Social Approach in WS - 1

Title: Social approach in preschool children with Williams syndrome: The role of the face.

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Abstract

Background

Indiscriminate social approach behaviour is a salient aspect of the Williams syndrome (WS) behavioural phenotype. The present study examines approach behaviour in preschoolers with WS and evaluates the role of the face in WS social approach behaviour.

Method

Ten preschoolers with WS (aged 3-6 years) and two groups of typically developing children, matched to the WS group on chronological or mental age, participated in an observed play session. The play session incorporated social and non-social components including two components that assessed approach behaviour towards strangers, one in which the stranger's face could be seen and one in which the stranger's face was covered.

Results

In response to the non-social aspects of the play session, the WS group behaved similarly to both control groups. In contrast, the preschoolers with WS were significantly more willing than either control group to engage with a stranger, even when the stranger's face could not be seen.

Conclusion

The findings challenge the hypothesis that an unusual attraction to the face directly motivates social approach behaviour in individuals with WS.

Williams syndrome (WS) is a neurodevelopmental disorder, caused by a microdeletion on chromosome 7, with a prevalence rate of between 1 in 7,500 and 1 in 20,000 (Martin, Snodgrass, & Cohen, 1984; Stromme, Bjornstad, & Ramstad, 2003). Alongside a mild to moderate intellectual impairment and medical complications, a striking aspect of the WS behavioural phenotype is atypical social behaviour. In contrast to autism, which is associated with social withdrawal, individuals with WS are described as overly-friendly, hypersociable and always the centre of attention (Gosch & Pankau, 1997; Jones et al., 2000; Sarimski, 1997). Research examining social behaviour in WS has highlighted two areas of interest. Firstly, individuals with WS appear atypical in their social approach behaviour, particularly toward strangers. This behaviour is epitomised by the phrase 'everybody in the world is my friend' (Doyle, Bellugi, Korenberg, & Graham, 2004). Secondly, a growing body of research suggests that individuals with WS have a tendency to look intensely at faces (Jones et al., 2004; Mervis et al., 2003; Riby & Hancock, 2008, 2009). These two aspects of WS social behaviour are often discussed together (e.g. Riby & Hancock, 2008; Mervis et al., 2003) and have also been explicitly linked (Frigerio et al., 2006), however, there is a lack of research addressing the role that this attraction or interest in faces plays in WS social approach behaviour. The present research addresses this important theoretical question within the first detailed observation study of social approach behaviour in WS.

To assess social approach in WS, a number of studies have relied on parentreport of behaviour. These studies have consistently found that individuals with WS, even as young as 13 months, are rated as more sociable towards strangers than typically-developing children and children with other developmental disorders (Doyle et al., 2004; Dykens & Rosner, 1999; Jones et al., 2000; Sarimski, 1997). As an

alternative to parent-report, other researchers have assessed social approach behaviour by asking individuals with WS themselves to rate the approachability of pictures of strangers. Using this method, Bellugi, Adolphs, Cassady and Chiles (1999) found that individuals with WS rated strangers as more approachable than typically developing children. More recent studies using a similar method have, however, challenged this conclusion and suggest that approach ratings vary according to the emotion displayed by the stranger (Frigerio et al., 2006) and the participants' accurate recognition of this emotion (Porter, Coltheart, & Langdon, 2007).

Whilst the research described has contributed to our understanding of social behaviour in WS, there are several limitations to both methodologies. Firstly, although parents are an invaluable source of information, there are a number of difficulties with relying on parent-report, for example, knowledge of the social phenotype associated with WS and parent's expectations of their child's behaviour are likely to influence responses on these questionnaires. Secondly, the approach ratings method relies on the participants estimations of their behaviour, rather than their actual behaviour, and also depends on the use of a Likert scale, which may be difficult for the participants to use reliably (see Hartley & Mclean, 2006 for a discussion). Thirdly, both methods examine behaviour indirectly and consequently have questionable ecological validity. Finally, neither method provides details regarding exactly how social behaviour is atypical in WS. For example, social interaction in WS may be unusually intense, or prolonged, alternatively they may simply initiate interaction sooner that typically developing children. To develop an understanding of the processes that underpin the abnormal social behaviour observed in WS it is essential that we have a clear understanding about the way in which social behaviour is abnormal in this population.

The atypical social approach behaviour observed in WS and the causal processes that underpin it have attracted significant research attention. A number of researchers have proposed hypotheses regarding the psychological and neurological processes that underpin this atypical social approach behaviour (Bellugi et al., 1999; Frigerio et al., 2006; Porter et al., 2007). Of particular interest for the present research is the hypothesis proposed by Frigerio et al. (2006). Following observations that participants with WS tended to look intensely at researchers during experimental and medical procedures (Jones et al., 2000; Mervis et al., 2003), Frigerio et al. (2006) proposed that individuals with WS have high 'social stimulus attraction' (p.258) and that this drives the social approach behaviour observed in WS. Recent research using eye-tracking technology has provided the first experimental evidence that participants with WS tend to look at faces for extended periods (Riby & Hancock, 2008; 2009). Interestingly, however, Riby and Hancock (2009) found no evidence to suggest that faces capture attention abnormally in WS, a finding that seems to contradict the idea that faces directly motivate social approach behaviour in WS. Consequently research examining the role that the face plays in driving social approach behaviour in WS is currently of particular interest.

If the hypothesis proposed by Frigerio et al., (2006) is accurate and attraction to the face directly motivates social approach behaviour in WS, it follows that when the face is not visible, social approach behaviour in WS should be reduced. Recent findings have not supported this prediction; no differences were found in the approach judgements of participants with WS when rating photographs of faces versus photographs of people with the faces blacked-out (Porter et al., 2007). However, this finding is based on the approach ratings method described previously and is, therefore, subject to the limitations discussed previously.

The present research utilises an observational paradigm to compare the social approach behaviour of preschool children with WS to that of typically developing children matched to the WS participants on mental age or chronological age. The method is based on a well-known paradigm that is used to assess typically developing children's behaviour toward strangers (e.g. Kagan, Reznick, & Gibbon, 1989; Rapee, Kennedy, Ingram, Edwards, & Sweeney, 2005). In order to compare the behaviour of participants across domains, the paradigm incorporates both social and non-social components. In the non-social components, the participants' willingness to engage in an unfamiliar environment is assessed. In the social components, the participants' willingness to engage with a stranger is assessed. In one of these social components the stranger's face is visible. In the other, an addition to the original paradigm, the stranger's face is entirely covered using a burka. In keeping with previous findings, it was hypothesised that the participants with WS would be more willing to engage with the stranger than both control groups when the strangers face was visible. In contrast, in keeping with Frigerio et al. (2006), it was hypothesised that there would be no differences between the WS and typically developing groups in willingness to engage with the stranger when the strangers face was covered. No group differences were hypothesised for the non-social components.

Method

Participants

Ten preschool children with WS (6 male, 4 female) aged 3-6 years were recruited through the WS associations in 4 Australian states; all of the children known to the Australian WS association within this age range participated. All participants had received a diagnosis of WS following a positive FISH test showing deletion of the elastin gene at 7q11.23 (Fryssira et al., 1997).

The preschool scale of the Woodcock-Johnson Test of Cognitive Ability – Revised (Woodcock & Johnson, 1989, 1990) was conducted with all participants with WS to assess mental age. This measure was chosen as it provides an estimation of mental age from 2 years upwards based on a range of cognitive skills including short-term memory, vocabulary, auditory processing and visual processing. Two participants scored at basal (i.e. a mental age of 2 years) on this measure, consequently the Vineland Adaptive Behavior Scale, 2nd edition (VABS; Sparrow, Cicchetti, & Balla, 2005) parent interview was conducted for these participants as an additional measure of mental age to ensure that the WJ-COG-R mental age estimation was reliable. The mental age estimations provided for these two participants on the VABS were 1 year 10 months and 2 years, respectively. For these two participants, these mental ages were used for matching and analysis. We recognize the limitation of combining different measures, however, in this instance, there was little difference in mental age as assessed on the two instruments and it seemed that the slightly lower mental age for one participant was perhaps more reliable..

Twenty typically-developing children, recruited through local day-care centres and mothers' groups, also participated. Ten of these children were matched to participants with WS on sex and chronological age, and ten were matched on sex and mental age. Age data for each group are displayed in Table 1. All of the participants were Australian and of European or Far Eastern heritage.

[Insert Table 1 here]

Procedure

The procedure outlined below is based on that used extensively in previous research (e.g. Kagan et al., 1989; Rapee et al., 2005).

Parents were asked to remain as neutral as possible during the play session and to respond appropriately but briefly to interaction initiated by their child. The room was equipped with unobtrusive video cameras and recording equipment. Each play session included four components, two non-social components designed to assess willingness to engage in an unfamiliar environment (free-play, cupboard task), and two social components, one designed to assess participants' willingness to engage with a stranger whose face was visible, and one to assess participants' willingness to engage with a stranger whose face was covered. These play session components are outlined below in the order in which they occurred. The procedure for each component is described initially and then followed by a list of outcome variables coded for that component. Two female confederates, aged 22 years and similar to each other in height and build, assisted the researcher by acting as strangers. The ethical aspects of this study were approved by the Macquarie University Human Ethics Committee. Each component is detailed below.

Play session components

Free play (unfamiliar environment).

This component was designed to assess the willingness of participants to engage in an unfamiliar environment. Initially, the child and their parent were taken into the play room by the experimenter and the child was told that they could play with anything they liked. The experimenter then left the room and gave the child 3 minutes to explore the play equipment. There were three pieces of equipment available: a long dark tunnel, a rope ladder and a black 'textures' box. The textures box had a hole at one end for the child to put their hand through. Inside the box were items selected for their unusual texture.

The outcome variables coded for this component were: time taken to engage with the equipment (if the child was already engaged with the equipment when the experimenter left the room, this was coded as zero), proportion of time spent engaged with the toys and proportion of time spent within arms length of parent. For a participant to be considered 'engaged' with the equipment, they had to be touching or actively playing with the equipment.

Interaction with stranger – face available.

This component was designed to assess the willingness of participants to engage with a stranger whose face could be seen. After the first 3 minutes of free-play, a 'stranger' entered the room and sat down on a chair located in the opposite corner to the child's parent. Initially, the stranger made no eye-contact and did not initiate interaction (prompt 0). After 30 seconds, the stranger made eye-contact with the child and said "hello" (prompt 1). After a further 30 seconds, the stranger brought some toys into the room and said to the child "I have some toys here, would you like to play with me?" (prompt 2). The stranger then played with the toys at a children's table for approximately 8 minutes. This later condition was included to examine whether group differences in social approach would be affected by the attraction inherent in the introduction of new toys. The participants' behaviour was, therefore, coded separately for the period of time prior to the toys being revealed and the period of time after the toys were introduced.

Before the introduction of the toys

The participants' behaviour was coded according to whether they engaged with the stranger at the two stages of the component: before the first prompt and after the first prompt but before the toys were introduced. The proportion of time the participants spent engaged with the stranger (before the toys were introduced) was

also coded. To be coded as 'engaged' with the stranger they had to be smiling/waving directly at the stranger, touching the stranger, talking to the stranger or playing with the stranger.

After the introduction of the toys

The participants' behaviour was coded according to whether they engaged with the stranger at this stage of the component using the operational definition of 'engaged' outlined above. The proportion of time spent engaged with the stranger was also coded.

Cupboard task (unfamiliar environment).

Following the structured play, the stranger asked the participant whether they would like to play hide and seek and suggested that a good hiding place might be a cupboard in the room. This component was added to the paradigm to assess how comfortable the participants felt in the unfamiliar environment. However, none of the participants showed reluctance to hide in the cupboard; consequently no analyses were conducted for this component.

Interaction with stranger – face covered

This component was designed to assess the willingness of the participants to engage with a stranger whose face was covered. After a few minutes, a second stranger entered the room with their face completely covered (including the eyes). A dark blue burqa was used to cover the face and eyes completely whilst allowing the confederate to see. The stranger sat down on the chair in the opposite corner to the child's parent without making any attempts at interaction (prompt 0). After 30 seconds, the stranger said "hello" to the child (prompt 1). The stranger then remained in the room for another 90 seconds and did not interact further unless the child initiated interaction.

The participants' behaviour during this component was coded according to whether or not the participant engaged with the stranger at the two stages of the component: before the prompt; after the prompt. The proportion of time the participants spent engaged with the stranger was also coded. As outlined previously, to be coded as 'engaged' with the stranger participants had to be smiling/waving directly at the stranger, touching the stranger, talking to the stranger or playing with the stranger.

In the face available component, the stranger introduced toys. However, because of the unusual nature of interacting with a stranger whose face is covered, it was decided that the face covered component should be as short as possible and no toys were introduced.

Coding

Using explicit coding guidelines the play session videos were coded by the experimenter and second coded by one of two research assistants who were blind to the research hypotheses and participant group membership. The coding was conducted from video tapes of the play sessions to allow coders to pause and replay sections of the session if required for accurate coding. The participants' behaviour was coded for the entire duration of each component. As outlined above, to be considered 'engaged' with the equipment, participants had to be touching or actively playing with the equipment and to be considered 'engaged' with the stranger, participants had to be smiling/waving directly at the stranger, touching the stranger, talking to the stranger or playing with the stranger.

The variables coded for each component are outlined in the component descriptions above. All the variables coded either time or the stage at which the participant engaged. Excellent inter-rater reliability was found for both types of

coding variable: the 'proportion of time' variables (Spearman's Rank Correlation Coefficient: r = 0.90, p < 0.001) and the 'stage' variables (KAPPA = 0.9, p < 0.001). Indicators of validity were also obtained¹.

Statistical Analysis

For each variable, comparisons were made between the WS group and the two control groups; no analyses were conducted between the control groups. Two-tailed tests were conducted for all variables. Due to violations of the assumption of normality, non-parametric tests were used for all analyses. As the small sample size was small, a p-value of 0.05 was used to indicate statistical significance. There was no missing data.²

Results

For consistency, the results for each component are presented in the order used in the methodology.

Free play (unfamiliar environment)

Table 2 shows the median and interquartile range for the three coding variables for this component. The Wilcoxon matched-pairs procedure was used to examine between-group differences on these variables. On all three coding variables, the CA and WS groups behaved similarly. However, some differences between the WS and MA groups emerged. Compared with the MA group, the WS group engaged with the equipment significantly faster (Z=-2.366, p=0.018), spent more time engaged with the equipment, although this difference was not significant (Z=-1.784, p=0.084) and spent significantly less time within arms length of their parent (Z=-2.073, p=0.039).

[Insert Table 2 here]

Interaction with stranger (face available)

Before the introduction of the toys

Figure 1a shows the percentage of participants from each group who had engaged with the stranger at the two stages of the component. It is clear from this that only participants with WS engaged with the stranger prior to the first prompt ('hello'). Following this prompt, some of the control participants engaged with the stranger, however, even at this stage, more WS participants had engaged with the stranger than either control group. McNemar tests were conducted to examine group differences at these two stages statistically. The findings indicated that significantly more participants with WS engaged with the stranger without any prompts than both the CA (p=0.031) and MA (p=0.031) control groups and that significantly more participants with WS engaged with the stranger prior to the toys being introduced than the MA comparison group (p=0.031) but not the CA control group (p=0.375).

Figure 1b shows the median, interquartile range and range for each group on the proportion of time spent engaged with the stranger (before the toys were introduced) variable. Wilcoxon matched-pairs tests indicated that the WS group spent significantly more time engaged with the stranger than either the CA (Z=-2.533, p=0.008) or MA (Z=-2.527, p=0.008) control groups.

[Insert Figure 1 here]

After the introduction of the toys

As shown in Figure 2, the majority of participants from all three groups spent a significant proportion of time engaged with the stranger once the stranger had revealed the toys. Using McNemar tests, no significant between-group differences were found in the number of participants who engaged with the stranger at this stage. Furthermore, using the Wilcoxon matched-pairs test, no significant differences were found in the proportion of time spent engaged with the stranger after the toys were

revealed. Close examination of Figure 2 does, however, indicate that there was much less intra-group variation in the WS group than the 2 control groups.

[Insert Figure 2 here]

Interaction with stranger – face covered

In keeping with the analyses conducted for the stranger - face available component, between-group differences at each stage of this component were also examined. Figure 3a shows the percentage of participants from each group who had engaged with the stranger at the two stages of the component. It is clear from this that more participants with WS engaged with the stranger prior to the first prompt ('hello') than either control group. Following this prompt, some of the control participants engaged with the stranger, however, even at this stage, more WS participants had engaged with the stranger than either control group. McNemar tests indicated that more participants with WS engaged with the stranger prior to the first prompt, than the CA control group (p=0.016) and the MA control group, although this did not reach statistical significance (p=0.07), and that more participants with WS engaged with the stranger at any stage than the MA comparison group (p=0.008) and the CA control group, although this later difference did not reach significance (p=0.062).

Figure 3b shows the median, interquartile range and range for each group on the proportion of time spent engaged with the stranger (face covered) variable. Wilcoxon matched-pairs tests indicated that the WS group spent significantly more time engaged with the stranger than either the CA (Z=-2.524, p=0.008) or MA (Z=-2.675, p=0.004) control groups.

Interaction with stranger face available vs face covered

To examine whether there were differences in participants' willingness to engage with the stranger in the face available (pre-toys) and face covered conditions,

further analyses were conducted. McNemar tests were used to examine whether participants were more likely to have engaged with the stranger in one of the two conditions. Analyses were conducted for each group separately and no significant differences were found (p>0.5). The proportion of time spent engaged with the stranger in the two components was also compared using Wilcoxon matched-pairs tests for each group separately. Again, no significant differences between the two conditions were found for any group (p>0.4).

[Insert Figure 3 here]

Discussion

The present study is the first detailed observation study of social approach in children with WS. As anticipated, the results provided clear evidence that preschool children with WS were more willing to engage with a stranger than were their MA or CA matched peers. Furthermore, the results suggest that a stranger's face does not need to be visible for this behaviour to be observed.

Examination of the participants' behaviour during the 'Interaction with stranger (face available)' component revealed interesting group differences. The results suggest that preschool children with WS differ from their typically developing peers specifically in their *initiation* of interaction with a stranger. Whilst nearly all of the participants engaged with the stranger once the toys had been introduced, only participants with WS initiated interaction with the stranger prior to the stranger acknowledging them by saying "hello" (see Figure 2). This result points to differences in the motivation of children with WS and typically developing children: typically-developing children may not be motivated to interact with a stranger unless the stranger has toys or has explicitly initiated interaction with them; in contrast, children with WS may be motivated simply by the sight of the stranger. This qualitative

difference in the willingness of children with WS to engage with a stranger has important practical implications with respect to the vulnerability and safety of individuals with WS.

By observing the approach behaviour of young children with WS when a stranger's face could and could not be seen, the role of the face in WS social approach behaviour was assessed. The findings did not support the hypothesis that attraction to the face directly motivates social approach behaviour in WS (Frigerio et al., 2006). The WS group required less prompts to engage with the stranger and spent more time engaged with the stranger than both control groups in both the face available and face covered components. This can be seen by comparing Figures 1 and 3 which clearly demonstrate the similarities in between-group differences across the face available and face covered conditions. Furthermore, no significant differences were found between the face available and face covered conditions for any of the participant groups, which suggests that the covering the stranger's face had little affect on initial interaction behaviours in any group.

The finding that atypical social approach behaviour is seen in WS even when a stranger's face cannot be seen is in keeping with previous research (Porter et al., 2007) and suggests that, although individuals with WS exhibit prolonged looking at the face (Riby & Hancock, 2008), this attraction to the face may not be the principal motivator of social approach behaviour in WS. This result appears to be consistent with the finding that faces do not capture attention abnormally in WS (Riby & Hancock, 2009). If faces directly motivated the social approach behaviour observed in WS then unusual attentional capture would be expected. Instead it is plausible that the atypical looking behaviour reported in WS is underpinned by a difficulty disengaging attention (Riby & Hancock, 2009) that may not be directly related to social approach

behaviour. An important point to consider here is that, whilst the present findings suggest that a stranger's face does not need to be visible for WS social approach behaviour to be observed, this does not rule-out the possibility that attraction to faces may play an important role in the *development* of social approach behaviour in WS. (see Fidler, Hepburn, Most, Philofsky, & Rogers, 2007 for a discussion). It has also been proposed that individuals with WS might have a decreased ability to inhibit social approach behaviour due to frontal lobe impairment (Porter et al., 2007). There is currently a lack of research directly examining this hypothesis but some support has been gained from neuroimaging research (Meyer-Lindenberg, Mervis, Faith Berman, 2006) and research suggesting executive inhibition deficits in WS (Mobbs et al., 2007). Although the present study was not designed to assess the predictions of the frontal lobe hypothesis, across the play session as a whole, the WS group tended to display greater exploratory behaviour than the typically-developing controls, particularly those matched on mental age. For example, in the free-play component, the WS group were faster to engage with the unfamiliar equipment than the typically developing controls, with most participants engaging before the experimenter had left the room. Furthermore, in the stranger face covered component, the WS group displayed a willingness to engage even though the stranger was wearing something highly unusual. Taken together, these behaviours seem consistent with the idea that individuals with WS are disinhibited.

One difficulty with the frontal lobe hypothesis is that other populations, for example children who have Attention Deficit Hyperactivity Disorder (ADHD), also have deficits in response inhibition, but these children do not typically display the social approach behaviour seen in WS. Consequently, as discussed by Frigerio et al.

(2006), the social approach behaviour observed in WS might be caused by a deficit in inhibition in combination with an unusual drive towards social stimuli.

Limitations and future research

An observational paradigm provides an ecologically valid assessment of behaviour and overcomes the difficulties associated with reliance on parent report or hypothetical judgements made by the individuals themselves. However, the need for the paradigm to be appropriate for the participants' chronological and mental age resulted in a restricted age range, which together with the rarity of WS, necessarily resulted in a small sample size. It is noteworthy that all participants aged 3-6 years known to the Australian WS Association participated. It may, however, be of interest for future research to use a paradigm that is designed specifically for older children or adults with WS to replicate these findings.

A second consideration regarding the present methodology is that, as initial approach behaviour towards strangers was of interest, the participants' behaviour was observed over short periods of time, which were consistent with those used in previous research (e.g. Kagan, et al., 1989). It is not possible, therefore, to draw conclusions about how the participants would have behaved had they been observed for longer periods. Finally, because the face-covered component was novel and participants' reactions were, therefore, unpredictable, this component was always conducted at the end of the play session. As such, the face available and face covered conditions were not counterbalanced. It is, therefore, possible that different patterns of behaviour may have been observed had the components been conducted in a different order.

Conclusion

The results suggest that preschool children with WS are atypical in their initiation of interaction with strangers and that attraction to the face may not be the principal motivator of this social approach behaviour. These findings raise a number of important questions for future research including the role of attention to faces, social drive and disinihibition play in the social approach behaviour observed in WS.

References

- Bellugi, U., Adolphs, R., Cassady, C., & Chiles, M. (1999). Towards the neural basis for hypersociability in a genetic syndrome. *Neuroreport*, 10, 1653-7.
- Bishop, G., Spence, S.H., & McDonald, C. (2003). Can parents and teachers provide a reliable and valid report of behavioural inhibition. *Child Development*, 74, 1899-1917.
- Doyle, T. F., Bellugi, U., Korenberg, J., & Graham, J. (2004). "Everybody in the world is my friend" Hypersociability in young children with Williams syndrome. *American Journal of Medical Genetics*, 124A, 263-273.
- Dykens, E. M., & Rosner, B. A. (1999). Refining behavioral phenotypes: Personality-motivation in Williams and Prader-willi syndromes. *American Journal on Mental Retardation*, 104, 158-169.
- Fidler, D.J., Hepburn, S.L., Most, D.E., Philofsky, A., & Rogers, S.J. (2007).

 Emotional responsivity in young children with Williams syndrome. *American Journal of Mental Retardation*, 112, 194-206.
- Frigerio, E., Burt, D., Gagliardi, C., Cioffi, G., Martelli, S., Perrett, D. I., & Borgatti, R. (2006). Is everybody always my friend? Perception of approachability in Williams syndrome. *Neuropsychologia* 44, 254-259.
- Fryssira, H., Palmer, R., Hallidie-Smith, K. A., Taylor, J., Donnai, D., & Reardon, W. (1997). Fluorescent In Situ Hybridisation (FISH) for hemizygous deletion at

- the elastin locus in patients with isolated Supravalvar Aortic Stenosis. *Journal* of Medical Genetics, 34, 306-308.
- Gosch, A., & Pankau, R. (1997). Personality characteristics and behaviour problems in individuals of different ages with Williams syndrome. *Developmental Medicine and Child Neurology*, *39*, 327-333. Hartley, S. L., & MacLean, W. E. (2006). A review of the reliability and validity of likert-type scales for people with intellectual disability. *Journal of Intellectual Disability Research*, *50*, 813-827.
- Jones, W., Bellugi, U., Lai, Z., Chiles, M., Reilly, J., Lincoln, A., & Adolphs, R. (2000). II. Hypersociability in Williams syndrome. *Journal of Cognitive Neuroscience*, 12, 30-46.
- Kagan, J., Reznick, J. S., & Gibbons, J. (1989). Inhibited and uninhibited types of children. *Child Development*, 60, 838-845.
- Martin, N.D.T., Snodgrass, G.J.A.I., & Cohen, R.D. (1984). Idiopathic infantile hypercalcemia: a continuing enigma. *Archives of Disease in Childhood*, *59*, 605-13.
- Mervis, C. B., Morris, C. A., Klein-Tasman, B. P., Bertrand, J., Kwitny, S.,
 Appelbaum, L. G., & Rice, C. E. (2003). Attentional characteristics of infants
 and toddlers with Williams syndrome during triadic interactions.
 Developmental Neuropsychology Vol 23(1-2) 2003, 243-268.
- Meyer-Lindenberg, A., Mervis, C.B., & Berman, K.F. (2006). Neural mechanisms in Williams syndrome: a unique window to genetic influences on cognition and behaviour. *Nature Reviews Neuroscience*, *7*, 380-393.
- Mobbs, D., Eckert, M. A., Mills, D., Korenberg, J., Bellugi, U., Galaburda, A. M., & Reiss, A. L. (2007). Frontostriatal dysfunction during response inhibition in Williams syndrome. *Biological Psychiatry*, 62, 256-264.

- Pober, B.R., & Dykens, E.M. (1996). Williams syndrome: An overview of medical, cognitive and behavioural features. *Child and Adolescent Psychiatric Clinics of North America*, *5*, 929-943.
- Porter, M. A., Coltheart, M., & Langdon, R. (2007). The neuropsychological basis of hypersociability in Williams and Down syndrome. *Neuropsychologia*, 45, 2839-2849.
- Rapee, R.M., Kennedy, S., Ingram, M., Edwards, S., Sweeney, L. (2005). Prevention and early intervention of anxiety disorders in inhibited preschool children. *Journal of Consulting and Clinical Psychology*, 73, 488-497.
- Riby, D.M., & Hancock, P.J.B. (2008). Viewing it differently: Social scene perception in Williams syndrome and autism. *Neuropsychologia*, 46, 2855-60.
- Riby, D.M., & Hancock, P.J.B. (2009). Do faces capture the attention of individuals with Williams syndrome or autism? Evidence from tracking eye movements.

 **Journal of Autism and Developmental Disorders, 39, 421-431.
- Sarimski, K. (1997). Behavioural phenotypes and family stress in three mental retardation syndromes. *European Child & Adolescent Psychiatry*, 6, 26-31.
- Sparrow, S. S., Cicchetti, D. V., & Balla, D. A. (2005). *Vineland-II: Vineland Adaptive Behavior Scales* (2nd ed.). Circle Pines, MN: AGS Publishing.
- Stromme, P., Bjornstad, P.G., & Ramstad, K. (2002) Prevalence estimation of Williams syndrome. *Journal of Child Neurology*, *17*, 269-71.
- Woodcock, R. W., & Johnson, M. B. (Eds.). (1989, 1990). Woodcock-Johnson psycho-educational battery revised. Itascam IL: Riverside.

Table 1

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

Group	N	Mental age	Chronological age	
		M (sd)	M (sd)	
WS group	10	2;8 (0;8)	4;6 (1;0)	
CA match controls	10	4;5 (0;10)		
MA match controls	10		2;8 (0;6)	

WS-Williams Syndrome; CA-Chronological age; MA-Mental age

Table 2 The median (m) and inter-quartile range (iq range) for each group on the coding variables of the non-social component

Coding Variable	WS	CA	MA
	m (iq-range)	m (iq-range)	m (iq-range)
Time taken to engage with equipment	0	0	12
(in seconds)	0 - 7	0-39	0 - 34
Proportion of time spent engaged with	71.5%	77.5%	42%
equipment	(51%-94%)	(66%-88%)	(21%-65%)
Proportion of time spent within arms	0%	0%	27%
length of parent	(0%-9%)	(0%-13%)	(1%-60%)

Footnotes

¹Spearman rank correlation coefficients were conducted to examine the relationship between the coding variables during interaction with the unmasked stranger (i.e. stage at which child engaged with stranger and proportion of time child spent engaged with stranger) and the 'adults' scale of the parent report Behavioural Inhibition Questionnaire (BIQ; Bishop, Spence, & McDonald, 2003) for the WS group. A Bonferroni corrected p-value of 0.025 (0.05/2) was used to indicate statistical significance. The 'adults' scale of the BIQ was significantly correlated with the number of prompts required to engage with stranger, r=0.716, *p*=0.020, and the proportion of time spent engaged with stranger, r=-0.757, *p*=0.011, variables.

² Spearman rank correlation coefficients were conducted to examine the relationship between chronological and mental age and each coding variable for the participants with WS. No correlations reached significance at p < 0.05.

Figures

Figure 1: Comparison of groups on the 'interaction with stranger – face available' component before the toys were introduced (a) Proportion of participants who had engaged before and after the stranger said "hello" (b) Proportion of time spent engaged with the stranger during component. (* p < 0.05).

Figure 2: Comparison of groups on the proportion of time spent engaged with the stranger after the toys were introduced. No significant differences were found (p>0.05).

Figure 3: Comparison of groups on the 'interaction with stranger – face covered component. (a) Proportion of participants who had engaged before and after the stranger said "hello". (b) Proportion of time spent engaged with the stranger during component. (* p < 0.05).