

Title: Interpretation of ambiguous situations: evidence for a dissociation between social and physical
threat in Williams syndrome

Running Head: Interpretation bias in WS

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Abstract

Williams Syndrome (WS) is associated with an unusual profile of anxiety, characterised by increased rates of non-social anxiety but not social anxiety (Dodd & Porter, 2009). The present research examines whether this profile of anxiety is associated with an interpretation bias for ambiguous physical, but not social, situations. Sixteen participants with WS, aged 13-34 years, and two groups of typically developing controls matched to the WS group on chronological age (CA) and mental age (MA), participated. Consistent with the profile of anxiety reported in WS, the WS group were significantly more likely to interpret an ambiguous physical situation as threatening than both control groups. However, no between-group differences were found on the ambiguous social situations.

Keywords: Interpretation bias, cognitive bias, Williams syndrome, anxiety, developmental disorders

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Interpretation of ambiguous situations: evidence for a dissociation between social and physical threat in Williams syndrome

Williams syndrome (WS) is a rare neurodevelopmental disorder caused by a microdeletion on chromosome 7 (Ewart et al., 1993). The WS phenotype is associated with a mild to moderate intellectual impairment, facial dysmorphology, medical complications and outgoing social behaviour (Bellugi, Lichtenberger, Jones, Lai, & St, 2000; Doyle, Bellugi, Korenberg, & Graham, 2004; Mervis & Klein-Tasman, 2000). In addition to these phenotypic characteristics, there is emerging evidence that individuals with WS are at increased risk for anxiety disorders and, interestingly, that the profile of anxiety in WS may be atypical, with a possible dissociation between risk for social and non-social anxiety (Dodd & Porter, 2009; Leyfer et al., 2006; Meyer-Lindenberg et al., 2005). A plethora of research has examined the role of cognitive processes, such as attention and attribution, in anxiety in typically developing individuals (e.g. Bar-Haim, Lamy, Pergamin, Bakermans-Kranenburg, & van Ijzendoorn, 2007; Bradley, Mogg, White, Groom, & de Bono, 1999; Calvo, Avero, Castillo, & Miguel-Tobal, 2003; Eysenck, Mogg, May, Richard, & Mathews, 1991). However, very little is currently understood about the cognitive processes associated with the anxiety profile observed in WS. The present research aims to address this gap in the literature by examining whether individuals with WS exhibit an interpretation bias, such that ambiguous situations are interpreted as threatening, and whether there is evidence for a dissociation between social and non-social anxiety in this interpretation bias.

A Dissociation between Social and Non-social Anxiety in Williams Syndrome

Early studies examining psychopathology in WS noted unusually high levels of anxiety, fears and worries (Einfeld, Tonge, & Florio, 1997; Udwin, 1990). These observations have since been supported by studies using diagnostic interviews validated against the Diagnostic and Statistical Manual for Mental

Disorders (DSM; American Psychiatric Association, 1994). In the most comprehensive assessment of clinical anxiety in WS conducted to date, Leyfer et al. (2006) assessed a large sample of children with WS and found that rates of Generalised Anxiety Disorder (GAD; 12%) and Specific Phobia (54%) were unusually high when compared to rates reported for typically developing children and those with intellectual disabilities¹. Importantly, Leyfer et al. (2006) found little evidence that the rate of Social Phobia (1.7%) was increased in WS. Recent research has subsequently replicated these findings in an independent sample of children and adults with WS (Dodd & Porter, 2009). The elevated rates of GAD and Specific Phobia in WS in the absence of a corresponding increase in Social Phobia is of interest because, in the typically developing population, Social Phobia is often comorbid with GAD (Kessler, Chiu, Demler, & Walters, 2005; Lewinsohn, Clarke, Seeley, & Rohde, 1994). These results therefore suggest that the profile of anxiety in WS may be atypical, with unusually high rates of non-social anxiety in the context of normal or low rates of social anxiety.

This psychological evidence for a dissociation between social and non-social anxiety in WS has also been supported by neuroimaging research. For example, Meyer-Lindenberg et al. (2005) found that, relative to typically developing controls, individuals with WS exhibited elevated amygdala activation in response to threatening non-social stimuli, but attenuated amygdala activation in response to threatening social stimuli. In interpreting their findings, Meyer-Lindenberg et al. (2005) suggested that a pattern of 'dissociated fear (decreased social fear and increased non-social fear)' is present in WS (p.993). Consequently, both psychological and neuroimaging research suggest that there may be a dissociation between social and non-social anxiety in WS. To date, however, very little research has examined the cognitive processes associated with anxiety in this population. It is currently unclear therefore, whether the dissociation between social and non-social anxiety is also apparent at the cognitive level.

Interpretation Bias

One cognitive process that has been studied in relation to anxiety in the typically developing population is interpretation bias, which refers to the tendency for anxious individuals to interpret ambiguous stimuli in a threat-related way (Chorpita, Albano, & Barlow, 1996; Hadwin, Frost, French, & Richards, 1997; Muris, Rapee, Meesters, Shouten, & Geers, 2003). This is predominantly assessed using one of two methods: by examining how participants interpret words that have a threat-related and a neutral meaning (homophones such as 'dye'/'die' or homographs such as 'stroke'); or by asking participants what they would think was happening in ambiguous situations such as 'you're lying in bed at night when you hear a big crash in the house'. In this latter situation, an example of a threatening interpretation would be that there was a robber in the house. In contrast, a non-threatening interpretation would be that someone dropped something. There is substantial evidence, based on research using both methodologies, that clinically anxious adults and children, and individuals high in trait anxiety, exhibit a tendency to interpret ambiguous stimuli in a threatening way (Creswell, Schniering, & Rapee, 2005; Eysenck et al., 1991; Hadwin, Frost, French, & Richards, 1997; Muris, Rapee, Meesters, Shouten, & Geers, 2003). In one such study, Barrett, Rapee, Dadds, and Ryan (1996) asked clinically anxious and non-anxious control children what they would think was happening in twelve ambiguous situations. The results indicated that the anxious children were significantly more likely to interpret ambiguous situations in a threatening manner than the non-anxious control children, providing clear support for an interpretation bias in the clinically anxious group.

The Relationship between Anxiety and Interpretation Bias

To date, the exact relationship between interpretation bias and anxiety remains unclear. There is some indication that interpretation bias may be a consequence of anxiety symptoms. For example, it has been demonstrated that interpretation bias is diminished following Cognitive Behavioural Therapy

to treat anxiety (Creswell et al., 2005; Waters, Wharton, Zimmer-Gembeck, & Craske, 2008). However, interpretation bias does not necessarily correlate with anxiety symptoms (e.g. Calvo et al., 2003), which is inconsistent with the idea that interpretation bias is simply a consequence of anxiety symptoms. Alternatively, interpretation bias may be related to anxiety vulnerability or play a role in the onset of anxiety. Recent research using training paradigms has provided initial evidence to support this hypothesis. For example, Wilson, MacLeod, Mathews & Rutherford (2006) used a training program to induce an interpretation bias towards non-threat and an interpretation bias towards threat in two groups of non-anxious participants and found that, following an emotional event, anxiety was increased in those participants who had received the training towards threat relative to those who had received training towards non-threat. Furthermore, Mathews, Ridgeway, Cook, and Yiend (2007) found that when participants were trained to make benign interpretations, this led to significant decreases in anxiety symptoms.

A number of authors have sought to examine whether interpretation biases are specific to an individual's concerns or indicative of a general underlying negativity. Research examining the specificity of interpretation bias has typically focused on the social/non-social threat distinction, where the feared outcome of social threat is negative evaluation and the feared outcome of non-social threat is physical harm (Campbell & Rapee, 1994). Using physically-based and socially-based ambiguous situations, Barrett et al. (1996) found that typically developing children who are generally anxious tend to interpret both physical and social ambiguous situations as threatening. In contrast, there is some evidence that Specific Phobia may be associated with interpretation bias for physical but not social ambiguous situations (Barrett et al., 1996) and that Social Phobia may be associated with interpretation bias for social, but not physical, ambiguous situations (Amir, Foa, & Coles, 1998; Miers, Blöte, Bogels, & Westenberg, 2008; Wilson & Rapee, 2005).

Summary, Aims and Hypotheses

The aim of the present research was to examine whether individuals with WS exhibit an interpretation bias and whether there is evidence for a dissociation between social and non-social threat in this interpretation bias. The ambiguous situations task used by Creswell et al. (2005) and Barrett et al. (1996) was used. In this task participants are asked what they think is happening in each of twelve ambiguous situations, six that are physically-based and six that are socially-based.

The WS group was compared to community samples of typically developing groups individually matched to the WS participants on mental or chronological age. It was anticipated that the WS group would be more likely to interpret the physical scenarios as threatening than both control groups but no differences were expected on the social scenarios. The relationship between threat interpretations and anxiety was examined within the WS group and the controls. As previous findings have been inconclusive, no predictions were made regarding this relationship.

Method

Participants

There were three groups of participants: a Williams syndrome group; a mental age matched control group; a chronological age matched control group. Descriptive data for the three groups are shown in Table 1.

[Insert Table 1 about here]

Williams syndrome group.

Sixteen individuals with WS (9 male, 7 female), aged between 13 years 0 months and 34 years 9 months, with a mean age of 21 years 0 months, participated. WS participants were recruited through

the Australian Williams Syndrome Association. All participants exhibited the typical WS phenotype (Bellugi et al., 2000; Dykens, 2003) and had received a diagnosis of WS following a positive fluorescent in situ hybridization (FISH) test showing deletion of the elastin gene at 7q11.23 (Fryssira et al., 1997). Participants were selected from a larger cohort of individuals with WS based on their mental age as assessed using the Woodcock-Johnson Test of Cognitive Ability – Revised (WJ-COG-R; Woodcock & Johnson, 1989, 1990). Due to the level of understanding and reasoning required to complete the ambiguous situations task, only individuals with a mental age of 6 years 6 months or above were invited to participate. The mental age of participants in the WS group ranged from 6 years 9 months to 10 years 6 months with a mean of 8 years 1 month. The overall level of impairment for all WS participants was in the mild to moderate range, typical of WS.

Current diagnostic status, according to DSM-IV criteria, was assessed for all WS participants through an interview with the primary caregiver using the Schedule for Affective Disorders and Schizophrenia for School-Age Children-Present and Lifetime Version (K-SADS-PL; Kaufman, Birmaher, Brent, Rao, & et al., 1997). The diagnostic data for these participants is included in a separate study (Dodd & Porter, 2009). The interviews were conducted by a postgraduate psychologist who had completed the training provided by the authors of the K-SADS-PL. A registered clinical psychologist supervised the interviews and was consulted regarding diagnoses. Seven of the WS participants met criteria for at least one anxiety disorder: five met criteria for a Specific Phobia; one met criteria for Generalised Anxiety Disorder; one met criteria for Obsessive Compulsive Disorder. There were no significant differences in chronological age or mental age between those WS participants who met criteria for an anxiety disorder (mean CA = 254.29 months (sd = 73.10); mean MA = 102.14 (sd=12.67)) and those who did not (mean CA = 250.33 months (sd = 79.63); mean MA = 93.00 (sd=11.31)), $p > 0.1$.

Chronological age comparison group (CA).

Sixteen typically-developing individuals (9 male, 7 female), individually matched to the WS participants on chronological age and sex, were recruited via a university-administered register of teenagers and young adults who are willing to participate in research. The CA group were well matched to the WS group on chronological age, $t(30)=0.027, p=0.979$.

Mental age comparison group (MA).

Sixteen typically-developing individuals (9 male, 7 female) whose chronological age and sex matched the mental age and sex of the WS participants, were recruited through local primary schools. Using chronological age as a proxy for mental age in this group, the MA group were well matched to the WS group on mental age, $t(30) = 0.143, p=0.887$.

All control participants were considered to be typically-developing. Children with a developmental disorder, clinical diagnosis, or any history of atypical development were not selected to participate.

*Materials**Ambiguous situations task.*

Participants' threat interpretations were assessed via the forced choice ambiguous situations task used by Creswell et al. (2005) and Barrett et al. (1996). Participants were given printed versions of twelve ambiguous situations (6 physical and 6 social) that could be interpreted in a threatening or non-threatening way². For each situation two interpretations (a threatening and a non-threatening interpretation) were provided and participants were asked to indicate the one that they would be most likely to make. The order in which the interpretations were presented was counterbalanced across the

task such that participants saw the threatening interpretation before the non-threatening interpretation in half the situations and vice versa for the other half. Non-threat responses were given a score of 0 and threat responses were given a score of 1. The total number of threat responses made in response to physical and social situations was calculated for each participant with a higher score reflecting more threat interpretations.

Spence Children's Anxiety Scale.

To assess current symptoms of anxiety, participants completed the self-report version of the Spence Children's Anxiety Scale (SCAS; Spence, 1998) and parents completed the parent-report version (Nauta et al., 2004). The parent and child versions of the SCAS have been developed for use with children aged between seven and eighteen years. The measures comprise 45 items loading to six scales: separation anxiety, social anxiety, fear of physical injury, obsessive-compulsive, panic and generalised anxiety. For the present research, the social anxiety, fear of physical injury (which assesses fears such as spiders and the dark) and generalised anxiety scales were of relevance to the physical and social theme of the ambiguous situations task. The SCAS has good internal consistency, with α coefficients of greater than 0.90 for the total score, adequate test-retest reliability over 6 months and good convergent and discriminant validity (Nauta et al., 2004; Spence, 1998). The child and parent versions of the SCAS have been used successfully in previous research with individuals with WS (Dodd, Schniering, & Porter, 2009).

Chronbach's alpha was calculated to assess the internal consistency of each scale of the self-report and parent-report measures for each group. Adequate internal consistency was found for the total anxiety score (alphas ranged from 0.82 – 0.93), the generalised anxiety scales (alphas ranged from 0.60 – 0.78) and the Social anxiety scales (alphas ranged from 0.53 – 0.87). However, the internal consistency of the fear of physical injury scale was not adequate for any of the groups on the parent report SCAS or for the WS and CA groups on the self-report SCAS (alphas <0.3), consequently, no further

analyses were conducted using this scale. The SCAS scores for each group are shown in Table 2. This shows that significant differences were found between the WS and CA groups on the SCAS parent report total anxiety scale, $t(15) = 3.676, p = .002$ and generalised anxiety scale, $t(15) = 2.429, p = .028$. Although group differences on the self-report SCAS did not reach significance, the WS group scored higher than both control groups on the total anxiety and generalized anxiety scales.

The SCAS was modified for participants who were no longer in school such that items that referred to school were edited to refer to work and items that referred to kids were edited to refer to people. For example the item 'I am popular amongst other kids my own age' was edited to read 'I am popular amongst other people my own age'. This was to ensure that the item content was appropriate for all participants.

[Insert Table 2 about here]

Procedure

Informed consent was obtained from the participants or their parents, as appropriate. The study was approved by the Macquarie University Human Ethics Committee. Participants completed both measures during a research session conducted either at their home or in a university laboratory. All participants completed the ambiguous situations task prior to the SCAS. After verbal instructions were given by the experimenter, participants were provided with printed versions of both measures. Where necessary, the items on both measures were read to the participants and their responses were recorded by the experimenter.

Results

The total number of threatening interpretations made for the physical (0-6) and social (0-6) situations was calculated for each participant. The median, interquartile range and range for each group are shown in Table 3. There was no missing data.

[Insert Table 3 about here]

As the data were ordinal, the non-parametric Wilcoxon matched pairs test was used to compare the WS group to both comparison groups. No analyses were conducted between the MA and CA comparison groups. The p -value used to indicate statistical significance was adjusted for each set of family-wise comparisons using the Bonferroni correction. Exact p -values and probability-based effect sizes are reported (A) as recommended for non-parametric data (Ruscio, 2008).

Threat Interpretation

Group comparisons.

A Bonferroni-corrected p -value of .0125 (0.05/4) was used to evaluate statistical significance. On the physical threat situations, the WS group made significantly more threat interpretations (median = 3.5) than both the MA (median = 1.5), $Z = -3.004$, $p = .003$ ($A = 0.81$), and the CA (median = 2), $Z = -2.542$, $p = .011$ ($A = 0.75$), comparison groups. In contrast, no significant differences were found between the WS group (median = 2) and either the MA (median = 1.5), $Z = -0.494$, $p = .621$ ($A = 0.053$), or the CA (median = 1), $Z = -0.856$, $p = .392$ ($A = 0.59$), comparison groups for the social threat situations.

Within-group comparisons.

Using a Bonferroni-corrected p -value of .017 (0.05/3), no significant differences were found between the number of threat interpretations made to physical and social situations for either the CA

(median for physical = 2, median for social = 2), $Z=-1.275$, $p=.202$ ($A=0.59$), or MA (median for physical = 1.5, median for social = 1), $Z=-0.91$, $p=.928$ ($A=0.50$), comparison groups. In contrast, the WS group made significantly more threat interpretations on the physical threat situations than the social threat situations (median = 3.5 for physical, median = 2 for social), $Z=-2.775$, $p=.006$ ($A=0.81$).

Relationship to anxiety.

As previous research has suggested that Specific Phobia may be associated with an interpretation bias for physical but not social situations, Table 3 shows the results for the WS group divided into those who met criteria for Specific Phobia and those who did not. These results suggest that the dissociation between physical and social interpretation bias was not limited to those WS participants who met criteria for Specific Phobia. In support of this, the main analyses were conducted again using only those WS participants who did not meet criteria for Specific Phobia and an identical pattern of results was found; significant differences between the WS group and both control groups were found for the physical threat ambiguous situations (MA: $Z = -2.448$, $p=.014$, $A=0.55$; CA: $Z=-2.209$, $p=.027$, $A=0.6$), but not for the social threat ambiguous situations (MA: $Z=-1.121$, $p=.262$, $A=0.85$; CA: $Z = -0.513$, $p=.608$, $A=0.85$), and a significant difference between the physical and social situations was found, $Z=-2.297$, $p=.022$ ($A=0.90$).

Figure 1 shows the total SCAS scores (self-report) and threat interpretation scores (for physical situations) for all participants, including those WS participants who met criteria for an anxiety disorder and those who did not. There is little evidence based on this figure that either anxiety diagnostic status or SCAS total score were associated with threat interpretation scores.

To examine this further, a Mann-Whitney U test was used to compare participants with WS who met criteria for an anxiety disorder with those who did not. No significant differences were found

between these groups on number of threat interpretations made for physical situations, $U=22.5$, $p=.354$ ($A=0.64$), or social situations, $U=22$, $p=.325$ ($A=0.58$).

[Insert Figure 1 about here]

To explore the relationship between anxiety symptoms and threat interpretations, Spearman Rank Correlation Coefficients were calculated between the self-report and parent-report SCAS scores (total score and scores for the social anxiety and generalised anxiety scales) and threat interpretations on the social and physical situations separately. For the typically developing controls and the WS group there were no significant correlations between SCAS scores and threat interpretations for either the physical or social situations ($p>.1$).

Bias, Age and Gender

To explore whether threat interpretations for ambiguous physical or social situations were related to chronological age or mental age, Spearman rank correlation coefficients were calculated. For the sample as a whole and each group in isolation, no significant correlations between number of threat interpretations made and either mental age or chronological age were found ($p>.05$). Further, Mann-Whitney U tests were conducted to examine the effect of gender on threat interpretation for ambiguous physical and social situations. No significant effect of gender was found for the entire sample or for any group in isolation ($p>.05$).

Discussion

The aim of the present research was to examine whether individuals with WS exhibit an interpretation bias such that ambiguous situations are interpreted as threatening. Both physical and social situations were included in order to examine whether the dissociation found between social and non-social anxiety at the diagnostic level is accompanied by a dissociation at the cognitive level. It was

hypothesised that, relative to typically developing controls matched on mental age or chronological age, the WS group would show an interpretation bias on the physical situations but not the social situations. The results provided clear support for this hypothesis. The WS group made significantly more threat interpretations on the physical situations than both control groups. In contrast, no group differences were found on the social situations. Furthermore, a within-group effect was evident for the WS group, who made significantly more threat interpretations on the physical situations than on the social situations, but not for either control group. These findings are highly consistent with the profile of anxiety reported in WS and suggest that the dissociation between social and non-social anxiety reported in previous research (Dodd & Porter, 2009; Leyfer et al., 2006; Meyer-Lindenberg et al., 2005) is accompanied by a dissociation at the cognitive level.

The Relationship between Interpretation Bias and Anxiety

As discussed, the findings of previous research have been mixed with regards the nature of the relationship between interpretation bias and current symptoms of anxiety; whether interpretation bias acts as a vulnerability factor, increasing the risk of subsequent anxiety, or whether interpretation bias is a consequence of anxiety. In the present research, there was little evidence for a relationship between interpretation bias scores and anxiety scores in the typically developing participants or in the WS group. Furthermore, there was no evidence that the interpretation bias found in the WS group differed between those participants who met criteria for an anxiety diagnosis and those who did not. In relation to WS, these findings provide some initial indication that the interpretation bias observed may be related to a general vulnerability to anxiety. These findings should, however, be treated as preliminary given the sample size of the present research.

The non-significant correlation between interpretation bias and anxiety in the typically developing control participants requires further consideration. Previous research has found associations

between anxiety and interpretation bias in typically developing children (e.g. Hadwin et al., 1997), although not consistently (Calvo et al., 2003). One possible explanation for these inconsistent findings is the anxiety measure used. In the present study, the SCAS was used to assess current symptoms of anxiety. However, previous research that has found significant relationships has typically used measures of trait anxiety. It remains possible, therefore, that a relationship between anxiety and interpretation bias may have been found had a trait measure of anxiety been used. It will be important, therefore, for future research to include a measure of trait anxiety to further explore the role of interpretation bias in anxiety in WS.

The results provide clear evidence for a dissociation between interpretation bias for physical and social ambiguous situations in WS. However, as discussed previously, there is some evidence for specificity of interpretation bias in anxious typically developing groups, particularly those with Specific Phobia (Barett et al, 1996). It is not clear from the present results, therefore, whether this dissociation is specific to WS or whether a similar pattern of results would be found in a typically developing sample with high rates of Specific Phobia. To investigate this indirectly, the main analyses were conducted again using only those WS participants who *did not* meet criteria for Specific Phobia. An identical pattern of results was found. This provides initial evidence that the dissociation found in the WS group was not due to differences in rates of Specific Phobia between the groups. However, to systematically examine whether the dissociation in interpretation bias found in the present research is unique to the WS population, future research should include a comparison group of typically developing individuals matched to the WS group on anxiety levels.

Implications for treatment of anxiety in WS

As discussed, there is extensive evidence that anxious typically-developing individuals are biased to interpret ambiguous stimuli as threatening (Creswell et al., 2005; Eysenck et al., 1991; Hadwin et al.,

1997; Muris et al., 2003). Given the interpretation bias evident for the WS group on the physical situations, the present findings suggest that the cognitive processes underpinning GAD and Specific Phobia in this atypical population may be similar to those that underpin anxiety in the typically developing population. It will, therefore, be of interest for future research to evaluate the efficacy of cognitive therapy in the treatment of anxiety in this population. Such treatments have been shown to be highly effective in the treatment of anxiety in the typically developing population. However, research examining the use of cognitive therapy to treat anxiety in WS remains in its infancy (Klein-Tasman & Albano, 2007; Phillips & Klein-Tasman, 2009).

Interpretation Bias and Social Behaviour in Williams Syndrome

Alongside an unusual profile of anxiety, WS is also associated with outgoing, social behaviour; individuals with WS are often described as behaving as if ‘everybody in the world is their friend’ and as lacking ‘stranger danger’ (Doyle et al., 2004; Jones et al., 2000; Sarimski, 1997). This atypical social behaviour has attracted significant research interest, particularly in recent years (e.g. Haas et al., 2009; Plesa-Skwerer et al., 2008; Riby & Hancock, 2008, 2009a, 2009b). In keeping with the methodology used in the present research, an interesting possibility is that the unusual social behaviour observed in WS may be related to a bias in the interpretation of social situations. The present results indicate that the WS group were less variable than the control groups in the number of threat interpretations made for ambiguous social situations but that the overall number of threat interpretations made by the WS group was similar to controls (see Table 3). As the ambiguous situations task used in the present research required participants to choose between only threatening and neutral interpretations, it remains possible that WS could be associated with a positive interpretation bias, such that individuals with WS are biased to interpret ambiguous social stimuli in a positive way. This will be an interesting question to explore in future research.

Methodological Limitations

The findings of the present research must be considered within the context of some limitations. Firstly, although a larger sample size would have been ideal, the sample size was restricted by both the rarity of WS and the cognitive demands of the task, which meant that only individuals with WS who had a mild to moderate intellectual impairment and a mental age of at least 6.5 years were able to participate. Participants were recruited nationally to ensure the sample size was as large as possible and the resulting participant numbers are comparable to many recent studies conducted with this population (e.g. Krajcsi, Lukacs, Igacs, Racsmany, & Pleh, 2009; Riby & Hancock, 2009a; Vicari, Bellucci, & Carlesimo, 2006). It is important to note that this study was not powered for comparisons between the WS participants who met criteria for an anxiety disorder and those who did not. These results must, therefore, be considered preliminary. Secondly, the ambiguous situations task was chosen because it has successfully been used in a number of previous studies (Barrett et al., 1996; Creswell et al., 2005) and is simple and easy to follow. However, the short situations and forced choice binary (threat/no threat) judgement may limit the ecological validity of the task and the response format may be susceptible to response bias. Other researchers have addressed this by using more detailed stories and allowed participants to outline their own interpretations (Bogels, Dongen, & Muris, 2003) or by using implicit cognitive tasks (Field, 2006). It will, therefore, be useful for future research to replicate the present findings using alternative paradigms.

Conclusion

This research is the first to examine interpretation bias in WS. The findings clearly indicate that individuals with WS are biased to interpret ambiguous situations as threatening when the situation is physically-based. In contrast, there was no evidence of an interpretation bias in WS for socially-based ambiguous situations. This pattern of results is highly consistent with the clinical presentation of

individuals with WS and suggests that the dissociation between social and non-social anxiety reported by psychological and neuroimaging research is mirrored by a dissociation at the cognitive level. It will be of interest for future research to examine whether interpretation bias plays a role in the onset and maintenance of anxiety in WS and also to consider whether interpretation bias may play a role in the outgoing, social behaviour observed in this unusual population.

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Footnotes

¹Chavira, Stein, Bailey, & Stein (2004) used the ADIS-IV to assess the prevalence of anxiety disorders in a large cohort of typically developing children and found that 3% met criteria for GAD, 10% met criteria for Specific Phobia and 6.8% met criteria for Social Phobia. Dekker and Koot (2003) reported that 17.5% of their sample of intellectually impaired children and young adults met criteria for a Specific Phobia, 0% met criteria for GAD and 2.5% met criteria for Social Phobia.

²Example physical threat situation: You are walking to a friend's house when a big dog comes towards you. What do you think the dog is going to do? Threat interpretation: The dog is going to bite you. Non-threat interpretation: The dog wants to sniff you and have a pat. Example social threat situation: You're having a party that starts at 3 o'clock, it's just after 3 o'clock and no one is there yet. What do you think? Threat interpretation: No-one wants to come to the party, Non-threat interpretation: They're running a bit late.

Table 1

Mean and standard deviation of age (years) and gender data for all groups

	N	Gender (M;F)	Chronological Age M (sd)	Mental Age M (sd)
Williams syndrome group	16	9;7	21.01 (6.19)	8.08 (1.04)
Chronological age matched group	16	9;7	21.06 (5.95)	
Mental age matched group	16	9;7	8.14 (1.23)	

Table 2

<i>Group means, standard deviations and effect sizes (Cohen's d) for group comparisons on the Spence Children's Anxiety Scale (SCAS) self-report and parent-report.</i> Scale	WS	CA	MA	<i>d</i>	<i>d</i>
	M (sd)	M (sd)	M (sd)		
Total anxiety – Self report	28.31 (12.85)	19.88 (14.50)	25.56 (12.14)	0.62	0.22
Social Anxiety – Self report	5.50 (3.08)	5.63 (4.27)	3.63 (2.55)	0.00	0.66
Generalised Anxiety – Self report	7.13 (3.96)	4.69 (2.85)	4.69 (2.57)	0.71	0.73
Total Anxiety – Parent report	20.88 (10.33)	10.31* (5.97)	15.13 (6.65)	1.25	0.66
Social Anxiety – Parent report	3.94 (2.17)	3.63 (1.75)	3.53 (1.81)	0.16	0.21
Generalised Anxiety – Parent report	4.88 (2.73)	2.75* (2.11)	3.13 (1.51)	0.87	0.79

Table 3

Group differences in Median and inter-quartile range on the number of threat interpretations made on social and physical situations.

	Social situations	Physical Situations
	Median	Median
	(Inter-quartile range)	(Inter-quartile range)
Williams syndrome group	2	3.5*
	(1-2)	(2-4)
<i>With Specific Phobia</i>	1.5	2.5
	(0-2.5)	(2 – 4.25)
<i>Without Specific Phobia</i>	2	4
	(1 – 2.25)	(2.75 – 4.25)
Chronological age matched group	1	2
	(0-2.75)	(1-3)
Mental age matched group	1.5	1.5
	(0-2)	(0 – 2.75)

* indicates significant between-group and within-group comparisons at $p < 0.01$.

Figure captions

Figure 1: The relationship between SCAS total score (self-report) and number of threat interpretations made on the physical threat situations (0-6) separated by group. WS = Williams syndrome; TD = typically developing; Anx = Anxiety.

TOP

