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Examining reports of mental health in adults with Williams syndrome

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ABSTRACT

Prior research suggests that individuals with Williams syndrome (WS) have a disposition towards anxiety. Information regarding this is typically derived from parents and carers. The perspectives of the individuals with WS are rarely included in research of this nature. We examined the mental health of 19 adults with WS using explicit (psychiatric interview) and implicit (modified Stroop task) measures and compared informant (parents/carers) and respondent (adults with WS) reports of psychiatric symptoms. Informants and respondents both reported more symptoms of anxiety ($n = 7$–9) than depression ($n = 2$). Strong positive correlations were found between informant and respondent reports of symptoms of mental health problems. Compared to informants, respondents reported significantly more symptoms overall and somewhat more symptoms of anxiety. Results from the Stroop task indicated that the adults with WS were more vigilant to anxiety-related words than to depression-related words. The adults with WS provided reliable information regarding their mental health, thus providing further evidence that anxiety is part of the behavioural phenotype of the syndrome.

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1. Introduction

Williams syndrome (WS) is a genetic disorder caused by a deletion of genes on one copy of chromosome 7 (Ewart et al., 1993). It is associated with mild-to-moderate intellectual disability (ID) and a distinctive pattern of cognitive abilities that comprises weaknesses in aspects of spatial and number cognition and strengths in receptive vocabulary and face processing (Ansari et al., 2003; Farran, Jarrold, & Gathercole, 2003; Howlin, Davies, & Udwin, 1998; Paul, Stiles, Passarotti, Bavar, & Bellugi, 2002). Many individuals with WS display a behavioural phenotype that includes hypersociability, impulsivity and emotional difficulties (Davies, Udwin, & Howlin, 1998; Einfeld, Tonge, & Florio, 1997; Udwin & Yule, 1991). One of the most notable features of this behavioural phenotype is a disposition towards anxiety, with authors reporting significantly higher rates of anxiety among individuals with WS compared to those with Autism, Prader–Willi syndrome, ID of mixed/unknown aetiology and typically developing children (Dimitropoulos, Ho, Klaiman, Koenig, & Schultz, 2009; Dykens, 2003; Einfeld et al., 1997).

In recent years, research groups have sought to categorise the emotional and behavioural difficulties of individuals with WS in terms of diagnosable psychiatric disorders (e.g., Cherniske et al., 2004; Kennedy, Kaye, & Sadler, 2006; Leyfer, Woodruff-Borden, Klein-Tasman, Fricke, & Mervis, 2006; Stinton, Elison, & Howlin, 2010). These studies have supported prior assertions of high rates of anxiety, indicating that up to 54% of participants meet ICD/DSM diagnostic criteria for anxiety disorders (e.g., agoraphobia, generalised anxiety disorder and specific phobia). In addition to anxiety, a small number of

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studies has reported on affective disorders (e.g., bipolar disorder, depression and hypomania) and psychotic disorders (e.g., schizophrenia), indicating rates of 3–25% and 1–2%, respectively (e.g., Cherniske et al., 2004; Dodd & Porter, 2009; Kennedy et al., 2006; Stinton et al., 2010).

In many cases, information regarding the emotional difficulties and psychological states of individuals with WS has been derived from third party reports (i.e., "informants"), often with no attempt made to obtain information from the individuals with WS themselves (i.e., "respondents"). For example, Dodd and Porter (2009) state that 'as is standard procedure when assessing an intellectually impaired population, only caregivers were interviewed' (p. 95). Yet, there is a wealth of evidence indicating that individuals with ID (especially those with mild-to-moderate ID) are able to understand and provide meaningful information about their emotional states and experiences (e.g., Cuthill, Espie, & Cooper, 2003; Deb, Thomas, & Bright, 2001; Douma et al., 2006; Emerson, 2005; Lindsay et al., 1994; Matson, Kazdin, & Senatore, 1984). In addition, disregarding the perspective of the individuals themselves is likely to result in underdiagnosis of mental health problems due, for example, to diagnostic overshadowing and the necessity for information that can only be derived from respondents such as autonomic symptoms (Bramston & Fogarty, 2000; Moss, Prosser, Ibbotson, & Goldberg, 1996; Reiss, Levitan, & Szyszko, 1982). In the case of people with WS, while their range of abilities is broad, the vast majority of individuals have a mild ID and functional communication skills (Howlin, Elison, & Stinton, 2010; Martens, Wilson, & Reutens, 2008). Further, several studies have demonstrated that interviews and questionnaires about emotional difficulties can be successfully carried out with both adults and children with WS (e.g., Cherniske et al., 2004; Dykens, 2003; Freeman, Williams, Farran, & Brown, 2009; Kennedy et al., 2006; Stinton et al., 2010). These studies have highlighted the importance of including both informant and respondent reports. For example, using the revised Fear Survey Schedule for Children (Ollendick, King, & Frary, 1989), Dykens (2003) found that individuals with WS reported significantly more fears than their parents and Stinton et al. (2010) reported that anxiety disorders were more commonly derived from psychiatric interviews conducted with adults with WS than with their parents/carers, with the opposite true for depressive disorders. While limited agreement between informant and respondent reports of difficulties is common, the simple presence of this difference cannot be assumed to indicate that respondent reports are less reliable than informant reports. Differences are likely to reflect their different perspectives.

The finding that different individuals provide information that might result in very different interpretations of symptoms presents a considerable challenge in research and clinical practice. To better understand this it is necessary to examine mental health using alternative methodologies. One way in which mental health can be examined experimentally is through the use of emotional Stroop tasks. In the original format of this task, participants were presented with a series of items (words or meaningless letter strings such as XXXX) printed in various colours and were asked to name the colour that the stimuli were printed in while attempting to ignore the stimulus itself. Stroop (1935) found that participants were slower to name items that were incongruent (i.e. the word red written in blue) than those that were congruent (i.e. the word green written in green). This effect has been consistently observed in both typically developing individuals and individuals with ID (Das, 1970; Ellis, Woodley-Zanthos, Dulaney, & Palmer, 1989; Williams, Mathews, & MacLeod, 1996). More recent research has demonstrated that it is not only incongruent word/colour items that produce interference. Colour naming is significantly slower for clinically relevant emotional words (e.g., scared) than for neutral words (e.g., square) among clinical groups (i.e. those with anxiety or affective disorders) but not among control groups who do not have the given disorder (Mathews, Mogg, Kentish, & Eysenck, 1995; Mitterschiffthaler et al., 2008). As such, the Stroop task presents an opportunity to measure emotional problems implicitly and hence without the sorts of biases that are evident in reports obtained via interviews and questionnaires.

The present study examines the mental health of adults with WS via both explicit (psychiatric interview) and implicit (Stroop) measures. The Stroop task included anxiety-related (e.g., "alone"), depression-related (e.g., "crying"), and neutral words (e.g., "paper"). Convergence between explicit and implicit measures completed by the adults with WS would indicate that their reports are accurate, whereas a significant degree of divergence would suggest that their reports are inaccurate. These reports by the adults with WS were also compared to informant reports by their parents or carers. Convergence between respondent and informant reports would further support the adults with WS's ability to provide meaningful information about their own emotional states and experiences.

2. Methods

2.1. Recruitment

Potential participants were identified via the Williams Syndrome Foundation's database of research participants ($n = 110$). Inclusion criteria were: the individual with WS was aged 18 years or older, their diagnosis had been confirmed by genetic testing, they had a parent or other person who knew them well, agreement to participate from both parties. Exclusion criteria were: inability to read simple words and colour blindness.

2.2. Participants

The response rate to recruitment letters was 28.2% ($n = 31$). Of these families, 12 (38.7%) were excluded from the study for the following reasons: the adult with WS was not able to provide sufficient information to questions regarding their mental health ($n = 2$), the adult with WS was unable to complete the Stroop task ($n = 5$), the family withdrew before data collection

began ($n = 4$). In addition, data from 1 participant was lost due to experimenter error. The final sample comprised 19 adults with WS (9 female, 10 male) and their parents ($n = 17$) or carers ($n = 2$). The mean age of the adults with WS was 32 years (range = 20–42 years).

2.3. Materials and design

2.3.1. Stroop task

Stimuli used for the Stroop task were derived from word lists generated by primary school aged children, a level comprehendible to most adults with WS (Howlin et al., 1998), and categorised according to their emotionality (Neshat-Doost, Moradi, Taghavi, Yule, & Dalgleish, 1999). Stimuli from the lists were chosen for 3 categories for the present study (anxiety-related, depression-related and neutral). In addition, words in each of the groups were matched by the number of letters and syllables they contained. A full list of the words used is provided in Appendix A.

2.3.2. Practice trials – colours

To examine whether the adults with WS were able to accurately identify the colours used in the experimental testing phase, they were presented with a practice trial consisting of four individual blocks of colours (blue, grey, red and yellow). They were asked to name the colours. The order of presentation of the colours was randomised by the experimenter.

2.3.3. Practice trials – words

To examine whether the adults with WS were able to read and comprehend the words used in the study, they were shown a selection of words (one at a time), which they were asked to read and to indicate what the word meant. Each participant was shown one word from each of the three word categories (anxiety, depression and neutral). Words were chosen and presented in a random order by the experimenter. Participants additionally completed another series of practice trials on a computerized Stroop task, as described next.

2.3.4. Experimental trials

Participants were presented with 12 practice trials followed by 84 experimental trials (3 word categories $\times$ 7 words $\times$ 4 colours) that were divided into 4 blocks. Practice trials included 2 words from each category, with each word presented twice in different colours, and all four colours appearing equally often. Experimental stimuli consisted of each of the 21 words presented in each of the four colours (i.e., blue, grey, red and yellow). These 84 experimental trials were presented in four blocks of 21 trials each. Six stimulus lists were created with six different random orders of stimuli, subject to the constraints that (a) each of the 21 words appeared once per block, and (b) each colour was approximately equally likely within each block. After each block of trials participants were offered an opportunity to have a break from testing. Participants were randomly assigned to stimulus lists.

Each trial began with a white fixation (+) centred on a black background. The fixation remained onscreen until the experimenter pressed a button on a computer mouse whenever the participant was ready to proceed. This initial mouse click was followed by a half-second delay, and then the target word appeared in the centre of the display in 36-point bold font in one of the four colours. Participants were asked to indicate, via a verbal response, the colour of the word that appeared on the screen. Responses were registered by the experimenter who pressed a button on the computer mouse as soon as a correct response was given. This approach reduced the task's memory load and avoided any response biases that might be caused by fine motor difficulties. Stimuli remained on the computer screen until a correct response had been registered. Participants were prompted if they produced an incorrect response, such as naming the word rather than the colour. The task was administered via E-Prime experimental software operating on a laptop computer with a 12-in. display.

2.3.5. Psychiatric assessment

Mental health was assessed using the Psychiatric Assessment Schedule for Adults with Developmental Disabilities (PAS-ADD; Moss, Goldberg, et al., 1996). The PAS-ADD is described in detail in (Costello, Moss, Prosser, & Hatton, 1997 and Moss et al. (1997)). In brief, the PAS-ADD is a semi-structured interview that is designed for use with adults with a range of IDs. Interviews are conducted separately with both the respondent and an informant, with symptoms and diagnoses derived either from the interviews in isolation or from a combination of the two. The PAS-ADD has good reliability (Costello et al., 1997) in terms of individual items ($\kappa = .65$), total score ($\kappa = 74$) and clinical significance of symptoms ($\kappa = 70$). Validity has been demonstrated in comparisons between PAS-ADD diagnoses and those of psychiatrists, which indicate agreement in 75% of cases (Moss et al., 1997).

In the present research, three subscales of the PAS-ADD were used: anxiety, depression and total score. Each of the scores was derived from the number of symptoms that were reported for each scale.

2.4. Procedure

Assessments took place in the homes of the participants and at the University of Warwick. The order of presentation of measures was: Stroop task, informant PAS-ADD, respondent PAS-ADD. This provided the adults with WS with a break between assessments. Informant and respondent assessments were conducted separately. The testing session lasted 1–2 h.

Table 1
Intercorrelations of anxiety, depression, and total psychiatric disorder among individuals with Williams syndrome, as rated by informants and respondents.

	1	2	3	4	5	6
1. Self anxiety	–					
2. Self depression	.71***	–				
3. Self total	.97***	.85***	–			
4. Carer anxiety	.78***	.74***	.82***	–		
5. Carer depression	.40†	.86***	.59**	.58**	–	
6. Carer total	.68***	.88***	.80***	.90***	.85***	–

† $p < .10$.
** $p < .01$.
*** $p < .001$.

2.5. Data analysis

Following standard procedures, the data from the Stroop colour naming task were trimmed and transformed prior to analysis. Response times from trials in which the participant responded incorrectly were removed from all analyses (1% of trials). Outlying response times that were more than 2.5 standard deviations beyond the mean, calculated separately for each participant and each condition, were also removed from all analyses (2% of trials). Finally, because response time distributions are typically skewed positively, the remaining trimmed correct response times were log transformed to minimize this skew. Error rates were also extremely skewed, with many participants committing no errors in some conditions. Because log and square root transformations failed to fully correct the positive skew, error rates were analyzed via nonparametric statistics.

3. Results

As shown in Table 1, the evaluations of anxiety, depression, and other psychiatric disorders by the individuals with WS and by their carers were strongly and positively intercorrelated. That is, individuals who rated themselves highly on anxiety also tended to rate themselves highly on depression and other disorders, and their carers also tended to rate them highly on those same symptoms. Conversely, individuals who rated themselves relatively low on one disorder also rated themselves low on other disorders, and their carers rated them similarly low. This strong multicollinearity, particularly that between the evaluations of the individuals with WS and the carers, suggests that the two parties generally converged in their evaluations of the emotional states of the individuals with WS.

The intercorrelations reported in Table 1 indicate convergence between the reports of the individuals with WS and carers' *relative* ratings of anxiety and depression, but they are uninformative of *absolute* ratings. For instance, a strong positive correlation between anxiety and depression ratings does not necessarily indicate that the two conditions are equally important or prevalent. Indeed, as illustrated in Fig. 1, anxiety ratings differed substantially from depression ratings, and the individuals with WS and their carers also diverged systematically in their evaluations. PAS-ADD scores were analyzed via a 2 (Rater: self, carer) × 3 (Scale: anxiety, depression, total) repeated measures analysis of variance. The main effect of Scale was significant, $F(2, 36) = 41.44$, $p < .001$, with highest scores on the total measure and lowest scores on depression. This observation is unsurprising, given that total scores are a sum of the anxiety, depression, and other disorder scores. More

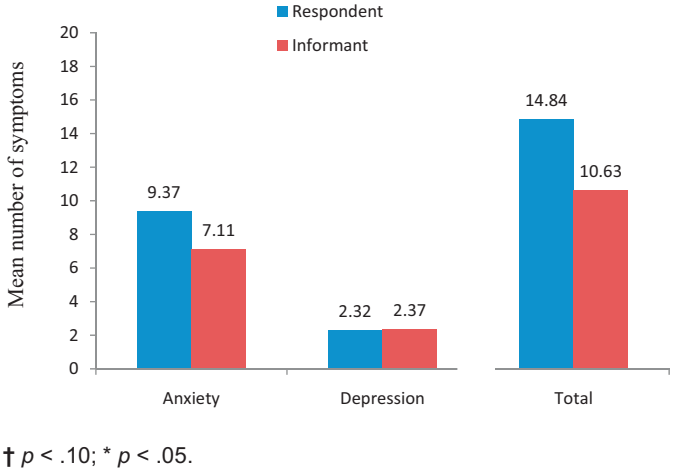

Fig. 1. Anxiety, depression, and total psychiatric disorder ($M + SE$) among individuals with Williams syndrome, as rated by informants and respondents.

importantly, the main effect of Rater was also significant, $F(1, 18) = 4.95$, $p < .05$, with higher scores overall among respondent ratings than among informant ratings. However, these main effects were qualified by a highly significant interaction, $F(2, 36) = 8.80$, $p < .001$. Relative to informant ratings, respondent ratings were marginally higher for anxiety [$t(18) = 1.91$, $p = .07$] and significantly higher for total scores [$t(18) = 2.83$, $p < .05$] but were virtually identical for depression ($p = .90$). Thus, individuals with WS and their carers agreed that those individuals experience relatively low levels of depressive symptoms, and they also agreed that the individuals experience relatively high levels of anxiety, but the individuals and their carers disagreed on just how severe the anxiety and other psychiatric disorders are. The individuals themselves reported more symptoms of anxiety and other psychiatric disorders. This finding corroborates prior demonstrations of differential evaluations by WS individuals and their carers (Dykens, 2003; Stinton et al., 2010), and in so doing, it also supports the representativeness of the current sample.

Performance on the Stroop task was generally good. Response times are illustrated in Panel A of Fig. 2. Planned comparisons revealed that anxiety-related words elicited significantly slower responses than depression-related words, $t(18) = 2.06$, $p = .05$. Response times to the anxiety- and the depression-related words did not differ significantly from the neutral words (both $p > .28$). Error rates, which are illustrated in Panel B of Fig. 2, were very low. Error rates were analyzed via the nonparametric Wilcoxon signed-rank test. As evident in the figure, the pattern of error rates (Fig. 2B) was qualitatively identical to the pattern of response times (Fig. 2A). Error rates were marginally lower for depression-related words than for both anxiety-related words ($Z = 1.63$, $p = .10$) and neutral words ($Z = 1.84$, $p = .07$), which did not differ from one another ($p = .95$). Thus, together the response times and error rates indicate that WS individuals were more vigilant for anxiety-related words than for depression-related words. This implicit measure of emotional processing (Fig. 2) therefore is consistent with our prior finding of greater symptoms of anxiety than depression among these individuals (Fig. 1).

We additionally tested whether any of the six explicit ratings (2 raters × 3 disorder scores) illustrated in Fig. 1 correlated significantly with any of the six implicit measures (3 word-types × 2 dependent measures) shown in Fig. 2. None of the ratings significantly correlated with any of the response times or error rates (all $p > .13$). Finally, we also examined via multiple linear regression whether the independent ratings (i.e., self-anxiety, self-depression, carer-anxiety, carer-depression)

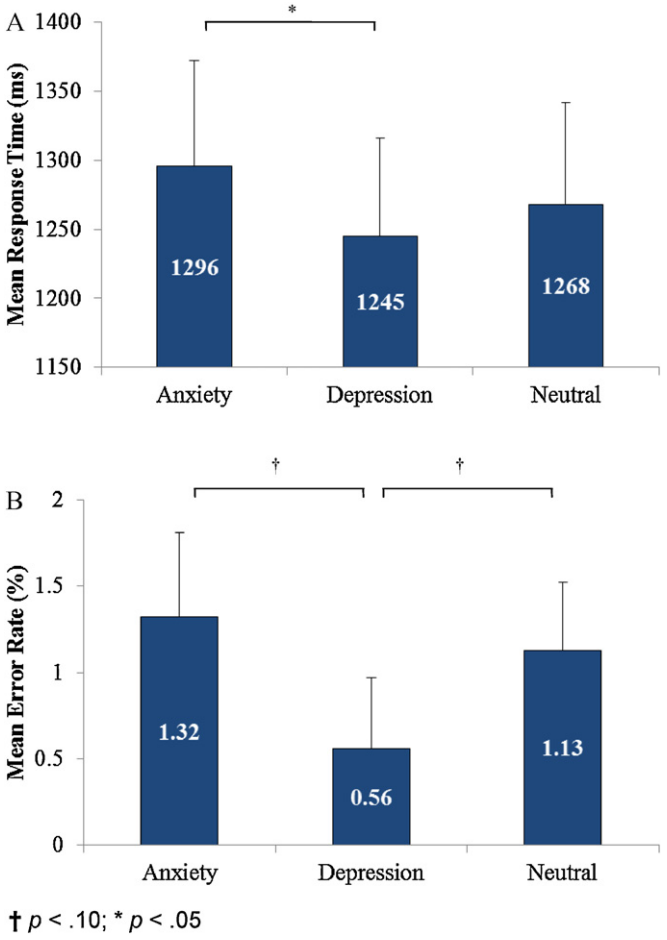

Fig. 2. Response times (Panel A) and error rates (Panel B) to anxiety-related, depression-related, and neutral words ($M + SE$) among individuals with Williams syndrome.

collectively predicted response times or error rates to anxiety-related or depression-related words. None of the four regression models approached significance (all $p > .15$). The null relationship between the explicit evaluations and the implicit behavioural measures might be due to low statistical power, so conclusions regarding this relationship should be considered with caution.

4. Discussion

4.1. Summary of research

This study adds to the limited knowledge of mental health in adults who have WS. It is one of few studies to include the experiences and perspectives of individuals with WS. It is also one of few to directly compare reports of carers/parents and those of individuals with WS themselves, and it is the first to examine mental health in WS using explicit and implicit methods. The results suggest several important conclusions. First, informant and respondent reports indicate generally similar evaluations of the emotional experiences of adults with WS. Second, the individuals with WS and their carers agreed that those individuals experience relatively lower levels of depression, and they also agreed that the individuals experience relatively high levels of anxiety, though the individuals and their carers disagreed on the severity of that anxiety and total psychiatric symptoms. And third, individuals with WS responded significantly more slowly and committed marginally more errors to anxiety-related words than to depression-related words, suggesting greater interference by anxiety-related stimuli.

4.2. Anxiety and depression in Williams syndrome

Anxiety is a frequently cited emotional difficulty of individuals with WS. At the uppermost limits, 89% of individuals with WS are reported to be anxious to some degree (Davies et al., 1998) while 54% meet diagnostic criteria for at least one type of anxiety disorder (Leyfer et al., 2006). Rates of anxiety disorders are far higher for people with WS than typically developing individuals or other people who have ID (e.g., Deb et al., 2001; Kessler et al., 1994). The present study supports this in several ways; all but 2 of the participants had at least 1 symptom of anxiety, the average number of symptoms reported was high (7–9), the number of symptoms of anxiety was 3–4 times higher than the number of symptoms of depression and, finally, anxiety-related words caused more interference on the Stroop task than depression-related words. The disposition towards anxiety observed in the present study is comparable to other studies that have administered the PAS-ADD. Costello et al. (1997) conducted psychiatric interviews with 40 adults who had an ID and who had been selected specifically because they were thought to have a psychiatric disorder. They reported a mean number of symptoms of 3.84 for depression and 8.58 for anxiety, with a total symptom score of 21.32. This compares to the self-reported symptom scores of 2.32 (depression), 9.37 (anxiety) and 14.84 (total) for adults with WS. The participants in our study were recruited via a support group on the basis of age and genetic confirmation of WS rather than via psychological services or on the basis of having emotional difficulties. Thus, a nonclinical sample of adults WS was more anxious than a sample of adults who were recruited specifically because they were suspected to have mental health problems.

Relatively to anxiety, other psychiatric disorders are much less common in WS. However, depressive disorders might also be an area of concern, especially during adulthood, with 3–25% of individuals with WS meeting diagnostic criteria (Cherniske et al., 2004; Dodd & Porter, 2009; Kennedy et al., 2006; Stinton et al., 2010). In the majority of cases, the rates of depressive disorders in WS are at or above those reported for other individuals with ID (6.6%, Cooper et al., 2007) or for the general population (10%; Kessler et al., 1994). The present study produced equivocal results regarding low mood in adults with WS. All but one of our participants reported one symptom or more of depression (of at least moderate severity), with the vast majority reporting between 1 and 3. However, only 2 participants reported any significant issues relating to mood and only 1 had ever been diagnosed with depression (giving a rate of depression in the sample of 5%).

There is some evidence to suggest that depression might be a greater issue during adulthood for people with WS. For example, while the rates of depression among children are low (3%), many more adults (9–25%) receive a diagnosis of a depressive disorder (Cherniske et al., 2004; Dodd & Porter, 2009; Stinton et al., 2010). It is unsurprising that depression might emerge as an issue during adulthood. In the typically developing population, children with clinically significant levels of anxiety are more likely than nonanxious children to be depressed as adults and are likely to have worse outcomes, e.g., the severity of their depression will be worse and depressive episodes will occur more frequently and last longer (Beesdo et al., 2007; Pine, Cohen, Gurley, Brook, & Ma, 1998). While longitudinal research is required to draw any firm conclusions, the high rates of depressive disorders seen in adults with WS relative to children might indicate a similar trajectory to that seen in the typically developing population. Given that anxiety of a clinical severity has been reported in WS from at least the age of 4 and that, for many people, persists throughout life, intervention at an early age should be seen as a priority.

4.3. Informant and respondent reports of mental health

In the majority of studies regarding the mental health of individuals with WS, information is obtained from parental reports, and individuals with WS are typically excluded from the process. Due consideration is rarely given to research regarding the importance of involving the individuals themselves in this process or to studies that have investigated self-reports by individuals with WS. A key argument against the inclusion of self-reports in research with people with ID is

the possibility of acquiescence biases (Heal & Selegman, 1995; Moss, Costello, & Prosser, 2000). Contrary to this argument, however, Dykens (2003) found little or no evidence of acquiescence by individuals with WS, and other studies have demonstrated that participation in research regarding mental health is possible for the vast majority of individuals with WS. For example, Cherniske et al. (2004) and Stinton et al. (2010) have reported that only 2–5% of their samples of adults with WS were unable to provide information about psychological states. These studies also highlight important differences between informant and respondent reports of mental health, with anxiety reported more frequently by respondents and depression reported more frequently by informants (Dykens, 2003; Stinton et al., 2010). This likely reflects the different access of the 2 parties to the different elements of mental health problems. For example, autonomic symptoms (a key component of anxiety disorders) are derived more readily from respondent interviews than from informant interviews (Moss, Prosser, et al., 1996). Anecdotal evidence of the difficulties that informants have in identifying autonomic symptoms of anxiety is provided by Stinton et al. (2010).

Our study broadly supports the findings of prior research. First, from an initial sample of 27, all but 2 (7%) were able to provide sufficient information to complete the psychiatric assessment (cf. Cherniske et al., 2004; Stinton et al., 2010). Second, respondents reported more symptoms of anxiety than informants did (cf. Dykens, 2003; Stinton et al., 2010). And third, informant and respondent reports were highly correlated (cf. Freeman et al., 2009). However, unlike Stinton et al. (2010), we found no significant difference between reports of symptoms of depression. This might be due to our small sample size. Overall, the present results clearly indicate that most individuals with WS are able to actively participate in research of this nature, and moreover, their self-reports accurately convey their emotional states.

4.4. Methodological limitations

This study has several limitations. The sample was relatively small. While this is typical of research regarding rare syndrome groups, a consequence is low statistical power. This might explain the null relationship between participants' explicit reports of mental health and their implicit measures of emotional processing, for instance. Another potential consequence is limited generalisability of these results to other individuals with WS, although this sample does appear to be representative of individuals with WS. This sample exhibited comparable rates of difficulties reading (Howlin et al., 1998) and assessing their own mental health (Cherniske et al., 2004; Stinton et al., 2010) as in prior studies of WS adults. Due to its difficulty, the Stroop task was a further limitation. To increase the potential for participation by less able adults with WS, alternative versions of this task could be employed. These might include using picture stimuli or emotional faces rather than words (e.g., Pishyar, Harris, & Menzies, 2004; Reinholdt-Dunne, Mogg, & Bradley, 2009).

4.5. Future directions

In research that aims to understand better the difficulties faced by individuals who have WS and ID, there is often an assumption that the information about internal states that is provided by carers is the 'gold standard'. Consequently, the individuals themselves are often not directly involved. Thus, such research is conducted *on* people with ID rather than *with* them. While it is clear that there is considerable heterogeneity of abilities among individuals who have WS (Porter & Coltheart, 2005) and that in some circumstances it will be necessary to rely on informant reports to obtain the most accurate information (Matson et al., 1984), efforts should be made to foster genuine inclusion and participation by individuals with WS. One possible approach to this is a line along those presented in the United Kingdom's Mental Capacity Act (2005). In this, there is an assumption that an individual has capacity and is able to make decisions for her or himself. Only when all practical steps have been taken to establish that they are not able to do so are decisions made on their behalf. Thus, for the purposes of research a more reasonable starting position should be that individuals with WS (and others with ID) are presumed to be able to actively participate and provide meaningful information about themselves rather than being automatically excluded. The onus, therefore, is on researchers to facilitate the inclusion of people with disabilities in research that seeks to elucidate their lives. This would involve employing appropriate assessment measures (of which there are many, see Unwin & Deb, 2008, for a review) and ensuring that researchers have undergone training in the skills required to enhance the participation of the individuals themselves.

5. Conclusions

This study demonstrated that regardless of the assessment measures used, problems related to anxiety are more common than other types of mental health problems for adults with WS. While explicit evaluations of mental health differed in terms of the severity of problems, both adults with WS and their parents and carers correctly identified the emotional difficulties of those participants with WS. This indicated that adults with WS are reliable reporters of their psychological states.

Acknowledgements

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Appendix A. Experimental stimuli

Anxiety	Depression	Neutral
Alone	Afraid	Ball
Dark	Bomb	Bird
Dies	Bully	Monkey
Down	Crying	Paper
Lonely	Dead	Pencil
Spider	Lost	Table
Upset	Worry	Tree

References

Ansari, D., Donlan, C., Thomas, M. S. C., Ewing, S. A., Peen, T., & Karmiloff-Smith, A. (2003). What makes counting count? Verbal and visuo-spatial contributions to typical and atypical number development. *Journal of Experimental Child Psychology, 85*, 50–62.

Beesdo, K., Bittner, A., Pine, D. S., Stein, M. B., Höfler, M., Lieb, R., et al. (2007). Incidence of social anxiety disorder and the consistent risk for secondary depression in the first three decades of life. *Archives of General Psychiatry, 64*, 903–912.

Bramston, P., & Fogarty, G. (2000). The assessment of emotional distress experienced by people with an intellectual disability: A study of different methodologies. *Research in Developmental Disabilities, 21*, 487–500.

Cherniske, E. M., Carpenter, T. O., Klaiman, C., Young, E., Bregman, J., Insogna, K., et al. (2004). Multisystem study of 20 older adults with Williams syndrome. *American Journal of Medical Genetics Part A, 131A*, 255–264.

Cooper, S. A., Smiley, E., Morrison, J., Williamson, A., & Allan, L. (2007). Mental ill health in adults with intellectual disabilities: prevalence and associated factors. *British Journal of Psychiatry, 190*, 27–35.

Costello, H., Moss, S., Prosser, H., & Hatton, C. (1997). Reliability of the ICD-10 version of the Psychiatric Assessment Schedule for Adults with Developmental Disability (PAS-ADD). *Social Psychiatry and Psychiatric Epidemiology, 32*, 339–343.

Cuthill, F. M., Espie, C. A., & Cooper, S-A. (2003). Development and psychometric properties of the Glasgow Depression Scale for people with a Learning Disability: Individual and carer supplement versions. *British Journal of Psychiatry, 182*, 347–353.

Das, J. P. (1970). Changes in Stroop test responses as a function of mental age. *British Journal of Social & Clinical Psychology, 9*, 68–73.

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.

Deb, S., Thomas, M., & Bright, C. (2001). Mental disorder in adults with intellectual disability. I: Prevalence of functional psychiatric illness among a community-based population aged between 16 and 64 years. *Journal of Intellectual Disability Research, 45*, 495–505.

Dimitropoulos, A., Ho, A., Klaiman, C., Koenig, K., & Schultz, R. (2009). A comparison of behavioral and emotional characteristics in children with Autism, Prader–Willi syndrome, and Williams syndrome. *Journal of Mental Health Research in Intellectual Disabilities* .

Dodd, H. F., & Porter, M. A. (2009). Psychopathology in Williams syndrome: The effect of individual differences across the life span. *Journal of Mental Health Research in Intellectual Disabilities, 2*, 89–109.

Douma, J. C. H., Jolanda, C. H., Dekker, M. C., Marielle, C., Verhulst, F. C., & Koot, H. M. (2006). Self-reports on mental health problems of youth with moderate to borderline intellectual disabilities. *Journal of the American Academy of Child and Adolescent Psychiatry, 45*, 1224–1231.

Dykens, E. M. (2003). Anxiety, fears, and phobias in persons with Williams syndrome. *Developmental Neuropsychology, 23*, 291–316.

Einfeld, S. L., Tonge, B. J., & Florio, T. (1997). Behavioral and emotional disturbance in individuals with Williams syndrome. *American Journal on Mental Retardation, 102*, 45–53.

Ellis, N. R., Woodley-Zanthos, P., Dulaney, C. L., & Palmer, R. L. (1989). Automatic-effortful processing and cognitive inertia in persons with mental retardation. *American Journal on Mental Retardation, 93*, 412–423.

Emerson, E. (2005, March). Use of the Strengths and Difficulties Questionnaire to assess the mental health needs of children and adolescents with intellectual disabilities. *Journal of Intellectual & Developmental Disability, 30*(1), 14–23.

Ewart, A. K., Morris, C. A., Atkinson, D., Jin, W. S., Sternes, K., Spallone, P., et al. (1993). Hemizygosity at the Elastin Locus in A Developmental Disorder, Williams-Syndrome. *Nature Genetics, 5*, 11–16.

Farran, E. K., Jarrold, C., & Gathercole, S. E. (2003). Divided attention, selective attention and drawing: Processing preferences in Williams syndrome are dependent on the task administered. *Neuropsychologia, 41*, 687–696.

Freeman, K., Williams, T., Farran, E., & Brown, J. (2009). Williams syndrome: The extent of agreement between parent and self-report of psychological difficulties. *European Journal of Psychiatry, 24*, 167–175.

Heal, L. W., & Selegman, C. K. (1995). Response biases of interviews with individuals with limited mental abilities. *Journal of Intellectual Disability Research, 39*, 245–250.

Howlin, P., Davies, M., & Udwin, O. (1998). Cognitive functioning in adults with Williams syndrome. *Journal of Child Psychology and Psychiatry, 39*, 183–189.

Howlin, P., Elison, S., & Stinton, C. (2010). Cognitive, linguistic and adaptive functioning in Williams syndrome: Trajectories from early to middle adulthood. *Journal of Applied Research in Intellectual Disabilities, 23*, 322–336.

Kennedy, J. C., Kaye, D. L., & Sadler, L. S. (2006). Psychiatric diagnoses in patients with Williams syndrome and their families. *Jefferson Journal of Psychiatry, 20*, 22–31.

Kessler, R. C., McGonagle, K. A., Zhao, S., Nelson, C. B., Hughes, M., Eshleman, S., et al. (1994). Lifetime and 12-month prevalence of DSM-III-R psychiatric disorders in the United States: Results from the national comorbidity survey. *Archives of General Psychiatry, 51*, 8–19.

Leyfer, O. T., Woodruff-Borden, J., Klein-Tasman, B. P., Fricke, J. S., & Mervis, C. B. (2006). Prevalence of psychiatric disorders in 4 to 16-year-olds with Williams syndrome. *American Journal of Medical Genetics Part B-Neuropsychiatric Genetics, 141B*, 615–622.

Lindsay, W. R., Michie, A. M., Baty, F. J., Smith, A. H., & Miller, S. (1994). The consistency of reports about feelings and emotions from adults with mild intellectual disability. *Journal of Intellectual Disability Research, 45*(2), 106–114.

Martens, M. A., Wilson, S. J., & Reutens, D. C. (2008). Research review: Williams syndrome: A critical review of the cognitive, behavioral, and neuroanatomical phenotype. *Journal of Child Psychology and Psychiatry and Allied Disciplines, 49*, 576–608.

Mathews, A., Mogg, K., Kentish, J., & Eysenck, M. (1995). Effect of psychological treatment on cognitive bias in generalized anxiety disorder. *Behavior Research and Therapy, 33*, 293–303.

Matson, J. L., Kazdin, A. E., & Senatore, B. (1984). Psychometric properties of the Psychopathology Instrument for Mentally Retarded Adults. *Applied Research in Mental Retardation, 5*, 81–90.

Mental Capacity Act. (2005). *Code of practice*. London: TSO. Accessed 22.08.11 www.dca.gov.uk/legal-policy/mentalcapacity/mca-cp.pdf.

Mitterschiffthaler, M. T., Williams, S. C. R., Walsh, N. D., Cleare, A. J., Donaldson, C., Scott, J., et al. (2008). Neural basis of the emotional Stroop interference effect in major depression. *Psychological Medicine, 38*, 247–256.

Moss, S., Costello, H., & Prosser, H. (2000). Detecting and assessing mental health problems. In G. Holt & N. Bouras (Eds.), *Mental health in learning disabilities: A training pack for staff working with people who have a dual diagnosis of mental health needs and learning disabilities* (2nd ed., pp. 27–36). Brighton: Pavillion Publishing.

Moss, S., Goldberg, D., Patel, P., Prosser, H., Ibbotson, B., Simpson, N., et al. (1996). *The psychiatric assessment schedule for adults with developmental disabilities*. Manchester, England: University of Manchester, Hester Adrian Research Centre.

Moss, S., Ibbotson, B., Prosser, H., Goldberg, D., Patel, P., & Simpson, N. (1997). Validity of the PAS-ADD for detecting psychiatric symptoms in adults with learning disability (mental retardation). *Social Psychiatry and Psychiatric Epidemiology, 32*, 344–354.

Moss, S., Prosser, H., Ibbotson, B., & Goldberg, D. (1996). Respondent and informant accounts of psychiatric symptoms in a sample of patients with learning disability. *Journal of Intellectual Disability Research, 40*, 457–465.

Neshat-Doost, H., Moradi, A., Taghavi, R., Yule, W., & Dalgleish, T. (1999). The development of a corpus of emotional words produced by children and adolescents. *Personality and Individual Differences, 27*, 433–451.

Ollendick, T. H., King, N. J., & Frary, R. B. (1989). Fears in children and adolescents: Reliability and generalizability across age, gender, and nationality. *Behaviour Research and Therapy, 27*, 19–26.

Paul, B. M., Stiles, J., Passarotti, A., Bavar, N., & Bellugi, U. (2002). Face and place processing in Williams syndrome: Evidence for a dorsal–ventral dissociation. *Neuroreport, 13*, 1115–1119.

Pine, D. S., Cohen, P., Gurley, D., Brook, J., & Ma, Y. (1998). The risk for early-adulthood anxiety and depressive disorders in adolescents with anxiety and depressive disorders. *Archives of General Psychiatry, 55*, 56–64.

Pishyar, R., Harris, L. M., & Menzies, R. G. (2004). Attentional bias for words and faces in social anxiety. *Anxiety, Stress & Coping: An International Journal, 17*, 23–36.

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

Reinholdt-Dunne, M. L., Mogg, K., & Bradley, B. P. (2009). Effects of anxiety and attention control on processing pictorial and linguistic emotional information. *Behaviour Research and Therapy, 47*, 410–417.

Reiss, S., Levitan, G. W., & Szyszko, J. (1982). Emotional disturbance and mental-retardation—Diagnostic overshadowing. *American Journal of Mental Deficiency, 86*, 567–574.

Stinton, C., Elison, S., & Howlin, P. (2010). Mental health problems in adults with Williams syndrome. *American Journal on Intellectual and Developmental Disabilities, 115*, 3–18.

Stroop, J. R. (1935). Studies of interference in serial verbal reactions. *Journal of Experimental Psychology, 28*, 643–662.

Udwin, O., & Yule, W. (1991). A cognitive and behavioural phenotype in Williams syndrome. *Journal of Clinical and Experimental Neuropsychology, 13*, 232–244.

Unwin, G., & Deb, S. (2008). Psychiatric and behavioural assessment scales for adults with learning disabilities. *Advances in Mental Health and Learning Disabilities, 2*, 37–45.

Williams, J. M. G., Mathews, A., & MacLeod, C. (1996). Emotional Stroop task and psychopathology. *Psychological Bulletin, 120*(1), 3–24.