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Review

'Too withdrawn' or 'too friendly': considering social vulnerability in two neuro-developmental disorders

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Abstract

In some neuro-developmental disorders, the combined effect of intellectual disability and atypicalities of social cognition may put individuals at increased vulnerability in their social environment. The neuro-developmental disorders Williams syndrome, characterised by 'hypersociability', and autism spectrum disorders, characterised by 'social withdrawal', are at two extremes of atypical social functioning in humans. In this article, we use Williams syndrome and autism spectrum disorders as exemplars to demonstrate how atypicalities of social cognition may contribute to social vulnerability in these populations. The lives of individuals with both these disorders are marred by an increased risk of social isolation, bullying, unsteady relationships, employment difficulties and abuse. While different behavioural interventions have been tried to improve social functioning in these populations, there has been great variability in their success. Finally, we discuss different issues regarding social independence of these individuals; including employment, safety and decision making.

Keywords autism, social, vulnerability, Williams syndrome

Introduction

Social vulnerability[1] is common in individuals with neuro-developmental disorders. A huge body of literature provides evidence for high rates of

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[1] Social vulnerability refers to the inability of an individual or a selective population group to withstand adverse effects of different stressors to which they are exposed. The term was first conceptualised to highlight how certain population groups, for example, minorities, refugees and elderly are at a greater risk of being affected by a natural disaster. In the context of this article, social vulnerability refers to the disadvantages faced by an individual while he or she endeavours to survive as a productive member of the society. Social isolation, unemployment, bullying, physical or sexual abuse, etc. are different experiences which may be considered under the umbrella term of 'social vulnerability'.

psychopathology (Emerson 2003; Emerson & Hatton 2007), physical and sexual abuse (Balogh *et al.* 2001; Reiter *et al.* 2007), suicidal ideation (Lunsky 2004) and aggressive/disruptive behaviours (Crocker *et al.* 2006; McCarthy *et al.* 2009) in this population. Social vulnerability in these groups is largely attributable to intellectual disability (ID). However, evidence suggests that additional cognitive factors may be implicated (Hove & Havik 2010). For example, rates of social problems and victimisation are higher than typical in individuals with autism who have IQ levels within the normal range (Hofvander *et al.* 2009); hence, by themselves intellectual factors do not explain the difference. Similarly, presence of autistic traits has been associated with increased bullying and social isolation at school in individuals with obsessive–compulsive disorder, a disorder in which intelligence and cognitive capacity are generally within the normal range (Bejerot & Mörtberg 2009).

An important factor which may contribute to social vulnerability in individuals with some neuro-developmental disorders and the focus of this article is how atypicalities of social cognition associated with these disorders may contribute to social vulnerability. Numerous neuro-developmental disorders are characterised by the presence of distinct and atypical social phenotypes (Feinstein & Singh 2007). Williams syndrome (WS), characterised by 'hypersociability' (Jones *et al.* 2000), and autism spectrum disorders (ASDs), characterised by 'social withdrawal' (American Psychiatric Association 1994), are placed at two extremes of atypical social functioning (Brock *et al.* 2008). In Table 1, we list these and other neuro-developmental disorders known to show atypicalities of social cognition. We speculate that atypicalities of social cognition, on top of generalised intellectual difficulties, might make an important contribution to social vulnerability in these populations.

Williams syndrome and ASD serve as important populations to study the effects of atypical socio-cognitive development in humans for a number of reasons. The social phenotypes associated with both these disorders are well characterised. There may be individual variability in the degree of socio-cognitive functioning within both disorders. However, the socio-cognitive atypicalities lie within a single spectrum: hypersociability in WS and social withdrawal in ASD (Table 1). The disorders manifest early in life and hence, the disruption to the typical pathway of socio-cognitive development may have long-lasting implications (Riby *et al.* 2011).

Here, we will use WS and ASD as exemplars to elaborate how atypical social cognition may make these individuals extremely vulnerable in their social environment. We will also consider the interventions to improve social functioning in these populations, and issues related to their independent living, which include employment, safety and decision making.

Do atypicalities of social cognition relate to social vulnerability in neuro-developmental disorders?

The nature of neuro-developmental disorders implies that the atypicalities of social cognition are likely to affect the basic development of social expertise in these populations, continuing through childhood, adolescence and into adulthood. Any disruption to the typical pathway of socio-cognitive development may result in atypical social behaviours and interaction styles. These individuals may find themselves in a situation of increased vulnerability because of these atypicalities. This appears to be the case for individuals with the two key disorders discussed in this manuscript, namely WS (Fig. 1) and ASD (Fig. 2).

Williams syndrome

The relatively rare neuro-developmental disorder of WS is characterised by mild–moderate intellectual deficits (Searcy *et al.* 2004), connective tissue abnormalities, cardiovascular anomalies, facial dysmorphology (Pober 2010) and a hypersociable phenotype (Jones *et al.* 2000). The disorder has an estimated prevalence between 1:7500 and 1:20 000 (Pober 2010) and is caused by the microdeletion of approximately 25 genes on the long arm of chromosome 7 (7q11.23; Donnai & Karmiloff-Smith 2000). One copy of elastin gene is deleted in almost 95% of individuals diagnosed with the disorder, confirmed by fluorescent *in situ* hybridisation (FISH; Korenberg *et al.* 2000). Genetic confirmation of this deletion has received widespread acceptance as a sophisticated diagnostic test to confirm phenotypic suspicion of WS (Martens *et al.* 2008).

Table 1 Neuro-developmental disorders with atypical social phenotypes

Neuro-developmental disorder	Social phenotype	Salient social atypicalities	Postulated neural underpinnings
Fragile X syndrome	Social withdrawal	Social anxiety, active avoidance of eye contact, increased time to initiate social interaction, inattention	Amygdala, insular cortex
Down syndrome	Strong social skills; a minority may show autistic features	Increased emotional responsiveness to music, increased smiling and engagement behaviours in infancy, selective deficits for 'sadness' on emotional attribution tasks	Amygdala, frontal lobes
Prader–Willli syndrome	Social withdrawal accompanied by compulsive hyperphagia	Social withdrawal, preservative speech, stereotyped behaviours, temper tantrums, deficits in social attribution	Frontal lobes, amygdala
Smith–Magenis syndrome	Hypersociability accompanied by frequent outbursts of aggression and self-injurious behaviours	Preference for adult contact, lack of adult attention leads to aggressive and self-injurious behaviours	Insular cortex, lenticular nucleus
Turner syndrome	Social withdrawal (more prominent in ring X Turner syndrome)	Fear recognition deficits, eye-tracking deficits, social anxiety	Amygdala, frontal lobes
22q11.2 deletion (velo-cardio-facial) syndrome	Social withdrawal; psychosis in adolescents	Attention deficits, social anxiety	Fusiform gyrus, frontostriatal connections, cerebellocortical connections
Williams syndrome	Hypersociability	Over-friendliness, prolonged gaze behaviours, atypical and indiscriminate approachability towards both familiar and unfamiliar faces	Amygdala, frontal lobes
Autism spectrum disorders	Social withdrawal	Lack of communication and social interactions, lack of eye contact during conversations, repetitive, stereotyped and restricted activities	Amygdala, frontal lobes, cerebellum, fusiform face area

Williams syndrome is characterised by a social phenotype that includes hypersociability – an exaggerated interest to engage in social encounters with both familiar and unfamiliar people (Jones *et al.* 2000; Frigerio *et al.* 2006). The majority of individuals with WS are described as 'people-oriented', 'affectionate', 'sensitive', 'empathetic' and 'friendly' (Tomc *et al.* 1990; Tager-Flusberg & Sullivan 2000; Klein-Tasman & Mervis 2003).

Neural substrates of hypersociability in Williams syndrome

Two neuro-anatomical foci have been proposed as central to the hypersociable WS phenotype: (i) the amygdale; and (ii) the frontal lobes.

A role for the amygdale. It is well recognised that the amygdala, a structure in the medial temporal lobe, is critical to the perception of facial expressions (Adolphs *et al.* 1994) and individuals with amygdala damage have difficulty interpreting social emotions (Adolphs *et al.* 2002). It has been suggested that impairments in amygdala structure and functioning may have an important contribution to hypersociability characteristics of WS (Jawaid *et al.* 2008).

Individuals with WS show abnormalities of amygdala structure and functioning, as detected by tensor-based morphometry, and functional magnetic resonance imaging (fMRI). In a study by Haas *et al.*, individuals with WS showed abnormal amygdala response to fearful expressions; this was paired with an increased tendency to approach

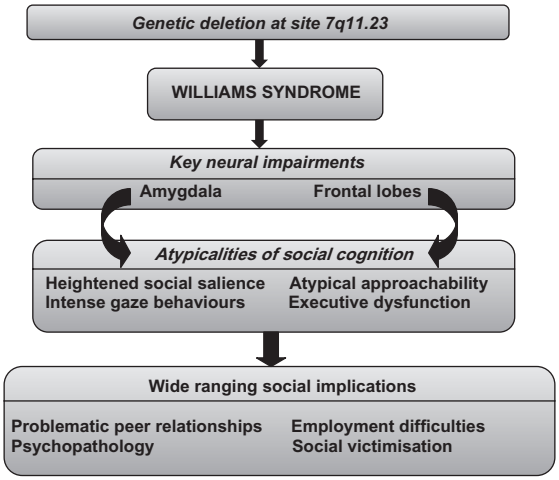

Figure 1 A descriptive account of the social domain of Williams syndrome.

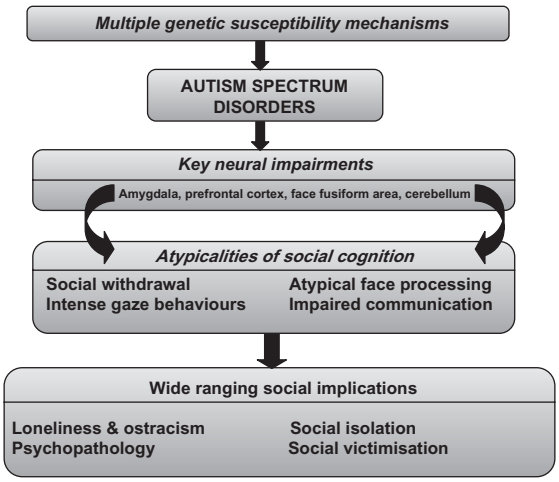

Figure 2 A descriptive account of the social domain of autism spectrum disorder.

strangers (Haas *et al.* 2010). Another study demonstrated an abnormal amygdala–prefrontal connectivity in WS individuals using fMRI techniques. When adults with WS attended to angry and fearful facial expressions, they elicited reduced amygdala activation compared to typically developing individuals (Meyer-Lindenberg *et al.* 2005).

Involvement of the amygdala can also be considered by comparison to other populations that are known to show atypicalities of amygdala functioning. The social phenotype of individuals with WS is closely related to those with bilateral amygdale damage. Individuals with WS, likewise those with bilateral amygdala damage, are characterised by hypersociability, an increased approachability towards unfamiliar people, and impaired recognition of facial emotions with preserved recognition of facial identity (Bellugi *et al.* 1999).

Involvement of the frontal lobes. It has been suggested that the hypersociable characteristics of individuals with WS result from an inability to suppress strong impulses towards social interaction. A fundamental aspect of this hypothesis is that individuals with WS may have intact emotional understanding; however, they exhibit hypersociability because they are unable to control their prosocial drive due to frontal lobe dysfunction (Porter *et al.* 2007).

Individuals with WS show abnormally increased grey matter volumes in frontal lobes on voxel-based morphometry. This may also be associated with problems of inattention and ratings of peer problems, which suggest important contributions of frontal lobe dysfunction to behavioural characteristics of the disorder (Campbell *et al.* 2009).

Atypicalities in frontal lobe functions have also been reported in WS. Porter *et al.* (2007) showed that WS individuals display impairment in response inhibition, a function of the frontal lobe (Mobbs *et al.* 2007; Porter *et al.* 2007). Furthermore, frontal lobe impairment, measured through the ability to control attention, has been reported in other domains of functioning in WS besides social behaviours. For example, Lincoln *et al.* (2002) found that adolescents and adults ($n = 3$) had problems shifting attention when they were required to alternate their response between an auditory target (a target tone) and a visual target (a coloured square). Similarly, Cornish *et al.* (2007) noted that infants and toddlers with WS could be dissociated from infants with other disorders of development (specifically Fragile X syndrome) due to problems disengaging attention from one location and shifting to another location within a visual search paradigm.

Atypicalities of social cognition in Williams syndrome

In experimental and observational settings, individuals with WS demonstrate a preference for faces when looking, prolonged gaze during social engage-

ment (Mervis *et al.* 2003; Riby & Hancock 2008, 2009), atypical approachability towards people without discrimination (Bellugi *et al.* 1999; Jones *et al.* 2000; Frigerio *et al.* 2006; Martens *et al.* 2009) and atypicalities in comprehension of emotional prosody (Pinheiro *et al.* 2011).

Research with WS toddlers revealed that during encounters with their geneticist nearly all (23 out of 25) showed atypically prolonged and intense gaze towards the geneticist's face (Mervis *et al.* 2003). This behaviour contrasted that of typically developing infants who were matched for age. An interest in looking at faces and holding prolonged gaze is also evident in older ages. Adolescents and adults with the disorder tend to fixate on faces in social scenes and movies for significantly longer than typically developing individuals (Riby & Hancock 2008, 2009).

Recent research has suggested that individuals with WS may have difficulties disengaging attention once it is captured by a face (Riby & Hancock 2008; Doherty-Sneddon *et al.* 2009). This may occur to a more exaggerated extent than seen for the disengagement of attention from other non-face objects (e.g. images of houses, butterflies; Riby *et al.* 2010).

Individuals with WS also show atypicalities in the way they process facial features and affect. They show a greater accuracy in matching the faces using upper facial features, and greater detection of eye than mouth modifications (Riby & Hancock 2009). However, their ability to perceive the affect from facial expressions is significantly lower than typically developing counterparts (Järvinen-Pasley *et al.* 2010b). It is plausible that the superior ability in facial configuration is at the expense of impaired affect processing, and the atypical attention mechanisms may be implicated here.

The WS social phenotype is also characterised by an increased rating of approachability to unfamiliar faces in tasks-employing rating scales. There have recently been a number of studies exploring approachability ratings by individuals with WS. While one study reports no atypicality of approach ratings when taking into consideration the emotional understanding of individuals with WS (Porter *et al.* 2007), others indicate abnormally increased approachability towards faces depicting negative expressions (Bellugi *et al.* 1999; Jones *et al.* 2000; Martens *et al.* 2009), or abnormally high ratings given to faces depicting positive expressions (Frigerio *et al.* 2006). There is little doubt that individuals with WS are likely to show some level of atypicality in the way that they consider other people as approachable (especially those who are unfamiliar to them). However, the exact nature of this atypicality entails further exploration.

The atypicalities in emotional processing are not limited to visual processing only in WS. It has been revealed that individuals with WS also show atypical processing of negative vocalisations (Järvinen-Pasley *et al.* 2010a). They also exhibit poor processing of emotional prosody, which may limit their ability to comprehend sarcasm, faux pas or other emotional connotations in the speech. It has been further suggested that the processing of speech prosody is atypical irrespective of the semantic content (Pinheiro *et al.* 2011).

Now, let us combine all these empirical findings, and incorporate them in an everyday life scenario; two strangers standing at a train station at dusk. The WS individual will readily look at the stranger's face, keep a prolonged gaze without being able to accurately judge the expressions of the other person, and will very likely approach the stranger owing to his or her heightened social salience. This atypical behaviour may put the individual at increased vulnerability in the ensuing scenario.

Social vulnerability in Williams syndrome

Although social vulnerability in WS is an understudied topic, the limited literature provides strong evidence for social vulnerability in the disorder, which may be a combined effect of mild–moderate ID and atypical social behaviours and interaction styles.

Individuals with WS experience overly problematic peer interactions and unstable relationships, despite their friendly demeanour. Almost 73% of adults with the disorder experience social isolation (Davies *et al.* 1998). Although not empirically demonstrated, it is speculated that the social isolation could be a result of atypical social behaviours of individuals with WS. For instance, companions may not be accommodating of intense gazing of individuals with WS and their indiscriminate approachability behaviours (Riby *et al.* 2011).

The characteristic over-friendliness and enhanced social salience have also been observed to interfere with occupational assignments for adults with the disorder. Davies *et al.* (1997) studied independence and adaptive behaviours in a moderately large ($n = 70$) cohort of individuals with WS, with a mean IQ of 62 (mild ID). They reported that only 30% were employed, with a vast majority on part-time or voluntary jobs. The range of employments was: kitchen assistants, office assistants, shop assistants, packer and nursery helper. The supervisors of the 21 employed individuals reported that almost all of them (86–100%) had problems which threatened their job stability. These included over-friendliness, anxiety, distractibility and inappropriate or excessive chatter (Davies *et al.* 1997).

The rate of sexual abuse (20%) is alarmingly high in individuals with WS (Rosner *et al.* 2004). Although there has not been a direct comparison, these rates appear to be higher than those reported for children and adolescents with generalised idiopathic mild–moderate ID (5–14%; Balogh *et al.* 2001; Pan 2007). This problem may be the combined effect of individuals with WS making inappropriate social evaluations of other people, as well as other people misinterpreting their over-friendly demeanour or intentions. It is noted that some individuals with WS readily engage in intimate behaviours with strangers including kissing, hugging and touching. This may be interpreted as a desire for further physical contact by others (Davies *et al.* 1997).

Mental health problems are common in individuals with WS. A study employing Psychiatric Assessment Scale for Adults with Developmental Disabilities (PAS-ADD) identified mental health problems in 24% individuals, the most common being anxiety (16.5%). It is important to note that the mental health problems were not associated with individuals' IQ or language disability (Stinton *et al.* 2010). This raises the possibility that the mental health problems could be related to socio-cognitive deficits in these individuals; however, this requires confirmation by longitudinal assessments.

Autism spectrum disorders

Autism spectrum disorders encompass a range of psychological conditions characterised by widespread abnormalities of social interactions and communication, as well as severely restricted interests and highly repetitive behaviours. Recent reviews estimate the prevalence 6/1000 for ASD (Newschaffer *et al.* 2007).

Individuals with ASD, by definition, have difficulties in communication and appropriate social engagement. Children with ASD show detectable deficits in both verbal and non-verbal communication skills as early as 2 years of age (Stone *et al.* 1999). Interactions with caregivers are particularly impaired from infancy, having implications for the development of communicative skills (e.g. joint attention, lack of eye gaze, atypicalities in pointing and requesting behaviours; Charman *et al.* 1997).

Neural substrates of social withdrawal in autism spectrum disorders

Neuroimaging studies in ASD have revealed widespread involvement of different brain regions. A voxel-based morphometry study in children with ASD showed grey matter reductions within frontostriatal, parietal, ventral and superior temporal grey matter. White matter was reduced in the cerebellum, left internal capsule and fornices (McAlonan *et al.* 2005). Similarly, in another study the grey matter volumes of the medial frontal gyri, left pre-central gyrus, right post-central gyrus, right fusiform gyrus, caudate nuclei and the left hippocampus were found to be larger in the autism group relative to age-matched controls. Regions exhibiting smaller volumes in the autism group were observed exclusively in the cerebellum (Rojas *et al.* 2006). With such widespread structural abnormalities, it is difficult to pinpoint any particular substrates of social withdrawal in ASD. However, it is noteworthy to mention that amygdala and its circuitry have received important consideration as being central to core symptomatology of autism over the last decade.

Amygdala theory of autism

Brothers *et al.* suggested amygdala as an important structure governing social intelligence in primates in 1990 (Brothers *et al.* 1990). Since then numerous studies have been performed to explore a role for the amygdala underlying the socio-cognitive deficits in autism. Baron-Cohen *et al.* showed that unlike

age-matched controls, individuals with ASD did not activate amygdala while judging mentalistic inferences from the eyes (Baron-Cohen *et al.* 1999). A recent fMRI study employing a trustworthiness scale for faces further showed that individuals with ASD have significantly reduced activation in right amygdala, fusiform face area and left ventrolateral prefrontal cortex while they made evaluation of faces (Pinkham *et al.* 2008). There is also a great overlap between observations from individuals with ASD and bilateral amygdala damage of developmental origin, both groups show failure to fixate eyes on faces, impaired recognition of emotions from faces, impaired theory of mind abilities and difficulties in regulating personal space distance. However, when standard scales are employed, individuals with bilateral amygdala damage do not fit the diagnostic criteria for ASD. It has been suggested that amygdala dysfunction in isolation is not sufficient to produce the core social withdrawal features of ASD. However, dysfunction of the broader circuitry between amygdala and other socially relevant structures (prefrontal cortex, cerebellum, fusiform face area, etc.) may underlie the socio-cognitive deficits of ASD (Paul *et al.* 2010)

Atypicalities of social cognition in autism spectrum disorders

In experimental and observational studies, individuals with ASD have been shown to demonstrate significant deficits in emotion recognition (Kätsyri *et al.* 2008), atypical social processing and response generation (Embregts & van Nieuwenhuijzen 2009), atypical gaze patterns (Wolf *et al.* 2008; Chawarska *et al.* 2010; Krebs *et al.* 2011) and deficits in their own communicative signals (Willemsen-Swinkels *et al.* 1998).

Unlike typically developing children, toddlers with ASD do not demonstrate an attentional preference for faces or other social stimuli (Chawarska *et al.* 2010). When they do look at faces, individuals with ASD demonstrate wide atypicalities in the way they process facial information. Preferential looking at the eyes of approaching adults is considered important for social development. However, ASD children as young as 2-year-olds demonstrate atypicality in this behaviour, and show an increased preference towards mouth (Jones *et al.* 2008). Furthermore, Wolf *et al.* showed that ASD individuals are impaired in their ability to recognise changes in orientation, expression and featureful information from faces. The ability to recognise configurational and featureful expressions was preserved for the mouth regions, but impaired for the eye regions (Wolf *et al.* 2008). It has also been suggested that children with ASD also show atypicalities in relating facial identity features with facial expression features. While typically developing children process facial expressions in interaction with facial identity, children with ASD process facial expressions and identity independent of each other (Krebs *et al.* 2011). Finally, it has been variably suggested that that children and adults with ASD show deficits in emotional recognition from faces (Kätsyri *et al.* 2008).

The atypicalities in emotional processing are not limited to facial processing only. A study by Philip *et al.* ($n = 23$) observed that individuals with autism showed impairment in emotion recognition in multiple domain of processing; facial processing, body movements and vocal stimuli. The ASD group in their study was also impaired in making social judgments as compared to the controlled group, which correlated with deficits in basic emotion recognition (Philip *et al.* 2010).

Individuals with ASD also demonstrate atypicalities in how they process socially relevant information from presented scenarios, as compared to their age- and IQ-matched typically developing counterparts. Their evaluations of socially relevant information, response decisions and responses are atypical to their level of intellectual functioning. There is an inclination towards negative emotions and responses generated are inappropriate (Embregts & van Nieuwenhuijzen 2009).

Recent research has also revealed that individuals with ASD use their own communicative signals atypically during social encounters; for examples, showing atypicalities in the timing of their gaze behaviours during social communication (Willemsen-Swinkels *et al.* 1998) and showing atypically increased gaze aversion when listening to people talking to them (Doherty-Sneddon *et al.* 2009). Furthermore, it has been shown that individuals with ASD show deficits in the formation of empathy similar to individuals with psychopathic tendencies. However, it is not clear whether it suggests a lack of concern likewise psychopathy or

simply a failure to understand the emotional state of the distressed (Jones *et al.* 2010).

Now, let us combine all these empirical findings, and try to incorporate them in an everyday life scenario, two strangers standing at a train station at dusk. The ASD individual will not look at the stranger's face, will likely not keep the eye contact if approached by the stranger, will not be able to understand the social context of the approach (which could be a polite request for help) and will likely do not offer an appropriate response even if being approached in a polite manner. This atypical interaction style combined with the lack of awareness by the other person may result in a scenario in which the ASD individual may have to face spurn or hostility.

Social vulnerability and the autism spectrum

The consequences of social deficits are far-reaching for individuals with ASD. In their social life, individuals with ASD face numerous problems which are largely attributable to their lack of social skills.

The peer relations of individuals with ASD are extremely problematic during childhood. A study ($n = 35$) of high-functioning children with autism showed that a majority reported high levels of loneliness, and only spent half the time interacting with their peers as compared to their age- and IQ-matched counterparts. Furthermore, the children reported a lower association between social interaction and loneliness, which suggests a poor understanding of sociability among the children (Bauminger *et al.* 2003). The isolation and loneliness continues into adolescence and adulthood. A cross-sectional study on high-functioning adults with autism showed that only 19% ever had a long-term relationship (Hofvander *et al.* 2009).

Children with ASD are often subjected to bullying and ostracism at school (Hofvander *et al.* 2009; Twyman *et al.* 2010). An American study by Montes & Halterman (2007) on nationally represented sample of 4- to 17-year-olds with ASD showed that 44% reported being subjected to bullying at school. Individuals with ASD could be vulnerable to bullying and victimisation for numerous reasons. Their lack of social interaction and limited communication skills often lead to social isolation, and hence they become 'easy targets' for exploitation. Furthermore, their lack of interaction may be interpreted as 'arrogance' by other individuals, which may bring out aggressive and condescending behaviours in them. There is also some evidence which suggests that individuals with ASD also lack the understanding of socially deviating behaviours, and may not even realise that they are being mistreated. In a study by van Roekel *et al.* (2010), it was shown that children with ASD not only have a higher prevalence of victimisation but some of them may also misinterpret bullying situations as non-bullying and vice versa (van Roekel *et al.* 2010).

The rates of sexual and physical victimisation are also alarmingly high in individuals with ASD. A cross-sectional study of individuals with autism showed that 16.6% of them had been victims of sexual abuse, whereas the prevalence of physical abuse was 18.5% (Mandell *et al.* 2005). Individuals with ASD do not usually engage in exaggerated social or physical advances, unlike individuals with WS. However, the high rates of abuse could be because of mild–moderate ID, inability to comprehend the intentions and advances of others, and an ability to effectively communicate their disapproval of physical advances. In future studies, it is imperative that demographics and behavioural characteristics of the victims are explored to determine if any particular traits are associated with an increased vulnerability to abuse in ASD.

Individuals with ASD also have increased prevalence of mental health problems, which may be attributable to their inability to tackle social stressors, isolation and victimisation. A study on adolescents and young adults with autism showed that 20% met criteria for major depressive disorder, 30% met criteria for generalised anxiety disorder and 50% had clinically significant suicidal ideation (Shtayermman 2007). Another study revealed a strong association between parent-reported child withdrawal and depression in 7- to 14-year-old children with autism (White & Roberson-Nay 2009).

Inefficient communication skills and disruptive behaviours may also hinder occupational functionality of individuals with ASD. Despite a high level of education (high school or above, related to their level of intellectual ability), more than half were reported to be unemployed or on long-term medical leave in a study (Hofvander *et al.* 2009). Even with intensive supported employment pro-

grammes, the employment rates can only be raised up to 68% in high-functioning adults with ASD. The jobs acquired usually are of technical or computing nature, and are highly dependant on individuals' IQ, educational achievements and language skills (Howlin *et al.* 2005). It has also been shown that as compared to other individuals with mild–moderate ID, usually individuals with autism work lesser hours, earn lower wages per week and are the most costly individuals to serve (Cimera & Cowan 2009).

Approaches to improve social functioning in Williams syndrome and autism spectrum disorders

The first question which obviously comes to mind is that is it possible to modulate the social phenotypes associated with these disorders. In Tables 2 and 3, we list different strategies which have been employed to enhance social skills and improve socially disruptive behaviours in ASD and WS respectively. We included studies which focussed on improving social and adaptive skills in both disorders. However, this list is not exhaustive and a publication bias is a valid possibility here.

Behaviourally based interventions, designed to improve communication skills and reduce interfering and non-functional behaviours, have been implemented and evaluated for at least the last seven decades for children with ASD. A review of literature identifies a large number of different intervention strategies applied to the range of deficits faced by individuals with ASD. Examples include applied behaviour analysis, communication-focussed interventions, developmental approaches, social skills intervention, sensory motor interventions, as well as some integrative approaches (Table 2). There is a great deal of variation in the success of each and every method currently used for intervention, partly due to the high level of individual variability in terms of functioning and degree of socio-cognitive impairment. There is also great heterogeneity in the nature and duration of the interventions, study designs and settings, number of subjects and outcome measures. These considerations make it difficult to directly compare the effectiveness of different behavioural interventions. Finally, an important issue that applies to most of the interventions is the lack of long-term evaluation, to determine if skills are maintained and if any appreciable reduction in social vulnerability ensues as a result of the intervention.

For individuals with WS, there have been fewer intervention attempts to compensate for areas of deficits. In fact, we are unaware of any behavioural intervention which has focussed specifically on moderating the hypersociability traits or atypical approachability behaviours of the individuals. There are, however, smaller case studies with a focus on obsessive–compulsive behaviours, etc., which are common in individuals with WS (Table 3). With clearer understanding of the cognitive and behavioural phenotypes that accompany WS, there is a need to examine the types of interventions that may be most beneficial to individuals with the disorder and their families (Klein-Tasman *et al.* 2009).

Combining the evidence from WS and autism, it is clearly important to integrate scientific/empirical findings with interventional approaches. The success of intervention strategies also appears somewhat limited and more avenues need to be explored.

Is social independence achievable in Williams syndrome and autism spectrum disorders?

The next important question is whether individuals with WS and ASD could live independently in the society. It is a very broad topic, which involves not only medical but also medicolegal and sociological perspectives. Here, we discuss three important issues with respect to independent living in individuals with WS and ASD: employment, victimisation and decision making.

Employment

Employment could be a very effective means of enhancing social independence in individuals with WS and ASD. In turn, this social independence may reduce social isolation difficulties and social vulnerability. In one study by García-Villamisar & Hughes (2007), sustained employment support of adults with autism was associated with significant

Table 2 Interventions to improve social behaviours and adaptive skills in individuals with autism spectrum disorders (ASD)

Intervention	Duration of intervention	Author and year	Results
Intensive behavioural treatment vs. eclectic treatment in children to reduce disruptive behaviours	1 year	Eikeseth *et al.* 2002	Decrease in disruptive behaviours in the behavioural treatment group
Traditional behavioural approaches vs. natural play interventions to enhance social interaction, compliance and communication abilities	10 weeks	Bernard-Opitz *et al.* 2004	Significant gains in play, attention, compliance and communication skills in both groups. Behavioural approaches were superior to play interventions in improving attention and compliance
Computer-based intervention to teach how to recognise and predict emotions	2 weeks	Silver & Oakes 2001	Significant improvement in ability to recognise and predict emotions
Interactive multimedia tool designed to improve recognition of complex emotions from faces and voice	10–15 weeks	Golan & Baron-Cohen 2006	Significant improvement in recognition of complex emotions in both groups
Repeated sessions of adult imitation vs. regular play to improve social behaviours	Three sessions	Field *et al.* 2001	Improvement in distal (e.g. attention) and proximal (e.g. touching) social behaviours with imitation
Adult imitation-based intervention vs. contingently responsive interaction group to improve social behaviours	Single session	Escalona *et al.* 2002	Contingency responsive interaction was more effective in facilitating distal social behaviours. Imitation was more effective in facilitating proximal social behaviours
Dyadic social communication intervention targeted at parent communication vs. routine care in children with autism	1 year	Aldred *et al.* 2004	The intervention group showed significant improvement compared with controls on reciprocal social interaction, communicative initiation and parent–child interaction
Social story intervention to decrease disruptive behaviours in children with autism	Single session	Scattone *et al.* 2002	Decrease in disruptive behaviours in all participants
Social story intervention to improve verbal greeting initiation in an 11-year-old child with high-functioning autism	Single session	Reichow & Sabornie 2009	No improvement in verbal greeting initiation
Auditory training intervention to improve school-appropriate behaviours	10 weeks	Smith *et al.* 1985	Improvement in attentiveness, school-appropriate behaviours and communication
Use of social adjustment enhancement psychoeducational curriculum for boys with ASD aged 8–12 years	20 weeks	Solomon *et al.* 2004	Significant improvements in facial recognition
Use of Lego blocks for increasing motivation and peer interaction	3 years	Legoff & Sherman 2006	Lego therapy produced significant gains on measures of social skills as compared to the control group

improvement in non-vocational outcomes. However, the social atypicalities of these disorders greatly impede job acquisition and sustainability, as we discussed previously. Yokotani (2010) demonstrated that while educational level may predict job acquisition in individuals with autism, it has little association with job sustainability at 1 year. It has been observed that simulation techniques along with

Table 3 Interventions to improve social behaviours and adaptive skills in individuals with Williams syndrome

Intervention	Author and year	Salient results
Behavioural intervention including escape extinction and differential reinforcement of each bite eaten to treat food refusal in a child with WS. The child was not allowed to leave the table for a predetermined period and was praised by the mother for each bite consumed	O'Reilly & Lancioni 2001	An increase in the food consumed and decrease in other inappropriate behaviours
Intensive short-term cognitive-behavioural therapy to treat OCD-like symptoms in a young adult with WS	Klein-Tasman & Albano 2007	Increase in patient's insight about his problems and decrease in OCD-like symptoms

WS, Williams syndrome; OCD, obsessive–compulsive disorder.

on-site training (Burke *et al.* 2010), performance cue systems (Lattimore *et al.* 2006) and occupational therapy (Capo 2001) may be beneficial in improving job performance in individuals with autism. We are not aware of any strategies which have been employed to enhance job performance in individuals with WS.

An additional step in this regard could be employing individuals diagnosed with these conditions in jobs appropriate to their social capabilities. Individuals with WS who show characteristics of hypersociability could be accommodated in people-oriented jobs. However, they should be under constant supervision, because while the individuals with WS have a friendly demeanour, they are prone to misinterpret social cues. Similarly, individuals with ASD should be engaged in jobs that require modest social interaction. Anecdotes suggest that people with high-functioning autism also do well in jobs which have a fixed routine and order (for example, as a library assistant that is in charge of maintaining the library catalogue).

Safety

Because of their atypical interaction styles and socio-cognitive deficits, individuals with WS and ASD become 'easy targets' for victimisation and abuse. Their lack of emotional understanding, as well as the lack of awareness about their characteristics in the community, combines to place them at heightened vulnerability to victimisation.

This may be of great relevance to socially over-demonstrative individuals of WS. The population has a very high rate (20%) of alleged sexual abuse. It may be attributable to their over-demonstrative nature combined with the fact that their family members, peers, co-workers, etc. may not be aware that their heightened social salience does not always imply a desire for further intimacy. It is important to translate the findings from experimental studies into practical approaches, and develop screening paradigms based on individuals' intellectual level as well level of socio-cognitive impairment.

Decision making

It is important to also consider whether individuals with WS and ASD are able to make rational decisions. Both the disorders are characterised by varying levels of ID, as well as deficits in the neural regions deemed to play a role in decision making (Table 1).

While the decision-making abilities of individuals with WS have not been explored, evidence does suggest atypicalities in the decision-making characteristics of individuals with autism. There is limited evidence for impaired motivational processing and altered autonomic responsiveness to 'gain' and 'loss' stimuli in individuals with Asperger's syndrome (Johnson *et al.* 2006). However, on the other hand, individuals with autism have been demonstrated to be more 'rational' in decision making as they fail to integrate emotional contextual cues into the decision-making process. This

results in a lesser susceptibility to be influenced by emotional prospects and consequences of decision making, and a reduced 'framing' effect (De Martino *et al.* 2008).

Future directions

In the future studies, it is essential to bridge the gaps between experimental observations and real-life experiences in WS and ASD. While there is sufficient evidence to suggest that individuals with these disorders are socially vulnerable, it is not clear whether the vulnerability is partially or wholly attributable to atypical interaction styles and socio-cognitive deficits. A recent study by Järvinen-Pasley *et al.* can serve as an example here. Indiscriminate approachability has been a consistent observation in individuals with WS. However, the exact basis for this atypical behaviour is not known. Järvinen-Pasley *et al.* showed an association between high self-reported approachability ratings and perceptual deficits in affect identification in individuals with WS (Järvinen-Pasley *et al.* 2010b). It not only provides clues towards the basis for this atypical behaviour, but also has implications with regards to screening and prevention. Studies which employ multiple modalities, that is imaging, neuropsychological assessment and self/teacher-reported scales on the same population, are likely to reveal neural and psychological correlates of atypical social behaviours in these disorders, which can be used to screen these individuals for rigorous intervention and supervision.

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