J Autism Dev Disord (2011) 41:341–351
DOI 10.1007/s10803-010-1060-5

ORIGINAL PAPER

Honing in on the Social Phenotype in Williams Syndrome Using Multiple Measures and Multiple Raters

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Published online: 8 July 2010

Abstract The behavioral phenotype of Williams syndrome (WS) is characterized by difficulties with establishment and maintenance of friendships despite high levels of interest in social interaction. Here, parents and teachers rated 84 children with WS ages 4–16 years using two commonly-used measures assessing aspects of social functioning: the Social Skills Rating System and the Social Responsiveness Scale. Mean prosocial functioning fell in the low average to average range, whereas social reciprocity was perceived to be an area of significant difficulty for many children. Concordance between parent and teacher ratings was high. Patterns of social functioning are discussed. Findings highlight the importance of parsing the construct of social skills to gain a nuanced understanding of the social phenotype in WS.

Keywords Williams syndrome · Social functioning · SRS · SSRS

Introduction

Williams syndrome is a relatively rare genetic neurodevelopmental disorder with an estimated prevalence rate of one in 7,500 births (Stromme et al. 2002), resulting from a microdeletion of approximately 20 genes on chromosome 7q11.23 (Hillier et al. 2003). In addition to a distinctive cognitive profile (see Mervis et al. 2000), there is also evidence of a distinct behavior and personality profile in individuals with Williams syndrome that includes a high prevalence of attention problems (Leyfer et al. 2006), sociability and empathy (Doyle et al. 2004; Klein-Tasman and Mervis 2003), and heightened anxiety and emotional difficulties (Dykens 2003; Einfeld et al. 2001; Leyfer et al. 2006). Children with Williams syndrome have great interest in interacting with others, even from a very young age (Mervis et al. 2003), and their affiliative nature is a hallmark of the personality profile.

While there has been considerable focus on the sociable nature of children with Williams syndrome, there is also mounting evidence that people with Williams syndrome have difficulties establishing and maintaining friendships. Research with adults with Williams syndrome has indicated that most are socially isolated, have never experienced a significant relationship with the opposite sex, and do not typically engage in social contact with peers (Udwin 1990). Furthermore, Udwin (1990) also found that 96% of all the individuals were characterized as experiencing difficulty in making friends and 76% were described as having few to no friends. Of the small proportion of individuals who reported having friends, they indicated that their friendship was based around qualities such as mutual trust and shared interests. A more recent follow-up by (Davies et al. 1998) examined the social, emotional and behavioral characteristics of 70 adults with Williams syndrome between the ages of 19 and 39 years using an interview-based method with the primary caregivers. Their results supported the earlier findings from (Udwin 1990) in that the vast majority of individuals with Williams syndrome were reported to have significant difficulties with establishing friendships (96%), social disinhibition (94%), and social isolation (73%). Comparisons to other children

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with intellectual disabilities have shown that children with Williams syndrome show poorer social competency compared to children with Down syndrome, but stronger than children with Prader-Willi syndrome (Rosner et al. 2004). The development of social skills in Williams syndrome appears to be delayed compared to normative data from same-aged peers (Mervis et al. 2001).

Difficulties with the pragmatics of communication and social interaction likely contribute to the social difficulties of people with Williams syndrome. Although a surface-level impression of Williams syndrome may indicate a relative proficiency in conversational skills (Bellugi et al. 1999), it is evident that individuals with Williams syndrome do exhibit marked impairments in this area. For example, in studies of conversational abilities in Williams syndrome, weaknesses have been observed in areas of communication, social reciprocity, and social inhibition (Stojanovik 2006; Stojanovik and James 2006). In comparison to individuals with Down syndrome and Specific Language Impairment, children with Williams syndrome have been rated as showing higher levels of difficulties in pragmatic language abilities (Laws and Bishop 2004). One potential explanation for the apparent mismatch between the fluency in speaking abilities and deficits in pragmatic language skills may be due to a reliance on their relatively good spontaneous verbal abilities which, on the surface, creates an impression of linguistic fluency (Garayzabal 2004; Tarling et al. 2006).

Difficulties with social cognition, including perspective-taking as measured by Theory of Mind-related (ToM) tasks, have also been observed, with some heterogeneity (Plesa-Skwerer et al. 2006; Sullivan et al. 2003; Tager-Flusberg and Sullivan 2000). Generally, people with Williams syndrome show about the same level of performance on various Theory of Mind tasks as other children with developmental disabilities (Plesa-Skwerer and Tager-Flusberg 2006), although there is evidence to suggest that the social-perceptual component of theory of mind, which refers to the ability to make an immediate judgment of a person's mental state (Plesa-Skwerer and Tager-Flusberg 2006) may be preserved (Sullivan and Tager-Flusberg 1999; Tager-Flusberg et al. 1998). Porter et al. (2008) examined theory of mind abilities among individuals with Williams syndrome using an entirely non-verbal task. Their results indicated that while the overall ToM abilities were low relative to their general cognitive functioning, there was also an indication of within-group heterogeneity in ToM abilities including false belief understanding and understanding social scripts. In other words, specific difficulties with ToM tasks were seen in a subset of individuals with Williams syndrome.

Relatedly, Sullivan et al. (2003) compared a group of adolescents with Williams syndrome, Prader-Willi syndrome, and non-specific mental retardation on their understanding of non-literal language (i.e., jokes). Their results indicated that the Williams syndrome group did not significantly differ from the other two groups in their ability to distinguish a lie from a joke, but that the performance of all three groups in their ability to distinguish a lie from a joke was similar to that of much younger typically developing children, indicating that this aspect of their socio-cognitive development was significantly impaired. The authors suggested that this difficulty in understanding lies from jokes could impair everyday social interactions with others, as the use of jokes and non-literal statements are a part of everyday socio-communicative functioning.

There is also evidence that while children with Williams syndrome may experience heightened sensitivity to emotional expressions of others, this does not seem to lead to strong social decision making skills in social interactions (Fidler et al. 2007). Fidler et al. compared young children with Williams syndrome and developmental disabilities of mixed etiology on a social-decision making task in which the children were exposed to a researcher who affected either a positive or negative reaction (facial expression and vocalization) after eating two different snack foods. Later, the children were given the opportunity to give one of the snack foods to the researcher. Although the children with Williams syndrome were much more likely to imitate the vocal and or facial expressions of the researcher, this was not consistently related to the child providing the researcher with the snack food for which a positive reaction was made. The authors concluded that children with Williams syndrome show a heightened level of sensitivity to the emotional reactions of others (as evinced by their high level of mimicry and imitation) but that this ability does not appear to be related to the ability to make good social decisions (i.e., providing a desirable as opposed to undesirable snack food). This difficulty in social decision-making skills may also be partially responsible for difficulties with maintain friendships with other people.

Furthermore, there is also indication that the socio-communicative functioning of some young children with Williams syndrome may overlap with that of children with autism spectrum disorders (ASD). There have been several case reports of comorbid autism diagnoses for children with Williams syndrome (Gillberg and Rasmussen 1994; Herguner and Motavalli Mukaddes 2006; Reiss et al. 1985). Similarities to children with ASD include hyperacusis, social isolation, distractibility, pragmatic deficits, indiscriminately approaching of strangers, and other types of social impairment (Gillberg and Rasmussen 1994). Philofsky et al. (2007) examined the pragmatic language abilities of children with Williams syndrome compared to a matched group of children with ASD. Their findings

indicated that children with Williams syndrome and ASD displayed similar communicative and pragmatic language abilities, although the Williams syndrome group showed better performance in specific domains related to sensitivity of the communicative partner's perspective, non-verbal and verbal cues, and stereotyped language. What this may indicate is that although children with Williams syndrome possess a natural interest in interacting with others, they may nonetheless exhibit significant socio-communicative difficulties that affect their ability to interact well with others.

In our recent research, we found that socio-communicative abnormalities were common in young children with Williams syndrome (Klein-Tasman et al. 2007), with half of children with Williams syndrome with limited language abilities showing difficulties on the Autism Diagnostic Observation Schedule (Lord et al. 1999) similar to those of children with Pervasive Developmental Disorder—Not Otherwise Specified (Klein-Tasman et al. 2009). Abnormalities in declarative pointing, giving objects to others, and showing objects were commonly seen for children with Williams syndrome. About half of the sample, however, showed additional abnormalities including sparse vocalizations directed toward others, lack of direction of a range of facial expressions to others, poorly modulated eye contact, and a paucity of joint attention initiations. Children with Williams syndrome showed distinctly fewer abnormalities than the children with autism. However, more difficulties in reciprocal social interaction were observed in comparison to a mixed etiology nonspectrum group. The participants with Williams syndrome were most similar in their socio-communicative abnormalities to a matched group of children with PDD-NOS.

The Current Study

The goal of the current study is to further examine the social functioning of children and adolescents with Williams syndrome, using parent and teacher ratings from two measures, the Social Skills Rating Scale (Gresham and Elliot 1990), which assesses prosocial behaviors (including cooperation, assertion, and self-control) and the Social Responsiveness Scale (Constantino 2002), which assesses areas of social functioning that are typically problematic for children with autism spectrum disorders (social cognition, social communication, social awareness, social motivation, autistic mannerisms). Multiple raters are included to gain a more comprehensive account of social functioning across settings, and to expand on the current body of research that relies almost exclusively on parental report. It is expected that while children with Williams syndrome may show few prosocial functioning difficulties, difficulties with social cognition and social communication will be observed. Relations to age, gender, and intellectual functioning will also be examined.

Methods

Participants

Participants were of parents of 84 children with genetically confirmed diagnoses of Williams syndrome (36 boys, 48 girls) between the ages of 4 and 16 years old (mean = 9.44, SD = 3.89). Participants were recruited through the University of Wisconsin-Milwaukee Child Neurodevelopment Research Lab and at the 2006 and 2008 National Williams Syndrome conventions. The native language of all participants and children is English. All children were administered the K-BIT-II as a brief measure of intellectual abilities. Mean KBIT-II Composite IQ score was 70.63 (SD = 13.86). Not all questionnaire measures were available for all participants. The participants were involved in a variety of research projects with slightly differing batteries of measures included, such that not all children were administered all measures. To maximize representativeness, the greatest number of participants available for each analysis is typically included; when parent and teacher report are directly compared, only participants with both reporters are included.

Measures

Kaufman Brief Intelligence Test, 2nd Edition (KBIT-II: Kaufman and Kaufman 2004)

The KBIT-II is a brief measure of verbal and nonverbal intelligence that is individually administered. It contains three subtests; two of these comprise the Verbal domain, with verbal knowledge and riddle sections, which measures crystallized knowledge and verbal abilities. One subtest comprises the Nonverbal domain, which measures fluid and nonverbal reasoning with the use of matrices subset. The KBIT-II provides standard scores, percentiles, and age equivalents for the Verbal and Nonverbal domains as well as an overall standard score and percentile. Normative data are available for individuals aged 4–90 years. The norming sample consisted of an even distribution of male and female participants and a relatively even distribution of participants across all ages. Adequate reliability and validity have been established (Kaufman and Kaufman 2004).

Social Skills Rating Scale

The Social Skills Rating Scale (Gresham and Elliot 1990) is a multi-rater assessment of an individual's social skills

functioning. Adequate internal consistency, test–retest reliability, and validity have been demonstrated (Gresham and Elliot 1990). Different forms are available for different age groups (Preschool for 3- to 5-year-olds, Elementary form for K-6th grade, Secondary form for 7th through 12th grade), and separate forms are available for parents and teachers. The SSRS is comprised of two main scales: Social Skills and Problems Behaviors. For this study, only the Social Skills Scale was used. This scale assesses the presence of positive social behaviors, with higher numerical values representing more positive social behaviors. Standard scores have a mean of 100 and a standard deviation of 15. Four subscales are available (Cooperation, Assertion, Responsibility and Self-Control); however, these subscales do not yield scaled scores, but rather include only interpretive categories indicating the extent to which an individual displays a particular behavior that is "average," "fewer" or "more" in frequency compared to the general population. These subscales were not used in this investigation.

Social Responsiveness Scale (Constantino and Gruber 2005)

The SRS is a 65-item rating scale that ascertains autistic symptoms across the entire range of severity in which they occur in natural social settings. The scale is completed by parents, teachers, or day care providers who have observed a child's social interactions, particularly with peers, in naturalistic social contexts. The SRS includes items that identify a child's social impairments, assessing social awareness, social information processing, capacity for reciprocal social communication, social anxiety/avoidance, and autistic preoccupations and traits. Ratings are given on a scale from 1 (*not true*) to 4 (*almost always true*) on the basis of a behavior's frequency of occurrence. The SRS generates a singular scale score that describes the severity of social deficits in the autism spectrum. Higher scores on the SRS indicate greater severity of social impairment. In addition, subscale scores measuring Social Awareness (SA), Social Motivation (SM), Social Cognition (SCog), Social Communication (SCom), and Autistic Mannerisms (AM) are available. The SRS is appropriate for use with children from 4 to 18 years of age.

Results

Social Skills Rating System Social Skills Scale Parent and Teacher Ratings

SSRS parent-report data were available for 74 participants (16 preschool, 41 elementary, 17 secondary) and SSRS teacher-report data were available for 51 participants (2 preschool, 36 elementary, 13 secondary). Both parent and teacher data were available for 50 participants (2 preschool, 35 elementary, 13 secondary). Internal consistency statistics (computed for all forms except the preschool Teacher form due to low n) indicated acceptable internal consistency for both parent (Preschool: alpha = .86, Elementary: .84, Secondary: .73) and teacher report (Elementary alpha = .88, Secondary alpha = .91).

Mean parent-report SSRS Social Skills score was 87.68 (SD = 13.85) and mean teacher-report SSRS score was 88.71 (SD = 12.98). Both parent and teacher ratings fell in the low average range. One sample t-tests were used to compare the Social Skills standard scores to the normative mean. For parent ratings ($n = 74$), mean standard scores were significantly below population norms ($t(73) = -7.66$, $p < .001$), as was also the case for teacher ratings ($t(50) = 6.21$, $p < .001$). Parent and teacher ratings did not significantly differ ($t(49) = .513$, $p = .61$) and were significantly correlated [$r = .33$, $p < .05$].

Frequency statistics were used to examine the proportion of individuals with Williams syndrome who exhibited social skills functioning considered delayed (standard score below 70), borderline (70–79), low average (80–89), and average (90 and above). Examination of the SSRS Parent data indicated that 41.9% of the sample ($n = 31$) was perceived by their parents as "average" in overall social functioning, 25.7% ($n = 19$) as "low average," 24.3% ($n = 18$) as borderline, and 8.1% ($n = 6$) as delayed. Examination of teacher data indicated that 51% of the sample ($n = 26$) was perceived by their teachers as "average" in overall social functioning, 27.5% ($n = 14$) as "low average," 13.7% ($n = 7$) as borderline, and 7.8% ($n = 4$) as delayed. Z-tests for proportions indicated no significant difference in the proportion of parents and teachers describing participants in the average range ($z = .82$, ns).

A small but significant correlation between age and social skills was found based on parental report ($r = .25$, $p < .05$), and a similar-sized correlation, that did not reach statistical significance, was observed based on teacher report ($r = .24$, $p = .09$). Neither parent nor teacher SSRS scores were significantly correlated with overall intellectual functioning ($r = .20$, ns and $r = .01$, ns respectively) or with nonverbal intellectual functioning ($r = .15$, ns, and $r = .09$, ns respectively). A small but significant correlation between SSRS scores and verbal intellectual functioning was observed for parent report ($r = .23$, $p < .05$) but not for teacher report ($r = -.04$, ns). Based on parental report, males showed somewhat stronger social skills than did females [$t(72) = 2.64$, $p = .01$], but there was no significant gender effect based on teacher report [$t(49) = 1.60$, $p = .12$].

While generally difficulties on the SSRS were relatively mild, an item analysis was conducted to explore the items most and least frequently endorsed by parents and teachers. Participants with both parent and teacher reports were included. This analysis was complex, as there are 3 age levels within the current study, and each age level is administered a slightly different form; very few preschool forms were completed, so this analysis was conducted for the elementary and secondary forms only. A rating of "never" showing a prosocial behavior was considered endorsement of an item as problematic. Initially, a criterion of more than 50% of participants endorsed as "never" showing a behavior was considered to reflect frequent difficulties within the group. However, very few items showed this level of endorsement, and a less stringent level of 40% was therefore used. For the children in kindergarten through 6th grade, only four items were rated by parents as problematic for more than 40% of the participants, all having to do with helping with household chores. Only six items were rated by teachers as problematic for more than 40% of the participants; these items tapped concentration and organization (e.g., keeping desk neat, ignoring distractions, using time well in class) as well as managing conflict (e.g., appropriately questioning rules that are unfair, compromising in conflict situations). For the children grades 7 through 12, only one item (keeping room neat) was endorsed as problematic by parents for more than 40% of the participants, and only one item, related to appropriately questioning rules that are unfair, was endorsed as problematic by teachers.

Items with especially low problem ratings were also examined. Initially, a criterion of a rating of "always" rating on prosocial behaviors for more than 80% of participants was used. None of the items reached this criterion for the children kindergarten through 6th grade. With a more liberal criterion of 60%, 7 items showed low rates of problem ratings based on parental report; these items included behaviors such as acknowledging praise, introducing self to others, congratulating others on accomplishments, self-confidence in social situations, and being well-liked by others. Only two items met this criterion based on teacher report; these included initiating conversations with peers and getting along with others who are different. For children grades 7 through 12, four items showed low problem ratings based on parent report with the more stringent 80% criterion; these included acknowledging praise from others, showing concern for friends, and beginning conversations appropriately. An additional 6 items met the 60% criterion; like the parent ratings, these included introducing self to others, being well-liked by others, and showing self-confidence. Only one item met the 80% criterion based on teacher report; 93% of the children were rated "always" for complying with teacher directions. An additional 4 items met the more liberal criterion based on teacher report; these included easily making transitions, controlling temper, initiating conversations with peers, and acknowledging praise.

Social Responsiveness Scale Parent and Teacher Ratings

SRS parent-report data were available for 82 participants and SRS teacher-report data were available for 49 participants. Internal consistency statistics were conducted for both the parent and teacher SRS ratings at the total SRS scale level as well as by domain. At the total scale level, internal consistency was excellent (alpha = .92 and .93 for parents and teachers respectively). Internal consistency at the scale level for parent report data was weaker, but still broadly acceptable (SA = .63, SCog = .65, SCom = .85, SM = .67, AM = .79). Internal consistency at the scale level for teacher report data was good (SA = .73, SCog = .78, SCom = .85, SM = .78, AM = .81).

Mean SRS *T*-scores at the domain level for parents and teachers are presented in Table 1. A significant effect of domain was present for both parent- [$F(4, 324) = 93.05$, $p < .001$] and teacher-report data [$F(4, 192) = 32.87$, $p < .001$)]. *T*-test comparisons (using a more conservative alpha level of .01 given the number of comparisons) indicated that for parents, Autistic Mannerisms were most problematic and were rated significantly higher than all scales except Social Cognition ($t(82) = 1.35$, $p = .181$). Social Cognition was rated significantly higher than Social Communication, Social Awareness, and Social Motivation. Social Communication was rated significantly higher than Social Motivation, and there was a trend toward greater difficulty than in Social Awareness ($t(81) = 2.12$, $p < .05$). Finally, significantly more difficulties in Social Awareness were present than for Social Motivation. *T*-test comparisons indicated that for teachers, difficulties with Social Cognition were most problematic and were rated significantly higher than all other scales. Next most

Table 1 Means (and standard deviations) of parent- and teacher-reported SRS domain scores

Rater	*N*	Social awareness	Social cognition	Social commun.	Social motivation	Autistic mannerisms
Parents	82	65.68 (13.33)	76.23 (11.75)	68.32 (12.35)	54.40 (11.25)	77.99 (15.82)
Teachers	49	61.67 (10.64)	67.84 (10.00)	60.08 (7.75)	53.12 (7.61)	63.49 (11.24)

affected were Autistic Mannerisms, which were significantly higher than Social Communication and Social Motivation. Social Awareness was rated significantly higher than Social Motivation. Finally, Social Communication was rated significantly higher than Social Motivation. Hence, for both parents and teachers, Autistic Mannerisms and Social Cognition difficulties were most common while difficulties with Social Motivation were least common.

According to the developers of the SRS, *T*-scores below 60 indicate no clinically significant difficulties; *T*-scores of 60–75 are representative of deficiencies in reciprocal social behavior that are clinically significant and result in mild to moderate interference in everyday social interactions; *T*-scores of 76 or greater indicate elevations in the severe range. For the overall parent-report SRS Total score, 39% ($n = 32$) showed severe range difficulties, 47.6% ($n = 39$) showed mild to moderate range difficulties, and only 13.4% ($n = 11$) showed no clinically significant difficulties. For the overall teacher-report SRS Total score, 8.2% ($n = 4$) showed severe range difficulties, 53.1% ($n = 26$) showed mild to moderate range difficulties, and 38.8% ($n = 19$) showed no clinically significant difficulties. The number of children falling within each of these ranges at the domain level, according to parents and teachers, is presented in Figs. 1 and 2 respectively. Results indicate that the majority of the participants showed at least mild to moderate elevations (*T*-score >60) on four of the five subscales, including social awareness, social communication, social cognition, and autistic mannerisms based on parental report. Difficulties with social motivation were less commonly reported.

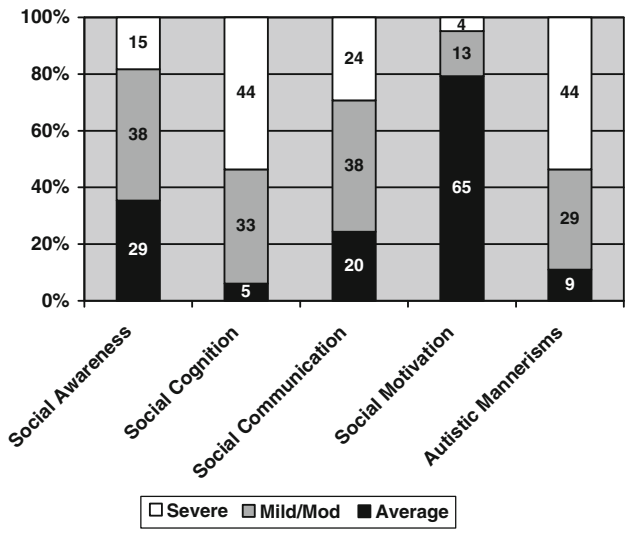

Fig. 1 Percent of participants falling in the average, mild/moderate, and severe ranges on the SRS parent-report (number of participants indicated)

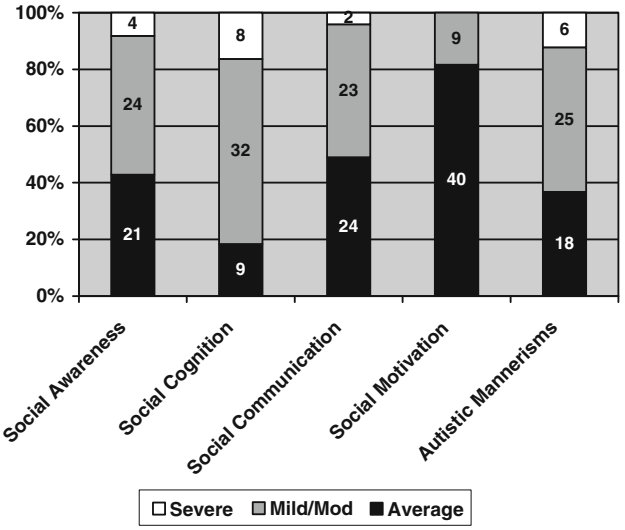

Fig. 2 Percent of participants falling in the average, mild/moderate, and severe ranges on the SRS teacher-report (number of participants indicated)

Correlational analyses were conducted to evaluate the relation between the SRS scores and intellectual functioning. No significant correlations with KBIT-II IQ (Overall, Verbal, or Nonverbal) were found for the teacher-report data. For the parent-report data, significant correlations with overall intellectual functioning were found for Social Awareness raw ($r = -.35$, $p < .001$), Social Cognition raw ($r = -.29$, $p < .01$), Social Communication raw ($r = -.41$, $p < .001$), Autistic Mannerisms ($r = -.29$, $p < .01$), and total raw SRS score ($r = -.39$, $p < .001$). At the *T*-score level, a significant correlation with Social Communication was observed ($r = -.34$, $p < .005$), and with Total SRS *T*-score ($r =$ -−.32, $p < .005$), and correlations with the remaining *T*-scores were observed only at a trend level ($p < .05$). No additional significant correlations were found with either verbal or nonverbal intellectual functioning.

Age was not significantly correlated with any parent-report ratings. Significant bivariate correlations between age and teacher Social Awareness raw ratings [$t(47) = -.36$, $p < .05$) and *T*-scores [$t(47) = -.41$, $p < .005$] were found, with improvements in social awareness found with age. Results of the independent samples t-tests indicate that females did not differ from males at the raw score level in Total SRS score based on either parent [$t(80) = 1.13$, $p = .26$] or teacher report [$t(47) = .37$, ns] or at the subscale level. At the *T*-score level, which compares the performance of participants to others of the same gender, females had significantly more difficulties than males in Total SRS score based on parent [$t(80) = 3.17$, $p < .005$] and teacher report [$t(47) = 2.64$, $p < .05$], and also in the areas of Social Communication based on parent [$t(80) = 4.04$, $p < .001$] and teacher report [$t(47) = 2.85$, $p < .01$],

Social Cognition based on parent [$t(80) = 2.89$, $p < .005$] and teacher report [$t(47) = 2.12$, $p < .05$], Social Motivation based on parent [$t(47) = 2.20$, $p < .05$] and teacher report [$t(47) = 2.40$, $p < .05$] and Autistic Mannerisms based on parent [$t(80) = 3.04$, $p < .005$] and teacher report [$t(47) = 3.04$, $p < .01$].

Examination of SRS data from participants with both parent- and teacher-report data ($n = 47$) indicated that Total SRS scores of parents and teachers were significantly correlated ($r = .42$, $p = .003$), as were ratings of Social Communication ($r = .44$, $p < .005$), Autistic Mannerisms ($r = .38$, $p < .01$), Social Awareness ($r = .37$, $p < .05$), and Social Cognition ($r = .34$, $p < .05$). Parent and teacher ratings of Social Motivation were not significantly correlated ($r = .22$, $p = .14$), which may relate to restriction of range, as difficulties in this domain were rare. T-tests comparing parent and teacher ratings indicated that parents reported higher levels of difficulties overall [$t(46) = 5.58$, $p < .001$], as well as in Social Cognition [$t(46) = 4.51$, $p < .001$], Social Communication [$t(46) = 4.62$, $p < .001$], and Autistic Mannerisms [$t(46) = 6.46$, $p < .001$], but did not differ significantly in ratings on Social Awareness [$t(46) = 1.83$, $p = .07$] or Social Motivation [$t(46) = .69$, $p = .49$].

An item analysis was conducted to examine the items most frequently endorsed by both parents and teachers. Data from participants with both parent and teacher reports were included. A rating of "often true" or "almost always true" was considered an endorsement (with some items reverse coded appropriately). Endorsement of an item by more than 50% of respondents was considered frequent endorsement. For parents, 50% of the Social Awareness and Social Cognition items, 25% of the Autistic Mannerism items, 9% of the Social Communication items, and none of the Social Motivation items were endorsed by more than 50% of the respondents as problematic. For teachers, 62.5% of the Social Awareness items, 50% of the Social Cognition items, 31.8% of the Social Communication items, 25% of the Autistic Mannerism items, and none of the Social Motivation items were endorsed as problematic by more than 50% of respondents. Items with high endorsement rates for both parents and teachers included those related to inappropriate physical proximity, potential for victimization, repetitive thoughts or statements, sensitivity to sound, play skills, and nonliteral understanding of conversations. Teachers also indicated difficulties communicating feelings and understanding others feelings, and difficulties offering comfort to others when they are sad, while these items were not especially highly endorsed by parents.

Items with low problem ratings were also examined. For parents, 54.5% of the items on the Social Motivation were endorsed by less than 20% of respondents, 27.2% of Social Communication items, 25% of Social Awareness items, and 8.3% of Social Cognition and Autistic mannerisms items. For teachers, 45.5% of the Social motivation items, 27.2% of the Social Communication, 25% of the Social Awareness, 16.7% of the Autistic Mannerisms, and 8.3% of the Social Cognition items were endorsed by less than 20% of respondents. Items endorsed by less than 20% of respondents included items related to failures to seek out social interaction and being uncomfortable in social situations.

Comparisons of SSRS and SRS Ratings

Both SSRS and SRS ratings were both available for parents of 72 children and teachers of 42 children. Correspondence between ratings on these forms was examined using correlational analysis. Parent SRS Total scores were significantly correlated with both parent ($r = -.56$, $p < .001$) and teacher SSRS ratings ($r = -.55$, $p < .001$). Teacher SRS Total scores were significantly correlated with teacher SSRS ratings ($r = -.38$, $p < .05$), but not with parent SSRS ratings ($r = -.24$, $p = .11$). Because the two measures use scales of opposite valences (high scores are better on the SSRS and high scores are worse on the SRS), SRS scores were reverse coded to enable comparison of ratings across measures. Both parent [$t(71) = 10.99$, $p < .001$] and teacher reports [$t(41) = 2.23$, $p < .05$) indicated significantly greater social difficulties on the measure of social reciprocity than on the measure of prosocial skills.

Discussion

Based on the literature, people with Williams syndrome are generally sociable but display difficulties in social reciprocity, social understanding, and social skills. To date, questionnaire investigations of the behavior of children with Williams syndrome have relied largely on parental report. In this study, the social functioning of children with Williams syndrome was examined using multiple measures from both parent and teacher perspectives, in order to gain a sense of the broad range of social behaviors as well as the cross-situational consistency in the behavioral phenotype. Difficulties with prosocial aspects of social functioning, including social motivation and social awareness, were less commonly observed than were difficulties with the social-cognitive aspects of social functioning such as social communication and social cognition. Significant difficulties with social communication and social cognition were observed. Additionally, the results of this investigation add to our current body of work examining the presence of autism spectrum (AS) symptomatology in children with Williams syndrome, as a large proportion of the sample

showed clear difficulties on a measure of social reciprocity designed to capture AS-related difficulties. Overall, good correspondence between parents and teachers was found related to social reciprocity behaviors in particular, and also for prosocial skills. While teachers reported milder difficulties, the characteristic pattern of social strengths and weaknesses typically seen for children with Williams syndrome is reflected in both parent and teacher ratings.

Social Profile

In terms of profile, children with Williams syndrome showed relative strengths on the Social Motivation subscale than on the other subscales of the SRS, and the number of children with marked social difficulties on the SSRS was relatively small. Hence, difficulties in prosocial skills, as measured by certain SRS scales and by the SSRS were also present but generally not pronounced. When prosocial difficulties were present for school-aged children, at the item level they often were in the realm of organization (within the classroom) or performance of household chores (within the home environment), likely reflecting difficulties with attention (Leyfer et al. 2006) and difficulties with adaptive functioning involving motor control (Mervis et al. 2001) that have been identified in the literature as characteristic of children with Williams syndrome. Difficulties with seeking out social interaction, self-confidence, initiating conversations, and being well-liked by peers were generally low based on both parent and teacher report. These results mirror previous findings that children with Williams syndrome are less reserved toward strangers, more approaching, more gregarious, overly friendly, and affectionate (Klein-Tasman and Mervis 2003; Udwin and Yule 1991).

Despite these prosocial strengths, many children with Williams syndrome showed abnormalities in social reciprocity, as measured by the overall score on the SRS and a number of its subtests. Children and adolescents with Williams syndrome were rated by their parents as having significant deficiencies of reciprocal social interaction as measured by the overall SRS score. Specifically, almost half of the participants fell into the mild to moderate range of severity in terms of overall social reciprocity difficulties. An additional 39% of participants displayed deficiencies in everyday social reciprocity that fell in the severe range. Teacher report indicated that more than half of children with Williams syndrome showed social reciprocity difficulties. These difficulties were most commonly seen in the areas of social cognition, social communication, and autistic mannerisms, while difficulties with social motivation were uncommon. These results confirm previous research characterizing people with Williams syndrome as having poor social skills and poor understanding of socially-relevant information (e.g., Fidler et al. 2007; Philofsky et al. 2007), and these difficulties may contribute to their challenges with sustained friendships. In addition to evidence supporting difficulties in social reciprocity in general, the results of this investigation also confirm findings of possible overlap with other symptomatology related to the autism spectrum.

Individual Differences: Gender, Age and Intellectual Functioning

When gender differences were found in the current study, they indicated greater difficulties for females with Williams syndrome compared to the general population than for males, using gender-specific norms. In other words, we find that the social behavior of girls with Williams syndrome is more different from same-aged peers of the same gender than is the social behavior of boys with Williams syndrome. This finding was also observed by Porter et al. (2009) in their examination of parental ratings of problem behaviors. These differences may reflect disparities in parents' and teachers' expectations for the social behaviors of boys and girls in that the types of behavioral problems typically seen in children with Williams syndrome may be more in-line with the types of more normative behavioral problems seen in typically developing boys but not girls, such that observations of behavioral difficulties in girls with Williams syndrome are more salient. These differences in behavior may also reflect an actual gender-based difference in the expression of behavioral difficulties within this population. Gender differences in psychopathology in Williams syndrome have been similarly observed in prior research, with girls generally showing more emotional difficulties (Dodd and Porter 2009; Leyfer et al. 2006; Porter et al. 2009).

In the current study, few consistent correlations with age and intellectual functioning were found. Both parents and teachers tended to report that older children with Williams syndrome have better prosocial skills; however, these ratings were not generally associated with intellectual functioning. With respect to ratings of deficits in social functioning, age did not appear to be a factor in either parent or teacher reports, and intellectual functioning was not related to teacher ratings. For parents, higher intellectual functioning was consistently associated with fewer deficits in social reciprocity and responsiveness.

Parent–Teacher Concordance

Overall, good concordance between parent and teacher ratings using both SSRS and SRS measures was observed. With respect to evidence of prosocial functioning, both parents and teachers reported that about half of the children

with Williams syndrome fell in the average range in terms of their levels of social skills on the SSRS. A moderate significant correlation between parent and teacher ratings was observed. In the area of social reciprocity and autism-related behaviors, SRS ratings of social functioning also indicated that parents and teachers agree that the presence of autistic mannerisms and difficulties in social cognition represented the two areas of greatest deficit in the children with Williams syndrome, and both groups of raters also agreed that social motivation was an area of least deficit. Additionally, parent and teacher ratings were significantly positively correlated. However, across all domains of the SRS, parents reported consistently higher indications of problematic behaviors than did teachers. Hence, it appears that the general pattern of social functioning of children with Williams syndrome is similar across the school and home contexts. The consistency also speaks to the characteristic nature of the Williams syndrome phenotype, as it is clearly evident in multiple settings.

Differences in ratings by observer were indeed seen and could be due to the variability in environmental contexts observed. Teachers' observations are tied to the behaviors of the children with Williams syndrome in the classroom, and it is possible that their ability to observe the full spectrum of problematic behaviors may be hampered by their need to attend to multiple children at the same time. Conversely, parents have the opportunity to observe their child in the relative calm of their own home, thus it is possible that they are able to pick up on a greater number of problematic behaviors. Another potential explanation pertains to social context under which these ratings were made. It is possible that the parents were assessing their child with Williams syndrome's social performance to typically developing siblings or to a personal standard of social behavior that may differ from parent to parent. Conversely, the type of classroom in which the teachers rated the children with Williams syndrome were mixed; some were in regular education classrooms and others in special education classrooms. Given their experience with a broad range of children, however, it is possible that the teachers were comparing the social behavior of the children with Williams syndrome to that of typically developing children, or more likely to that of children with other forms of developmental delay which may have skewed their perceptions of "average" as opposed to "below average" social functioning. However, there were areas in which teachers actually saw more difficulties. For example, teachers were more likely than parents to report difficulties communicating and understanding feelings and offering comfort to others.

Item content may also have contributed to the findings. For the SSRS, while there was significant item overlap, different items were indeed present on the parent and teacher measures, contributing to lack of agreement between parents and teachers. For example, parents have greater opportunity to observe difficulties with household tasks, while teachers have greater opportunity and context in which to observe difficulties with organization and concentration in group settings. This item variability does not contribute to the findings on the SRS because this measure has the same items across respondents.

It is notable that discrepancies in perceptions of prosocial functioning between parents and teachers have also been found in other bodies of research. For example, Macintosh and Dissanayake (2006) examined the social skills profile of children with High Functioning Autism (HFA), children with autism spectrum disorders (ASD), and typically developing children using the parent and teacher versions of the SSRS. Similar to the findings of the current study, teachers reported fewer social difficulties than did parents for the children with ASD.

Limitations and Future Directions

There are several notable limitations in the design of the current study. First, although the current study utilized data from multiple measures and raters to describe the social profile of individuals with Williams syndrome, there is no direct comparison to children with other developmental disabilities or children with autism spectrum disorders. Second, only questionnaire measures are used in this investigation; direct observation would likely provide a more comprehensive sense of social functioning. Finally, the quality of peer friendship is not examined and would enable examination of the extent to which the variations in the social profile of individuals with Williams syndrome contribute directly or indirectly to differences in their ability to formulate and maintain meaningful friendships.

In sum, the current study provided additional description of the social profile of individuals with Williams syndrome using a multi-measure, multi-rater assessment approach. While the results regarding the social profile of individuals with Williams syndrome appears to be consistent across raters, the findings support the presence of two different domains of social functioning capability in children with Williams syndrome. Despite their outwardly sociable and gregarious nature and willingness to engage with others, problematic social behaviors are present, including difficulties with social communication and perspective-taking. These difficulties likely play a part in challenges establishing and maintaining friendships and contribute to the often poor social outcomes observed in this population.

Acknowledgments This research was supported by grant NIMH R03 MH069400, a University of Wisconsin-Milwaukee Graduate

School Research Committee Award, and a Research Scholar grant from the Center for Addition and Behavioral Health Research. We would also like to thank Frank Gallo and Kristin Phillips for assistance with data collection and to Molly Barton, Amy Sietenzahl, and Jesse Behagen scoring and checking of data. Portions of this research were included in Kirsten Li-Barber's Dissertation and in Erin Magargee's Senior Thesis and were presented at the Society for Research in Child Development, the International Neuropsychological Society, and Association for Psychological Sciences meetings.

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