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Williams syndrome hypersociability: A neuropsychological study of the amygdala and prefrontal cortex hypotheses

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ABSTRACT

Individuals with Williams syndrome display indiscriminate approach towards strangers. Neuroimaging studies conducted so far have linked this social profile to structural and/or functional abnormalities in WS amygdala and prefrontal cortex. In this study, the neuropsychological hypotheses of amygdala and prefrontal cortex involvement in WS hypersociability was explored using three behavioral tasks – facial emotional recognition task, a social approach task and a go no/go task. Thus, a group 15 individuals with Williams syndrome was compared to two groups of normal developing individuals – a group of 15 individuals matched for chronological age (CA) and 15 individuals matched for mental age (MA), and sex. Individuals with WS present a specific impairment in recognizing negative facial expressions and do not display impairments in response inhibition when compared with typically developing groups. Although these findings partially support the amygdala contribution to WS hypersociability, we found that general cognitive functioning predicted this performance. Additionally, individuals with WS did not differ from both CA and MA groups in the recognition of angry facial expressions, a finding suggesting that they are actually able to identify stimuli associated with social threat. Overall, the results seem to indicate that this social profile must be understood within a developmental framework.

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1. Introduction

Williams syndrome (henceforth WS) is a rare neurodevelopmental disorder caused by a hemideletion on chromosome 7q11.23 (Peoples et al., 2000). One of the most striking features of individuals with WS is their distinct social-affective profile, characterized by high sociability, disinhibition, over-friendliness (Bellugi, Adolphs, Cassady, & Chiles, 1999; Klein-Tasman & Mervis, 2003) and strong empathy (Klein-Tasman & Mervis, 2003).

The precise etiology of WS hypersociability is still unknown; however, neuroanatomical, genetic/molecular and behavioral/neuropsychological studies have been providing important clues regarding the neural and genetic mechanisms underlying WS social phenotype. Of the different brain areas, the amygdala and prefrontal cortex have emerged more consistently as the two main neuroanatomical regions that can be hypothesized to be associated with WS hypersocial phenotype.

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Amygdala, a component of the neural network underlying social cognition, plays an important role in processing emotional and social-related stimuli (Adolphs, 1999) and is related to monitoring environmental events such as danger (Amaral, 2002). These findings have led some researchers to propose that an abnormal amygdala processing may be responsible for the hypersocial behavior typical of WS phenotype (e.g., Galaburda & Bellugi, 2000; Reiss et al., 2004). However, structural magnetic resonance neuroimaging has produced inconsistent results regarding volume or gray matter density of amygdala in WS (Chiang et al., 2007; Jernigan, Bellugi, Sowell, Doherty, & Hesselink, 1993; Martens, Wilson, Dudgeon, & Reutens, 2009; Reiss et al., 2004). Functional studies found that people with Williams syndrome showed greater amygdala activation in response to threatening scenes than to threatening faces (Meyer-Lindenberg et al., 2005). Taking into account that amygdala plays a key role in monitoring danger; these authors proposed that its reduced activation in individuals with WS in response to threatening faces might contribute to their reduced fear of strangers and consequent social disinhibition. Moreover, recent fMRI studies also found an abnormal activation of the amygdala during face processing (Haas et al., 2009; Paul et al., 2009), providing additional evidence indicating that failure to recruit this area during face processing may underlie WS high sociability. Indeed, this hypothesis of an amygdala involvement in WS hypersociability is also congruent with data showing that individuals with lesions in this area are abnormally friendly towards others (Adolphs, 1999), suggesting impairment in evaluating the potential threat of objects or organisms in the environment prior to approaching them. Also, these individuals fail to recognize emotions such as fear, anger, disgust and sadness (Adolphs, 1999) and present a positive bias when evaluating trustworthiness and approachability of faces in natural poses (Adolphs, Tranel, & Damasio, 1998).

Together with abnormal patterns of amygdala activation during face processing, evidence of an abnormal connection between the amygdala and the prefrontal cortex (in particular the orbitofrontal cortex), suggests also a prefrontal involvement in the WS social phenotype (Meyer-Lindenberg et al., 2005). Thus, an abnormal reactivity of prefrontal areas has been implicated in WS social knowledge. For instance, in the study of Meyer-Lindenberg and colleagues (2005), WS individuals (as opposed to controls) showed a task-invariant pattern: orbitofrontal (OFC) cortex was not differentially activated, and both medial prefrontal cortex (MPFC) and dorsolateral prefrontal cortex (DLPFC) were equally responsive during face and scene matching. However, the demanding nature of those tasks was not similar (face matching task was more difficult than the non-social scene matching task). To overcome this limitation, Muñoz et al. (2009) used the same group of participants but using a more cognitively exigent task (Muñoz et al., 2009). Again, results were consistent with the previous findings; specifically, they evidenced increased amygdala reactivity to non-social relevant aversive stimuli in individuals with WS, regardless of cognitive load; and no prefrontal activation differences between conditions (contrasting with a greater prefrontal reactivity specifically in left DLPFC, left OFC and MPFC in the control group as a function of task difficulty). The results provide further evidence of disruption in amygdala-prefrontal circuitry in individuals with WS (Muñoz et al., 2009). Additionally, Mobbs et al. (2007) proposed that an abnormal frontostriatal circuit, in conjunction with abnormal connectivity between the amygdala and OFC, might be associated with the hypersocial profile characteristic of individuals with WS. The hypothesis of a prefrontal impairment being associated with the hypersociability found in WS has also been proposed by a set of behavioral studies (Porter, Coltheart, & Langdon, 2007; Rhodes, Riby, Park, Fraser, & Campbell, 2009) and is also congruent with specific structural brain patterns found in WS in terms of volumetric difference in the frontal lobes specifically, volumetric changes (Ewart et al., 1993; Gothelf et al., 2008; Meyer-Lindenberg et al., 2004; Reiss et al., 2004) and abnormal patterns of gyrification (Gaser et al., 2006; Schmitt et al., 2002).

Taken together, these characteristics suggest that abnormalities in amygdala and prefrontal cortex regulation may contribute to WS hypersocial profile. Whereas amygdala dysfunction is possibly related to an incapacity to recognize threat in the environment and thus to properly determine the trustworthiness and approachability of unfamiliar individuals (consistent with a positive bias in evaluating trustworthiness and approachability of faces in natural and emotional poses (Jones et al., 2000; Martens et al., 2009)), proposal of prefrontal cortex abnormalities is based upon clinical similarity with patients with prefrontal lesions (e.g., disinhibition in several circumstances, including social contacts, and displaying rigid and inflexible behaviors) (Semel & Rosner, 2003) and the fact that, despite the non-rewarding nature of their social relationships (e.g., problems in making and sustaining relationships and are often rejected by their peers), individuals with WS are continuously attracted to social stimuli, thus revealing some insensitivity to punishing and negative experiences.

Thus, the objective of the present study is to test amygdala and prefrontal cortex involvement in WS hypersociability with a neuropsychological approach, using three different behavioral tasks: an emotional recognition task; a social approach task; a judgment and response inhibition task. The two following hypotheses were tested:

(a) The amygdala hypothesis – if people with WS display manifestations of amygdala dysfunction similar to those with acquired damage to this structure, they should display impairment in recognizing negative emotions (specifically, anger, fearful and sadness). Also, they should judge photos of unfamiliar individuals as highly approachable, not rating happy expressions as more approachable than negative ones (especially those displaying in anger).
(b) The prefrontal hypothesis – if the abnormal social approach in WS is consistent with patterns of acquired frontal lobe impairment, individuals with WS should display impaired response inhibition on go/no-go tasks (Liddle, Kiehl, & Smith, 2001).

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