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Can individuals with Williams syndrome interpret mental states from moving faces?

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ABSTRACT

The Williams syndrome (WS) social phenotype is characterised by a high level of social engagement, heightened empathy and prolonged attention to people's faces. These behaviours appear in contradiction to research reporting problems recognising and interpreting basic emotions and more complex mental states from other people. The current task involved dynamic (moving) face stimuli of an actor depicting complex mental states (e.g., worried, disinterested). Cues from the eye and mouth regions were systematically frozen and kept neutrally expressive to help identify the source of mental state information in typical development and WS. Eighteen individuals with WS (aged 8–23 years) and matched groups of typically developing participants were most accurate inferring mental states from whole dynamic faces. In this condition individuals with WS performed at a level predicted by chronological age. When face parts (eyes or mouth) were frozen and neutrally expressive, individuals with WS showed the greatest decrement in performance when the eye region was uninformative. We propose that using moving whole face stimuli individuals with WS can infer mental states and the eye region plays a particularly important role in performance.

1. Can individuals with Williams syndrome interpret mental states from moving faces?

In a social environment it is critical that we have the skills for successful interpersonal communication to allow smooth running interactions with the people around us. One skill that is particularly important is the ability to interpret information from the faces of our companions. The human face provides a wealth of information that is central to social communication (Haxby, Hoffman, & Gobbini, 2000). The face provides cues to identity, the importance of which is emphasised by the social interaction difficulties experienced by individuals with prosopagnosia (e.g., Yardley, McDermott, Pisarski, Duchaine, & Nakayama, 2008). Equally important are communicative signals; such cues provide insights into emotions, thoughts and intentions that must be interpreted to allow us to adapt our behaviour appropriately. Individuals with some neuro-developmental disorders have problems interpreting communicative signals from faces, which may in turn have implications for social interaction styles. One such case is the genetically based neuro-developmental disorder Williams syndrome.

Williams syndrome (WS) has a prevalence of 1 in 20,000 and is caused by a micro-deletion of approximately 25 genes on chromosome 7 (7q11.23; Korenberg et al., 2000). There has been a recent surge of interest in the social phenotype associated with WS and the nature of social interaction styles. The WS social profile is characterised by a strong interest or even propulsion towards social engagement (Frigerio et al., 2006; Jones et al., 2000). In research exploring the personality characteristics of children with WS, Klein-Tasman and Mervis (2003) found that the majority of children with the disorder (96% of their sample) showed behaviours associated with being 'people-oriented' and 'sensitive'. The WS social profile is clearly very different to that associated with functioning on the autistic spectrum, although social behaviour in neither group can be considered 'typical' (Brock, Einav, & Riby, 2008). Researchers have been particularly interested in the role of the face in the social approach style of individuals with WS. Social interest may manifest itself as intense attention towards human faces with prolonged face gaze (Mervis et al., 2003; Riby & Hancock, 2008, 2009), high levels of empathy towards people in distress (Tager-Flusberg & Sullivan, 2000) and an increased tendency to rate unfamiliar people as highly approachable (Jones et al., 2000). It is widely reported that individuals with WS rate unfamiliar faces as approachable that typically developing individuals would deem unapproachable (see Frigerio et al., 2006; Jones et al., 2000) and this type of behaviour has been referred to as atypical social approach. Researchers have made an explicit link between the types of approach behaviour used by individuals with WS and their interest in attending to faces of other people. Recent research has suggested that such behaviours are

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unlikely to be due to 'social stimulus attraction' (Frigerio et al., 2006). There is little evidence that faces capture attention in an abnormal manner (Riby & Hancock, 2009). Rather, such behaviours have been linked with frontal lobe impairment and poor social response inhibition (Porter, Coltheart, & Langdon, 2007). Indeed frontal lobe impairments are likely to contribute not only to the social behaviours that typify the disorder but also extrapolate to the cognitive phenotype of WS (Rhodes, Riby, Park, Fraser, & Campbell, 2010).

Given the highly social behaviours that are associated with WS, alongside claims of empathetic behaviour and an interest in attending to faces, it might be predicated that WS individuals would perform well on tasks involving expression perception. However, in studies of explicit emotion recognition ability, researchers have repeatedly found deficits and performance at a level characteristic of individuals with other forms of intellectual disability (Gagliardi et al., 2003; Plesa Skwerer, Faja, Schofield, Verbalis, & Tager-Flusberg, 2006a; Plesa Skwerer, Verbalis, Schofield, Faja, & Tager-Flusberg, 2006b). When discriminating basic expressions of emotion from schematic faces (happy, sad), children with WS perform at a level comparable to mental age matched typically developing children (Karmiloff-Smith, Klima, Bellugi, Grant, & Baron-Cohen, 1995, Experiment 1), even when completing a task designed for much younger individuals. Slightly more complex tasks, such as sorting faces based on emotional expressions (Tager-Flusberg & Sullivan, 2000, Experiment 3) and recognising expressions of emotion from dynamic faces (Gagliardi et al., 2003) prove even more challenging. Finally Porter et al. (2007) report that individuals with WS perform at a level comparable to mental (rather than chronological) age when recognising emotions (happy, sad, angry, scared) from faces, voices and body postures. All these tasks have focused on perception of the six basic emotions: happiness, sadness, fear, anger, surprise and disgust (Ekman, 1993). Claims of empathetic behaviour appear not to be entwined with strength at interpreting these basic facial expressions.

More complex emotions (those of the 'non-basic' variety) can be considered as ones that involve cognitive parameters alongside the physiological correlates of basic emotions (see Zinck & Newen, 2008 for discussion). Examples include: worried, disinterested, jealous, amongst many others. The ability to ascribe such 'mental states' to others based on facial depictions of complex emotions is considered key to understanding others' minds (see Zinck, 2008). Together the ability to ascribe mental states ('basic mentalising') and the more complex skill of verbally attributing propositional attitudes to another person ('theory of mind') allow sophisticated interpersonal interactions and social understanding (Zinck, 2008). Real life social communication rarely relies solely upon basic expressions of emotion and often involves a range of much more complex mental states. The interpretation of complex emotional or mental states has been found to be problematic for individuals with some neurodevelopmental and clinical disorders (e.g., autism, Baron-Cohen, 1995; schizophrenia, Kington, Jones, Watt, Hopkin, & Williams, 2000). Difficulties interpreting such cues are likely to be central to observed social communication styles. There have been a small number of previously published studies exploring the interpretation of complex mental states by individuals with WS.

In an exploration of socio-perceptual (basic mentalising) skill, Tager-Flusberg, Boshart, and Baron-Cohen (1998) assessed how well adults with WS could label photographs of faces depicting complex mental states. The task was derived from the 'Reading the Mind from the Eyes' assessment (Baron-Cohen, Wheelwright, & Jolliffe, 1997) where only the eye region was available for viewing. The stimuli showed a strip of the face that included the eyes and were presented in black and white. This task has been widely applied to participants with autistic spectrum disorders (e.g., Baron-Cohen et al., 1997; Baron-Cohen, Wheelwright, & Hill, 2001) and has also been used with patients who have amygdala dysfunction (Adolphs, Baron-Cohen, & Tranel, 2002). The task used by Tager-Flusberg et al. (1998) involved WS participants deciding which mental state was depicted in the eye region of the face stimuli (e.g., disinterested, worried). Participants were required to choose between two semantically opposite (for example sympathetic/unsympathetic) target descriptions for each trial (50% chance). Adults with WS ($n = 13$) were compared to another population with reported social deficits; namely Prader–Willi syndrome. Although individuals with WS performed more accurately than those with Prader–Willi syndrome only 6 of the 13 participants scored within a range predicted by age matched typically developing individuals. The authors suggested that their study illustrated a relative 'sparing' of the cognitive capacity to mentalise, however the choice of comparison group, number of mental states and participants numbers make generalisations from this study somewhat difficult. The 'Reading the Mind from the Eyes' task was revised in 2001 (Baron-Cohen et al., 2001) and that version of the task, using four options for each answer and being more sensitive to subtle deficits of mental state perception, was used with a larger sample of individuals who had WS ($n = 43$). Plesa Skwerer et al. (2006) tested adults with WS who were compared to typically developing individuals and those with intellectual difficulties. The same participants completed the mental state task alongside a task involving moving faces for the interpretation of more basic emotional expressions. In Experiment 1 where participants interpreted mental states from the eye region, participants with WS performed at a level comparable to individuals with intellectual difficulties. In Experiment 2 where participants recognised basic expressions (happy, sad, fearful, angry, disgust, surprise) from moving faces shown in 5 s movie clips, individuals with WS again performed at a level comparable to their mental age. The authors concluded that when interpreting both basic expressions of emotion and more complex mental states individuals with WS perform at a level predicted by their mental (rather than chronological) age.

There is a clear difference between the performance levels reported by Plesa Skwerer et al. (2006b, Experiment 1) and Tager-Flusberg et al. (1998) for the interpretation of mental state information. This difference is likely to relate to changes in procedures between studies (e.g., number of mental states assessed, number of answer options, sample size, choice of comparison group). The importance of adapting task design has also been shown by the fact that deficits inferring mental states from the eyes by individuals on the autistic spectrum have failed to be replicated across tasks that vary the procedures and stimuli that are used (e.g., Back, Ropar, & Mitchell, 2007; Ponnet, Roeyers, Buysse, de Clercq, & van der Heyden, 2004). Importantly, changing the task requirements will inherently change the way that information is perceived and processed.

Relating to the previously reported behavioural performance, researchers have also attempted to pinpoint the neuropsychological underpinnings of social behaviour and emotional understanding in WS. Neuropsychological evidence has suggested that amygdala impairment may play a central role in the emotion deficits associated with WS (e.g., Martens, Wilson, Dudgeon, & Reutens, 2009). It is well recognised that the amygdala is critical to the perception of facial expressions (Adolphs, Tranel, Damasio, & Damasio, 1994) and individuals with amygdala damage have difficulty interpreting complex emotions from the eye region on the 'Reading the Mind from the Eyes' task (Adolphs et al., 2002). Individuals with WS also show abnormalities of amygdala structure and function (Haas et al., 2009). The role of the amygdala in the social phenotype associated with WS has attracted a great deal of attention (e.g., Jawaid, Schmolck, & Schulz, 2008; Jawaid et al., in press; Martens et al., 2009). Research has considered how behaviours that are associated with the WS social phenotype relate to the