



Approachability and the amygdala: Insights from Williams syndrome

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ABSTRACT

Williams syndrome (WS) is a genetic neurodevelopmental disorder in which hypersociability is a characteristic feature. Given that the amygdala has been identified as an integral component of the neural system underlying sociability, researchers have suggested that the abnormal amygdala volumes found in individuals with WS may play a role in their hypersociability. The aim of this study was to examine the relationship between amygdala volume and hypersociability, as measured by approachability ratings, in 22 individuals with WS and 22 normal controls matched on chronological age, sex, and handedness. We confirmed previous findings of increased amygdala volumes and higher approachability ratings of both 'positive' and 'negative' faces in individuals with WS. A positive relationship between right amygdala volume and approachability ratings was found in individuals with WS, particularly ratings of 'negative' faces. The results unexpectedly revealed that individuals with WS report using features other than the eyes and mouth to determine approachability, particularly when they are younger. These findings support the theory that amygdala dysfunction in WS is related to their hypersociability. Furthermore, we propose that individuals with WS use atypical cognitive strategies compared to controls to determine approachability.

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1. Introduction

Williams syndrome (WS) is a genetic neurodevelopmental disorder caused by a hemideletion of approximately 28 genes in chromosome 7 (band 7q11.23) (Ewart et al., 1993), resulting in a mild to moderate intellectual delay and an uneven cognitive and behavioral profile. The cognitive profile is characterized in part by relative strengths in certain aspects of language and auditory memory (Mervis & Klein-Tasman, 2000) and specific deficits in visuospatial skills (Farran & Jarrold, 2004; Porter & Coltheart, 2006). Individuals with WS display more distractible behaviors and anxieties, particularly in non-social situations, than other groups with intellectual disabilities (Dykens, 2003; Einfeld, Tonge, & Florio, 1997; Gosch & Pankau, 1994; Laws & Bishop, 2004; Rosner, Hodapp, Fidler, Sagun, & Dykens, 2004). Although they struggle to maintain peer relationships, a salient behavioral feature displayed by individuals with WS is their hypersociability, characterized by an excessive desire to meet people and a lack of stranger anxiety (Bellugi, Adolphs, Cassady, & Chiles, 1999; Doyle, Bellugi, Korenberg, &

Graham, 2004; Frigerio et al., 2006; Jones et al., 2000). In fact, there is evidence that individuals with WS of all ages have a stronger social drive than others with similar levels of intellectual disability (Doyle et al., 2004).

Compared to mental age (MA)-matched controls, infants with WS gaze at the faces of strangers more intensely (Mervis et al., 2003) and young children with WS spend more time focused on faces than on objects (Laing et al., 2002). Increased social gaze has also been noted in eye-tracking studies, in which individuals with WS exhibit a prolonged gaze at human faces when viewing still and moving pictures (Riby & Hancock, 2008, 2009). Young children with WS also display fewer negative facial expressions and less vocal distress when separated from their mothers than do chronological age (CA) or MA-matched controls (Jones et al., 2000), which is a marker of early social behavior.

The hypersociability displayed by individuals with WS has also been evaluated using approachability tasks in which photographs of unfamiliar faces are rated to determine how approachable they appear. The results of studies using these measures have produced conflicting findings depending on the nature of the task stimuli and whether or not the faces intentionally displayed specific emotions. Individuals with WS rated 'positive' (trustworthy and approachable) faces and 'negative' (untrustworthy and unapproachable) faces as more approachable than either CA- or MA-matched controls (Bellugi et al., 1999; Jones et al., 2000). When viewing faces

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depicting specific positive and negative emotions, such as happiness, anger, or fear, individuals with WS rated only the happy faces as more approachable (Frigerio et al., 2006). Porter, Coltheart, and Langdon (2007), however, noted that Frigerio and colleagues did not consider the impact of emotion recognition on this approachability task and suggested that impaired emotional recognition abilities may influence approachability ratings in individuals with WS.

In addition to examining behavioral measures of approachability among individuals with WS, there is an effort to understand the neuroanatomical basis of their hypersociable behavior. One hypothesis is that the heightened approachability response may be related to impaired response inhibition by the frontal lobes. Mobbs et al. (2007) proposed that individuals with WS are less able to inhibit their social drive due to reduced activation within the frontostriatal region. Porter et al. (2007) also suggested that abnormal social approach behaviors in WS may be associated with frontal lobe dysfunction based on impaired response inhibition on the Shape School Test (Espy, 1997). Others have proposed that the neural basis of hypersociability in WS may be linked to abnormalities within the structure and/or function of the amygdala and its connections to the orbitofrontal cortex (Haas et al., 2009; Meyer-Lindenberg et al., 2005; Reiss et al., 2004). Previous research strongly suggests that the amygdala plays a key role in social behavior and the perception of social stimuli (Adolphs, 2002, 2003; Gloor, 1997; Phelps, 2006; Phelps & LeDoux, 2005; Rolls, 1992). Although some studies have demonstrated that specific amygdala lesions in rodents, cats, and nonhuman primates alter social responsiveness (Bunnell, Sodetz, & Shalloway, 1970; Dicks, Meyers, & Kling, 1969; Schreiner & Kling, 1956) and produce socially disinhibited behavior (Amaral, 2002), other studies suggest that animals with amygdala lesions demonstrate appropriate or even reduced social interactions (Emery et al., 2001; Kling & Brothers, 1992). Some of these discrepant findings may be in part due to the age at which the amygdalae were lesioned. Bauman, Lavenez, Mason, Capitanio, and Amaral (2004) found that neonatal nonhuman primates with bilateral amygdala lesions demonstrated increased social fear but decreased non-social fear. Interestingly, this is the opposite of the WS phenotype.

Human studies have also indicated that the amygdala is central to the perception of facial emotional expression (Adolphs, 2003; Herba & Phillips, 2004). Humans with bilateral amygdala damage tend to judge 'negative' unfamiliar faces as more approachable than normal controls (Adolphs, Tranel, & Damasio, 1998), although a larger study indicated that while some individuals with amygdala damage are significantly impaired in recognizing facial expressions of fear, others demonstrate normal fear recognition (Adolphs et al., 1999). Abnormally large amygdalae have also been associated with dysfunctional fear recognition and atypical social behaviors. Notably, individuals with X-chromosome deletion disorders, such as Turner syndrome, have difficulty recognizing fearful facial expressions and demonstrate impairments in social skills despite abnormally large amygdalae (Good et al., 2003).

Functional magnetic resonance imaging (fMRI) has also been utilized to demonstrate that the amygdala is involved in the perception of emotional facial expressions (Breiter et al., 1996; Yang, Menon, Reid, Gotlib, & Reiss, 2003) and in response to faces considered to be untrustworthy (Winston, Strange, O'Doherty, & Dolan, 2002). Meyer-Lindenberg et al. (2005) examined amygdala activation in individuals with WS and found diminished activation in response to threatening faces, but increased activation for non-social threatening scenes. Haas et al. (2009) also found evidence of disparate functions within the amygdala of individuals with WS, with evidence of reduced reactivity in response to fearful expressions and heightened reactivity in response to happy expressions.

The results of volumetric studies of the amygdala in individuals with WS have produced conflicting findings, possibly due to differences in methodology and samples across studies. Jernigan

Table 1

Demographic characteristics of the Williams syndrome and control participants.

Variable	Williams syndrome (N = 27)	Control participants (N = 27)
Gender		
Male	14	14
Female	13	13
Chronological age (years)		
Mean (SD)	16.9 (7.2)	16.9 (7.3)
Range	8–41	8–41
Handedness ^a		
Right	22	22
Left	5	5
IQ ^b		
Full Scale (SD) ^c	53.5 (9.3)	109.2 (11.2)
Range	40–74	86–133

^a Handedness was based on the Edinburgh Handedness Inventory (Oldfield, 1971).

^b Full Scale IQ was based on either the Wechsler Intelligence Scale for Children-Third Edition or the Wechsler Adult Intelligence Scale-Third Edition.

^c $p < .01$.

and Bellugi (1994) noted that in individuals with WS, the absolute volume of the limbic region, including the amygdala, was the only regional volume not significantly reduced compared to controls. Reiss et al. (2004) used both volumetric analysis and voxel-based morphometry to examine amygdala volume in a relatively large sample of individuals with WS and found that amygdala volume and gray matter density was disproportionately increased in WS compared to CA-matched controls. In contrast, Meyer-Lindenberg et al. (2004) examined amygdala volumes using voxel-based morphometry in a select group of individuals with WS who have normal IQ and found no increase in gray matter density within the amygdala.

Given the strong association between the amygdala and various measures of sociability, the aim of this study was to examine the relationship between amygdala volume and approachability ratings, as one measure of sociability, in individuals with WS and CA-matched controls. We hypothesized that amygdala volumes in individuals with WS would be disproportionately increased compared to controls and that individuals with WS would score higher than controls on a standardized measure of sociability (Adolphs' approachability task). We also hypothesized that there would be a positive correlation between amygdala volumes and approachability ratings in individuals with WS.

2. Materials and methods

2.1. Participants

Twenty-seven individuals with WS and 27 neurologically normal controls individually matched on chronological age, sex, and handedness were initially recruited for the study. See Table 1 for demographic data of the participants. Twenty-five of the individuals with WS had been clinically diagnosed with WS and genetic confirmation was obtained with fluorescent *in situ* hybridization (FISH). Two participants declined FISH and were diagnosed on the basis of the clinical phenotype. Informed consent was obtained from the parents/guardians of all participants as well as the adult control participants themselves. Ethics approval was received by the University of Melbourne, Australia, and the Austin Hospital, Australia.

2.2. Stimuli

The WS and control participants completed the modified version of Adolphs' approachability task previously used with individuals with WS (Bellugi et al., 1999). Forty black and white photographs of unfamiliar faces were taken from an original set of 100 stimuli, 20 of which comprised the most positive ratings ('positive faces') and 20 of which comprised the most negative ratings ('negative faces') obtained from normal controls in Adolphs' previous study (Adolphs et al., 1998). The photographs were individually displayed on a personal computer in random order. The participants were required to rate the approachability of each face using a 5-point Likert type scale. In addition to verbal descriptors, in this study the Likert scale was uniquely designed using colors to accommodate the individuals with WS who were illiterate. All participants were also administered a standardized IQ test (Wechsler, 1991, 1997) and the Judgment of Line Orientation task (Benton, des Hamsher, Varney, & Spreen,

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