Research in Developmental Disabilities 34 (2013) 4466–4476

Contents lists available at ScienceDirect

Research in Developmental Disabilities

Looking and thinking: How individuals with Williams syndrome make judgements about mental states

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ARTICLE INFO

Article history:
Received 7 August 2013
Received in revised form 13 September 2013
Accepted 13 September 2013
Available online 18 October 2013

Keywords:
Williams syndrome
Mental states
Emotion
Eye gaze
Eye tracking

ABSTRACT

Individuals with the neuro-developmental disorder Williams syndrome (WS) are characterised by a combination of features which makes this group vulnerable socially, including mild-moderate cognitive difficulties, pro-social drive, and indiscriminate trust. The purpose of this study was to explore a key socio-communicative skill in individuals with WS, namely, mental state recognition abilities. We explored this skill in a detailed way by looking at how well individuals with WS recognise complex everyday mental states, and how they allocate their attention while making these judgements. Participants with WS were matched to two typically developing groups for comparison purposes, a verbal ability matched group and a chronological age matched group. While eye movements were recorded, participants were shown displays of eight different mental states in static and dynamic form, and they performed a forced-choice judgement on the mental state. Mental states were easier to recognise in dynamic form rather than static form. Mental state recognition ability for individuals with WS was poorer than expected by their chronological age, and at the level expected by their verbal ability. However, the pattern of mental state recognition for participants with WS varied according to mental state, and we found some interesting links between ease/difficulty recognising some mental states (worried/do not trust) and the classic behavioural profile associated with WS (high anxiety/indiscriminate trust). Furthermore, eye tracking data revealed that participants with WS allocated their attention atypically, with less time spent attending the information from the face regions. This challenges the widely held understanding of WS being associated with prolonged face and eye gaze, and indicates that there is more heterogeneity within this disorder in terms of socio-perception than previous reports would suggest.

1. Introduction

Individuals with Williams syndrome (WS) are known for their friendly, sociable and outgoing nature (Jones et al., 2000; Mervis & Klein-Tasman, 2000); but this characterisation hides an array of atypicalities of social perception, social cognition and struggles with everyday functioning. WS is a genetic disorder caused by a hemizygous deletion of around 1.6 megabase, containing 26–28 genes on chromosomal 7q11.23 (Eisenberg, Jabbi, & Berman, 2010). The disorder is characterised by a

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http://dx.doi.org/10.1016/j.ridd.2013.09.026

unique pattern of relative strengths and difficulties of cognition; for instance, more severe difficulties with visuo-spatial processing compared to a relative proficiency of verbal skill, all against a backdrop of mild-moderate intellectual impairment and cognitive heterogeneity (Porter & Coltheart, 2005; Searcy et al., 2004). Although the cognitive phenotype has attracted attention of cognitive scientists since the 1990s, much of the recent focus on WS have been on the social characteristics of individuals with the disorder (e.g. for a review see Jarvinen-Pasley, Korenberg, & Bellugi, 2013).

It has been proposed that many individuals with WS are highly motivated towards social engagement and that they exhibit a 'pro-social' drive (Frigerio et al., 2006) that may be difficult to inhibit (e.g. Little et al., 2013) and which increases social approach behaviours to both familiar and unfamiliar people (e.g. Jones et al., 2000). It has also been proposed that once individuals with WS engage in an interaction with others they show an array of subtle social engagement atypicalities, such as prolonged attention to a person's face at the expense of attending other information in their environment. This prolonged attention to faces has been observed in both toddlers with the disorder (Mervis et al., 2003) and has been shown experimentally with older individuals who have WS using eye tracking (Riby & Hancock, 2008). This prolonged attention to a person's face does not, however, lead to an increase or heightened proficiency at interpreting information from that face, such as cues as to how the person is feeling (Plesa-Skwerer, Faja, Schofield, Verbalis, & Tager-Flusberg, 2006) or where they might be attending (Riby, Hancock, Jones & Hanley, 2013). This is particularly important, as increased drive for social interaction (Jones et al., 2000), a lack of understanding of the dangers associated with interactions with unfamiliar people (e.g. Riby et al., 2013) and reduced ability to interpret sophisticated social cognitive cues once in that interaction, in parallel with a general reduction in intellectual capacity can leave individuals with WS highly vulnerable (see Jawaid, Riby, Owens, White, Tarar & Schulz, 2012 for discussion). We aim to provide a detailed investigation of how well individuals with WS make judgements about mental states (complex emotions) from unfamiliar faces because this type of judgement can be important during an interaction and the ability to make this type of judgement requires socio-cognitive capacity. As an important extension to current knowledge, we aim to explore the information that individuals with WS use when making this type of socio-cognitive judgement by using eye tracking techniques (thus also exploring social perception and linking these skills).

1.1. Looking at faces

Looking at faces is critical for social development as they provide a wealth of social and communicative cues that are central to social interaction (Haxby, Hoffman, & Gobbini, 2000). The role that looking at faces plays in the WS social profile has attracted attention partly because the pattern of atypicality appears to be syndrome-specific. Due to the link between looking at faces and social behaviour in WS, comparisons are often made between WS and autism (Riby & Hancock, 2008, 2009a, 2009b; Tager-Flusberg, Plesa Skwerer, & Joseph, 2006). Individuals functioning on the autism spectrum display a very different, but equally atypical, social profile to those with WS, both in the way people with autism look at faces (Hanley, McPhillips, Mulhern & Riby, in press), and in the nature of their everyday social behaviours, often characterised by social withdrawal. It has been proposed that both over- and under-attending to faces, as illustrated by these two disorders of development, has a negative consequence throughout development for learning to interpret social information and developing social expertise (Klin, Jones, Schultz & Volkmar, 2003; Riby et al., 2013).

The first observational evidence of atypical attention to the faces of others came from Mervis and colleagues (2003) who suggested that toddlers with WS stared for longer than toddlers with other developmental disorders at the faces of their paediatrician. Experimental evidence from older children and adolescents was provided by Riby and Hancock in a series of studies that showed atypically prolonged attention to faces of people within social scenes (Riby & Hancock, 2008) and to actors within movies (Riby & Hancock, 2009a). On average, individuals with WS spent longer than typical looking at the eye region within the faces of actors in the stimuli (Riby & Hancock, 2009a) and this finding has since been replicated using faces illustrating basic expressions of emotion (Porter, Shaw, & March 2010). Therefore it has been proposed that the individuals with WS (i) show atypically prolonged attention to faces and (ii) within faces individuals with WS show atypically prolonged attention to the eye region. However, these previous eye tracking studies have used matched-group designs and reported the mean pattern for the WS group compared to their matches and have not explored within-group variability. This is important as we know that there is within-syndrome heterogeneity of both cognition (Porter & Coltheart, 2005) and social behaviour (Little et al., 2013). The current study will explore the within-syndrome variability of attention allocation to faces and face regions.

1.2. Processing emotions in WS: basic and complex expressions

Many individuals with WS seem highly sensitive to the emotions of others but perform relatively poorly on measures of emotion recognition in experimental settings. Several studies have provided evidence to show that individuals with WS perform at a level expected for their mental age (but not as accurately as expected by chronological age) when recognising basic expressions of emotion (see various studies, Gagliardi, Figerio, Burt, Cazzaniga, Perrett, & Borgatti, 2003; Karmiloff-Smih, Klima, Bellugi, Grant & Baron-Cohen, 1995; Plesa-Skwerer et al., 2006; Porter et al., 2010). For example, Plesa-Skwerer et al. (2006) compared the emotion recognition abilities of 47 individuals with WS to individuals with a learning disability (LD) and TD individuals. Participants were shown static images of basic emotions (happy, sad, angry, fearful) and both the WS and LD groups were significantly less accurate for recognition than the TD group. Group differences were particularly