‘Too withdrawn’ or ‘too friendly’: Considering Social Vulnerability in two Neuro-developmental Disorders

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Annotation Article

‘Too withdrawn’ or ‘too friendly’: Considering Social Vulnerability in two Neuro-developmental Disorders

Ali Jawaid¹, Deborah M Riby², James Owens³, Susan W. White⁴, Tarfa Tarar⁵, Paul E Schulz⁶

¹ Institute of Neuropathology, University Hospital Zurich, Zurich 8091, Switzerland
² School of Psychology, Newcastle University, Newcastle upon Tyne, NE1 7RU, UK
³ Department of Neurology, Baylor College of Medicine, Houston, TX 77030, USA
⁴ Department of Psychology, Virginia Polytechnic Institute and State University, Blacksburg, VA 24061, USA
⁵ Mailman School of Public Health, Columbia University, New York, NY 10032, USA
⁶ Department of Neurology, University of Texas Health Science Center, Houston, TX, USA

Short Title: Social vulnerability in neuro-developmental disorders

Correspondence

Ali Jawaid, MD
Institute of Neuropathology
Schmelzbergstr. 12
University Hospital Zurich
Zurich 8091, Switzerland
Phone: +41 (78) 617 9971
Email: alijawaid84@gmail.com
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 disorder Williams syndrome, characterized by ‘hypersociability’ and Autism Spectrum Disorders, characterized 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.

Key Words: vulnerability; social; autism; Williams syndrome.
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Introduction

Social vulnerability\(^1\) is common in individuals with neuro-developmental disorders. A huge body of literature provides evidence for high rates of psychopathology (Emerson 2003; Emerson & Hatton 2007), physical and sexual abuse (Balogh et al. 2001; Reiter et al. 2007), suicidal ideation (Lunsky et al. 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. However, evidence suggests that additional cognitive factors may be implicated (Hove & Havik 2010). For example, rates of social problems and victimization 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 & Mortberg, 2009).

\(^1\) 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 conceptualized 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’.
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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 characterized by the presence of distinct and atypical social phenotypes (Feinstein & Singh 2007). Williams syndrome (WS) characterized by ‘hypersociability’ (Jones et al. 2000), and autism spectrum disorders (ASDs) characterized by ‘social withdrawal’ (APA 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 generalized intellectual difficulties, might make an important contribution to social vulnerability in these populations.

WS 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-characterized. 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 in Press).

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 (Figure 1) and ASD (Figure 2).

**Williams syndrome (WS)**

The relatively rare neuro-developmental disorder of WS is characterized 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 1:7,500 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 hybridization (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).
WS is characterized 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’ (Tager-Flusberg & Sullivan 2000; Klein-Tasman & Mervis 2003; Tomc et al. 1990).

**Neural Substrates of Hypersociability in Williams syndrome**

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

**A role for the Amygdala**

It is well recognized 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 (Adolph 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 Hass et al., individuals with WS showed abnormal amygdala response to fearful expressions; this was paired with an increased tendency to approach strangers (Haas et al. 2009). 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 amygdala damage. Individuals with WS likewise those with bilateral amygdala damage, are characterized 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 hyper-sociable 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 pro-social 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 (Porter et al. 2007, Mobbs 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 behaviors. 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 engagement (Mervis et al. 2003; Riby & Hancock 2008; Riby & Hancock 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 behavior 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 et al. 2009). However, their ability to perceive the affect from facial expressions is significantly lower than typically developing counter-parts (Jarvinen-Pasley et al. 2010). 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 characterized 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 Williams syndrome. It has been revealed that individuals with Williams syndrome also show atypical processing of negative vocalizations (Jarvinen-Pasley et al. 2010). 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/her heightened social salience. This atypical behavior 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 intellectual disability and atypical social behaviours and interaction styles.
Individuals with WS experience overly problematic peer interactions and unstable relationships, despite their friendly demeanor. 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 (Ribly et al 2011 *in press*).

The characteristic overfriendliness 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 intellectual disability). 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 Williams syndrome (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 intellectual disabilities (5-14%; Pan 2007; Balogh et al. 2001). 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 demeanor or intentions. It is
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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). The 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 (ASD) encompass a range of psychological conditions characterized 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 (Newshchaffer 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 two 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 ASD

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 fronto-striatal, 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. 2007). With such widespread structural abnormalities, it is difficult to pin-point 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 sociocognitive 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 (Baren-Cohen et al. 1999). A recent fMRI
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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 sociocognitive deficits of ASD (Paul et al. 2010)

Atypicalities of Social Cognition in ASD

In experimental and observational studies, individuals with ASD have been shown to demonstrate significant deficits in emotion recognition (Katsyri et al., 2008), atypical social processing and response generation (Embregts & van Nieuwenhuijzen 2009), atypical gaze patterns (Chawarska et al. 2010, Wolf et al. 2008, Krebs et al. 2010), 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.
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1 Preferential looking at the eyes of approaching adults is considered important for social
development. However, ASD children as young as two-year olds demonstrate atypicality in this
behavior, and show an increased preference towards mouth (Jones et al. 2008). Furthermore,
2 Wolf et al. showed that ASD individuals are impaired in their ability to recognize changes in
orientation, expression, and featureful information from faces. The ability to recognize
3 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
4 atypicalities in relating facial identity features with facial expression features. While typically
5 developing children process facial expressions in interaction with facial identity, children with
6 ASD process facial expressions and identity independent of each other (Krebs et al. 2010).
7 Finally, it has been variably suggested that that children and adults with ASD show deficits in
8 emotional recognition from faces. (Katsyri et al. 2008).
9
10 The atypicalities in emotional processing are not limited to facial processing only. A study by
11 Philip et al. (n=23) observed that individuals with autism showed impairment in emotion
12 recognition in multiple domain of processing; facial processing, body movements, and vocal
13 stimuli. The ASD group in their study was also impaired in making social judgments as
14 compared to the controlled group, which correlated with deficits in basic emotion recognition
15 (Philip et al. 2010)
16
17 Individuals with ASD also demonstrate atypicalities in how they process socially relevant
18 information from presented scenarios, as compared to their age- and IQ- matched typically
19 developing counter-parts. Their evaluations of socially relevant information, response decisions,
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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. 2010). 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.
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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 counter-parts. 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 4-17 year olds with ASD showed that 44% reported being subjected to bullying at school. Individuals with ASD could be vulnerable to bullying and victimization 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 behaviors in them. There is also some evidence which suggests that individuals with ASD also lack the understanding of socially deviating behaviours, and may
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not even realize that they are being mis-treated. In a study by van Roekel et al. (2009), it was shown that children with ASD not only have a higher prevalence of victimization but some of them may also mis-interpret bullying situations as non-bullying and vice versa (van Roekel et al., 2009).

The rates of sexual and physical victimization 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 intellectual disability, 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 victimization. A study on adolescents and young adults with autism showed that 20% met criteria for major depressive disorder, 30% met criteria for generalized anxiety disorder, and 50% had clinically significant suicidal ideation. (Shteyerman O. 2007). Another study revealed a strong association between parent-reported child withdrawal and depression in 7-14 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 programs, 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 intellectual disabilities, usually individuals with autism work lesser hours, earn lower wages per week, and are the most costly individuals to serve (Cimera 2009).

Approaches to Improve Social Functioning in WS and ASD

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 focused on improving social and adaptive skills in both disorders. However, this list is not exhaustive and a publication bias is a valid possibility here.

Behaviorally based interventions, designed to improve communication skills and reduce interfering and non-functional behaviors, 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.
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Is Social Independence Achievable in WS and ASD?

The next important question is whether individuals with WS and ASD could live independently in the society. It is a very broad topic, with involves not only medical but also medico legal and sociological perspectives. Here we discuss three important issues with respect to independent living in individuals with WS and ASD, employment, victimization, 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 Garcia-Villamisar & Hughes (2007), sustained employment support of adults with autism was associated with significant 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 one year. It has been observed that simulation techniques along with 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 mis-interpret 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 sociocognitive deficits, individuals with WS and ASD become ‘easy targets’ for victimization 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 victimization.

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. An important consideration here is that legally it is very difficult to decide whether there was an element of agreement for the sexual act from the victim. 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 sociocognitive impairment.
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1 Decision Making

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

3 While the decision-making abilities of individuals with Williams syndrome 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

4 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 sociocognitive deficits. A recent study by Jarvinen-Pasley et al. can serve as an example here. Indiscriminate approachability has been a consistent observation in individuals with Williams syndrome. However, the exact basis for this
atypical behaviour is not known. Jarvinen-Pasley et al. showed an association between high self-reported approachability ratings and perceptual deficits in affect identification in individuals with WS (Jarvinen-Pasley et al. 2010). 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; i.e., 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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Table 1: Neuro-developmental disorders with atypical social phenotypes.

<table>
<thead>
<tr>
<th>Neuro-developmental disorder</th>
<th>Social Phenotype</th>
<th>Salient Social Atypicalities</th>
<th>Postulated Neural Underpinnings</th>
</tr>
</thead>
<tbody>
<tr>
<td>Fragile X syndrome</td>
<td>Social withdrawal</td>
<td>Social anxiety, active avoidance of eye contact, increased time to initiate social interaction, inattention.</td>
<td>Amygdala, Insular cortex.</td>
</tr>
<tr>
<td>Down syndrome</td>
<td>Strong social skills; a minority may show autistic features</td>
<td>Increased emotional responsiveness to music, increased smiling and engagement behaviours in infancy, selective deficits for ‘sadness’ on emotional attribution tasks.</td>
<td>Amygdala, Frontal lobes.</td>
</tr>
<tr>
<td>Prader-Willi syndrome</td>
<td>Social withdrawal accompanied by compulsive hyperphagia</td>
<td>Social withdrawal, preservative speech, stereotyped behaviours, temper tantrums, deficits in social attribution.</td>
<td>Frontal lobes, Amygdala</td>
</tr>
<tr>
<td>Smith-Magenis syndrome</td>
<td>Hypersociability accompanied by frequent</td>
<td>Preference for adult contact, lack of adult attention leads to</td>
<td>Insular cortex, Lenticular nucleus</td>
</tr>
<tr>
<td>Disorder</td>
<td>Social Withdrawal</td>
<td>Fear recognition deficits, eye-tracking deficits, social anxiety</td>
<td>Amygdala, Frontal lobes.</td>
</tr>
<tr>
<td>----------------------------------</td>
<td>--------------------------------------------------------</td>
<td>-----------------------------------------------------------------</td>
<td>--------------------------</td>
</tr>
<tr>
<td>Turner syndrome</td>
<td>Social withdrawal (more prominent in ring X Turner syndrome)</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Williams syndrome</td>
<td>Hypersociability</td>
<td>Over-friendliness, prolonged gaze behaviours, atypical and indiscriminate approachability towards both familiar and unfamiliar faces</td>
<td>Amygdala, Frontal lobes</td>
</tr>
</tbody>
</table>
**Table 2: Interventions to improve social behaviours and adaptive skills in individuals with ASD.**

<table>
<thead>
<tr>
<th>Intervention</th>
<th>Duration of Intervention</th>
<th>First Author, Year</th>
<th>Results</th>
</tr>
</thead>
<tbody>
<tr>
<td>Intensive behavioural treatment vs. eclectic treatment in children to reduce disruptive behaviours</td>
<td>1 year</td>
<td>Eikeseth, 2002</td>
<td>Decrease in disruptive behaviours in the behavioural treatment group</td>
</tr>
<tr>
<td>Traditional behavioural approaches vs. natural play interventions to enhance social interaction, compliance and communication abilities</td>
<td>10 weeks</td>
<td>Bernard-Opitz, 2004</td>
<td>Significant gains in play, attention, compliance and communication skills in both groups. Behavioural approaches were superior to play interventions in improving attention and compliance.</td>
</tr>
<tr>
<td>Computer-based intervention to teach how to recognize and predict emotions.</td>
<td>2 weeks</td>
<td>Silver, 2001</td>
<td>Significant improvement in ability to recognize and predict emotions.</td>
</tr>
<tr>
<td>Interactive multimedia tool designed to improve recognition of complex emotions from faces and voice</td>
<td>10-15 weeks</td>
<td>Golan, 2006</td>
<td>Significant improvement in recognition of complex emotions in both groups.</td>
</tr>
<tr>
<td>Repeated sessions of adult imitation vs. regular play to improve social behaviours</td>
<td>Three sessions</td>
<td>Field, 2001</td>
<td>Improvement in distal (e.g., attention) and proximal (e.g., touching) social behaviours with imitation.</td>
</tr>
<tr>
<td>Adult imitation based intervention vs. contingently responsive interaction</td>
<td>Single session</td>
<td>Escalona, 2002</td>
<td>Contingency responsive interaction was more effective in facilitating distal social behaviours. Imitation was more effective</td>
</tr>
</tbody>
</table>
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<thead>
<tr>
<th>Intervention</th>
<th>Duration</th>
<th>Authors</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>Dyadic social communication intervention targeted at parent communication vs. routine care in children with autism.</td>
<td>1 year</td>
<td>Aldred, 2004</td>
<td>The intervention group showed significant improvement compared with controls on reciprocal social interaction, communicative initiation and parent-child interaction.</td>
</tr>
<tr>
<td>Social story intervention to decrease disruptive behaviours in children with autism.</td>
<td>Single session</td>
<td>Scattone, 2002</td>
<td>Decrease in disruptive behaviours in all participants.</td>
</tr>
<tr>
<td>Auditory training intervention to improve school-appropriate behaviours.</td>
<td>10 weeks</td>
<td>Smith, 1985</td>
<td>Improvement in attentiveness, school-appropriate behaviours and communication.</td>
</tr>
<tr>
<td>Use of social adjustment enhancement psychoeducational curriculum for boys with ASD aged 8-12 years.</td>
<td>20 weeks</td>
<td>Solomon, 2004</td>
<td>Significant improvements in facial recognition.</td>
</tr>
<tr>
<td>Use of lego blocks for increasing motivation and peer-interaction</td>
<td>3 years</td>
<td>Legoff, 2006</td>
<td>Lego therapy produced significant gains on measures of social skills as compared to the control group.</td>
</tr>
</tbody>
</table>
Table 3: Interventions to improve social behaviours and adaptive skills in individuals with Williams syndrome.

<table>
<thead>
<tr>
<th>Intervention</th>
<th>Author, Year</th>
<th>Salient Results</th>
</tr>
</thead>
<tbody>
<tr>
<td>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.</td>
<td>O’ Reilly, 2001</td>
<td>An increase in the food consumed and decrease in other inappropriate behaviours.</td>
</tr>
<tr>
<td>Intensive short term cognitive-behavioural therapy to treat OCD like symptoms in a young adult with WS.</td>
<td>Klein-Tasman, 2007</td>
<td>Increase in patient’s insight about his problems and decrease in OCD like symptoms.</td>
</tr>
</tbody>
</table>
WILLIAMS SYNDROME

Key neural impairments
- amygdala
- frontal lobes

Atypicalities of social cognition
- heightened social salience
- Intense gaze behaviours
- atypical approachability
- executive dysfunction

Wide ranging social implications
- problematic peer relationships
- psychopathology
- employment difficulties
- social victimization
Multiple genetic susceptibility mechanisms

AUTISM SPECTRUM DISORDERS

Key neural impairments
amygdala, prefrontal Cortex, face fusiform Area, cerebellum

Atypicalities of social cognition
social withdrawal
Intense gaze behaviours
atypical face processing
impaired communication

Wide ranging social implications
loneliness & ostracism
psychopathology
social isolation
social victimization