Individual differences in social behavior predict amygdala response to fearful facial expressions in Williams syndrome

Brian W. Haas, Fumiko Hoeft, Yvonne M. Searcy, Debra Mills, Ursula Bellugi, Allan Reiss

Center for Interdisciplinary Brain Sciences Research (CIBSR), Stanford University School of Medicine, 401 Quarry Road, Stanford, Palo Alto 94305-5795, CA, USA
Laboratory for Cognitive Neuroscience, Salk Institute for Biological Studies, USA
School of Psychology, Bangor University, United Kingdom

ABSTRACT

Williams syndrome (WS) is a genetic condition often paired with abnormal social functioning and behavior. In particular, those with WS are characterized as being relatively hypersocial, overly emotional/empathic, and socially uninhibited or fearless. In addition, WS is associated with abnormal amygdala structure and function. Very little is known however about the relationship between specific social behaviors and altered amygdala function in WS. This study was designed to compare three models that relate abnormal social behavior with amygdala function in WS (indiscriminate sociability, emotional and empathic sociability and social fearlessness). We used a social behavior assessment procedure (Salk Institute Sociability Questionnaire), functional magnetic resonance imaging and an implicit emotion face processing task to test these models. Our findings provide support for a model of abnormal social fearlessness by showing that in WS, abnormal amygdala response to fear is paired with an increased tendency to approach strangers. Specifically, individuals with WS that exhibited less amygdala response to fearful facial expressions (compared to neutral) also exhibited an increased tendency to approach strangers. These findings contribute to our understanding of social and emotional functioning in neurodevelopmental conditions and provide evidence that in WS, amygdala response to fear modulates social behavior.

1. Introduction

Williams syndrome (WS) is a neurodevelopmental condition caused by a hemizygous microdeletion on chromosome 7q11.23. WS is often paired with a distinctive, abnormal social and emotional phenotype. In particular, those with WS are often described as being relatively hypersocial (Bellugi, Adolphs, Cassady, & Chiles, 1999), overly emotional/empathic (Klein-Tasman & Mervis, 2000) and socially uninhibited or fearless (Gosch & Pankau, 1994; Meyer-Lindenberg, Mervis, & Berman, 2006). In terms of the brain, evidence suggests that alterations of the amygdala may in part contribute to the observed abnormal social and emotional phenotype in WS (Haas et al., 2009; Martens, Wilson, Dudgeon, & Reutens, 2009; Meyer-Lindenberg et al., 2005). Recently, brain-imaging studies have begun to explore the neural correlates of individual differences of social behavior in WS. For example, Martens and colleagues (2009) demonstrated that in WS, individual differences in social behaviors, such as with approachability biases, are associated with alterations in amygdala volume. Although studies have indicated that functional abnormalities of the amygdala occur in WS (relative to healthy controls) and that individual differences in social behaviors are associated with amygdala structure in WS (Martens et al., 2009), very little is known regarding the relationship between individual differences in social behavior and amygdala function in WS. This study was designed to investigate the relationship between social behavior and amygdala function in WS. We used functional Magnetic Resonance Imaging (fMRI) and an implicit emotion face processing task to test three models that relate abnormalities of social behavior with amygdala function in WS (Fig. 1).

One model posits that individuals with WS tend to display abnormal indiscriminate sociability (Doyle, Bellugi, Korenberg, & Graham, 2004; Einfeld, Tonge, & Florio, 1997). This model describes those with WS as being abnormally social and driven towards social interaction independent of emotional valence or arousal and is supported by studies showing that relative to mental and age-matched controls, individuals with WS tend to be rated as generally more “overly-friendly” (Mervis & Klein-Tasman, 2000), people-oriented and gregarious (Klein-Tasman & Mervis, 2003). In addition, as compared to controls, those with WS rate facial expressions as more
were collected within the 2000 ms following the onset of each stimulus. Participants were instructed to judge if each stimulus was either male, female or scrambled as quickly and as accurately as possible. Behavioral responses

Fig. 2. Examples of stimuli used in the experimental paradigm. Participants were presented with photographs of faces conveying fearful, happy, neutral expressions and scrambled images. Participants were instructed to judge if each stimulus was either male, female or scrambled as quickly and as accurately as possible. Behavioral responses were collected within the 2000 ms following the onset of each stimulus.

approachable (Bellugi et al., 1999) and tend to exhibit greater gaze duration towards socially relevant scenes (Riby & Hancock, 2008) and faces (Riby & Hancock, 2009). Some studies however, have also found normal approachability towards both negative and positive expressions in WS (Porter, Coltheart, & Langdon, 2007). Taken together, several studies provide evidence that WS is associated with abnormalities in overt social behavior and attention towards socially relevant information such as facial expressions. We predicted that if functional abnormalities of the amygdala are associated with indiscriminate sociability in WS, then we would observe a relationship between social approach towards strangers and amygdala response to fearful facial expressions compared to scrambled images (Fig. 1A and Fig. 2).

A second model posits that individuals with WS tend to display abnormal emotional and empathic sociability. This model describes those with WS as being highly emotionally responsive and particularly tuned to the affective states of others and is supported by studies showing that relative to controls, those with WS are rated higher in empathy (Klein-Tasman & Mervis, 2003), temperamental intensity (Tomc, Williamson, & Pauli, 1990) and tend to be relatively over-affectionate (Davies, Udwin, & Howlin, 1998). In addition, as compared to controls, those with WS are more emotionally responsive during social interaction (Fidler, Hepburn, Most, Philofsky, & Rogers, 2007), rate happy facial expressions as more approachable (Frigerio et al., 2006) and utilize more emotionally expressive language (Jones et al., 2000; Losh, Bellugi, Reilly, & Anderson, 2000). Other studies however, have demonstrated reduced expressive language in WS (Laws & Bishop, 2004). Together, several studies provide evidence that WS is associated with abnormalities in emotional and empathic processing. We predicted that if functional abnormalities of the amygdala are associated with the tendency to display emotional and empathic sociability in WS, we would observe a positive relationship between individual differences in emotional and empathic approach related behaviors and amygdala response to emotional (both fearful and happy) facial expressions (compared to neutral faces) (Fig. 1B and Fig. 2).

Lastly, a third model posits that individuals with WS are socially uninhibited with strangers and that they display abnormal social fearlessness. This model describes those with WS as being relatively unresponsive to social fear paired with an inappropriate tendency to approach strangers and is supported by studies showing that relative to controls, those with WS exhibit an inability to detect and respect social danger signals (Meyer-Lindenberg et al., 2006) and are less reserved towards strangers (Gosch & Pankau, 1994). In addition, as compared to controls, those with WS exhibit lower amygdala response to fearful facial expressions (Haas et al., 2009; Meyer-Lindenberg et al., 2005) and are less able to perceive negatively valenced emotional facial and vocal expressions (Plesaskwerer, Faja, Schofield, Verbals, & Tager-Flusberg, 2006). Taken together, these studies provide evidence that WS is associated with an abnormal social fear response and an increased tendency to approach strangers. We predicted that if functional abnormalities of the amygdala are associated with the tendency to display social fearlessness in WS, we would observe a relationship between individual differences in social approach related behaviors towards strangers and amygdala response to fearful facial expressions (compared to neutral) (Fig. 1C and Fig. 2).

2. Methods

2.1. Participants

Twelve adult participants with WS (8 females; mean (M) age = 29.46, standard deviation (SD) = 8.07, range = 18.03 – 43.58, 8 right handed) were recruited for this study. Subjects were excluded if they reported any current use of mood-altering medication, substance abuse during the 6 months prior to scan or any standard MRI contraindications. Each participant was recruited as part of an ongoing multiscenter
دریافت فوری
متن کامل مقاله

امکان دانلود نسخه تمام متن مقالات انگلیسی
امکان دانلود نسخه ترجمه شده مقالات
پذیرش سفارش ترجمه تخصصی
امکان جستجو در آرشیو جامعی از صدها موضوع و هزاران مقاله
امکان دانلود رایگان ۲ صفحه اول هر مقاله
امکان پرداخت اینترنتی با کلیه کارت های عضو شتاب
دانلود فوری مقاله پس از پرداخت آنلاین
پشتیبانی کامل خرید با بهره مندی از سیستم هوشمند رهگیری سفارشات