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Heterogeneity of social approach behaviour in Williams syndrome: The role of response inhibition

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ABSTRACT

The developmental disorder of Williams syndrome (WS) is associated with an overfriendly personality type, including an increased tendency to approach strangers. This atypical social approach behaviour (SAB) has been linked to two potential theories: the amygdala hypothesis and the frontal lobe hypothesis. The current study aimed to investigate heterogeneity of SAB in WS by exploring whether subgroups of SAB profiles could be identified using cluster analytic techniques. Twenty-five children with WS aged 6–15 years completed three behavioural tasks tapping (i) social approach behaviour, (ii) emotion recognition ability and (iii) response inhibition. Cluster analyses revealed preliminary evidence of WS subgroups based on SAB profiles and indicated that response inhibition ability was the key differentiating variable between SAB cluster profiles. The findings provide tentative support for the frontal lobe hypothesis of SAB in WS and highlight the importance of investigating SAB at a heterogeneous level.

1. Introduction

Williams syndrome (WS) is a neurodevelopmental disorder, with estimated prevalence rates ranging from 1:20,000 (Morris & Mervis, 2000) to 1:7.500 (Stromme, Bjømstad, & Ramstad, 2002) and is caused by the deletion of 25–28 genes on the long arm of chromosome 7 (7q11.23; Donnai & Karmiloff-Smith, 2000). The disorder is characterised by distinct, yet variable cognitive, physical and behavioural profiles (Hepburn, Fidler, Hahn, & Philofsky, 2011). Most individuals with WS have mild to moderate intellectual difficulties (Searcy et al., 2004); with verbal processing (Morris & Mervis, 1999) and certain aspects of language (Mervis & Klein-Tasman, 2000) identified as relative strengths within their cognitive profile. Specific areas of deficit include nonverbal processing and visuospatial skills (Farran & Jarrold, 2004). Individuals with WS have also been reported to display distractible behaviours (Dykens, 2003) and higher levels of anxiety than typically developing (TD) children and other groups with intellectual disabilities (Einfeld, Tonge, & Florio, 1997).

Hypersociability is frequently cited to be a defining feature of the social phenotype associated with WS (Järvinen-Pasley et al., 2010) and has been described as a 'general presentation of extreme happiness' (Levine & Wharton, 2000; p.364); being 'unusually sociable, friendly and empathic' (Jones et al., 2000, p. 30), an excessive interest in others and a distinct lack of inhibition with regard to approaching others in social contexts (Bellugi et al., 2007; Jones et al., 2000). Individuals with WS

Abbreviations: SAB, social approach behaviour.

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appear hypersociable from an early age (Doyle, Bellugi, Korenberg, & Graham, 2004), demonstrating an eagerness to make eye contact with and to approach strangers (Mervis & Klein-Tasman, 2000). An interest in looking at faces remains in childhood and into young adulthood (Riby & Hancock, 2008). Parents of children with WS often report concerns regarding the subsequent increased vulnerability and risk of exploitation that their children are exposed to as a result of their overfriendly behaviour and drive to approach strangers (Jones et al., 2000). This is especially relevant when considered alongside the developmental delay experienced by many individuals with the disorder (for a discussion of issues of social vulnerability, see Jawaid et al., 2012). Developing an understanding of social approach behaviour (SAB) has been increasingly prioritised over recent years and two hypotheses have been proposed: the amygdala hypothesis and the frontal lobe hypothesis (Porter, Coltheart, & Langdon, 2007). However, the literature on SAB in WS is fractionated by conflicting findings.

1.1. Amygdala hypothesis

The amygdala hypothesis suggests that atypically large amygdala volumes and subsequent amygdala dysfunction play a role in the aetiology of atypical SAB in WS (Bellugi, Adolphs, Cassady, & Chiles, 1999; Martens, Wilson, Dudgeon, & Reutens, 2009). The amydgala is a limbic structure that guides socio-emotional behaviour, plays a role in the identification of facial emotional expression (Adolphs & Spezio, 2006), and is required for accurate social judgment of individuals on the basis of their facial expressions (Adolphs., Tranel, & Damasio, 1998). Haas et al. (2009) reported findings of disparity in the amygdala functioning of individuals with WS compared to typical controls. The WS group demonstrated reduced amygdala reactivity in response to threatening faces and a heightened reactivity to happy expressions. It is suggested that the decreased amygdala activation to threatening faces evident in individuals with WS indicates a reduced reaction to social danger and helps explain the social disinhibition and reduced fear towards strangers observed in this population (Bellugi et al., 1999; Martens et al., 2005). Amygdala volume as well as functionality is likely to be related to social behaviours in WS and likely to be atypical (Martens et al., 2009). Martens et al. (2009) investigated the relationship between amygdala volume and approachability ratings in individuals with WS compared to TD controls. The findings revealed a significant relationship between increased volumes and higher approachability ratings in WS to both 'negative' faces and 'positive' faces which supports this hypothesis. However, Frigerio et al. (2006) and Porter et al. (2007) found that individuals with WS rated only the 'positive' faces as more approachable than controls whilst 'negative' faces were rated as less approachable. They concluded that individuals with WS are able to discriminate the approachability of individuals and their SAB was not a function of underlying emotion recognition difficulties.

1.2. Frontal lobe hypothesis

The frontal lobe hypothesis postulates that the atypical SAB in WS may result from impairment in response inhibition subsequent to frontal lobe dysfunction (Porter et al., 2007). Porter et al. (2007) describes the similarities in the atypicalities of SAB in WS and the SAB of patients with frontal lobe damage, and state that both groups seem to demonstrate a dissociation between 'knowing' and 'doing' which is reflected by their tendency to approach strangers in day-to-day life. Porter et al. suggest that individuals with frontal lobe damage 'know' that they shouldn't talk to or approach strangers but still go ahead and do so due to poor impulse control. Furthermore, several studies report neurological evidence to suggest that frontal lobe abnormalities do exist in WS (Meyer-Lindenberg et al., 2005; Mobbs et al., 2007) and behavioural tasks show evidence of executive functioning difficulties similar to those seen in individuals with ADHD (e.g. Rhodes, Riby, Matthews, & Coghill, 2011). Mobbs et al. (2007) used functional magnetic resonance imaging (fMRI) to investigate frontal lobe activation and found that the WS group demonstrated reduced frontostriatal activation compared to TD controls during a Response Inhibition Task. They suggest that individuals with WS display a generalised deficit in response inhibition which subsequently impacts upon their behaviour in social situations.

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Porter et al. (2007) investigated SAB in WS in relation to the theories discussed. They found that WS participants displayed emotion recognition abilities that were appropriate to their general level of cognitive functioning and did not display atypical responses on the social approach task. However, performance on a Response Inhibition Task was well below the level expected on the basis of their mental age or level of intellectual functioning. They therefore concluded that the tendency for WS individuals to approach strangers in everyday life may be due to poor response inhibition.

1.3. Heterogeneity of social behaviours in Williams syndrome

Research to date has focussed on describing SAB in WS as a homogenous construct. However, Porter et al. (2007) and Järvinen-Pasley et al. (2010) observed substantial variability in the approachability ratings given by individuals with WS