The Interplay between Social Functioning in Anxiety in Williams Syndrome

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Abstract

Background: The developmental disorder Williams syndrome (WS) has been associated with an atypical social profile of hyper-sociability and heightened social sensitivity across the developmental spectrum. In addition, previous research suggests that both children and adults with WS have a predisposition towards anxiety. **Methods:** The current research aimed to explore the profiles of social behaviour and anxiety across a broad age range of individuals with the disorder (n=59, ages 6-36 years). We used insights from parental reports on two frequently used measures, the Spence Children's Anxiety Scale (SCAS-P) and the Social Responsiveness Scale (SRS). **Results:** Severity of anxiety was correlated with a greater degree of social dysfunction as measured by the SRS in this group. We split the group according to high or low anxiety as measured by the SCAS-P and explored the profile of social skills for two groups. Individuals high and low in anxiety differed in their social abilities. **Conclusions:** The results emphasise the need to address anxiety issues in this group and to consider how components of anxiety might relate to other features of the disorder.

Keywords: Williams syndrome, Social, Anxiety

Abbreviations: SRS, Social Responsiveness Scale; SCAS-P, Spence Children's Anxiety Scale Parent Measure.

The Interplay between Social Functioning in Anxiety in Williams Syndrome

Williams Syndrome (WS) is a developmental disorder, prevalence of 1:20,000 (Korenberg, Bellugi, Salandanan, Mills, & Reiss, 2003, but see also 1:7,500 Strømme, Bjørnstad, & Ramstad, 2002), caused by the micro-deletion of 25-28 genes on chromosome 7q11.23 (Blomberg, Rosander, & Andersson, 2006) and with an equal gender distribution. The disorder is characterised by a hyper-sociable personality, and cognitive, social, and behavioural difficulties (Martens, Wilson, & Reutens, 2008). Although there is considerable variation within WS (Porter & Coltheardt, 2005), individuals generally function within the mild-moderate range of intellectual impairment, with relatively more proficient verbal than nonverbal skills (Mervis et al., 2000). Considering emotional and psychological characteristics, research has indicated high levels of anxiety and fear (Dykens, 2003). There is considerable evidence that individuals with WS are at an elevated risk for anxiety disorder compared to individuals who are developing typically (Rodgers, Riby, Janes, Connolly, & McConachie, 2012).

Anxiety

Anxiety is reported to be one of the most common psychopathologies for both children and adults with WS (Porter, Dodd, & Cairns, 2009; Stinton, Elison & Howlin, 2010). For example, in research on mental health in adults with WS, anxiety has been reported to be the most significant challenge (Stinton et al., 2010) and has been reported to be more prevalent than problems with depression (Stinton, Tomlinson, & Estes, 2012). Within an anxiety profile, the most prevalent subcategories associated with the disorder are specific phobia (30-53.8%), generalised anxiety disorder (GAD; 10-18%), separation anxiety disorder (SAD; 4-6.7%) and obsessive compulsive disorder (OCD; 2-2.5%) (Dodd & Porter, 2009; Dykens, 2003; Leyfer, Woodruff-Borden, Klein-Tasman, Fricke, & Mervis, 2006).

It is useful to explore whether individuals with WS are at more or less risk of anxiety than individuals who have other neurodevelopmental disorders. Dykens, Rosner, Ly and Sagun (2005) addressed this

question and reported that 8- to 47-year olds with WS (n=31) had higher levels of anxiety than those of comparable chronological age with Prader-Willi Syndrome (PWS) and Down syndrome (see also Graham, Rosner, Dykens & Visootsak, 2005). Dimitropoulos, Ho, Klaiman, Koenig, and Schultz (2009) reported a non-significant trend for individuals with WS (n=20; 4-19 years) to show more anxious behaviours than those with PWS and Autism Spectrum Disorder (ASD). Individuals with WS showed a trend towards more fears and generalised anxiety than the other two groups. Giving a different insight, research involving 6-15 years olds has suggested that anxiety is lower in WS than ASD (Rodgers et al., 2012). Certainly there is a wealth of literature to suggest that individuals with ASD are highly susceptible to anxiety (White, Oswald, Ollendick, & Scahill, 2009). Although the existing research emphasises that individuals with WS are at an elevated risk for anxiety disorder compared to individuals who are developing typically, and even in comparison to individuals with other developmental disorders, previous research has not yet explored how highly anxious individuals with WS differ from low-anxious individuals with the disorder. The current evidence suggests that further exploration of anxiety in children and adults with WS is required to ascertain the aetiology, trajectory and phenomenology of this distressing condition.

Social functioning

The WS social phenotype has been linked to hyper-sociability or an extreme drive towards social engagement with both familiar and unfamiliar people (Jones et al., 2000). There is also mounting evidence that across the developmental spectrum individuals with WS have difficulties functioning socially. These social difficulties include decreased social independence, problems using appropriate social communication strategies, difficulties using complex socio-cognitive skills and problems maintaining friendships. Even by adulthood the vast majority of individuals with the disorder have not developed the sophisticated social skills required to establish and maintain friendships and over 70% of WS adults suffer social isolation (Udwin, 1990, Davies et al., 1998). Problems functioning socially, especially when having difficulty deciding who to trust (Riby et al., 2013), who to approach (Jones et al., 2000) and having difficulty using appropriate social cues once in an interaction (e.g. holding

atypically prolonged face gaze, Mervis et al., 2003), alongside decreased intellectual capacity (Searcy et al., 2004) can lead to social vulnerability (Jawaid et al., 2012). Importantly, although individuals with WS might seem at first blush to be outgoing and friendly, their social behaviours are far from 'typical' and many individuals with the disorder struggle with functioning socially. In fact there has been a suggestion of overlap between the types of difficulties associated with WS and ASD. Although the prototypical characterisation of WS has been a hyper-social personality, there are significant impairments of social skills that share overlap with the autism spectrum. Klein-Tasman et al. (2007) reported that half of their young WS children (2-5 years old) showed difficulties on the Autism Diagnostic Observation Schedule similar to those of children with Pervasive Developmental Disorder—Not Otherwise Specified (Klein-Tasman et al., 2009). Using the Social Responsiveness Scale (SRS) van der Fluit, Gaffrey and Klein-Tasman (2012) reported the majority of their 8-15 year olds with WS were classified by parents in the mild-moderate range of social difficulties. Only 17% of their sample (3 individuals) was classified within the 'normal' range of social functioning. Klein-Tasman and colleagues (2011) went one step further and explored ratings within the SRS for 4-16 year olds with WS and emphasised that the most significant deficits of social functioning fall in the area of social cognition as opposed to social motivation.

Anxiety and social functioning

Research with neurodevelopmental disorders such as ASD indicates that difficulties with social functioning are associated with the presence of anxiety (Sukhodolsky et al., 2008). Indeed it has been postulated that anxiety may increase during the adolescent years as teenagers with ASD are faced with ever more challenging social situations (Klin, Volkmar, & Sparrow, 2000) and a developing awareness of their lack of skills in relation to maintaining friendships and interacting effectively in the social world (Tantam, 2003). There is evidence therefore of an association between social impairment and anxiety in people with ASD whereby social deficits might predict the development of anxiety in children with the disorder (Bellini, 2006). ASD traits have also recently been reported in children with anxiety disorders (van Steensel et al., 2013). Finally, some cognitive behaviour therapy

(CBT) interventions targeting anxiety in ASD incorporate social skills training as part of the programme (White et al., 2009).

Establishing the correlates of anxiety is critical in understanding both resilience and vulnerability factors and in guiding the development of appropriate interventions to reduce distress for individuals with WS. Given an atypical social profile coupled with high levels of anxiety for some individuals with the disorder it is important to examine whether the presence of social functioning impairments may be associated with anxiety. It is unclear whether the WS social phenotype (hyper-sociable behaviour or relatively good expressive language skill) acts as a 'protective' factor against the development of anxiety, or whether hyper-sociability may mask anxiety. This is especially important when we know that there is significant heterogeneity of cognition (Porter & Cotlheart, 2005), social behaviour (Little et al., 2013) and most likely, anxiety within WS. Only with this in mind can we understand the constellation of behaviours and experiences of individuals with WS and provide targeted assessment and intervention.

The current study

The aim was to explore the relationship between parental reports of anxiety and social functioning within WS. We were not aiming to explore the typicality of this relationship (e.g. by involving typical controls) or the syndrome-specificity of the relationship (by including other disorders) but we were specifically interested in the relationship between these features in this disorder of development. This is important because anxiety is the most common aspect of psychopathology associated with WS and the social phenotype of the disorder is a defining but variable feature of the group. We hypothesise that a high percentage of individuals with WS will show significant anxiety levels as shown in previous research. We also hypothesise that on the SRS the majority of individuals with WS will show at least mild-moderate impairments of social functioning. However, we have no specific hypotheses relating to the link between anxiety and social functioning as this is a first exploration of this relationship in this disorder. Therefore, once we have identified individuals experiencing anxiety

problems, the next step is to explore how a group with high reported anxiety might differ from those with WS with lower reported anxiety. This type of within-disorder exploration is important to try to address the needs of individuals with the disorder, especially if there is heterogeneity of skills, behaviours and needs.

Method

Participants

Participants were the parents of 59 individuals with WS aged 6- and 36-years old (mean age 17years; 30male, 29female) who had been recruited to participate in a multi-site programme of research on cognition, behaviour and psychopathology in WS[1]. Individuals had previously been diagnosed with WS based on clinical manifestations and 51[2] (86%) had previously had their diagnosis confirmed with genetic testing. We measured verbal ability using the British Picture Vocabulary Scale II (BPVS II; Dunn, Dunn, Whetton & Burley, 1997) and nonverbal ability using the Ravens Coloured Progressive Matrices (RCPM; Raven, Court & Raven, 1990) for 51 of the 59 individuals with WS (86% of the sample). The group had a mean BPVS Raw Score of 94 (standard deviation 28.52; min score 51, max score 150) indicating an average verbal mental age of 9years 7months. On the RCPM the group had a mean score of 17 (standard deviation 4.70; min score 9, max score 34 out of a possible 36).[3]

Measures

[1] All were biological parents of the individuals with WS and all WS individuals lived with their parents. In all cases the same respondent answered both questionnaires.

[2] The 8 individuals who had not previously had their diagnosis confirmed with fluorescent in situ hybridisation testing to detect one deletion of the elastin gene were all older adults (>20years) who were diagnosed prior to routine genetic testing for the disorder. The pattern of results remains consistent if these individuals are removed from the sample and therefore to allow us to analyse all available data these individuals have been left in the analysis.

[3] The pattern of results remains consistent if we only analyse the data for those participants for whom we have verbal and nonverbal ability measures.

Anxiety

The *Spence Children's Anxiety Scale – Parent Version* (SCAS-P; Spence, 1998) was completed by all parents of individuals with WS. This measure has previously been used to give a parental report of anxiety in WS for young children (6-15 years, Rodgers e al., 2012) and adults (12-28 years, Dodd et al., 2009). The SCAS-P is a 38-item parent-report scale providing an overall measure of anxiety and has been reported to have high internal consistency of .92 (Spence, Barrett, & Turner, 2003), indeed for our sample internal consistency has .93. There are six subscales that relate to panic/agoraphobia, separation anxiety, physical injury fears, social phobia, obsessive compulsive disorder, and generalised anxiety disorder (Yule, 1997). Parents rate the items on a four-point Likert scale (ranging from *never*, *sometimes*, *often*, to *always*). The Total Score was used for analysis. Due to the intellectual capabilities and wide age range of the participants it would not have been possible to use self-report and insights from parents were essential.

Some items were adapted for use with parents whose children were over the age of 18years. This involved replacing the words 'school' and 'kids' to 'work' and 'people' respectively throughout the questionnaire to make the content appropriate. This exact replacement has previously been used in published research with adults who have WS (e.g. up to 28years, Dodd et al., 2009). Indeed when adults with WS have been studied in relation to anxiety it has been common for child measures to be adapted for older individuals with this disorder (e.g. when using the Child Behavior Checklist designed for 6-18 year olds but used with 6-48 year olds in research by Porter et al., 2009 and when using the Multidimensional Anxiety Scale for Children with individuals aged 8-47 years in research by Dykens et al., 2005).

Social Reciprocity

The *Social Responsiveness Scale* (SRS; Constantino & Gruber, 2005) was completed by parents to assess social reciprocity abilities/impairments. This measure has previously been used with

individuals who have WS as a parent report of social functioning (Faye van der Fluit et al., 2012; Klein-Tasmin, Li, Barber & Margaree, 2011). The measure consists of 65 items that give an overall score for social ability / functioning (reported internal consistency.93, Constantino & Gruber, 2005), for the current sample the measure showed internal consistency of .91. There are also 5 subscales that relate to social awareness, social cognition, social communication, social motivation and autistic mannerisms. The total score and the subscale scores can be converted to T scores to determine if behaviour is within the 'normal' range, the 'mild – moderate impairment' range or shows 'severe impairments' that impact on everyday functioning and ability. For the purposes of the correlation analyses we have used the raw scores, as T scores implement a top cut off of 90 and therefore minimise the possible spread of scores, especially for this clinical sample where the scores can be extreme and thus more variable when raw scores are used. We also used raw scores for consistency, as T scores are not available for the SCAS-P. An adult version which corresponded item-by-item to the child and adolescent version was administered to parents of individuals with WS aged 18 years and above (Constantino & Todd, 2005). This version differed in that it included a number of items (specifically items 9, 15, 20, 21, 23, 36, 37, 43, 45, 49, 57, 58 & 63), which differed by wording or content in accordance with what is developmentally appropriate for adults. Internal consistency has been calculated at .95 (Constantino & Todd, 2005).

Procedure

Parents of individuals with WS were recruited from our existing research databases for individuals with the disorder. Parents chose to opt in to this specific phase of our larger research programmes, which had been given favourable ethical opinion by the local ethics committee. Parents provided their consent prior to participation. Parents were sent questionnaire packs containing the SRS and the SCAS-P scales to complete in their own time and to return to the researchers in a pre-paid envelope. For the direct testing (the BPVS and RCPM tasks) participants were tested either in their home, school or at the University where the research was being conducted. Participants were provided with an update about the project, including information sheet, at the end of the study.

Results

Parental insights of anxiety and social functioning

Raw scores are used in this analysis for both the SCAS-P and the SRS. There was a significant correlation between total score on the SCAS-P, as indication of overall anxiety, and social functioning as measured by the SRS total raw score (r=.362, $p<.01$). This positive correlation indicated that greater severity of anxiety was associated with a higher degree of social difficulty.

Chronological age was not significantly correlated with total anxiety score on the SCAS-P ($p=.77$). There was no significant difference in anxiety scores for the male and female individuals with WS ($t(57)=.30$, $p=.79$, $d=.08$). There was no significant association between verbal ability or non-verbal ability and anxiety scores (both p's >.05).

Chronological age was associated with SRS total raw score with greater chronological age associated with more proficient social functioning (lower SRS total score; r=-.47, $p<.001$[4]). There was no significant difference in SRS score between genders ($t(57)=.22$, $p=.84$, $d=.06$[5]) and SRS total score was not significantly correlated with verbal or non-verbal ability (both p's >.05).

For the sample as a whole, 10 individuals with WS were reported by parents to show social functioning within the normal range (17% of the sample). Fifteen individuals (25%) were classified in the mild-moderate deficits of reciprocal social behaviour that are clinically significant. Thirty-four individuals (58%) were classified by parents as showing severe deficits of reciprocal social interaction behaviour that will significantly impair everyday social functioning.

[4] This correlation remains significant if T scores are used instead of SRS raw scores r=-.49, $p<.001$

[5] We also calculated the gender difference using the T scores for the SRS as these have gender-specific norms and transformations. There was no difference between genders when using the T scores for the SRS Total $t(57)=1.05$, $p=.30$, $d=.27$

High-anxious versus low-anxious individuals and their social profiles

We split our WS sample according to their Anxiety score on the SCAS-P (see Table 1). Although there is no formal clinical cut-off for the SCAS-P, a score of 24 or above has been suggested as an indicator of clinical caseness, and has been used in previous research as an indication of the presence of anxiety in WS (Rodgers et al., 2012). A score of 24 is one standard deviation above the mean anxiety score of a typical community sample (Nauta et al., 2004; mean 14.2, SD 9.7). Using this cut-off, we classified 32 individuals (54%) as low-anxious and 27 as high-anxious (46% of the sample). Anxious and non-anxious groups did not differ significantly on chronological age ($t(57)=.296$, $p=.77$, $d=.07$). For those individuals for whom we had verbal ability scores (BPVS Raw Score, n=51) and nonverbal ability scores (RCPM, n=51) there was no difference between groups ($t(49)=.246$, $p=.81$, $d=.07$ and $t(49)=.421$, $p=.92$, $d=.03$ respectively). These data are presented in Table 1 showing the characteristics of the high- and low-anxious groups.

[Table 1]

We were particularly interested in the social functioning profile of our low-anxious and high-anxious individuals with WS. Total score on the SRS was significantly higher in the high-anxious group indicating greater severity of impairments in social functioning in those who had higher levels of anxiety (difference between groups $t(57)=2.23$, $p<.05$, $d=.57$; Table 1). This does not mean that the low-anxious individuals did not have social impairments. In the low-anxious group only 28% (9 individuals) were classified within the normal range for social functioning (Figure 1).

[Figure 1]

The high-anxious group showed significantly more impairment than the low-anxious group on social awareness ($t(57)=2.08$, $p<.05$, $d=.53$), social cognition ($t(57)=3.01$, $p<.01$, $d=.70$), and social

communication (t(57)=2.33, $p<.05$, $d=.57$). Importantly, the mean score for both groups across all categories was in the range indicative of impairment, yet where the groups differed was in the severity of the impairment. Groups did not differ on social motivation (t(57)=.981, $p=.33$, $d=.21$) or autistic mannerisms (t(57)=1.572, $p=.12$, $d=.47$). Figure 1 shows the higher proportion of individuals in the low-anxious group who fall within the normal range of functioning across the subscales (T scores are used here to indicate 'severity' of impairment).

Discussion

Anxiety

Parent reports indicated that 46% of the sample experienced high-anxiety. This is important in the context of a lack of current intervention options for anxiety associated with the specific needs of individuals with WS. The level reported here is similar to that reported elsewhere (Dodd & Porter, 2009; Dykens, 2003; Leyfer, Woodruff-Borden, Klein-Tasman, Fricke, & Mervis, 2006), emphasising that many individuals with WS are susceptible to high-anxiety, and that susceptibility is not related to age or intellectual level. Our mean level of anxiety in the high-anxious WS group of children and adults is above that of clinically anxious children (Nauta et al., 2004; mean 31.8 – our mean for high-anxious group 37.62, 19 WS individuals scored 32 or more, which is significantly different t(26)=2.46, $p<.05$[6]), indicating a clinical need comparable to clinically anxious individuals without WS. This level of anxiety is also similar to that reported elsewhere for children (but not adults) with ASD using the same measure (Rodgers et al., 2012, mean 35.07 child ASD, not significantly different $p=.49$; Gillott & Standen, 2007, mean 27.50 adult ASD, which is significantly different t(26)=4.78, $p<.01$).

[6] We note caution due to the developmental difference between our sample and those reported elsewhere against which we are testing.

These levels emphasise the need to use diagnostic measures to clearly evaluate anxiety needs in WS and develop clinical interventions to reduce high levels of anxiety. There has been a recent flurry of work on treatment and intervention options for anxiety associated with ASD, for example focusing on the use of CBT (Wood, Drahota, Sze, Har, Chiu, & Langer, 2009) which has highlighted the importance of the relationship between anxiety and social functioning in neurodevelopmental disorders. Recent exploratory clinical trials have suggested that CBT may help lessen anxiety symptoms in children with ASD (Sofronoff et al., 2005), but there has been no acknowledgement of a similar need of WS individuals. We acknowledge that formally diagnosing anxiety disorders in WS may be complicated by level of intellectual capacity, communication skills and deficits of social and emotional understanding, in fact these constraints are similar to those seen in ASD. The current work emphasises the necessity for targeted anxiety interventions in this group.

Social Behaviour

These data emphasise that social reciprocity is far from typical in WS. Many of these individuals have significant problems with social functioning which will impact upon their quality of life. Only 17% scored within the 'normal' range irrespective of anxiety level. Interestingly, a previous study using the SRS with 8-15 year olds with WS, also reported 17% of their sample to be in the normal range (Klein-Tasman et al., 2011). There is mounting evidence that across the developmental spectrum the vast majority of individuals with WS (here over 80%) show some form of difficulty with social functioning.

Klein-Tasman et al. (2011) have previously suggested that the SRS could be split into pro-social (social motivation and awareness) and social-cognition domains (communication and cognition). Their sample of 8-15 years olds with WS had more problems with social-cognition than pro-social functioning. This pattern is mirrored in the current data. A lack of socio-cognitive capacity linked with high social motivation tendencies (even when it is not 'appropriate' to engage in a social interaction) could be a combination that increases the social vulnerability of this group (Jawaid et al.,

2011) especially given their inappropriate social approach to unfamiliar people and lack of 'stranger danger' awareness (Riby, Kirk, Hanley, & Riby, 2013).

Importantly, despite any pro-social tendencies many individuals with WS struggle to function socially or to use appropriate reciprocal social behaviours. The strength of the wide age range used here is that social functioning difficulties are less pronounced in the older individuals. However, it is important to note that although we report a decrease in social difficulties with age, the social skills of adults with WS are still unlikely to be comparable to those of individuals with the same chronological age that are developing typically. Only 35% of adults in the current sample had social functioning in the normal range supporting this argument.

Finally, it is noted that over half (64%) of the individuals with WS in this sample fall in the severe range of the 'autistic mannerisms' subscale of the SRS (see Figure 1). This scale includes stereotypical behaviours and highly restrictive interests that are associated with autism, but interestingly are also seen in WS (Rodgers, Riby, Janes, Connolly, & McConachie, 2012; Janes, Riby & Rodgers, 2013). This subscale should be investigated further as these behaviours map onto the overlap between the clinical groups.

Linking anxiety and social behaviour in WS

The key question for the current study was whether there was a difference in the social profile of WS individuals with high- and low-anxiety. The current data suggest that high and low-anxious individuals with WS differ of aspects of social awareness, social motivation, and social communication (but not on social motivation or autistic mannerisms). Information of this nature is important when considering how best to support needs of individuals with the disorder. Although it isn't surprising that the split groups differ on general social functioning when these constructs are correlated in the current study, the pattern of differences across aspects of social expertise is particularly important. The data suggest a significant difference in social functioning between high-

anxious and low-anxious individuals with WS and that the groups differ on the problems they have with social abilities. Although there was variability of social functioning within both the high- and low-anxious groups, those who experienced higher anxiety, on average, showed more severe social dysfunction.

Future longitudinal studies examining the developmental trajectory of the anxiety in WS are clearly warranted and would allow consideration of the direction of the relationship between these aspects of behaviour. Indeed the wide age range of the current study is an advantage as we are able to see how anxiety and social behaviour might vary with age, however longitudinal research would be the optimal approach. Therefore targeted intervention strategies require consideration of the full WS anxiety profile and need to take a holistic view of the individual and their needs.

Limitations and future research

Our assessments of anxiety and social functioning are limited in that they rely on parent reports rather than formal clinical assessment. However, in formal clinical assessment it is frequently parental report across various measures that inform the assessment. One limitation of using parental insights on more than one measure, in the manner with which they are used here, is that there may be shared variance across the measures. This is a compromise when it is difficult to obtain other insights into these constructs in clinical populations. Considering the importance of who provides insight on questionnaire measures, recent research by Klein-Tasman and colleagues (2011) has shown that when exploring social behaviours in WS using the SRS and combining parents and teacher ratings, agreement is generally high, with no significant difference in ratings from these two sources. Indeed we are confident that the parents of the individuals with WS in our sample were able to adequately evaluate their everyday capabilities. Although some older individuals with WS might be able to give insights into their capabilities in some domains of functioning and in relation to their mental health (e.g. see information from WS adults, Stinton et al., 2012) the large age range meant that self-report would not have been possible across the whole cohort. Reassuringly though, in research on mental

health issues in adults with WS, Stinton and colleagues (2012) report a strong positive correlation between self-reports and informant reports (parents / carers) for adults with WS concerning their mental health, including anxiety. Therefore, although we acknowledge arguments that parental understanding of childhood anxiety may be incomplete (e.g. Hurtig et al., 2009) the links between the current findings and previous reports in the literature on anxiety and social functioning associated with WS provide reassurance. Future research may consider how best to combine these parental reports with every day (observational / clinical) insights into social / anxious behaviours.

One further issue regarding measurement is the use of the SCAS-P and the SRS with adults who have WS. Both these measures have been designed for use with children and not purposefully designed for use with adults. However, there is an inadequacy of alternative measures available to capture the issues addressed here with a population of adults who have intellectual difficulties. This is a problem inherent in research on intellectual disability and measures designed for use with children are frequently used with adults who have intellectual difficulties (relating directly to measurement of anxiety in adults with intellectual difficulty see the following studies that have used parent report on child measures, Dodd et al., 2005; Porter et al., 2009). Although the SCAS-P has been designed for use with children, the measure has previously been used with adults who have WS (Dodd et al., 2009) as well as for children with WS (Rodgers et al., 2012). Although the SRS has previously been used for young children with WS (Klein-Tasman et al., 2011) it has not, to our knowledge, been used with adults who have the disorder. The inclusion of the adult data (using parental reports on the adult version of the SRS and the parental reports using the SCAS-P) is important in the current study because adults with the disorder are often overlooked. Critically, there are no parallel measures of anxiety for adults and children with developmental disability that would have allowed us to use an alternative to that included in the current study. Furthermore, there are no appropriate self-report measures of either anxiety or social functioning that are appropriate for adults and children with reduced intellectual capacity as involved here. Therefore, we acknowledge that it is a compromise to use measures ideally suited to a different demographic than that we are researching.

Conclusions

Nearly half of the individuals with WS in the current study were classified as highly anxious by parent reports and over 80% showed deficits in social functioning. Individuals in the high-anxious group were significantly more impaired in their social skills than those in the low-anxious group. The current data emphasise the need for targeted anxiety intervention in WS that considers the whole individual, for example the relationship between anxiety and the WS social phenotype. Longitudinal research is required to adequately explore the profile of abilities across the developmental trajectory for individuals with WS who are high or low in anxiety.

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Table 1:

Participant characteristics for WS individuals with high-anxiety and low-anxiety (group mean with standard deviation in brackets)

	Low-Anxious (N=32)	**High-Anxious (N=27)**
Demographics		
Age	20.00 (11.49)	18.83 (7.52)
Gender (M:F)	17:15	13:14
BPVS Raw Score ^	93.38 (31.85)	95.35 (26.55)
RCPM Raw Score ^	17.38 (5.08)	17.00 (4.76)
SCAS-P		
Total Score	14.23 (5.53)	37.62 (9.82)
Panic	1.16 (1.39)	5.07 (1.39)
Separation anxiety	2.59 (1.91)	6.67 (3.06)
Physical injury fears	3.19 (2.22)	6.85 (2.66)
Social phobia	1.75 (1.68)	5.56 (3.07)
OCD	1.28 (1.90)	3.33 (2.24)
GAD	3.84 (2.34)	7.81 (2.90)
SRS Raw Scores		
Total Score	73.05 (38.99)	92.83 (28.83)
Social awareness	9.33 (4.85)	11.61 (3.64)
Social cognition	16.33 (7.12)	20.91 (5.85)
Social communication	22.05 (14.06)	29.09 (10.28)
Social motivation	9.77 (7.41)	11.30 (6.60)
Autistic mannerisms	15.57 (10.06)	19.91 (7.96)
SRS T Scores		
Total Score	71.91 (16.01)	81.33 (10.72)
Social awareness	63.94 (14.16)	70.96 (11.46)
Social cognition	74.75 (14.73)	83.04 (9.72)
Social communication	68.19 (14.06)	77.74 (12.58)
Social motivation	62.50 (13.04)	66.14 (15.15)
Autistic mannerisms	75.18 (14.71)	83.37 (10.97)

^ *BPVS and RCPM available for 51 individuals. (26, high-anxious, 25 low-anxious)*

Figure 1:

Distribution of classifications for the subscales of the SRS according to group status for anxiety using T scores to classify the severity of difficulty in each domain