Attrition of negative intention in Williams syndrome

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A B S T R A C T

People with Williams syndrome (WS) are said to have sociable and extremely trusting personalities, approaching strangers without hesitation. This study investigated whether people with WS are less likely than controls to attribute negative intent to others when interpreting a series of ambiguous pictures. This may, at least partially, explain their hypersociability toward strangers.

Twenty-seven individuals with WS and 54 typically developing controls (27 matched to WS participants on sex and chronological age and 27 matched on sex and mental age) viewed 10 ambiguous pictures, where one person in the picture may be seen as having a negative objective. Participants were asked to describe what was happening in the picture. Responses were scored for negative intention attribution (NIA).

NIA was reduced in WS individuals relative to typically developing controls of the same chronological age, but was similar to typically developing controls of the same mental age. Findings are discussed in relation to possible underlying neurological and cognitive mechanisms and practical implications for understanding and teaching stranger danger to people with WS.

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1. Introduction

Williams syndrome (WS) is a rare neurodevelopmental disorder caused by a microdeletion on chromosome 7 (Ewart et al., 1994). The microdeletion is associated with medical complications, a specific set of facial features and abnormal brain and cognitive development, including a mild to moderate intellectual impairment. In addition, and of direct relevance to the current study, WS individuals are known for their remarkably outgoing and extremely trusting personalities, they are also described as hypersociable – approaching people, even strangers without hesitation (Bellugi, Liclitenerberger, Jones, Lai, & George, 2000; Doyle, Bellugi, Korenberg, & Graham, 2004; Jones et al., 2000; Mervis & Klein-Tasman, 2000).

As a manifestation of their hypersociability and extremely trusting personalities, one of the biggest social concerns reported by parents of people with WS is their seemingly limited awareness of stranger danger or their indiscriminant behavior toward strangers and familiar others. In one anecdote, a young girl with WS was at the beach and overheard a stranger telling her children they were going to a fast food outlet. According to the girl’s mother, the girl “went over and got in the lady’s van, got in the backseat, buckled up, and was waiting to be taken to Dairy Queen with that family” (Spiegel, 2010).

Hypersociability in WS has also been studied empirically. Questionnaire-based data exploring social approach in daily life shows that children with WS are more hypersociable than children with autism (Jones et al., 2000), Down syndrome (Doyle et al., 2000; Mervis & Klein-Tasman, 2000), and typically developing controls (Burack, Ladd, & Bornstein, 2000). A small number of studies have also explored this using attribution tasks (Jones et al., 2000; Mervis & Klein-Tasman, 2000; Godbee & Porter, 2013). The current study extends these findings and further investigates whether the attribution of negative intention towards stranger danger is affected in WS compared to a typically developing control group.

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et al., 2004; Jones et al., 2000), developmental disabilities of mixed etiologies (Klein-Tasman & Mervis, 2003), typically developing children matched for chronological age (TDCA) (Doyle et al., 2004; Jones et al., 2000; Tomc, Williamson, & Pauli, 1990), and typically developing children matched for mental age (TDMA) (Tomc et al., 1990). For example, Jones et al. (2000) administered the Salk Institute Sociability Questionnaire (SISQ) to the parents of children with WS, Down syndrome or typically developing controls. SISQ items were specifically designed to provide a global measure of sociability and two particular aspects of sociability: social approach behavior and social emotional behavior. Social Approach items can be further differentiated into approaching familiar others and approaching strangers. Results indicated that the WS group significantly exceeded the control group on all SISQ scales and the WS group exceeded the Down syndrome group on all but one subscale – approaching familiar others, indicating high social approach and high social emotional behaviors in WS. Similarly, Klein-Tasman and Mervis (2003) administered the Children’s Behavior Questionnaire (CBQ) and the Multi-dimensional Personality Questionnaire (MPQ) – Parent Version to explore hypersociability and other behavioral characteristics in WS and in children with a developmental disability of unknown etiology. The CBQ contains over three hundred statements about child behaviors in typical situations; parents are asked to rate how typical a particular behavior is of their child using a seven point scale. On the MPQ, parents are presented with a list of 34 adjectives and accompanying descriptions of individuals with high and low traits on each adjective and are asked to rate their own child on a four point scale. In line with the authors’ predictions, on the CBQ, the WS group rated significantly higher on empathy and approach and significantly lower than the mixed etiology group on shyness. On the MPQ, individual items were found to best differentiate the WS group from the mixed etiology group, including: Gregarious, People-Oriented, Tense, Sensitive and Visible.

Laboratory-based studies of social approach in WS, which look at children’s ratings of how much they would like to approach someone based on their facial expression (trustworthy/untrustworthy or happy, sad, angry/fearful expressions), have found that participants with WS display abnormally high positive approach ratings when compared to TDCA controls (Bellugi, Adolphs, Cassady, & Chiles, 1999; Frigerio et al., 2006) and TDMA controls (Frigerio et al., 2006; Jones et al., 2000; Porter, Coltheart, & Langdon, 2007), but not compared to individuals with Down syndrome (Porter et al., 2007). This research is discussed in more detail below.

To date, three explanations have been proposed to help explain this intriguing phenomenon: (1) The Amygdala Hypothesis; (2) The Social Salience Hypothesis and (3) The Frontal/Disinhibition Hypothesis (Mobbs et al., 2007; Porter et al., 2007). Of course, these explanations are not mutually exclusive and each may contribute, in part, to the hypersociability that characterizes WS. The aim of the present study was to investigate whether people with WS have a reduced tendency to attribute negative intentions to others. That is, when they see an ambiguous picture, are they less likely to attribute negative intentions to the people in the scene when compared to typically developing controls – are they less suspicious? If negative intention attribution (NIA) is reduced in WS relative to typically developing controls, this may also help to explain their striking lack of caution when it comes to approaching strangers.

1.1. Three hypotheses to explain hypersociability in Williams syndrome

The Amygdala Hypothesis suggests that the hypersociability in WS may be driven by social perception difficulties secondary to amygdala dysfunction, in particular, a difficulty in evaluating threat. On the social approach rating task, where patients are asked to rate using a 5 point Likert scale how likely it is that they would approach faces based on how trustworthy they look, Bellugi et al. (1999) found that their group of individuals with WS displayed abnormally high positive approach ratings when compared to TDCA controls. Moreover, they report that WS ratings were similar to ratings of patients with acquired bilateral amygdala damage (e.g. see Adolphs, Tranel, & Damasio, 1998; Hayman, Rexer, Pavol, Strite, & Meyers, 1998). Based on this finding, Bellugi et al. proposed that the hypersociability observed in WS may be secondary to amygdala impairment.

In support of the amygdala hypothesis, Martens, Wilson, Dudgeon and Reutens (2011) found a positive relationship between right amygdala volume on MRI and approachability ratings, particularly ratings of negative faces, using a similar approachability rating paradigm to that of Bellugi et al. (1999). Similarly, Capitão et al. (2011) found a specific impairment in recognizing negative facial expressions in WS, although this effect was no longer observed when they controlled for level of cognitive functioning.

Not all empirical evidence supports the Amygdala Hypothesis. In Bellugi et al. (1999) study, unlike the amygdala patients, the WS group rated trustworthy faces as significantly more approachable than untrustworthy faces; thus showing the normal rank order of approach. In addition, in their social approach paradigm, Frigerio et al. (2006) used pictures of various facial expressions rather than trustworthy and untrustworthy faces and found that the abnormally high positive approach ratings in WS were specific to the ratings of happy facial expressions. There were no abnormally high positive approach ratings for the ‘non-happy’ expressions. Likewise, Capitão et al. (2011) found similar approachability ratings across their WS and control groups using an array of positive, negative and neutral emotional stimuli. Like Frigerio et al., Porter et al. (2007) also found that the unusual approach ratings in WS were specific to happy expressions and did not occur for threatening (angry or fearful) faces. The above findings suggest that the hypersociability in WS is not due to a lack of knowledge regarding threat.

Frigerio et al. (2006) proposed a second explanation for this strong drive to approach people, which they named the Social Salience Hypothesis. Frigerio et al. suggest that social stimuli are particularly and abnormally salient in WS. Faces, in particular, appear to be more salient for WS individuals compared to typically developing controls. WS individuals are drawn
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