Parent insights into atypicalities of social approach behaviour in Williams syndrome

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Abstract

Background: Individuals with Williams syndrome have been reported to show high levels of social interest and a desire to interact with others irrespective of their familiarity. This high social motivation, when combined with reduced intellectual capacity and a profile of atypical social behaviour, is important in terms of social vulnerability of individuals with the disorder. Therefore social approach to unfamiliar people and the role of this behaviour within the WS social phenotype warrants further research to inform social skills intervention design.

Methods: The current study used parent interviews (n=21) to probe aspects of social behaviour and interactions with strangers, as well as the impact of such behaviour on the family. Using thematic analysis, it was possible to explore themes that emerged from the interviews, offering qualitatively rich insight into the variability of social approach behaviour in WS.

Results: Thematic analysis confirmed a significant desire to interact with strangers as well as a lack of awareness of appropriate social boundaries. However, parental reports about their child's social approach behaviour varied considerably. The within-syndrome variability of the sample was emphasised in parental reports of their child's personality characteristics (e.g. levels of impulsiveness), as well as the level of parental supervision employed.

Conclusions: These in-depth parent insights can help target the needs of individuals with WS and emphasise that an individual approach to intervention will be essential due to the heterogeneity of the WS social profile.

Keywords: Williams syndrome, social approach

Abbreviations: SRS, Social Responsiveness Scale

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Williams syndrome (WS) is a genetic neurodevelopmental disorder with a prevalence of approximately 1:20,000 and is caused by the micro-deletion of 25-28 genes on chromosome 7 (7q11.23; Hiller et al., 2003). Alongside mild-moderate levels of intellectual impairment (Searcy et al., 2004), individuals with WS have been reported to display a paradoxical cognitive profile of relative strengths in verbal processing and relative weaknesses of spatial ability (Mervis et al., 2000). Although, it is acknowledged that even the relative strengths in verbal processing are select (Paterson et al., 1999; Laing et al., 2002; Mervis et al., 1999), as language acquisition often follows an atypical developmental pathway, showing deficits in areas such as past tense formation and atypical phonological representations (Thomas et al., 2001). Most relevant to the current study, the social phenotype of WS has attracted significant research attention, largely due to claims of hyper-sociability (Jarvinen et al., 2013). This translates as an extreme pro-social drive to approach and interact with other people, irrespective of whether the person is known to them or not (Jones et al., 2000).

Several studies have explored and characterised social approach behaviours in WS by asking individuals to rate faces for approachability using a Likert scale, when given a hypothetical situation of whether they would like to talk to the presented face or not. However, such studies have produced conflicting findings, which can typically be accounted for by the type of task used and the emotional expression of the faces that have been presented (Porter et al., 2007). For example, in some studies individuals with WS report higher approachability ratings for trustworthy and untrustworthy faces compared to their typically developing (TD) peers (Jones et al., 2000; Martens et al., 2009). Yet, other studies have shown that individuals with WS only give high approachability ratings to happy faces, rather than those expressing negative emotions such as anger or fear (Frigerio et al., 2006). The role of emotion in social approach decisions is thought to be further complicated by impairments in emotion recognition (Porter et al., 2007). Thus the exact nature of social approach behaviour in WS remains unclear, but the issue remains of great importance because of the social vulnerability status associated with increased approach to unfamiliar people (for a discussion of vulnerability issues see both Jawaid et al., 2012 and Lough et al., 2015a). This vulnerability is heightened by considering the increased social approach in addition to the previously

mentioned intellectual impairments (Searcy et al., 2004) and an abundance of social functioning atypicalities such as staring at faces (e.g. Riby & Hancock, 2008) and an inability to make accurate socio-cognitive judgements (Tager-Flusberg & Sulluivan, 2000). Social vulnerability warrants further exploration using multiple methods to gain complimentary insights into the social approach behaviours and underlying issues that could be tackled as part of a social skills training programme.

Riby and colleagues (2014) approached the issue of stranger danger awareness in WS in a novel way by conducting a qualitative analysis of discussions with young people with WS that stemmed from stranger danger video vignettes. Based on the qualitative data produced, it was clear that young people with WS showed heightened vulnerability compared to typically developing individuals. Crucially, 73 per cent of the answers given by the young people with WS (mean 12 years) failed to show an appropriate knowledge or awareness of any risks of interacting with unfamiliar adults. This compared to an average 40 per cent of the responses given by a younger group of typically developing children (mean age 7 years). Riby and colleagues recommended that further qualitative data were needed from a variety of sources on the issue of social approach behaviour and stranger danger awareness, especially based on the within-syndrome variability observed in the responses for the WS group (also captured by Little et al., 2013). This work would allow us to tailor training programmes to compliment individual differences of social approach and stranger danger awareness in WS.

The importance of individual differences was again highlighted in recent work by Ng, Jarvinen and Bellugi (2014). They emphasized the impact that the WS personality profile could have in explaining maladaptive social behaviours. They outlined the case of atypical social motivation in WS. Individuals with WS (both children and adults) were driven by a desire for social closeness in their social interactions, which was underpinned by their "gregarious, people-orientated and affectionate personality features" (p1844) whereas their typically developing peers sought social power driven by "persuasive, dominant and visible personality attributes" (p1844). They argued that identifying the role that personality traits play in the elevated levels of social drive seen in WS could allow us to target interventions towards these areas. Further research on individual social motivation and the underlying mechanisms of social motivation in WS is clearly warranted.

So why do individuals with WS struggle to make appropriate social judgements? There have been several theories proposed to explain the WS social phenotype. Specifically, the neural systems underpinning this social behaviour have attracted significant interest. The amygdala hypothesis proposes atypicalities in amygdala structure and function of individuals with WS. It suggests that those with WS have an atypically enlarged amygdala volume which is linked to the atypical social approach behaviours (Martens, 2009). According to Haas et al. (2009), individuals with WS show decreased amygdala activation in response to threatening faces, which the authors suggest could explain the disinhibited approach behaviour. Therefore both structure and function of the amygdala appear critical. An alternative has been proposed by the frontal lobe hypothesis (e.g. Porter et al. 2007). According to this theory, individuals with WS show similarities of social approach behaviour to individuals who have experienced frontal lobe damage. Both groups share deficits in response inhibition, which leads to atypical approaches, such as approaching strangers. This occurs in spite of 'knowing' that this type of approach behaviour is not appropriate. It could therefore be that inhibitory control is key (Little et al., 2013). However, the proposed theories are far from mutually exclusive. In reality, most researchers acknowledge that these theories are unlikely to be absolute, and rather each makes a partial contribution to our understanding of social approach behaviours in WS (Gaser et al., 2006; Meyer-Lindenberg et al., 2005).

Indeed, recent work has noted considerable variability in areas such as frontal lobe functioning, social functioning, anxiety and social approach behaviours in WS (e.g. Porter et al., 2007; Little et al., 2013; Jarvinen-Pasley et al., 2010; Riby et al., 2014). Little et al. (2013) proposed the notion of sub-groups within WS based on social approach. Through cluster analysis of children's responses on Adolphs Approachability Task (Adolphs et al., 1998), an emotion recognition task and a response inhibition task (the Sun-Moon Stroop Task; Archibald & Kerns, 1999), they noted substantial variability of approach desires. They argued that WS subgroups could be identified based on the social approach profile of an individual, with inhibition being the strongest indicator of subgroup membership. This highlights the need to look at social approach behaviour in a manner that captures individual differences and without reliance on group 'means'. This is especially important for accurately evaluating intervention needs.

The methods employed to investigate social approach behaviour have been discussed. Recent work by Fisher, Mello and Dykens (2014) highlighted a discrepancy between self-report and parental reports of social approach behaviour in adults with WS. They found that the responses given by individuals with WS in a number of different tasks (e.g. self-report approachability scale, self-report faces task) suggested that they displayed much lower levels of abnormal social approach behaviour compared to the levels reported by their parents. Indeed, behavioural observation in a community setting showed it was parent report responses which were more consistent with observations of social behaviour in a natural setting, suggesting that parents could more accurately report their child's social approach behaviour towards strangers. This may be something that individuals with WS find very hard to reflect upon, especially during childhood.

Parent report has been used in the existing literature on social approach behaviour, however, it has predominantly been in the form of questionnaire responses (e.g. Doyle et al., 2004). Considering the value attached to parental reports, and the current discrepancy of findings in WS, the current study aims to extract more in-depth, rich, qualitative data through semi-structured parent interviews. Using the Social Responsiveness Scale (SRS; Constantino & Gruber, 2005) and the Spence Children's Anxiety Questionnaire – Parent Version (SCAS-P; Spence, 1998) we will gain insight in to the general social and anxiety profile of this group (previous research has shown anxiety levels to be high in WS; Riby et al., 2014; Rodgers et al., 2012) as well as establishing whether there is heterogeneity and thus within-syndrome variability in the parental accounts given. This will provide a novel and valuable insight into the social competence of the group, their patterns of social approach behaviour and within-syndrome variability.

Method

Participants

The parents of twenty-one children with WS (range 6 – 15 years; mean age 9.8 years; SD3.2; 10 males, 11 females) were recruited through the Williams Syndrome Foundation. Their child must have had a formal WS diagnosis which had been confirmed through positive genetic florescent *in situ* hybridisation testing. We used the Wechsler Intelligence Scale for Children (WISC-IV; Wechsler, 2003) which generated an overall intellectual ability mean of 54.14 (SD 7.57; Full Scale Intelligence Quotient; FISQ). The sample had a mean verbal IQ (VIQ) score of 63.62 (SD 9.93) and a performance IQ (PIQ) of 51.29 (SD 6.86). For two families, both parents took part in the interview, and for the remaining 19 families, the mother was interviewed. The ethnicity of the cohort was entirely white British. Participants who had a co-morbid diagnosis of an Autism Spectrum Disorder were excluded from the study. The study received favourable ethical approval from the local ethics committee. Informed consent was obtained from parents who took part in the interview.

Materials and Procedure

Social Responsiveness Scale

The Social Responsiveness Scale (SRS; Constantino & Gruber, 2005) is a 65-item parent report questionnaire that measures the normality / abnormality of a child's social functioning. It was originally designed as a screener for Autism Spectrum Disorders, and has since been used to detail the social profile of a variety of typical and atypical populations including with individuals who have Williams syndrome (see Lough et al., 2015b; Riby et al., 2014; Klein-Tasman et al., 2011; Van der Fluit et al., 2012; Channell et al., 2015). Each item on the SRS is coded on a scale of 0 – 3, which generates scores across five sub-domains - social awareness, social cognition, social communication, social motivation and autistic mannerisms, as well as an overall T score as a degree of severity of social abnormality. Higher scores represent greater deficits of everyday social functioning. Previous research using the SRS has suggested that only a small percentage of individuals with WS are likely to be classified as showing 'normal' social behaviours; far more are likely to show either mild-moderate or severe impairments that impact on daily functioning. For example, van der Fluit, Gaffrey, and Klein-Tasman (2012) reported only 17% of individuals with WS (total n= 24) were classified

within the 'normal' range and this was corroborated by Riby et al. (2013) who also reported 17% of their sample to fall within this range (sample size n=59). In that same study, 58 % were classified by parents as showing severe deficits of reciprocal social interaction behaviour that would significantly impair everyday social functioning and 25% showed mild deficits of social behaviour (Riby et al., 2014).

Spence Children's Anxiety Questionnaire – Parent Version

The Spence Children's Anxiety Questionnaire – Parent Version (SCAS-P; Spence, 1998) was completed by 18 of the parents (86%) in the sample. The SCAS-P has previously been used in the literature to measure anxiety in children with WS (e.g. Rodgers et al., Riby et al., 2014) and in relation to the link between social behaviour and anxiety in this population. It is a 38-item measure, on which parents must rate statements on a four point Likert scale, which correspond to the options *never, sometimes, often* and *always*. This measure provides an overall indication of anxiety levels, as well as scores in six subdomains: separation anxiety, physical injury fears, social phobia, obsessive compulsive disorder, and generalised anxiety disorder.

Social Approach Behaviour Interview

A bespoke semi-structured interview was developed by the authors and completed with parents of children with WS. The interview had four modules; auditory sensitivity, social approach behaviour, understanding of emotion and anxiety; of which the social approach behaviour module is explored here (see Appendix A for interview schedule). Relating to the child's social behaviour, the questions covered themes such as interest in social situations, confidence around strangers, and knowledge not to approach strangers.

The researchers met with the parent individually to complete the SRS, the SCAS-P and the semi-structured interview. The interviews were conducted in the homes of families, and the

whole interview (including the social approach / social behaviour module) took approximately 60 minutes.

Data analysis strategy

Thematic analysis was used to systematically analyse the data in line with the suggestions of Braun and Clarke (2006). The interviews were transcribed and initial codes and conceptualisations were generated from line-by-line coding of the accounts given by parents. These codes were analysed and developed into themes which were deemed to fit the data as closely as possible. These themes were processed and reprocessed until final themes were generated and could be reviewed.

Results

Social Responsiveness Scale

The mean SRS T score for the sample showed that the group as a whole experienced severe levels of impairment in their social functioning (mean T score = 79.6), although there were high levels of variability within the sample (SD = 13.5). There was no significant difference in the overall T scores of males (M = 77.9, SD = 15.02) versus females (M = 81.18, SD = 12.45; t(19) = 1.77, $p = 0.59$), and there was no significant correlation between total SRS T score and FSIQ (r = 0.19, $p = 0.41$), VIQ (r = 0.04, p = 0.88) or PIQ (r = 0.27, $p = 0.24$). Figure 1 shows that 72% of the group had overall T scores in the severe range, whilst 14% scored within the mild-moderately impaired range, and 14% within the normal range of social functioning. It is worth noting that the proportion of WS participants being classified as having mild-moderate and severe social deficits is similar to previous reports with larger WS samples using the SRS (van der Fluit, et al., 2012; Riby et al., 2014).

There was a significant correlation between the age of the participants and their total T score (r = -0.56, $p<0.01$). There was also a significant correlation between age and scores on 4 out of the 5 sub-domains of the SRS (awareness: r = -0.52, $p<0.05$; cognition: r = -0.63, $p<0.01$; communication: r = 0.64, $p<0.01$; mannerisms: r = -0.58, $p<0.01$), suggesting that the most socially impaired were, on average, younger. However, this was not the case for the sub-domain of social motivation (r = 0.21, $p = 0.37$), indicating that atypicalities in social

motivation do not differ with age. Of further interest, all of the sub-domains of the SRS were significantly correlated with each other (all at $p<0.05$), with the exception of social motivation and awareness (r = 0.3, $p = 0.19$), and social motivation and cognition (r = 0.23, $p = 0.31$). Social motivation is something that is clearly atypical in WS across ages and that has been captured in the WS literature to date as an identifying aspect of the WS social phenotype (e.g. Doyle et al., 2004; Frigerio et al., 2006; Jawaid et al., 2012).

[Figure 1]

Spence Children's Anxiety Scale – Parent Version

The mean raw score for overall anxiety was 20.23 (SD 12.18), suggesting the sample experience low levels of anxiety (Rodgers et al., 2012). From this it can be proposed that this will have limited influence on social approach behaviours (Riby et al., 2014). The mean sub-scale scores are shown in Table 1. Interestingly, participants scored highest on the GAD subscale of anxiety, with low scores on the OCD, social phobia and panic subscales. There was no significant correlation between total SCAS scores and total SRS T scores (r = 0.38, $p = 0.12$), age (r = 0.19, $p = 0.45$) or IQ (FISQ: r = 0.05, $p = 0.84$; VIQ: r = 0.05, $p = 0.83$; PIQ: r = 0.2, $p = 0.42$).

[Table 1]

Social approach behaviour interview

The thematic map shown in Figure 2 depicts the themes that arose from the semi-structured interviews with parents of children with WS.

[Figure 2]

Naivety to danger and a lack of social boundaries were prominent themes in the accounts of parents with children with WS. However, there were qualitative differences in the nature of their social behaviour, personality traits and the level of parental supervision employed, reinforcing the heterogeneous nature of social approach behaviour in WS.

The parents talked about the naivety of their children, in particular to dangerous or potentially risky situations and as seen by the ages of the illustrations below, this was an issue across ages:

"She can't understand why she can't talk to people she doesn't know she will say they're nice and she liked them so she doesn't understand why that's bad" (female, 8 years)

"I just know for a fact anyone could come up in a car and say come on Natalie and she would climb in and go with them" (female, 6 years)

"I think he's too trusting particularly of adults … he would be very easily lead" (male, 15 years)

"I picked her up because she was sick and we crossed the road and a man walked past and she just starts waving and says hello as he got closer, asking him what his name was" (female, 6 years)

They also frequently highlighted the difficulties experienced by their children with regards to understanding and respecting social boundaries:

"She will ask private questions she will tell things about herself which are just not appropriate" (female, 9 years)

"She's not got boundaries … if she was going to talk to someone she would put her hand on their knee or arm she would break that personal space and not understand that it wasn't right" (female, 8 years)

"… he will hold hands and try and hug people whether he knows someone or not is irrelevant" (male, 9 years)

"She has no concept of personal space … if someone has a nice necklace she will touch it and tell them she likes it, she can get that close to them" (female, 6 years)

These observations seemed to be tied in with the extreme outgoing and gregarious behaviour reportedly displayed by some of the children. Several parents described their children as highly impulsive in their social approach behaviour:

"I don't know, you can tell her until you're blue in the face but it's like it is inbuilt it's something that she can't stop" (female, 12 years)

"It's just an instinct for her it's part of her genetic make-up its spontaneous it's not something she thinks" (female, 9 years)

"She doesn't ever really think about what she's doing" (female, 8 years)

Parents were also concerned about the longevity of this behaviour, and many shared their concerns for the future:

"I don't know that she will ever be aware that you don't approach strangers" (female, 14 years)

"I keep saying she will never be in a situation on her own, but she's going to get older and you don't know what's going to happen" (female, 14 years)

However, there was a notable amount of variability in the accounts, as not all parents reported these impulsive behaviours. Some parents discussed the reserved personality of their child, which they saw as serving to minimise inappropriate social approach behaviour:

"He wouldn't like to be the centre of attention or to stand up and talk in front of lots of people so I think he would be more comfortable in familiar surroundings with people he knows" (male, 15 years)

"I see him hold back sometimes if he doesn't like someone" (male, 13 years)

"She doesn't actively seek others out, she's quite quiet" (female, 15 years)

The above quotes begin to illustrate the heterogeneity in the accounts given. With differing degrees of social approach behaviour, as well as distinctly different personality traits, it is perhaps unsurprising that the level of parental supervision employed was also varied. Some of the parents referenced the high level of parental supervision they employed to ensure that their children were safe around strangers.

"I think because he doesn't go out by himself I don't really worry about strangers" (male, 15 years)

"I think what holds him back most of all is … he's very restricted by having to need us to be there or take him somewhere so I think that has stunted his social life"(male, 9 years)

The first of the above quotes is interesting given the age of the individual with WS and the likelihood that if they were typically developing this is an age (15 years) when we would expect social independence to be evident. It seems that such a high level of parental supervision has curtailed opportunities for social approach, but at a cost to their level of independence. The primary driving force when parents are considering this equation was their need to protect their child.

"If we weren't there, she would be easy picking" (*female, 6 years*)

For other parents, they have been able to build up confidence and trust in their child, allowing them less parental supervision, and greater autonomy:

"At first I was worried because I'm a mum and he was going up talking to people he doesn't know, but now I've got confidence in him and knowing his own mind." (male, 13 years)

Perhaps unsurprisingly, many families spoke about the impact that their child's social approach behaviour had on the family unit and daily living:

"His sister gets embarrassed. She's younger … and she will start to talk to people as well because she sees him doing it" (male, 10 years)

"We have to do holidays different my husband would love to do an all-inclusive somewhere but I can't possibly go somewhere where she can pester the same people through breakfast at the pool through the afternoon and at dinner as well so for holidays we always go self-catering and we always go to the same places so we know our containment areas." (female, 6 years)

Discussion

By analysing interviews conducted with parents of children with WS, the current study identified impaired social competence and high levels of social approach behaviour across the sample. We also noted considerable heterogeneity of social approach behaviours in this clinical group, consistent with previous research (e.g. Little et al., 2013; Porter et al., 2007). Based on our findings, it would seem that this variability cannot be predicted solely by age or IQ. Indeed there were some themes in the parent interviews that were evident for all parents irrespective of the age of their child. As expected, all of the children were reported to display inappropriate social behaviour, and to be naïve to danger, but crucially their personality traits (e.g. their level of impulsiveness) as well as familial factors (e.g. level of parental supervision) influenced the nature of this behaviour. This is in line with previous research (e.g. Porter et al., 2007; Ng et al., 2014) and compliments findings by Porter and Colheart (2005) on the heterogeneity of cognitive strengths and weaknesses in WS. The differences found in the qualitative interview data are likely to help shape the individual atypical social profiles of these children, and impact upon the effectiveness of interventions which assume a homogenous WS social behaviour profile. Furthermore the individual nature of the social approach profiles in these children will impact upon the way that such behaviours influence family life in each of these family units.

Based on the interviews, and the data obtained from the questionnaire rating items, it is clear that the children with WS in this study showed an interest in social situations and as evident across both the SRS and the interview data, were strongly socially motivated (in line with Frigerio et al., 2006); however only some were reported to be especially confident and disinhibited around strangers. When considering the theoretical explanations offered by the amygdala hypothesis and the frontal lobe hypothesis, this heterogeneity proves problematic. The frontal lobe hypothesis centres on difficulties with response inhibition, yet not all of the participants were reported to experience this, or indeed not to the same extent. Unfortunately we do not have cognitive or behavioural inhibition data for this sample of children but it would be interesting to explore the role of the cognitive heterogeneity in WS with the social heterogeneity reported here. Furthermore, it may be that individual differences in personality factors could play an important mediating role in pro-social WS drive as recently suggested

(Ng et al., 2014). Certainly some parents suggested that their children were outgoing and extraverted, whereas others emphasised the reserved nature of their child. This issue suggests an interesting area for further exploration. Finding an appropriate theoretical framework for social approach behaviours in WS is dependent on acknowledgement of the heterogeneity and subgroups that exist within the disorder and the role of both cognitive and social profiles. Therefore taking an in-depth individual / holistic approach to understanding such issues is crucial for both theory and practice.

These findings offer a novel insight into the vulnerability status of some individuals with WS. Given that individuals with WS struggle to form and maintain peer relationships (Davies et al., 1998), experience high levels of anxiety (Riby et al., 2014; Rodgers et al., 2012) and lack stranger danger awareness (Riby et al., 2014), the increased social approaches of some individuals with WS is of particular concern. These individuals may be targeted for intervention. Indeed the qualitative data provided by parents in this study allows us to delve deeper into the social approach profile of individuals with the disorder than face rating tasks used previously (e.g. Jones et al., 2000). The work can have a significant impact by highlighting the heterogeneity of social approach in WS, but also by emphasising the impact of the atypicalities of social behaviour and social approach on the wider family unit. Parents noted this in their responses as highlighted in a number of quotes in the Results section. Therefore supporting these family needs is important.

The limitations of the current study merit consideration. The qualitative interview data have provided us with a rich insight in to how parents view their child's social approach behaviour. However, these data do not allow for analysis of the link between SRS scores, SCAS scores and social approach behaviour. Therefore, whilst these measures are useful in outlining the profile of the sample, the relationship between social functioning, anxiety and social approach remains unclear. Furthermore, although we have outlined the impact that age and IQ has on social functioning and anxiety in our sample, it is not clear how these factors relate to the social approach behaviour described in the interviews. It seems likely that age will have an effect on social approach, although it is worth noting that quotes about abnormal social approach behaviour were provided by parents of children of varying ages, implying that it could transcend age boundaries. Finally, as parental report offers an indirect measure

of social approach, it is important that it is considered alongside other methodologies, in order to adopt a multi-informant approach to understanding social approach behaviour.

The findings from this study open up numerous avenues for future research. First, the developmental trajectory of social approach behaviours in WS remains unclear, and in particular whether the heterogeneity reported here persists into adulthood. Furthering our knowledge on this area is particularly important when considering the increased levels of independence associated with adulthood, and the potential impact of social approach on social vulnerability (e.g. Lough et al., 2014). Secondly, as the literature base on heterogeneity in WS begins to build, future research should look to bridge the gap between the reported heterogeneous social profile, and the heterogeneous cognitive profile, in order to generate more comprehensive ideas on how to define these subgroups. This could be invaluable in helping to tailor support and avoid a one size fits all approach to intervention. Finally, the current study emphasises the importance of considering social approach behaviours and subsequent issues of vulnerability at the individual level, moving away from reliance on group means in order to formulate effective interventions.

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Conflict of Interests Statement

The authors report no conflict of interests.

References

Adolphs., R., Tranel, D., & Damasio, A. R. (1998). The human amygdala in social judgment. *Nature Reviews Genetics*, 393, 470–474.

Archibald, S. J., & Kerns, K. A. (1999). Identification and description of new tests of executive functioning in children. *Child Neuropsychology*, 5, 115–129.

Barttfeld, P. Amoruso, L., Ais, J., Cukier**,** S., Bavassi**,** L., Tomio, A. et al. (2013). Organization of brain networks governed by long-range connections index autistic traits in the general population. *Journal of Neurodevelopmental Disorders*, 5, 16.

Braun, V., & Clarke, V. (2006). Using thematic analysis in psychology. *Qualitative research in psychology*, *3*(2), 77-101.

Channell**,** M. M., Phillips**,** B. A., Loveall**,** S. J., Conners**,** F. A., Bussanich, P. M. & Klinger, L. G.(2015). Patterns of autism spectrum symptomatology in individuals with Down syndrome without comorbid autism spectrum disorder. *Journal of Neurodevelopmental Disorders*, **7**:5.

Constantinno, J., Gruber, C. (2005). The social responsiveness scale: Western Psychological Services.

Davies, M., Udwin, O., & Howlin, P. (1998). Adults with Williams syndrome. Preliminary study of social, emotional and behavioural difficulties. *British Journal of Psychiatry, 172***,** 273–276.

Doyle, T. F., Bellugi, U., Korenberg, J. R., & Graham, J. (2004). ‘‘Everybody in the world is my friend’’ hypersociability in young children with Williams syndrome. *American Journal of Medical Genetics Part A*, 124A(3), 263–273.

Frigerio, E., Burt, D. M., Gagliardi, C., Cioffi, G., Martelli, S., Perrett, D. I., et al. (2006). Is everybody always my friend? Perception of approachability in Williams syndrome. *Neuropsychologia*, 44(2), 254–259.

Gaser, C. Luders, E., Thompson, P. M., Lee, A. D., Dutton, R. A. Geaga, J. A. *et al.* (2006). Increased local gyrification mapped in Williams syndrome. *NeuroImage*, 33(1), 46–54.

Haas, B. W., Mills, D., Yam, A., Hoeft, F., Bellugi, U., & Reiss, A. (2009). Genetic influences on sociability: Heightened amygdala reactivity and event-related responses to positive social stimuli in Williams syndrome. *Journal of Neuroscience*, 29, 1132–1139.

Hillier, L.W., Fulton, R.S., Fulton, L.A., et al. (2003). The DNA sequence of chromosome 7. *Nature*, 424:157–164.

Järvinen A., Korenberg J. R., Bellugi U. (2013). The social phenotype of Williams syndrome. *Current Opinion in Neurobiology*. 23, 414–422.

Jarvinen-Pasley, A., Adolphs, R., Yam, A., Hill, K. J., Grichanik, M., Reilly, J., et al. (2010). Affiliative behavior in Williams syndrome: Social perception and real-life social behavior. *Neuropsychologia*, 48(7), 2110–2119.

Jawaid, A., Riby, D., Owens, J., White, S., Tarar, T., & Schulz, P. (2012). 'Too withdrawn' or 'too friendly': considering social vulnerability in two neuro-developmental disorders. *Journal of Intellectual Disability Research*, *56*(4), 335–350.

Jones, W., Bellugi, U., Lai, Z., Chiles, M., Reilly, J., Lincoln, A., & Adolphs, R. (2000). Hypersociability in Williams syndrome. *Journal of Cognitive Neuroscience*, 12, 30-46.

Klein-Tasman, B. P., Li-Barber, K. T., & Magargee, E. T. (2011). Honing in on the social phenotype in Williams syndrome using multiple measures and multiple raters. *Journal of autism and developmental disorders*, *41*(3), 341-351.

Little, K., Riby D. M., Janes, E., Clark, F., Fleck, R. & Rodgers, J. (2013). Hetrogeneity of social approach behaviour in Williams syndrome: The role of response inhibition. *Research in Developmental Disabilities*, 34, 959 – 967.

Lough, E., Flynn, E. & Riby, D. M. (2015a). Mapping real-world to online vulnerability in young people with developmental disorders: Illustrations from autism and Williams syndrome. *Review Journal of Autism and Developmental Disorders* 2(1), 1-7.

Lough, E., Hanley, M., Rodgers, J., South, M., Kirk, H., Kennedy, D. P., & Riby, D. M. (2015b). Violations of Personal Space in Young People with Autism Spectrum Disorders and Williams Syndrome: Insights from the Social Responsiveness Scale. *Journal of autism and developmental disorders*, 1-8.

Mervis, C., Robinson, B., Bertrand, J., Morris, C., Klein-Tasman, B., Armstrong, S. (2000).The Williams syndrome cognitive profile. *Brain and Cognition*, *44*, 604-628.

Martens, M. A., Wilson, S. J., Dudgeon, P., & Reutens, D. C. (2009). Approachability and the amygdala: Insights from Williams syndrome? *Neuropsychologia*, 47(12), 2446–2453..

Ng, R., Jarvinen, A. & Belludi, U. (2014). Toward a deeper characterization of the social phenotype of Williams syndrome: The association between personality and social drive. *Research in Developmental Disabilities,* 35: 1838 – 1849.

Porter, M. A., & Coltheart, M. (2005). Cognitive heterogeneity in Williams syndrome. *Developmental Neuropsychology*, 27(2), 275–306.

Porter, M. A., Coltheart, M., & Langdon, R. (2007). The neuropsychological basis of hypersociability in Williams and Down syndrome. *Neuropsychologia*, 45(12), 2839–2849.

Riby, D. M. & Hancock, P. J. B. (2008).Viewing it differently: social scene perception in Williams syndrome and autism. *Neuropsychologia*, 46, 2855 – 2860.

Riby, D M, Hanley, M, Kirk, H, Clark, F, Little, K, Fleck, R. et al. (2014). The Interplay between Anxiety and Social Functioning in Williams Syndrome. *Journal of Autism and Developmental Disorders* 44(5), 1220-1229.

Riby, D. M., Kirk, H., Hanley, M., & Riby, L. M. (2014). Stranger danger awareness in Williams syndrome. *Journal of Intellectual Disability Research, 58*(6), 572-582.

Rodgers, J., Riby, D. M., Janes, E., Connolly, B., & McConachie, H. (2012). Anxiety and repetitive behaviours in autism spectrum disorders and Williams syndrome: A cross-syndrome comparison. *Journal of Autism and Developmental Disorders*, 42, 175–180.

Searcy, Y.M., Lincoln, A., Rose, F., Klima, E., Bavar, N., & Korenberg, J.R. (2004). The relationship between age and IQ in adults with Williams syndrome. *American Journal on Mental Retardation, 109*, 231–236.

Spence, S. H. (1998). A measure of anxiety symptoms among children. Behaviour Research and Therapy, 36, 545–566.

Tager-Flusberg H. & Sullivan K. (2000).A componential view of theory of mind: evidence from Williams syndrome. *Cognition* 76, 59 – 90.

van der Fluit, F., Gaffrey, M. & Klein-Tasman, B.P. (2012). Social cognition in Williams syndrome: Relations between performance on the social attribution task and cognitive and behavioral characteristics. *Frontiers in Developmental Psychology*, 3:197

Wechsler, D. (2003a). *Wechsler Intelligence Scale for Children* (4th ed.). San Antonio, TX: The Psychological Corporation.

Appendix A

Social approach behaviour interview schedule

To begin with, I would like to find out about _____level of interest in social interaction with other people in general. Some children really enjoy social interaction with others and actively seek out opportunities for this to happen, whereas other children do not show this level of interest.

How would you describe _____'s behaviour in this area? (Are they interested in social interaction? How do you know? What do they do?)

Does _____ show more interest in social interaction with certain people?

Does _____show more interest in social interaction with children or with adults? What makes you say this

Does _____show more interest in social interaction with familiar people or with unfamiliar people? What makes you say this?

Now, I would like you to think about how _____behaves around people they don't know (a stranger). Children vary in how confident they feel around people they don't know. Some children are very confident and will approach them without hesitation, whereas other children feel less confident and are quite cautious

Can you tell me a little bit about how _____ behaves around people he/she doesn't know? (How do they respond to strangers? What do they do?)

Do you think that the setting _____ affects how they behave around strangers? In what way? (e.g. is it the same at home/school?)

Does _____ seem more **confident** around strangers in familiar or unfamiliar settings? What makes you say this?

Do you think that _____**knows** that they shouldn't approach a stranger? What makes you say this?

To what extent do you think that _____**knows** that they shouldn't approach a stranger?

How **likely** it is that _____would approach a stranger? What makes you say this?

Could you describe an example in the last month when _____has approached a stranger? What happened? (Get specific detail) Including: What exactly happened before, during and after. Why do they think child approached stranger? How did parent respond? What did child do following parent's response?

How does the way _____behaves around strangers make you feel?

Sometimes parents report feeling **worried** about the way their child behaves around strangers. Do you ever feel worried about the way _____behaves around strangers? What makes you say this?

Sometimes parents report feeling **stressed** about the way their child behaves around strangers. Do you ever feel stressed about the way _____behaves around strangers? What makes you say this?

Sometimes parents report feeling **embarrassed** about the way their child behaves around strangers. Do you ever feel embarrassed about the way _____behaves around strangers? What makes you say this?

Does the way _____behaves around strangers have any impact upon family life? (Do you have to make any changes as a family to accommodate this behaviour?)

Does the way _____behaves around strangers ever make you feel that you need to be more protective of him/her?

Does the way _____ behaves around strangers ever cause you to avoid going to certain places?

Does the way _____ behaves around strangers make you feel that you need to prepare before going somewhere?

Now, I am interested in finding out about how much _____thinks about what they are doing in a social situation. I am also interested in how well you think they can stop themselves from doing something they know they shouldn't do in a social situation. Sometimes children can find this difficult and tend to behave without thinking about the potential consequences or risks. For example; they may say inappropriate things to other people, or look through someone's bag/possessions without asking.

Can you describe _____'s behaviour in this area? (e.g. does your child tend to behave without thinking? Do they often take risks?)

To what extent do you think that _____ **thinks** about what he/she is doing in a social situation? What makes you say this?

To what extent do you think that your child acts on impulse in a social situation? What makes you say this?

Can you describe a specific incident in the last month when your child has done something they know they shouldn't do in a social situation? (e.g. saying something inappropriate/looking through someone's bag) What happened?

Figure 1: Levels of impairment shown for total SRS scores and scores on the five sub-domains

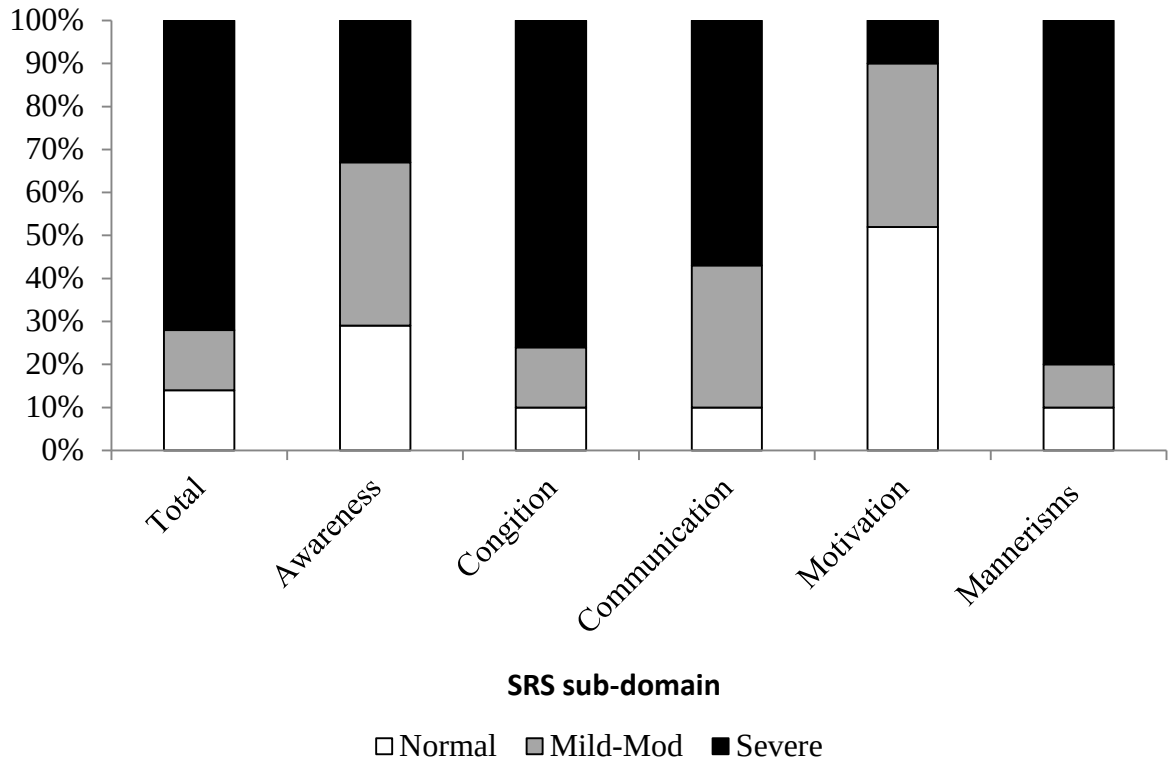

Figure 2: Thematic map for parent interviews on social approach behaviours

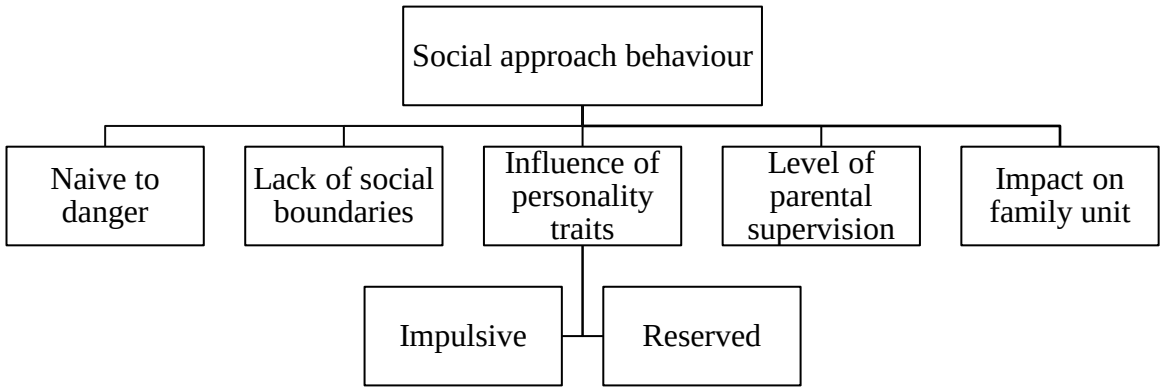

Table 1: Mean SCAS-P total score and sub-scale scores

SCAS-P	T scores
Total score	20.23 (12.18)
Panic/Agoraphobia	2.22 (2.51)
Separation anxiety	4.83 (4.08)
Physical injury fears	4.17 (2.79)
Social phobia	2.11 (2.14)
OCD	1.89 (1.99)
GAD	5.06 (3.19)