The other end of the spectrum? Social cognition in Williams syndrome

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Introduction

In typical development, social cognitive abilities are, by definition, predictable based on the child's chronological age. As a consequence, it is difficult to know whether associations between different social cognitive skills and other capabilities reflect underlying causal relationships or mere maturational coincidence. In atypical development, this association between age and ability is disrupted and, in many cases, social cognitive functions appear to develop 'out of synch' with one another or with the child's general developmental level. By looking at developmental disorders and trying to determine the reasons why specific skills may be relatively more impaired in one disorder (or individual) compared with another, it may be possible to tease apart hypothetical causal mechanisms and determine the factors that act as constraints upon development. The preceding chapters in this book have illustrated how the study of social cognitive deficits in autism can illuminate the processes involved in typical social development. In this chapter, we review the social-cognitive capabilities of individuals with Williams syndrome – a rare genetic disorder that is often seen as representing the opposite of autism.

Williams syndrome is caused by the deletion of about 25 genes in the 7q11.23 region of chromosome seven (see Donnai & Karmiloff-Smith, 2000) and is associated with a number of medical and physical characteristics including cardiac anomalies, excessive blood calcium levels, and an unusual 'elfin' facial profile (see Morris, 2006). Although there is considerable individual variation, the overwhelming majority of people with Williams syndrome are characterized as having mild to moderate intellectual disability (see e.g., Howlin, Davies, & Udwin, 1998). Crucially, however, the cognitive profile is somewhat uneven: visuo-spatial and number skills are particularly weak (e.g., Farran & Jarrold, 2003; Paterson, Girelli, Butterworth, & Karmiloff-Smith, 2006), whereas, by comparison, language and face-processing skills are considered to be much less severely affected.

Of particular relevance to this book and chapter, individuals with Williams syndrome are often described as having a characteristic 'hypersociable' personality, behaving 'as if everyone is their friend' (Jones et al., 2000). Parents of individuals with Williams syndrome rate their children as being more empathetic, sensitive, and gregarious than do parents of typically developing children or individuals with other developmental disorders such as Down syndrome, autism, or intellectual delay of mixed aetiology (Doyle, Bellugi, Korenberg, & Graham, 2004; Dykens & Rosner, 1999; Gosch, & Pankau, 1997; Jones et al., 2000; Klein-Tasman & Mervis, 2003). Researchers have also reported increased use of social engagement devices and emotion inferences (Reilly, Harrison, & Klima, 1995; Reilly, Losh, Bellugi, & Wulfeck, 2004; see also Jones et al., 2000) and an increased tendency to react empathetically towards another person's distress (Tager-Flusberg & Sullivan, cited in Tager-Flusberg & Sullivan, 2000). Moreover, in initial experimental cognitive studies, individuals with Williams syndrome were found to perform well on formal tests of theory of mind and emotion recognition that individuals with autism typically fail (Karmiloff-Smith, Klima, Bellugi, Grant, & Baron-Cohen, 1995; Tager-Flusberg, Boshart, & Baron-Cohen, 1998). Such findings led Bellugi and colleagues to conclude that individuals with Williams syndrome "exhibit a striking contrast to the social and language profiles of individuals with other disorders such as autism" (Bellugi, Lichtenberger, Mills, Galaburda, & Korenberg, 1999, p. 200; see also Bellugi, Wang & Jernigan, 1994). In a similar vein, Baron-Cohen and Hammer (1997) argued that, whereas individuals with autism have extreme 'male brains', with better spatial skills than social skills, those with Williams syndrome show the reverse pattern and, as such, may be characterised as having extreme 'female brains'.

This view of Williams syndrome and autism as diametric opposites has, however, proven to be

somewhat simplistic (Tager-Flusberg, Plesa-Skwerer, & Joseph, 2006). Despite their sociable and empathetic personalities, individuals with Williams syndrome are often reported as having high levels of social anxiety (Dykens, 2003; Udwin, Yule, & Martin, 1987). Children with Williams syndrome typically prefer adult company to mixing with their own age group, and have great difficulty making and sustaining friendships (Einfeld, Tonge, & Florio, 1997; Rosner, Hodapp, Fidler, Sagun, & Dykens, 2004; Udwin et al., 1987). The two disorders also overlap clinically. Leyfer, Woodruff-Borden, Klein-Tasman, Fricke, and Mervis (2006) reported that 7% of children with Williams syndrome met DSM criteria for autism spectrum disorders – considerably higher than in the general population. Similarly, Leekam, Burt, & Arnott (2006) noted that, although individuals with Williams syndrome were less impaired than those with autism on the socialization and repetitive behaviour scales of the Diagnostic Interview for Social and Communication Disorders (Wing et al., 2002), ratings of communication skills were in fact comparable across the two groups.

In the past decade or so, experimental research on Williams syndrome has also moved on considerably and this is the main focus of the present chapter. We aim to provide a comprehensive review of recent findings in relation to social cognition in Williams syndrome and, where possible, make direct comparisons between studies of Williams syndrome and studies that have employed similar methodologies to investigate autism. We begin by briefly reviewing the language capabilities of individuals with Williams syndrome, with particular reference to pragmatic skills. We then consider the performance of individuals with Williams syndrome on formal tests of theory of mind and its potential precursors in joint attention, before moving on to look at various aspects of face-processing. Finally, we review recent evidence concerning the neural mechanisms that potentially underlie the hypersociable personalities of people with Williams syndrome.

Language abilities in Williams syndrome

Research on language in Williams syndrome has focused primarily on the structural aspects of language (syntax, phonology, and semantics). It has been widely claimed that language abilities in Williams syndrome are 'intact' (e.g., Bellugi, Marks, Bihrle, & Sabo, 1988; Pinker, 1999) but this description appears to be well wide of the mark. Moreover, while few researchers would disagree that the language of individuals with Williams syndrome is relatively good when contrasted with their own visuo-spatial deficits (e.g., Mervis, Robinson, Rowe, Becerra, & Klein-Tasman, 2003), there is little evidence for a dissociation between language and other non-spatial abilities (Brock, 2007). One exception is that individuals with Williams syndrome do appear to perform relatively well on receptive vocabulary tests, in which they are required to match a spoken word to one of a number of pictures (e.g., Vicari et al., 2004). The reason for this is unclear, but it is notable that, on other measures of vocabulary knowledge, individuals with Williams syndrome tend not to show this advantage (e.g., Clahsen, Ring, & Temple, 2004).

A further issue concerns the extent to which language development in Williams syndrome, though clearly delayed, is subject to the same constraints as in typical development. Studies with young infants have indicated that joint attention (discussed in greater detail below), category concepts and speech segmentation skills are relatively weak when compared with the level of vocabulary knowledge obtained (e.g., Laing et al., 2002; Nazzi & Karmiloff-Smith, 2002; Nazzi et al., 2003). In each case, the relevant skills resemble those of much younger typically developing children; however, given that these skills may all play an important role in vocabulary learning, relative delays in these areas may indicate that alternative mechanisms are involved in language acquisition. Researchers have argued that there may be 'residual' abnormalities in the language of adolescents and adults with Williams syndrome that stem from early deviations from the normal developmental trajectory (see Thomas & Karmiloff-Smith, 2003), but attempts to replicate early findings supporting this view have almost universally failed (see Brock, 2007 for a review).

This focus on structural aspects of language in Williams syndrome has unfortunately meant that, at least until recently, there has been relatively little research on the pragmatic use of language as a social communicative tool. Pragmatic skills are severely impaired in autism, even among high-functioning individuals (see e.g., Tager-Flusberg, Paul, & Lord, 2005), but have often been described as a particular strength in Williams syndrome (Karmiloff-Smith et al., 1995; von Armin & Engel, 1964). Recent studies have, however, challenged such claims. For example, in qualitative analyses of conversational interactions, individuals with Williams syndrome have been noted to display an over-familiar manner with the experimenter (Udwin & Yule, 1990) and show poor turn-taking and topic maintenance (Meyerson & Frank, 1987; but see Stojanovik, 2006). Stojanovik and colleagues (Stojanovik, 2006; Stojanovik, Perkins, &

Howard, 2001) have reported difficulties interpreting the literal or inferential meaning of experimenter's utterance and providing adequate information in responses: conversations involve little exchange of information and speech is often heavily parasitic on the experimenter's contributions.

Of particular note is a questionnaire study by Laws and Bishop (2004), who found that parents of individuals with Williams syndrome reported significant impairments in conversational coherence, appreciation of conversational context, and development of conversational rapport, as well as tendencies towards stereotyped conversations and inappropriate initiation of conversations. In comparison with a group of younger typically developing children, those with Williams syndrome were rated as having greater overall pragmatic difficulties despite equivalent syntactic abilities. Similarly, when compared with individuals with Down syndrome or specific language impairment, they showed significantly better syntax but significantly greater impairments on the stereotyped conversation and inappropriate initiation subscales.

To summarise, despite initial claims of 'preserved' language abilities, formal testing of individuals with Williams syndrome reveal phonological, semantic, and grammatical skills that are broadly in line with overall mental age. In other words, the majority of people with Williams syndrome have significant language difficulties relative to their own typically developing peers. Given the importance of language for the development of social relationships and the difficulties with peer interaction that are often faced by children with more specific language impairments (see e.g., Conti-Ramsden, Simkin, & Botting, 2006), this is important to bear in mind when considering the origin of social difficulties in Williams syndrome. Moreover, many tests of social competence involve verbal comprehension and production skills – researchers who assume that individuals with Williams syndrome do not have language difficulties run the risk of ignoring potentially important confounds in their studies.

Of greater direct relevance to this chapter, there is also growing evidence that pragmatic skills are not intact in Williams syndrome and may even be relatively impaired with respect to structural aspects of language development; however, further research is obviously required in this area. In particular, it is unclear the extent to which the pragmatic difficulties associated with Williams syndrome are in any way comparable to those experienced by individuals with autism, or perhaps a subgroup of individuals on the autistic spectrum. Research addressing this issue would have important practical consequences as well as being of considerable theoretical interest. Pragmatic deficits in autism are often attributed to impaired theory of mind (e.g., Baron-Cohen, 1988), and it is to this issue that we now turn.

Theory of mind

'Theory of mind' refers to the ability to understand and predict behaviour in terms of underlying mental states. Since the seminal work of Baron-Cohen, Leslie, and Frith (1985), researchers have been interested in the idea that individuals with autism lack a theory of mind and that this deficit may explain many of the social difficulties that they experience. As noted in the introduction, Karmiloff-Smith et al. (1995) reported that, in contrast to those with autism, individuals with Williams syndrome performed well on a series of theory of mind tests; however, these findings may be misleading because participants ranged in age from 9 to 23 years – much older than the age at which children typically pass such tasks. Tager-Flusberg and Sullivan therefore conducted a series of studies of theory of mind capabilities in younger individuals with Williams syndrome. Their performance was compared with that of age-matched children with either non-specific intellectual delay or Prader-Willi syndrome – a rare genetic disorder that, like Williams syndrome, is also associated with mild learning difficulties (see e.g., Whittington, Holland, Webb, Butler, Clarke, & Boer, 2004).

First-order false belief tasks require participants to discount their own knowledge of the true state of affairs and deduce the beliefs of another uninformed character. For example, in the classic false location task, participants have to infer that a character who has not seen an object being moved will be under the misapprehension that the object was still in its original location. Such tasks are typically passed by the age of four or five. However, Tager-Flusberg and Sullivan (2000; see also Tager-Flusberg, Sullivan, & Boshart, 1997) reported a pass rate of only 24-29% (depending on the question asked) among four- to nine-year-old children with Williams syndrome. This was significantly lower than the pass rate for children in either of the two comparison groups, despite the fact that the children with Williams syndrome had somewhat higher verbal mental ages.

A similar picture emerges for 'second-order' false belief tasks, which require participants to reason about a character's false belief about another person's false belief (e.g., Molly's father thinks that

she doesn't know what her surprise present is, but in fact she has already seen it). Sullivan and Tager-Flusberg (1999) reported broadly equivalent performance on such tasks among 8- to 17-year-olds with Williams syndrome and those with Prader-Willi syndrome. Pass rates among children with unspecified intellectual delay were significantly lower than in the other two groups, but these individuals also had somewhat lower verbal mental ages. In a subsequent study with similar groups, Sullivan, Winner, and Tager-Flusberg (2003) presented stories in which a child protagonist made a false statement and participants were required to decide whether the child was lying or joking. Children with Williams syndrome showed a trend towards poorer performance on second-order knowledge questions and, in attempting to justify their interpretations of the stories, made significantly fewer references to mental states than children in either of the other two groups (see Reilly et al., 2004 for similar results).

Together, the available evidence suggests that, although many individuals with Williams syndrome eventually acquire sophisticated theory of mind and understanding of false belief, the developmental process is delayed and is certainly no more advanced than general cognitive development. Tager-Flusberg and Sullivan (2000) argued, therefore, that Williams syndrome demonstrates the extent to which theory of mind is constrained by more general cognitive and linguistic delay. In a similar vein, De Villiers (2000) has proposed that, in typical development, a representational understanding of mind is closely connected with comprehension of sentential complements (e.g., 'John said that Mary went shopping'). A number of studies have hinted at particular difficulties with the comprehension and production of such complex grammatical structures in Williams syndrome (Grant, Valian, & Karmiloff-Smith, 2002; Zukowski, 2004), suggesting a potential common cause of theory of mind difficulties.

Tager-Flusberg and Sullivan (2000) further proposed that a distinction should be made between 'the online immediate judgement of mental state' (termed 'social *perception*') and what is traditionally considered to be 'theory of mind' or social reasoning. Their suggestion was that in Williams syndrome (but not autism) social perception is relatively intact, but that theory of mind is constrained by more general reasoning abilities. Thus, despite being highly sociable and empathetic in nature, individuals with Williams syndrome have difficulties with more complex social reasoning and therefore struggle to maintain social relationships.

In support of their account, Tager-Flusberg and Sullivan (2000) cited evidence that individuals with Williams syndrome perform well on tests of emotion perception and this is discussed below. However, it is fair to say that there have been no studies to date that have really addressed 'on-line' social cognition in Williams syndrome. An example of the kind of approach that might be taken is provided by studies looking at children's eye-movements as they complete variations of the false location task. Young typically developing children tend to look towards the location in which the uninformed protagonist thinks the object remains, indicating an 'implicit' awareness of false belief, even when they immediately then give the incorrect verbal response that the protagonist will look in the object's current location (Clements & Perner, 1994). Ruffman, Garnham, and Rideout (2001) suggested that individuals with autism demonstrate the opposite pattern, looking in the wrong location, even if they can subsequently reason their way to the correct verbal response. If Tager-Flusberg and Sullivan (2000) are correct, then we would predict that children with Williams syndrome would show the pattern evidenced by young typically developing children as opposed to that shown by children with autism.

Joint attention

While it remains to be determined whether or not the relatively unimpressive performance of individuals with Williams syndrome on theory of mind tests is simply a consequence of the tasks' cognitive and linguistic demands, researchers have also explored the possibility that social cognitive difficulties in Williams syndrome may be related to earlier deficits in engaging in joint attention during infancy. Joint attention refers to the three-way or 'triadic' coordination of attention between an infant, his or her caregiver, and an object of potential interest. It has been argued that episodes of joint attention are early evidence of the infant's emerging understanding of others as intentional or mental agents (Tomasello, 1995) and that delays in the emergence of joint attention in autism (see Charman, 2003) are the early manifestation of impairment in this representational capacity (Baron-Cohen, 1995; Leslie, 1987).

A number of studies have noted joint attention difficulties in Williams syndrome. Using a standardised experimental procedure, Laing et al. (2002) found that infants with Williams syndrome, aged between 17 and 55 months, were impaired at both initiating and responding to joint attention bids in comparison to mental-age-matched typically

developing children. Specifically, they were poorer at using pointing to engage in a triadic interaction with their partner and a toy object. This finding is consistent with an earlier study by Mervis and Bertrand (1997), who reported that children with Williams syndrome did not respond appropriately to adults' pointing gestures.

One possibility is that, as postulated in the case of autism, deficits in joint attention in Williams syndrome are an early indicator of theory of mind difficulties. However, there appear to be subtle differences in the joint attention deficits associated with the two disorders. Individuals with autism are relatively unimpaired in *instrumental* triadic interactions that function as a request (i.e. to direct the partner's behaviour); but have severe difficulties in declarative triadic interactions, which serve to share awareness, or the experience, of an event or object (Baron-Cohen, 1989; Mundy & Sigman, 1989). By contrast, Laing et al (2002) found that children with Williams syndrome were impaired in their use of both instrumental and declarative gesturing.

An alternative explanation, therefore, is that infants with Williams syndrome fail to engage in triadic joint attention episodes simply because they find objects far less interesting than faces. Indeed, Mervis et al (2003) found that, compared to typically developing infants and those with developmental delay, young infants with Williams syndrome (aged 8-35 months) exhibited extended looking behaviour towards strangers' faces (see also Jones et al., 2000). A similar point was recently made by Triesch, Teuscher, Deak, and Carlson (2006), who proposed that infants learn to follow a care-giver's gaze because it predicts the location of interesting visual stimuli in the environment. According to their model, infants with autism show little gaze-following behaviour because they do not look at the face in the first place (cf. Swettenham et al., 1998); by contrast, infants with Williams syndrome find the face much more rewarding than other objects and so have no motivation to follow the direction of gaze. It is important to point out that, thus far, research on infants with Williams syndrome has focused primarily on the production and comprehension of pointing as opposed to gaze following and that there is no evidence for gaze-following difficulties among older children and adults with Williams syndrome (Gyori, Lukacs, & Pleh, 2004). What remains to be determined is the extent to which the development of such capabilities is delayed and the consequences of this delay for social cognitive development. Longitudinal research (cf. Charman, 2003; Charman et al., 2000) may help address this issue.

Recognition of facial identity

The fascination that individuals with Williams syndrome appear to have with faces could have further important consequences for their social development. The ability to recognise facial expressions of emotion is discussed in depth below; however, until recently, most of the research on face processing in Williams syndrome focussed on processing of facial identity and the claim that individuals with Williams syndrome have 'intact' face-processing abilities. Such claims were based almost exclusively on studies using the Benton Faces task (Benton, VanAllen, Hamsher, & Levin, 1994) – a standardised assessment that involves the simultaneous matching of one or more exemplars of a face across different viewpoints or lighting conditions. Individuals with Williams syndrome typically perform better on this task than might be expected on the basis of their visuo-spatial skills or mental age and performance is often described as being in 'the normal range' (Bellugi, Lichtenberger, Mills, Galaburda & Korenberg,1999; Bellugi et al., 1994; Gagliardi et al., 2003; Plesa-Skwerer, Verbalis, Schofield, Faja, & Tager-Flusberg, 2006). Critically, however, ceiling effects on this task make it difficult to evaluate claims of 'age-appropriate' performance (Farran & Jarrold, 2003) and on other similar measures, the performance of individuals with Williams syndrome is poorer than that of age-matched typically developing controls (Deruelle et al., 1999; Karmiloff-Smith et al., 2004; Mills et al., 2000; Riby, Doherty-Sneddon, & Bruce, 2006a; Tager-Flusberg, Plesa-Skwerer, Faja, & Joseph, 2003) and is typically no better than predicted by mental age (Deruelle et al., 1999; Riby et al., 2006a; although see Udwin & Yule, 1991). Indeed, the Benton Faces test does not appear to be as sensitive to face-processing deficits (prosopagnosia) as other tasks and can be completed successfully by comparing individual features rather than processing the face as a whole (Duchaine & Nakayama, 2004; Duchaine and Weidenfeld, 2003).

Karmiloff-Smith (1997) suggested that individuals with Williams syndrome perform well on this task by adopting such a 'piecemeal' strategy (see also Deruelle et al., 1999). A number of research groups have investigated this claim by comparing recognition of upright and inverted faces. Typically, adults' ability to discriminate between faces is severely disrupted when the faces are presented upside down, but the same manipulation is less disruptive when faces can be identified on the basis of individual features (e.g., Yin, 1969). It has been widely reported that individuals with Williams

syndrome fail to show this inversion effect, indicating a reliance on featural processing (Deruelle et al., 1999; Karmiloff-Smith et al., 2004; Rossen, Jones, Wang, & Klima, 1995), although normal effects of inversion have been reported in several other studies (Mills et al., 2000; Rose et al., 2006; Tager-Flusberg et al., 2003).

In fact, the inversion effect can arise for a number of reasons so its presence or absence is not necessarily diagnostic of face-processing strategy (Maurer, Grand, & Mondloch, 2002). An alternative research strategy, adopted in more recent studies, is to investigate the ability to discriminate between upright faces that have been manipulated in different ways. Tager-Flusberg et al. (2003) reported that, in contrast to those with autism (Joseph & Tanaka, 2003), individuals with Williams syndrome showed a normal advantage for discriminating between features in the context of a whole face as opposed to in isolation, indicating that they are affected by the facial gestalt when processing features.

Karmiloff-Smith and colleagues et al. (2004) have argued, however, that individuals with Williams syndrome *are* relatively insensitive to the configural information in faces, specifically the distances between features. These authors reported difficulties in detecting differences between photographs of faces that had been manipulated by changing the position of the features, although this finding may reflect a response bias rather than differences in accuracy and was not replicated by Riby, Doherty-Sneddon and Bruce (2006b). Karmiloff-Smith et al. (2004; see also Deruelle et al., 1999) also reported that individuals with Williams syndrome had particular difficulties detecting configural changes in schematic faces (i.e., faces made up of geometric shapes), even when performance on the Benton Faces task was controlled for by covariation. This pattern of performance was apparent even among the oldest and most proficient performers in the Williams syndrome group, whereas typically developing children showed a gradual developmental progression towards a more configural approach.

The evidence, although far from conclusive, suggests that individuals with Williams syndrome may have an atypically immature strategy towards face-processing (cf. Carey & Diamond, 1977). Given that, unlike children with autism, those with Williams syndrome appear to be fascinated by faces, this cannot be put down to a lack of experience with faces. So why do they fail to develop configural face-processing strategies? One possibility is that atypical face-processing is related to more general visual-perceptual impairments associated with Williams syndrome.

This view is supported by studies looking at the neural basis of face perception in Williams syndrome. Mills et al. (2000) recorded event related potentials (ERPs) while participants viewed pictures of faces. Early ERP components (N100 and P170) were smaller than normal in adults with Williams syndrome and, unlike in typical adults, these components were not affected by orientation, even though individuals with Williams syndrome showed normal inversion effects in terms of their behavioural accuracy. The authors linked their findings to evidence for subtle structural abnormalities in brain regions involved in visual perception and reported preliminary data showing similar group differences with non-social stimuli (cars).

Complementary results were reported by Mobbs et al. (2004) using functional magnetic resonance imaging (fMRI). Adults with Williams syndrome showed normal levels of activation in two regions, the fusiform gyrus (see also Meyer-Lindenberg et al., 2004) and the superior temporal sulcus, that have been linked to face-processing in typical individuals (see e.g., Haxby, Hoffman and Gobbini 2000). However, these individuals showed reduced activation of primary and secondary visual cortex and, conversely, increased activation within right prefrontal, anterior cingulate cortex, thalamic, striatal areas, hippocampus, and middle temporal gyrus, perhaps reflecting compensatory activity related to the increased difficulty of the task for individuals with Williams syndrome.

If individuals with Williams syndrome do have difficulty processing configural information then, by implication, they must rely heavily on featural information. A further issue, then, is whether they focus on the same features as typically developing children. Adults and children typically show a preference for looking at the eyes and mouths of human faces (Yarbus, 1967). There is some evidence to suggest that individuals with autism focus less on the eyes and perhaps more on the mouth region of faces (e.g., Klin, Jones, Schultz, Volkmar, & Cohen, 2002); however, findings are inconsistent and appear to vary depending on the specific paradigm used. Only two studies have investigated this issue in Williams syndrome and both indicate that individuals with Williams syndrome recognise people from their eyes and mouths in much the same way as typically developing children. Tager-Flusberg et al. (2003) reported that, like typically developing individuals (and in contrast to individuals with autism; Joseph & Tanaka, 2003), those with Williams syndrome were better at detecting changes to the eyes

region of the face than changes to the mouth region. Similar findings were reported by Riby et al. (2006b), who also found that, like typically developing children and unlike children with autism (see also Langdell, 1978), those with Williams syndrome were better at matching faces using upper rather than lower facial features.

Recognition of facial expressions of emotion

As noted above, faces provide important information about the emotions and mental states experienced by other people. Given reports of unusually empathetic responses in Williams syndrome, one might expect that individuals with Williams syndrome would perform relatively well on emotion recognition tasks. Indeed, as discussed earlier, Tager-Flusberg and Sullivan (2000) proposed that social *perception* is a definite strength in Williams syndrome. This hypothesis was based in part on the findings of an earlier study by Tager-Flusberg et al. (1998), who presented participants with black and white photographs of the eye region and asked them to decide which of two labels best described the emotion in the eyes. Even high-functioning adults with autism find this task difficult (Baron-Cohen, Wheelwright, & Jolliffe, 1997). In contrast, although adults with Williams syndrome performed worse than age-matched typically developing controls, they outperformed age-matched controls with Prader-Willi syndrome. Tager-Flusberg et al., (1998) concluded, therefore, that "adults with Williams syndrome are quite good at reading both simple and more complex mental state information from the eye region" (p.635).

A potential concern with this study, however, is that, in the original version of the task, the correct answer was always paired with its semantic opposite. Consequently, individuals with Williams syndrome may have been able to deduce the correct answer on many trials simply on the basis of valence (i.e., by determining whether the eyes were 'happy' or 'unhappy'). Indeed, Plesa-Skwerer et al. (2006) found that, using a modified version of the eyes task that avoided this problem, children with Williams syndrome performed no better than a group of children with non-specific learning disabilities, despite being matched on age, vocabulary knowledge, and IQ, and performing significantly better than these controls on the Benton Faces task.

Other studies using whole face stimuli (rather than just the eye region) have similarly found that emotion 'reading' in Williams syndrome is no better than that of comparison groups matched on mental age. Findings are consistent across a range of paradigms, including; matching a face to another face with the same emotion (Meyer-Lindenberg et al., 2005; Riby et al., 2006a; Tager-Flusberg & Sullivan, 2000); choosing a face that matches a spoken emotion word (Riby et al., 2006); and choosing a word to describe the emotion expressed in a static face (Plesa-Skwerer et al., 2006), in a short video clip (Plesa-Skwerer et al., 2005), or in an image that morphs from a neutral face into an expressive face (Gagliardi et al., 2003). Plesa-Skwerer et al. (2006) further noted that all participants were generally better at recognising happy faces compared with other emotions. This, the authors suggested, was because all other emotions were negative so were less discriminable. Thus, although individuals with Williams syndrome are able to categorize basic emotions by valence, they may have difficulties further differentiating between emotional expressions (cf. Adolphs, 2002).

Given these findings, we might question the idea that social perception is after all a relative strength in Williams syndrome. One possible mitigating factor is that performance on some of these tasks may be mediated by language abilities, thus disguising any Williams syndrome advantage (Tager-Flusberg & Sulliivan, 2000). Alternatively, recognition of facial expressions may be impaired by more general difficulties in processing configural information, as discussed above. Contrary to this view, however, Plesa-Skwerer et al. (2006) recently found that individuals with Williams syndrome were also no better than mental age-matched controls at recognising *vocal* expressions of emotion. Perhaps then individuals with Williams syndrome are just not especially good at differentiating between emotions. They may still be able to react empathetically in an appropriate way because they can pick up on whether someone is happy or sad even if they have greater difficulty interpreting more complex emotions. All we can say at present is that there is very little evidence to suggest that individuals with Williams syndrome have good emotion recognition skills.

Hypersociability and the amygdala

In this final section, we consider studies of face-processing that have attempted to uncover the origins of hypersociability in Williams syndrome. Bellugi, Adolphs, Cassidy, and Chiles (1999) noted that 'social disinhibition' is characteristic, not only of individuals with Williams syndrome, but also of individuals with acquired lesions of the amygdala. These authors explored this similarity by asking individuals with Williams syndrome to rate black-and-white photographs of faces according how much they would like to go up and begin a

conversation with them. Like individuals with bilateral (but not unilateral) amygdala damage (cf. Adolphs, Tramel, & Damasion, 1998), those with Williams syndrome gave unusually high approachability ratings to all the faces. More recently, Frigerio et al. (2006) attempted to replicate this finding using face stimuli that were rated (by typically developing adults) as being of similar approachability to those used by Bellugi and colleagues, as well as faces that were considered much less approachable. Participants with Williams syndrome were more likely than typically developing controls to give extreme negative as well as positive ratings, perhaps, as the authors suggested, reflecting their sociable yet socially anxious personalities. However, given the difficulties that individuals with developmental disorders often have in using rating scales appropriately (cf. Hartley & Maclean, 2006), the findings from these two studies should be treated with caution. Having said that, recent fMRI and psychophysical studies reviewed below do appear to support the idea that amygdala functioning is atypical in Williams syndrome.

Meyer-Lindenberg and colleagues (2005) conducted an fMRI study in which participants were presented with pictures of angry or afraid faces. Relative to typically developing controls, individuals with Williams syndrome showed reduced amygdala activation; however, they showed a relative increase in amygdala activation to non-social threatening stimuli (e.g., spiders, car crashes). The authors suggested that this latter finding may be related to the high levels of non-social anxiety and phobias associated with the syndrome (see e.g., Dykens, 2003; Leyfer et al., 2006). Meyer-Lindenberg and colleagues also noted that, whereas typically developing controls showed increases in activation of various prefrontal regions (dorso-lateral and medial prefrontal cortex and orbitofrontal cortex) when viewing faces as compared to non-social scenes, this effect was not present in the Williams syndrome group. These prefrontal regions are highly interconnected with the amygdala and have been implicated in regulation of amygdala function (Adolphs, 2003). Path analysis of the fMRI data indicated atypical interactions between frontal regions and the amygdala in Williams syndrome, suggesting that the unusual social profile associated with Williams syndrome may be a reflection of a reduced modulating effect of prefrontal regions on amygdala function.

In typical individuals, amygdala activation is increased by direct eye contact. Unlike many children with autism, those with Williams syndrome do not appear to find eye-contact aversive. Indeed, as noted above in relation to joint attention, infants with Williams syndrome appear to be transfixed by faces, and several authors have commented upon the unusual intensity of eye contact they exhibit (Jones et al., 2000; Mervis et al., 2003). Riby (2006) reported a relatively low level of autonomic arousal in children with Williams syndrome, as measured by galvanic skin responses. This suggests the possibility that, when individuals with Williams syndrome view faces, the resultant increase in arousal does not become uncomfortable and they are able to maintain fixation for longer than normal. These findings are only preliminary but provide an interesting contrast with studies of autism (e.g., Hutt, Hutt, Lee, & Ousted, 1964) and suggest a possible link between amygdala functioning and eye-contact in Williams syndrome. It should be noted that Mobbs et al. (2004) reported normal activation of the amygdala when viewing faces with either direct or indirect gaze. Unfortunately, however, the design of the study was such that it was impossible to compare different trial types. Clearly, further research is needed in this area.

Conclusions

Williams syndrome has often been contrasted with autism and there are certainly some interesting points of difference between the two disorders, as discussed below. However, it is clear that a straightforward dissociation between impaired social cognition in autism and 'intact' functioning in Williams syndrome is untenable. In particular, there is very little evidence that any aspects of language, theory of mind, or face-processing are in any sense 'preserved' in Williams syndrome. Phonological, semantic and grammatical language abilities are broadly in line with overall intelligence; pragmatic language skills are, if anything, poorer than structural language abilities; and the same can also be said of performance on theory of mind tests. Although individuals with Williams syndrome perform well on one specific face-matching measure, in general, their ability to match faces on either identity or emotional expression is no better than mental-age-based predictions.

Although social and linguistic skills in Williams syndrome are perhaps less impressive than one might be led to believe, the theoretical importance of the disorder remains. In particular, Williams syndrome provides a contrast with other disorders in which these abilities are severely and specifically impaired. Brock (2007) highlighted the contrast between language abilities in Williams syndrome and Down syndrome, arguing that there is clearly some factor (perhaps related to phonological

processing) that prevents structural language development in Down syndrome reaching the level that is achieved by individuals with Williams syndrome of comparable nonverbal intelligence. In a similar vein, it is clear that there are some developmental constraints that prevent individuals with autism achieving the levels of social cognitive competence one would predict on the basis of their overall intelligence. For most individuals with Williams syndrome, these additional constraints do not appear to be in operation – social cognitive abilities are only constrained by more general verbal and nonverbal abilities (cf. Tager-Flusberg & Sullivan, 2000).

It is, nevertheless, important to make a distinction here between constraints on the process of social cognitive development and confounding factors that might constrain performance on measures of social cognitive functioning. For example, as noted earlier, language difficulties may impact upon theory of mind development in Williams syndrome but are also likely to make conventional theory of mind tasks difficult. Only by carefully designing studies to control for these difficulties or minimize their impact will this issue be resolved.

Further consideration should also be given to the more subtle patterns of similarities and differences between autism and Williams syndrome. For example, despite superficially similar impairments in joint attention, subsequent social capabilities are clearly less impaired in Williams syndrome than in autism. This suggests that subtle group differences in the pattern of joint attention capabilities and their origins may illuminate the causal pathways leading from joint attention to social cognitive deficits. Similarly, when considering face-processing, in some respects, individuals with Williams syndrome resemble those with autism despite clearly having extensive experience of processing faces. Again, direct comparison of the two disorders may prove instructive. Finally, both autism and Williams syndrome have been linked to abnormalities of the amygdala and its interaction with prefrontal cortex. Comparing the neural basis of social functioning in autism and Williams syndrome should refine theories of both disorders.

In sum, the answer to the question posed in the title is that, no, Williams syndrome is not simply the 'other end of the spectrum' to autism. While there is still much work to be done, it is clear that the differences between the disorders are far more subtle and, potentially, far more interesting.

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