows in place of one stimuli. Attention bias is measured as the time it takes the participant to select the probe with a computer mouse. If the individuals consistently choose a certain type of stimuli faster (e.g., a threatening stimuli), they are described as having

an attention bias to that type of stimuli. Derryberry and Reed (2002) found slow disengagement (shifting attention from a stimuli) from threat-related stimuli in anxious individuals, and that individuals with both high anxiety and poor attention had the most difficulty disengaging from the negative stimuli. It has also been shown that highly anxious children have a greater bias to threatening words than non-anxious controls (Vasey, El Hag & Daleiden, 1996). An additional study found that youth with generalized anxiety disorder, social phobia, and/or separation anxiety disorder have a greater bias to threatening face images than peers (Roy et al., 2008). Highly anxious adults given a dot-probe task have a greater bias to threatening pictures than neutral pictures, and increased vigilance (tendency to continue looking to a stimuli) to higher threat images (Yiend & Mathews, 2001). A 2007 meta-analysis showed similarity in threat related bias in children and adults across anxiety disorders (Bar-Haim, Lamy, Pergamin, Bakermans-Kranenburg, & van IJzendoorn, 2007). Programs that manipulate attention bias, known as attention bias reduction (ABR), have shown promise in reducing anxiety in typical individuals.

A research review of ABR suggests that ABR treatments are a source of dramatic symptom reduction, and have advantages over traditional anxiety treatments (Bar-Haim, 2010). In adult individuals with social anxiety disorder, a program encouraging attention disengagement to disgusted faces (by having the probe repeatedly follow nonthreatening faces) resulted in lower social and trait anxiety. Importantly, these results continued through a four month follow up (Schmidt, Richey, Bukner, & Timpano, 2009). Adult individuals with generalized anxiety disorder using an ABR program demonstrated a decrease in anxiety and attention bias compared to controls with generalized anxiety disorder (Amir, Burns, Beard, & Bomyea, 2009). Attention bias reduction has also shown promise in anxious children. In a recent study of ABR training in

anxious children, 50% of participants who completed training no longer met clinical criteria for their diagnosis (Waters, Pittaway, Mogg, Bradley, & Pine, 2013). A home-based attention training program using a dot-probe task found that individuals had lower trait anxiety scores and attenuated state anxiety scores when dealing with a stressful life event following the training (See, Macleod, & Bridle, 2009).

To date, two studies on attention bias have been conducted in individuals with WS, although no studies have evaluated the effectiveness of reducing their attention bias to threatening stimuli. Dodd and Porter (2010) found that compared to controls, individuals with WS had greater attention bias to happy faces, but had similar threat bias to negative faces. In a later study, Dodd and Porter (2011) found that individuals with WS had increased bias to threatening nonsocial images, such as animals, disasters and medical procedures, compared to controls, and this bias was greater in those meeting criteria for an anxiety disorder. These studies support Dykens (2003) finding that individuals with WS have high levels of nonsocial anxiety, not social anxiety. However, the use of ABR training has not been investigated in individuals with WS.

The aim of the current experiment was twofold. Given that individuals with WS have previously shown anxiety toward thunderstorms (Dykens, 2003), the purpose of Study 1 was to compare the ratings of lightning images to the ratings associated with other environmental stimuli in individuals with WS. It was predicted that individuals with WS would have strong negative feeling toward the lightning images. The information gained from this experiment was used in the design of Study 2, which utilized a dot-probe task and eye tracking equipment to determine if individuals with WS show greater attention bias to images of lightning compared to TD control participants, and whether their attention bias can be altered. It was predicted that

individuals with WS would have a greater bias to lightning stimuli before ABR training, and a greater bias to nonthreatening stimuli after ABR training. This prediction is based on previous studies with TD populations that showed manipulating bias through ABR reduced anxiety symptoms. The information gained will be used in future studies that focus on anxiety reduction in individuals with WS.

### **Methodology: Study 1**

#### **Participants**

The study participants included 24 individuals with WS who were registered with the WS Association's Patient and Clinical Research Registry or from a local WS support group. Ages of participants ranged from 9 to 26 years of age ( $M=15.6$ ); 20.8% of participants ( $n=5$ ) chose to not disclose their age. Participants were 42.1% male ( $n=8$ ), and 57.8% female ( $n=11$ ); 20.8% of participants ( $n=5$ ) chose to not disclose their sex. Two participants were excluded because they did not complete the survey. An additional participant was excluded for lack of understanding the task, evidenced by his selection of the same survey answer 29 times (out of 30) despite the variety of images. Therefore, data was analyzed on 21 participants. The study was given IRB approval and consent was obtained by the participants or their guardians.

#### **Design**

Participants completed a survey to assess their response toward environmental stimuli. The survey was created for the current study and completed online. It consisted of demographic questions, sample survey items, and test items. An email containing directions and 4 separate hyperlinks were sent to participants. The first link included/asked for demographic information (i.e. age/sex) and a training set of 3 items to ensure understanding of the survey. The remaining three links contained ten survey items each, totaling 30 test items. Survey items (both sample and



test items) were presented on the computer screen one at a time. Each item consisted of a picture of an environmental stimulus (e.g., lightning, peaceful scenes), with a picture Likert scale underneath, where individuals selected how they felt about the environmental stimulus presented. The majority of these pictures, 60%, were of lightning. The scale used six different face images corresponding to the words “*Very happy*,” “*Somewhat happy*,” “*Just okay*,” “*A little sad*,” “*More sad*,” and “*Very sad or upset*.” The items were counterbalanced, half began with “*Very happy*,” and the other half began with “*Very sad or upset*.” There was no time limit for participants to rate the pictures.

## Results

On a Likert rating scale ranging from 1 (*Very happy*) to 6 (*Very sad or upset*), individuals with WS rated pictures of lightning as significantly more upsetting ( $M=4.92$ ) than pictures of other nature scenes ( $M=1.44$ ,  $t(19)=19.80$ ,  $p<.001$ ). To ensure that lightning ratings did not change as participant age increased, additional analysis was conducted. No relationship was found between age and the lightning image ratings ( $r=-.296$ ,  $p=.284$ ). These findings confirmed that pictures of lightning were a threatening stimulus for individuals with WS. Because of this finding, pictures of lightning were used as a stimulus for Study 2.

## Methodology: Study 2

### Participants

This study included a total of 24 participants: 10 individuals with WS and 14 TD participants. The study was given IRB approval and informed consent was obtained by the participants or their guardians. The WS group consisted of 10 individuals with aged 10-35 years ( $M=17.58$ ,  $SD= 7.51$ ). WS participants were recruited through a WS support group and through the WS Association’s Patient and Clinical Research Registry. The control group consisted of 14

participants aged 6-34 years ( $M=12.71$ ,  $SD=7.40$ ), and were recruited through friends and family members of WS participants, and through community fliers.

## **Materials**

**Pictures.** Copyright free images were purchased or obtained with the authors' consent for this study. The pictures had a resolution of 72 pixels per inch (ppi) and were 5 ½" wide and 4" high. Threatening stimuli included a total of 65 lightning pictures, and 11 other threatening pictures, such as an alligator or a spider. Nonthreatening stimuli included 55 pictures, such as peaceful nature scenes. The stimulus presented to participants consisted of one negative picture and one nonthreatening picture presented simultaneously.

**Dot probe task.** The dot probe task was programmed using E-Prime software and presented to participants on Tobii computer equipment. Tobii 1750 eye-tracking hardware and software were used to measure the participants' eye gaze movements throughout the study and to present the dot probe task. The dot probe task included a total of 90 trials (stimulus presentations), divided into 3 Blocks. Each trial consisted of one lightning or other threatening picture and one nonthreatening picture, side-by-side with their location (left and right) counter-balanced. After each stimuli presentation, a star (the probe) followed in the location of one of the stimuli. Block 1 was the pre-training condition and consisted of 20 trials with the star location randomized (following on the side of either image after the stimulus presentation). Block 2 was the ABR training condition and consisted of 50 trials, with the star always following the nonthreatening image. Block 3 was the post-training condition and consisted of 20 trials with the star location randomized after the stimulus presentation.

**KBIT-2.** The Kaufman Brief Intelligence Test Second Edition (KBIT-2) was administered to gain a measure of Verbal IQ, Nonverbal IQ, and Composite IQ (Kaufman & Kaufman 2004).

### **Procedure**

Participants were seated approximately 65cm from the screen and their eyes were calibrated using Tobii eye tracking equipment. Seating and the computer height were adjusted as necessary for calibration. Each trial began with a black fixation cross in the center of a white background, which remained until participant eye gaze had been detected. The fixation point disappeared and the two picture stimuli appeared on the screen. The images remained on the screen for 4000ms, then they disappeared and a star probe immediately followed in the location of one of the images. Participants used a computer mouse to click on the star as quickly as possible. If the star was not clicked, the response was not included in the data analysis. When the participant clicked on the star probe, music would play and a box that said “click here when ready” would appear. Once the box was clicked using the computer mouse, the fixation point again appeared and the next trial began. Reaction time (RT) data, the amount of time it took participants to click on the star, and eye gaze data were collected for each trial.

The study took approximately 10 minutes to complete, and breaks were offered before each of the Blocks. After all Blocks were completed, researchers administered the KBIT-2 to participants.

## **Results**

### **KBIT-2**

Individuals in the WS group had a mean verbal IQ of 71.0 ( $SD=13.08$ ), a mean nonverbal IQ of 66.3 ( $SD=21.82$ ), and a mean composite IQ of 65.1 ( $SD=17.85$ ). The WS group composite

IQ results reflected a mild intellectual delay. Individuals in the control group had a mean verbal IQ of 117.7 ( $SD=10.68$ ), mean nonverbal IQ of 110.6 ( $SD = 15.97$ ), and a mean composite IQ of 116.2 ( $SD=12.87$ ). The control group composite IQ was in the high average range.

### Reaction Time

A mixed design ANOVA was used to analyze the RT data, which is shown in Table 1. A significant main effect for group was found, as individuals with WS had a slower reaction time than controls [ $F(1,813)=31.906, p<.001$ ]. A significant main effect for block was also found, as both groups had a faster RT in Block 3 than in Block 1 [ $F(1,813)=22.602, p<.000$ ]. A significant interaction was demonstrated between group and picture type [ $F(1, 813)=6.24, p=.013$ ]; individuals with WS had a faster RT to negative stimuli, while control individuals had a faster RT to nonthreatening stimuli. There was no significant interaction between group and block, or between group, picture type, and block.

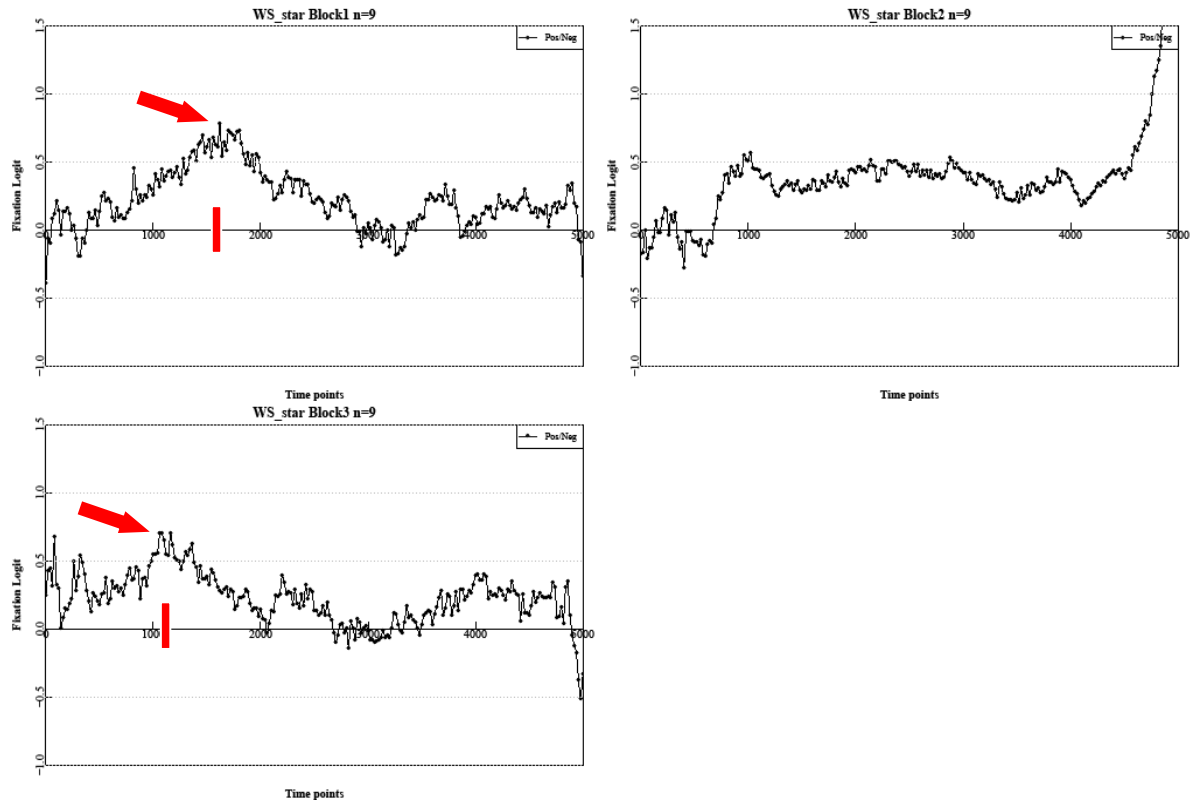
**Table 1** Reaction time in ms of participants by block and picture type

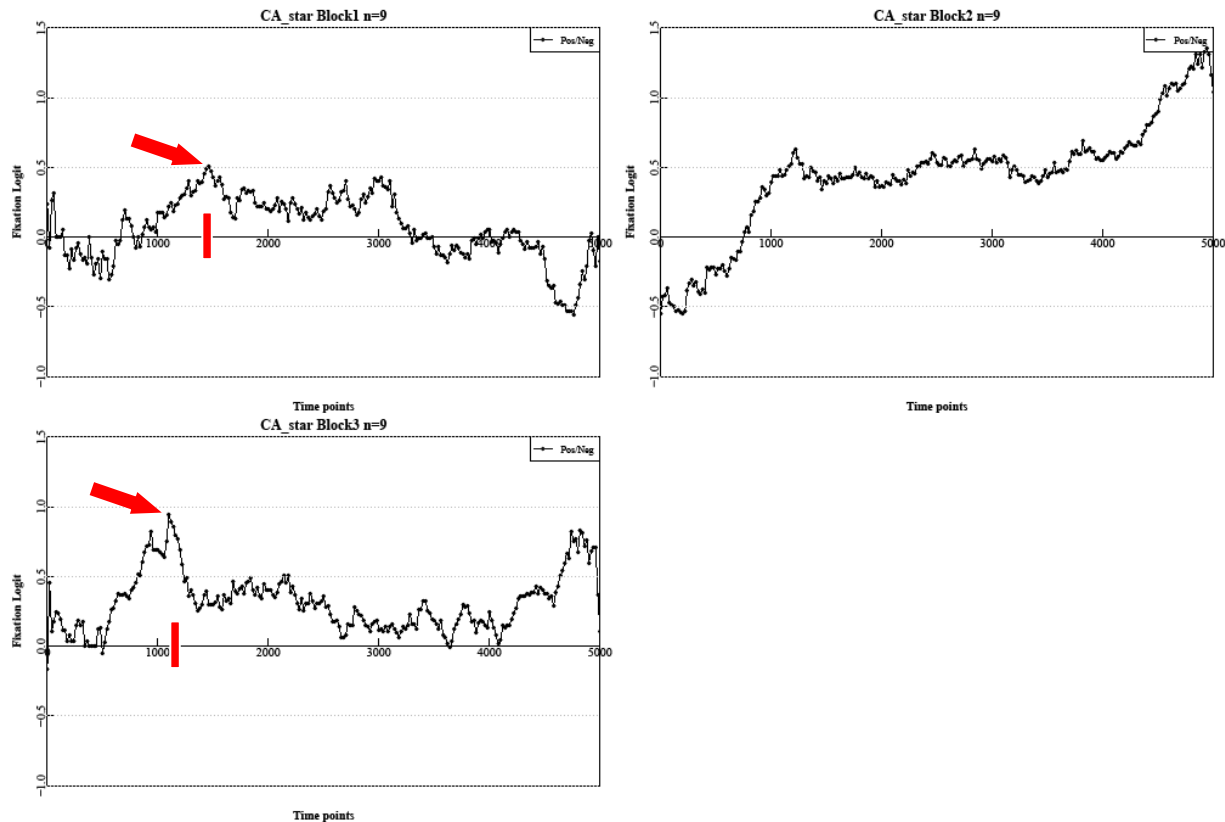
	Picture Type	WS	Control
		Mean( $SD$ ) in ms	Mean( $SD$ ) in ms
Block 1	Negative	958.67(418.39)	852.52(278.834)
	Nonthreatening	1063.44(499.92)	815.39(231.93)
Block 3	Negative	802.71(298.54)	757.58(260.03)
	Nonthreatening	908.15(442.58)	765.03(262.95)
Blocks 1 and 3 combined	Negative	882.13(371.73)	805.84(273.36)
	Nonthreatening	982.29(476.31)	798.78(248.94)

## Eye Tracking

Linear mixed modeling was used to analyze the eye tracking data. Each model included block and group as fixed predictor factors, with subject intercept and effects of predictor factors as subject random slope. The models analyzed the effects of block and group factors on the logit of fixation ratio for four time windows (1000 ms each).

There were no main or interaction effects of the two predictor factors in windows 1, 3, and 4 ( $p > .05$ ). In Window 2 (1000-2000ms), the individuals with WS gazed at the nonthreatening image more than the control group across Blocks 1 and 3 combined (beta=0.272,  $t=5.176$ ,  $p=.011$ ). There was a marginal interaction between group and block (beta=-0.355,  $t=-3.38$ ,  $p=.089$ ); specifically, the WS group fixated on the nonthreatening images more in Block 1 than in Block 3, while the direction was opposite in the control group. Further analysis of the 1000-2000ms window confirmed that WS group looked more at the nonthreatening images in Block 1 compared to Block 3 (beta=-0.306,  $t=3.523$ ,  $p=.048$ ). However, a trend was noticed in that both groups reached a peak toward the nonthreatening stimuli faster in Block 3 than in Block 1, which is the point of greatest group gaze allocation to the nonthreatening stimulus. These results suggest that each group tended to look at the nonthreatening stimulus more quickly after ABR training (See Figure 1).





*Figure 1.* Williams syndrome (top half) and control (bottom half) eye tracking data. Red arrow and line represent peak toward nonthreatening stimuli.

## Discussion

Individuals with WS have high levels of anxiety (Stinton et al., 2010; Woodruff-Borden et al., 2010), particularly toward threatening environmental stimuli, such as thunderstorms (Dykens, 2003). Typically developing individuals with high rates of anxiety show an attention bias to threatening stimuli, and show reductions in anxiety after completing ABR programs (Bar-Haim et al., 2007; Yiend & Mathews, 2001). Individuals with WS also show attention bias toward threatening stimuli (Dodd & Porter, 2011), but the effectiveness of ABR training on individuals with WS has not yet been investigated. The present study hypothesized that individuals with WS would find images of lightning significantly more upsetting than other

environmental stimuli (Study 1) and predicted that individuals with WS would demonstrate a bias to lightning stimuli, which would be reduced following ABR training (Study 2).

The results of Study 1 confirmed that individuals with WS, regardless of age, find pictures of lightning to be very upsetting. These results support Dykens, (2003) finding that individuals with WS have a strong fear of thunderstorms. Therefore, lightning is an ecologically valid stimuli to investigate the effectiveness of ABR in individuals with WS.

The results from Study 2 were analyzed using RT and eye tracking data. The results of the RT data indicated that overall, individuals with WS displayed a faster RT to the lightning pictures than the nonthreatening pictures, suggesting an attention bias to threatening stimuli, while the control participants responded more quickly to the nonthreatening pictures. However, the RTs for the nonthreatening stimuli did not decrease in individuals with WS following ABR training, as had been predicted. The results of the eye tracking data (measured at 1000ms, 2000ms, 3000ms, and 4000ms), did not indicate the presence of an attention bias to the lightning stimuli in individuals with WS or a change in eye gaze following ABR training, although there was a trend for both groups' eye gaze to reach a peak toward the nonthreatening pictures more quickly following ABR training.

The current results found, not surprisingly, that individuals with WS had slower RTs than typically developing controls. Previous research has demonstrated that individuals with WS have impaired processing of visual-spatial and location information (Farran, 2008; Menghini, Addona, Costanzo, & Vicari, 2010) and slower RTs when using a computer mouse (Martens, Hasinski, Andridge, & Cunningham, 2012). The increased RT of individuals with WS compared to controls could reflect these impairments.



Previous research on attention bias (based on dot probe RT) shows that bias to threat can be shifted away from threat with ABR training (Amir et al., 2009). An important difference between the present ABR study and previous ABR studies is the length of training for participants. Bar-Haim (2010), in a review of literature on ABR, noted that the number of sessions in studies had varied from 1 to 10 sessions and the total number of trials varied from 160 to 7500. In the current experiment, Study 2 was comprised of only one session and consisted of only 90 trials. Therefore, this shortened exposure to ABR training most likely limited its effectiveness. The stimuli were presented for a longer duration to account for visual-spatial impairments, therefore the number of trials were decreased to offset the overall duration of each session.

The duration of the stimulus may have also impacted the attention bias results. The length of the stimulus presentation for Study 2 was 4000ms, rather than a faster rate more commonly used in other dot probe studies (typically 500ms). It was anticipated that individuals with WS would need more time to complete the task based on impaired visual-spatial abilities and anticipated slower reaction times using the computer mouse. The longer duration was also utilized to collect additional data on the gaze allocation of individuals with WS.

Research by Cooper and Langton (2005) suggests that attention in a dot probe task (at least in face image stimuli) is allocated differently based on stimulus duration. In Cooper and Langton's study, participants were placed into one of two groups that viewed stimuli at either 100ms or 500ms. Using RT data, the researchers found that the typical 500ms stimulus presentation did not reflect the bias demonstrated using the 100ms presentation rate. The RT data from Study 2 utilized only one presentation time that was longer than the typical duration, and may not give full insight to initial bias.

This study was the first to use eye tracking to study attention bias in individuals with WS. In both the WS and control group, there was a trend for a faster establishment of nonthreatening peak after ABR training. Future studies may wish to increase the sample size and consider more strict participation criteria, as Study 2 found younger individuals and those with greater cognitive impairment were found to have difficulty with the ABR program. It was also found that the length of the stimulus presented a problem in keeping the attention of some participants, so shortening the duration of the stimulus presentation could be beneficial. Future research should also consider increasing the number of sessions and trials of ABR, and minimizing the complexity and variety of the nonthreatening pictures.

A recent study utilized fMRI technology during a dot probe task and found impaired hippocampal response to threat in TD individuals with high levels of anxiety (Price et al., 2014). fMRI studies in individuals with WS have also shown hippocampal abnormalities (Meyer-Lindenberg, et al., 2005). Future research in WS and attention bias could combine fMRI technology and ABR training to better understand the applicability of ABR training as a means of anxiety reduction in individuals with WS.

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