



Linking social behaviour and anxiety to attention to emotional faces in Williams syndrome



Hannah E. Kirk^{a,*}, Darren R. Hocking^a, Deborah M. Riby^b, Kim M. Cornish^{a,*}

^aSchool of Psychology and Psychiatry, Monash University, Australia

^bDepartment of Psychology, Durham University, UK

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ABSTRACT

The neurodevelopmental disorder Williams syndrome (WS) has been associated with a social phenotype of hypersociability, non-social anxiety and an unusual attraction to faces. The current study uses eye tracking to explore attention allocation to emotionally expressive faces. Eye gaze and behavioural measures of anxiety and social reciprocity were investigated in adolescents and adults with WS when compared to typically developing individuals of comparable verbal mental age (VMA) and chronological age (CA). Results showed significant associations between high levels of behavioural anxiety and attention allocation away from the eye regions of threatening facial expressions in WS. The results challenge early claims of a unique attraction to the eyes in WS and suggest that individual differences in anxiety may mediate the allocation of attention to faces in WS.

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Williams syndrome (WS) is a rare (1:7500 to 1:20,000) neurodevelopmental disorder resulting from a microdeletion of approximately 25–28 genes on chromosome 7q11.23 (Donnai & Karmiloff-Smith, 2000; Stromme, Bjornstad, & Ramstad, 2002). At the cognitive level, individuals with WS have mild to moderate intellectual difficulties with relative proficiency in language and short term verbal memory (Mervis et al., 2000) alongside more profound weaknesses in visuospatial and visuomotor abilities (Bellugi, Lichtenberger, Jones, Lai, & St George, 2000; Hocking, Bradshaw, & Rinehart, 2008). At the behavioural level, WS is associated with a distinctive profile of increased social drive, non-social anxiety, and empathic and gregarious emotional personalities (Plesa-Skwerer, Sullivan, Joffre, & Tager-Flusberg, 2004). Of particular interest is the social phenotype of compulsion to engage and approach both familiar and unfamiliar people referred to as hypersociability or prosocial drive (Frigerio et al., 2006; Jones et al., 2000). Once individuals with WS are engaged in social interactions they have been shown to exhibit further atypicalities such as an unusual attraction to the human face, in particular an intense focus of attention to the eye region (Porter, Shaw, & Marsh, 2010; Riby & Hancock, 2009a, 2009b). Despite a characteristic social profile for WS that has been emphasised in the extant literature, there is considerable variability in genetic, cognitive and social functioning that suggests a more nuanced approach is warranted.

Although individuals with WS might show prolonged attention to faces within their environment, they are far from experts at processing and interpreting information from them. There may be a breakdown between attention allocation to faces, perception of appropriate facial cues, and the more sophisticated and cognitive interpretation of faces. Atypical strategies have been unveiled during face processing in WS, with visual focus being directed to specific areas within the face (e.g. eyes and the mouth), and the use of a piecemeal structural encoding strategy rather than a typical configural encoding strategy (e.g. Isaac & Lincoln, 2011). Studies that have employed eye tracking techniques have provided evidence that

* Corresponding author at: School of Psychology and Psychiatry, Faculty of Medicine, Monash University, Building 17, Clayton, VIC 3800, Australia. Tel.: +61 03 99050230.

E-mail address: hannah.kirk@monash.edu (H.E. Kirk).

individuals with WS demonstrate a reduced ability to disengage attention from the eye region of a face when compared to mental-age (MA) matched controls (Porter et al., 2010; Riby & Hancock, 2009a, 2009b). In addition, emotional expressions of faces have been shown to influence attention allocation, with positive (happy) emotions yielding greater visual attention than negative emotions (fearful, angry) in faces (Dodd & Porter, 2010; Plesa-Skwerer, Faja, Schofield, Verbalis, & Tager-Flusberg, 2006; Santos, Silva, Rosset, & Deruelle, 2010). The findings of atypical attention to positive emotional faces, specifically the eye region of faces in WS, are consistent with previous evidence of the role of the amygdala in directing visual attention (Gamer & Buechel, 2009; Gamer, Zurowski, & Buechel, 2010). Functional imaging (fMRI) studies have revealed that WS individuals exhibit anomalous amygdala activation—that is, both reduced amygdala activation for threatening fearful faces (Meyer-Lindenberg et al., 2005), and heightened amygdala reactivity to happy facial expressions (Haas et al., 2009). It is possible that amygdala abnormalities may offer an explanation for the indiscriminate social approach behaviours associated with WS. However, atypical amygdala responses to emotional faces have also been shown in the neurodevelopmental disorder autism, which is characterised by social withdrawal and reduced visual attention to the eye region of faces, coupled with heightened amygdala responses to direct gaze (Riby & Hancock, 2009a, 2009b). These disorders clearly have opposing socio-cognitive profiles associated with anomalous amygdala reactivity; however the nature of the relationship between visual attention to faces and social behavioural characteristics in WS is yet to be fully characterised.

Despite the opposing profiles of atypical social functioning between WS and autism, existing studies have revealed considerable heterogeneity in social functioning, in particular, elevated rates of reciprocal social difficulties and socio-communicative abnormalities in a large proportion of young children with WS suggestive of some overlap with autism spectrum disorders (Klein-Tasman, Li-Barber, & Magargee, 2011; Klein-Tasman, Mervis, Lord, & Phillips, 2007). As such anxiety has been implicated in atypical social interaction in both groups (e.g. Klein-Tasman et al., 2011), and higher levels of social anxiety have been associated with greater amygdala activation to emotional faces in ASD (Kleinhans et al., 2010). Thus it is possible that eye-gaze allocation to emotional facial expressions may be related to variation in levels of anxiety and social impairments in WS. In recent years, evidence has suggested that alongside increased sociability exists a strikingly opposing profile of increased non-social anxiety in WS (Dodd, Schniering, & Porter, 2009; Leyfer, Woodruff-Borden, Klein-Tasman, Fricke, & Mervis, 2006). In particular, high levels of generalised anxiety disorder (GAD) as well as disorder specific fears and phobias have been identified in individuals with WS (Blomberg, Rosander, & Andersson, 2006; Rodgers, Riby, Janes, Conolly, & McConachie, 2012). Existing studies have suggested that individuals with WS may use their hypersociable behaviours to mask any underlying anxiety in social interactions (Dykens, 2003); however, the previously reported heterogeneity in the socio-cognitive profile has often been overlooked, (but see Little et al., 2013). Indeed a more recent study showed that atypical attention to face regions was associated with variation in social functioning during mental state recognition (Hanley et al., 2013). Although research has sought to probe the nature of visual attention allocation to emotional facial expressions in WS, the interactive influences of behavioural characteristics such as anxiety and social reciprocity are yet to be fully understood.

1. Current study

The purpose of the present study was to examine attentional allocation to regions of emotional facial expressions, and the interactive influences of anxiety and social reciprocal behaviours in WS. Thus the aims of the current study were threefold: firstly, we aimed to examine the interplay between visual attention and emotional facial expressions in adolescents and adults with WS. Consistent with previous studies showing greater attention for happy faces in WS (Dodd & Porter, 2010; Porter et al., 2010); we hypothesised that individuals with WS would show greater visual attention to happy facial expressions relative to both mental age (MA) and chronological age (CA) matched controls. Secondly, we aimed to investigate visual attention in relation to specific regions within emotionally expressive faces (e.g. mouth and eye regions). Based on previous studies showing atypical attention to eye regions of faces (e.g. Porter et al., 2010; Riby & Hancock, 2008), we hypothesised that individuals with WS would allocate more attention to the eye region of emotional faces when compared to both MA and CA matched controls. Thirdly, we aimed to explore the relationship between behavioural aspects of anxiety and reciprocal social behaviour and attentional allocation to emotional expressions and face regions in WS. On the basis of previous studies showing that anxiety mediates amygdala response to direct eye gaze to threatening faces in ASD (Kliemann, Dziobek, Hatri, Baudewig, & Heekeren, 2012), we expected that individuals with WS who display higher levels of anxiety and social reciprocity difficulties would show reduced attention to the eye region of threatening facial expressions.

2. Method

2.1. Participants

Thirteen individuals with a diagnosis of WS were recruited via the Williams Syndrome Family Support Group (Victoria), as well as an existing research database. All participants with WS has previously been diagnosed phenotypically and had also previously had their diagnosis confirmed with positive fluorescent in situ hybridisation testing detecting the absence of one copy of the elastin gene. Participants with WS were individually matched to two typically developing groups based on gender and on either (i) chronological age (CA) or (ii) verbal ability (VMA; see Table 1). The typically developing CA and VMA groups were recruited through social networks and via online and print advertisements. Verbal ability was assessed using

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