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Discrepancies in Parent and Teacher Ratings of Social-Behavioral Functioning of Children with Chromosome 22q11.2 Deletion Syndrome: Implications for Assessment

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Abstract

Children with 22q11.2 deletion syndrome exhibit high rates of social-behavioral problems, creating an area in need of intervention. This study obtained parent and teacher ratings on the CBCL/TRF of 67 children with 22q11DS and 57 controls. Results indicated significant differences in social-behavioral functioning of children with 22q11DS compared to controls, depending on rater type. Parents reported greater internalizing, withdrawal, and social problems in children with 22q11DS while teachers perceived few differences between groups. Correlational analyses indicated weak concordance between parent and teacher reports, with no significant correlations on three summary scales. The findings support the use of multiple informants when evaluating the social-behavioral functioning of children with 22q11DS, and suggest that interpretations based on one informant/setting should be made cautiously.

Keywords

22q11 deletion syndrome; velocardiofacial syndrome; DiGeorge syndrome; behavior; social skills

Introduction

Chromosome 22q11.2 deletion syndrome (22q11DS), also known as velocardiofacial syndrome, or DiGeorge syndrome, is estimated to affect approximately 1 in 1,600 to 4,000 individuals (Shprintzen, 2008; Tezenas Du Montcel, Mendizabai, Ayme, Levy, & Phillip, 1996). The medical problems commonly associated with 22q11DS include cardiac anomalies, velopharyngeal incompetence, cleft palate, immune deficiencies, and characteristic facial features (McDonald-McGinn et al., 1999; McDonald-McGinn et al., 1997; Shprintzen, 1978). In addition to these physical disabilities, research involving children and young adults with 22q11DS have revealed neuropsychological deficiencies including impaired intellectual abilities and executive functioning, and a higher incidence of psychiatric disorders. Cognitive abilities of children with 22q11DS extend from the

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moderately impaired range to the low average range with a mean IQ ranging from 70 to 89 (Lajiness-O'Neill et al., 2006; Moss et al., 1999; Niklasson, Rasmussen, Óskarsdóttir, & Gillberg, 2001; Niklasson, Rasmussen, Óskarsdóttir, & Gillberg, 2002; Sobin, Kiley-Brabeck, Hadley Monk, Khuri, & Karayiorgou, 2009; Swillen et al., 1997;Woodin et al., 2001). In addition, numerous reports have documented discrepancies among cognitive domains, with better performance on tasks of verbal IQ than tasks of performance IQ (Lajiness-O'Neill et al., 2006; Moss et al., 1999; Niklasson et al., 2002; Swillen et al., 1997). This cognitive profile is often described as a nonverbal learning disability (NVLD; Moss et al., 1999), although the exact alignment with the 22qDS cognitive profile has been questioned (De Smedt et al., 2007; Schoch et al., in press).. Numerous studies have also reported that individuals with 22q11DS are at increased risk for developing psychotic disorders in adulthood, particularly affective bipolar disorders and schizophrenia (Gothelf & Lombroso, 2001; Murphy, Jones, & Owen, 1999; Niklasson et al., 2002; Papolos et al., 1996; Shprintzen, Goldberg, Golding-Kushner, & Marion, 1992).

Social-Behavior Difficulties in Children with 22q11DS

Children with 22q11DS often exhibit psychiatric and behavioral challenges that negatively impact their learning and social development. Although parents of affected children report fewer concerns regarding the presence of externalizing symptoms, internalizing problems are quite common (Jansen et al., 2007; Woodin et al., 2001). Previous investigations by our team and others have reported that the behavior difficulties reported in the 22q11DS population appear to be largely unrelated to individuals' intellectual abilities and medical complications, suggesting the presence of a behavioral phenotype of the disorder (Jansen et al., 2007; Shashi et al., 2011). Common psychiatric conditions comorbid in children with 22q11DS include ADHD, withdrawal, specific phobias, anxiety disorders, depressive disorders, obsessive-compulsive features, and features of autism, with ADHD and anxiety disorders being the most prevalent (Lewandowski, Shashi, Berry, & Kwapil, 2007; Sobin et al., 2009; Swillen et al., 1997; Woodin et al., 2001). Additionally, children with 22q11DS have been reported to experience weaker social competence and more social problems than their unimpaired peers (Shashi et al., 2011). (Sobin et al., 2009; Swillen et al., 1997; Woodin et al., 2001). Unfortunately, few affected children appear to be receiving treatment for their psychosocial challenges, despite 67% meeting diagnostic criteria for at least one psychiatric condition (Young, Shashi, Schoch, Kwapil, & Hooper, 2011).

All reports on the behavior problems in 22q11DS thus far have relied solely on parent behavior ratings, which may only provide a partial picture of the child's overall behavior and functioning. Little is known about how the behavioral difficulties of children with 22q11DS are manifested across settings. Parents are valuable respondents because they observe their child's behaviors across multiple settings. In contrast, teachers have the unique ability to compare one child's functioning with that of a large group of age-matched peers. Based on the literature in the general population however, it is evident that clinicians often have difficulty integrating reports from multiple informants due to inconsistent results (Verhulst & Akkerhuis, 1989). Across studies, most research teams have documented low to moderate agreement between parent and teacher behavior ratings, with parents typically reporting more problems than teachers and teachers identifying externalizing behaviors more

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frequently than internalizing problems (Cai, Kaiser, & Hancock, 2004; Verhulst & Akkerhuis, 1989; Youngstrum, Loeber, & Stouthamer-Loeber, 2000). Since children with 22q11DS often exhibit internalizing symptoms, teacher ratings may overlook the presence and/or the severity of internalizing behaviors. Although examined in the general child population, agreement between parent and teacher behavior ratings has not been investigated within the 22q11DS population. Developing a greater understanding of children's behavior, as measured by parent and teacher reports, could indicate how the social-behavioral phenotype of 22q11DS may be impacting children's learning and social-emotional development. This information could have implications for treatment of emotional and behavioral problems in 22q11DS by guiding intervention efforts in targeted settings.

In that regard, the current investigation was designed to obtain information regarding parent and teacher ratings of the social-emotional behavior of children with 22q11DS as compared with their unimpaired peers. Two specific questions were examined. First, the degree to which parents and teachers report social-behavioral difficulties of children with 22q11DS relative to age-matched controls. It was hypothesized that parents *and* teachers of children with 22q11DS would be more likely to report the presence of internalizing forms of social-behavioral difficulties than parents or teachers of typical children. Second, we examined the relationship between parent and teacher ratings of social-behavioral functioning within the 22q11DS group. Based on results from the general population, we hypothesized that agreement between respondents would be weak to moderate, with higher concordance between parents and teachers on externalizing types of behaviors than on internalizing behaviors.

Methods

Participants

The participants in the present study were included as part of a larger comprehensive study investigating the neuropsychological and psychiatric functioning of children with 22q11DS. The current investigation included 67 children and adolescents with 22q11DS and 59 control children. Control participants were recruited through pediatric practices in the community and the public school system and were age- and gender-matched to the 22q11DS subjects. Participants in the control group were not excluded based on the presentation of ADHD. As they were recruited for an investigation of risk for psychosis, none of the subject or control participants presented with psychosis at the time of the evaluation. Specific sociodemographic characteristics of the sample can be seen in Table 1.

Measures

Data were collected regarding parent and teacher perceptions of children's social-emotional functioning through the use of two complementary behavior rating scales, the *Child Behavior Checklist* (CBCL) and the *Teacher Report Form* (TRF). Both instruments have well-established psychometric properties, including good reliability and validity, particularly for the Total Problem, Internalizing and Externalizing composites (Achenbach, 1991a; Achenbach, 1991b; Sattler &Hoge, 2006). Both instruments are widely used in research and clinical practice (Gothelf et al., 2007; Jansen et al., 2007; Lajiness- O'Neill et al., 2006;

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Swillen et al., 1997). Responses on the CBCL and TRF yield eight clinical syndrome subscales (Anxious/Depressed, Withdrawn, Somatic, Social problems, Thought problems, Attention problems, Rule-breaking/Delinquent, and Aggressive), which contribute to three summary scales (Internalizing Problems, Externalizing Problems, and Total Problems). CBCL and TRF scores are presented as T-scores, which have a mean of 50 and standard deviation of 10, with higher scores on both instruments indicating greater impairment.

Data Analysis

Preliminary data analysis examined the similarities and differences between groups on selected demographic variables such as chronological age, race/ethnicity, gender, and socioeconomic status, with t-tests being used for continuous data and the Fishers Exact Test being used for the categorical data. Variables that contributed to group differences would be considered as covariates in subsequent group comparisons.

To address the first research question, which compared parent and teacher ratings for participants with 22q11DS and control participants, we employed two separate multiple analyses of covariance (MANCOVA) for parent and teacher scales. With a significant MANCOVA, follow-up univariate procedures were conducted. Selected covariates were included in the analyses.

For the second question examining the concordance between parent and teacher ratings, we conducted Pearson Product Moment Correlations between the two scales for the 22q11DS sample, with medication status and the type of teacher that completed the rating (regular education versus special education) as covariates. Additionally, we utilized Bland-Altman Plots for significant correlations in order to determine the level and pattern of correlations between the different raters.

Results

Parent Ratings

Preliminary data analyses revealed no differences between the 22q11DS and control groups on socioeconomic status, t (109) = -1.03, $p < .31$; chronological age, t (123) = -1.60, $p < .12$; and gender, Fisher's exact test p=.725. The groups were significantly different on Full Scale IQ, t (121) = -11.12, $p < .00$, with the control group being higher; and ethnicity, X^2 (4) = 18.53, $p < .001$, where there was a higher proportion of African Americans in the control group. This finding regarding ethnicity likely reflects the demographics of the population in the area around Duke University Medical Center, one of the two sites of the study. Table 1 illustrates the demographics of the samples in our study. For subsequent analyses, the two groups were covaried on ethnicity. The need to statistically control for the significant difference in IQ between the two groups posed an interesting decision, however. Given the lower intelligence scores due to the genetic microdeletion in individuals with 22q11DS, Dennis et al. (2009) have argued that controlling for IQ would eliminate some of the variance inherent in the disorder, thus minimizing or eliminating potential important group differences. In contrast, a number of studies have documented the importance of level of functioning, as defined by intelligence, to subsequent behavior (Rutter, Graham, & Yule,

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1970). Consequently, controlling for differences in intelligence would be essential to determine the presence of group differences in social-behavioral functioning. Since we were interested in how the 22q11DS group would compare to the controls on the parent ratings of social-behavior, we conducted the subsequent analyses examining the data without IQ as a covariate, and then conducted secondary analyses including IQ as a covariate.

CBCL Parent Summary Scales—Controlling for ethnicity, the MANCOVA showed an overall significant group difference in parental reports of the social-emotional behavior of children with 22q11DS as compared to controls, F (3, 117) = 5.33, $p < .01$. As can be seen in Table 2, follow-up univariate comparisons showed that parents of children with 22q11DS reported significantly greater Internalizing problems, F (3, 121) = 19.1, $p =<.01$, and Total problems, F (3, 121) = 27.78, $p < .001$, than did parents of control children. Medium and large effect sizes were present for both findings. There was no significant difference between the groups on the CBCL Externalizing problems scale, F (3, 121) = 2.88, $p < .09$.

A second MANCOVA was conducted which included the same variables as the original analysis; however, IQ was included as a covariate. The results did not change when IQ group differences were accounted for. When controlling for ethnicity and IQ, the MANCOVA again showed a significant group difference for the CBCL parent summary scales, F (3, 117) = 5.33, $p < .01$. As in the original analysis, follow-up univariate comparisons showed the 22q11DS and control groups to be significantly different on the Internalizing problems scale, F (1, 119) = 8.53, $p < .004$, and the Total problems scale, F (1, 119) = 6.25, $p < .01$. There was no difference between the groups on the CBCL Externalizing Problems scale, F (1, 119) = 0.11, $p < .74$.

CBCL Parent Clinical Scales—Initial examination of the T-scores for the clinical scales showed the 22q11DS group to have scores that were elevated more than one standard deviation past the mean on five of the eight scales (Table 2). For the CBCL clinical scales, again controlling for ethnicity, the MANCOVA showed a significant overall group difference, F (8, 116) = 8.81, $p < .000$. As can be seen in Table 2, the 22q11DS group received significantly higher ratings (i.e., greater impairment) on all of the CBCL clinical scales, except Delinquent and Aggressive. For the scales with significant differences, the effect sizes ranged from small (Attention, Anxious/Depressed) medium (Withdrawn, Somatic, Thought Problems) to large (Social Problems).

Entering ethnicity and IQ into the MANCOVA as covariates produced a significant group difference, F (8, 112) = 2.73, $p < .009$. Follow-up group comparisons showed the 22q11DS group to receive higher (i.e., more impaired) ratings on Somatic Problems, F (1, 119) = 7.46, $p < .007$; Social Problems, F (1, 119) = 8.61, $p < .004$; and Attention Problems, F (1, 119) = 3.84, $p < .05$. None of the other parent clinical scales showed group differences.

Teacher Ratings

Since we collected teacher rating scales from fewer subjects (22q11DS=54 and controls=44) than those who provided parent CBCL reports, we further examined the demographics of the subjects for which teacher ratings were obtained. Preliminary data analyses revealed significant group differences only on IQ, t (96) = 10.71, $p < .000$, with the control group

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being higher in functioning. The groups were not different on SES, t (88) = 0.94, $p < .34$; chronological age, t (97) = 0.76, $p < .44$; gender, Fisher's exact test p= 0.73; or ethnicity, X^2 (3) = 2.90, $p < .41$. As with the parent ratings, the teachers' ratings were examined with and without IQ as a covariate.

Teacher Rating Form Summary Scales—The overall MANCOVA showed no significant overall group difference, F(3, 93)= 1.0, p=>0.05. Furthermore, none of the summary scales were significantly elevated. Table 3 presents the differences in teacher ratings of the social-emotional behavior of children with 22q11DS relative to controls. Controlling for IQ, the MANCOVA did not show a significant overall group difference, F (3, 93) = 1.02, $p < .39$; consequently, no follow-up univariate comparisons were conducted.

Teacher Rating Forms Clinical Scales—Initial examination of the individual clinical scales for the 22q11Ds group showed only one to have a significant rating that was more than one standard deviation above the mean (Social Problems). The MANCOVA showed a significant difference between the groups on the TRF clinical scales, F (8, 89) = 5.51, $p < .000$. As can be seen in Table 3, the follow-up univariate analyses revealed the groups to be significantly different on two scales: Withdrawn Problems, F (1, 96) = 6.48, $p < .01$, and Social Problems, F (1, 96) = 24.07, $p < .000$. While a small effect size was present for Withdrawn Problem, η_p^2= .06, a large effect size was seen for Social Problems, η_p^2= .20. None of the other TRF clinical scales showed significant differences between children with 22q11DS and unimpaired peers.

When IQ was entered into the model as a covariate, there continued to be a significant difference between groups on the MANCOVA, F (8, 87) = 2.23, $p < .03$. When examining the follow-up univariate comparisons, however, none of the individual clinical scales were significantly different between the groups, with all of the effect sizes falling within the small range.

The Relationship between Parent and Teacher Ratings of Social-Behavioral Functioning

Table 4 presents the partial correlations between parent and teacher CBCL summary scores and Table 5 for the clinical scales. In these analyses it was decided to control for medication status and type of teacher (i.e., regular education or special education). The effect of teacher type on the findings was controlled as it was found that special education teachers were more likely to report problematic behaviors in our study. When examining the magnitude of the correlations, teacher and parent reports of social-behavioral functioning of children with 22q11DS were largely discordant (Table 4). No significant inter-rater correlations were found for any of the three summary scales; however, significant moderate correlations occurred for parent and teacher reports on Withdrawal, r(40) = 0.48, p = 0.001; Thought Problems, r(40) = 0.43, p = 0.005; and Anxious/Depressed, r(40) = 0.34, p = 0.026. These results indicated that, for these domains, teachers and parents were identifying similar patterns of behavior for children with 22q11DS. Furthermore, when the T-scores for each of these three scales were examined, both parents and teachers were reporting mild to moderate concerns for these scales (i.e., the same severity levels). Interestingly, teacher report of thought problems was also significantly correlated with a number of other parent report

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scales, including Rule Breaking/Delinquent Behavior, $r(40) = 0.42$, $p = 0.006$ and Aggressive Behavior, $r(40) = 0.35$, $p = 0.025$). Of note, correlations between teacher and parent report for summary and clinical scales were considerably stronger for children in the control group than for those with 22q11DS (Table 5).

In addition to calculating partial correlations between respondents' ratings of behavior, agreement between raters on significant CBCL scales for both groups was calculated using Bland-Altman plots. In contrast to measures of correlation, which reflect which variables are moving in the same direction, Bland-Altman Plots illustrate how similar the respondents were in what they were reporting. Thus, agreement is often considered to be a more reliable indicator of concordance between multiple measures. Regarding the Internalizing problems summary scores, Bland-Altman found modest agreement between parent and teacher responses for 22q11DS participants, with 8.6% of the variance in teacher report being explained by parent report (R^2=0.086). Agreement was stronger for teacher and parent Withdrawn scores, explaining 28% of the variance (R^2=0.279). When rating control participants, teacher and parent agreement was moderate for the Internalizing problems summary scale (R^2=0.412) and the Withdrawn clinical scale (R^2=0.324). These results are presented in Figure 1A-D.

Discussion

This study is the first to evaluate and compare parent and teacher perceptions of the behavior and social-emotional functioning of children with 22q11DS and their unimpaired peers. Previous research has shown that children with 22q11DS are at increased risk for internalizing symptoms, such as anxiety, depression, withdrawal, obsessions, compulsions, and social skill impairments (Jansen et al., 2007; Woodin et al., 2001; Shashi et al., 2011). Results of the present study provide further evidence for this, particularly in the areas of internalizing behavior, social problems, and thought problems. This was especially true in the home setting where parents of children with 22q11DS were significantly more likely to report internalizing problems, than were parents of control children. In contrast, teachers indicated perceiving few significant differences between the functioning of 22q11DS children and their typically developing classmates. Teacher responses did corroborate significant difficulties with social problems and withdrawal in children with 22q11DS, as compared to their typical peers; however, teachers did not note significant differences between 22q11DS and control groups regarding thought problems or overall internalizing problems.

The discordancy noted between parent and teacher reports in our study is consistent with similar findings in the general population (Cai et al., 2004; Verhulst & Akkerhuis, 1989; Youngstrum, Loeber, &Stouthamer-Loeber, 2000) and among children with intellectual disabilities (Tassé & Lecavalier, 2000). Indeed, correlational analyses in the present study concluded that parents and teachers are providing disparate reports about the emotional and behavioral functioning of children with 22q11DS. In fact, within the 22q11DS group, teacher and parent reports were discrepant for all three summary scales of the CBCL: Internalizing problems, Externalizing problems, and Total problems. The only areas of agreement between teacher and parent report were withdrawal, thought problems, and

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anxious/depressed. Bland-Altman plots revealed significantly high levels of agreement between parents and teachers on withdrawn scores and less agreement on the internalizing symptoms in the 22q11DS group. This result suggests that parents and teachers rarely reported similar concerns for the same child.

The lack of significant correlations between parent and teacher report on the three summary scales of the CBCL in the 22q11DS group is consistent with previous research in the general population and among children with disabilities concluding that parents and teachers tend to exhibit low to moderate correlations when rating children's social-emotional behavior, with parents reporting more overall problems (Cai et al., 2004; Verhulst & Akkerhuis, 1989; Youngstrum et al., 2000). This discordance is strikingly apparent for internalizing symptoms, such as anxiety (Cai et al., 2004; Verhulst & Akkerhuis, 1989). This finding has particular relevance to the emotional/behavioral assessment of children with 22q11DS, as affected individuals are more likely to experience internalizing than externalizing difficulties. Consequently, teachers and parents may be particularly discordant in evaluating the behavior of children with 22q11DS.

An interesting finding in the present study was that teachers' reports of thought problems were significantly correlated with parent report of rule breaking/delinquent behavior and aggressive behavior. As measured by the CBCL, thought problems may include exhibiting strange or repetitive behaviors, difficulty getting one's mind off of a topic, having strange ideas, and seeing things that others cannot. This relationship suggests that children with 22q11DS who are exhibiting problematic externalizing symptoms at home, such as disobeying rules, noncompliance, and aggression, are being seen as exhibiting thought problems in the classroom.

The discrepancy between parent and teacher behavior ratings in the 22q11DS group may be explained by situational differences in children's behavior, with children exhibiting greater behavioral problems in the home setting than in the classroom (Cai et al., 2004; Verhulst & Akkerhuis, 1989), in part due to differing behavioral contingencies across settings. Or, a teacher with many students who demonstrate challenging behaviors may underreport a child's social-emotional difficulties, particularly given the internalizing nature of the behavior displayed by most children with 22q11DS. Not surprisingly, the emotional and behavioral difficulties experienced by children with 22q11DS may often be eclipsed by the more salient and disruptive behavior of classmates who exhibit aggressive, hyperactive, or conduct behaviors. Interestingly, agreement was higher between teachers and parents for internalizing symptoms in the control group, reflecting our findings of better correlations between parents and teachers for the control group. Although the latter finding may be reflective of a restricted range of ratings for the control group, it raises concerns for how social-behavioral assessments are conducted for children with 22q11DS. In general, the need for multi-raters in multiple settings using multiple instruments is critical for a thorough clinical assessment. Otherwise, in cases where eligibility for treatment or educational services is decided solely based on teacher report of behavior, children with 22q11DS may be denied necessary interventions.

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Young and colleagues (2011) reported that, despite compelling evidence that children with 22q11DS display social-emotional deficits that exceed those of their unimpaired peers, they are less likely to receive intervention (both medical and behavioral) for these challenges. The current study yielded evidence that teachers may not be sensitive to the behavioral difficulties of children with 22q11DS. Thus, the lack of treatment utilization in this population may be a result of discrepant behavior ratings across respondents, with teachers reporting minimal differences between the classroom behavior of 22q11DS children and their classmates. Children may not be deemed eligible for special education services or referred for psychosocial therapy because their teachers are not reporting clinically significant social-emotional behavior problems. In order to maximize participation in treatment by this group of children, clinicians assessing the needs of individuals with 22q11DS should collect behavioral data from multiple informants using multiple methods. Specifically, semi-structured interviews, classroom and home observations of behavior, projective testing, and other sociometric scales of behavior can be used to increase the reliability and validity of emotional and behavioral assessments.

Lastly, the current study results are relevant to the assessment and prevention of psychosis in individuals with 22q11DS. Given the high prevalence rates of psychosis in this population, particularly schizophrenia and schizophrenia spectrum disorders, numerous research studies have attempted to identify early risk factors associated with the development of psychosis later in life. Gothelf and colleagues (2007) assessed the psychiatric and adaptive functioning of children with 22q11DS and matched controls at baseline. At the five year follow-up, 32.1% of 22q11DS participants had developed psychotic disorders, as compared with 4.3% of control participants. Among affected individuals, baseline symptoms of anxiety or depression predicted 61% of the variance in severity of psychosis at follow-up. These findings emphasize the importance of accurately assessing internalizing features, such as anxiety and depression, in youth with 22q11DS.

Limitations

Results of the current study should be interpreted within the framework of a few limitations inherent to the research design and methodology. In particular, the primary data collection instruments were parent and teacher behavior rating scales. As such, they measure only those behaviors and abilities that are represented by the scales. A semi-structured interview may have allowed the investigators to assess for a wider range of social-emotional challenges, while concurrently obtaining more specific information about setting events for problematic behaviors and the specific nature of fears and anxieties. This type of information, in part, may have increased the explained inconsistencies between parent and teacher reports.

Furthermore, the control group that was included in the investigation did not exclude children with ADHD. Because attention deficits are also particularly common in the 22q11DS population, it is likely that a large degree of overlapping behavioral symptoms were present across groups, making it difficult to detect group differences in emotional/behavioral functioning. Despite this, we were still able to identify significant differences in emotional and behavioral functioning between the two groups.

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Lastly, preliminary analyses indicated that special education teachers in the sample were more likely to report problem behaviors than were general education teachers. However, not every child receiving special education services was rated by his/her special education teacher. Therefore, the data collected may underestimate the behavioral difficulties of children with 22q11DS who were receiving educational support.

Conclusion

The current investigation provides compelling evidence that parents are more likely to report emotional/behavioral problems in children and adolescents with 22q11DS than for their unaffected peers. In particular, parents reported elevated internalizing problems, such as withdrawal, social problems, and thought problems. In contrast, teachers perceive few significant differences in the social-emotional functioning of 22q11DS children, as compared with their unaffected classmates and there is little concordance between parent and teacher reports in the 22q11DS group as compared to the control group. This report provides a first exploration of the differential perception between parents and teachers of the social-emotional problems in children with 22q11DS. Ongoing education of teachers and other professionals on the features of the disorder appear warranted. Further, it will be important for evaluators to use multiple methods of social-behavioral assessments across different settings and informants to increase the chances of identifying specific problems in children and adults with 22q11DS.

References

Achenbach, TM. Manual for the Child Behavior Checklist/4-18 and 1991 profile. Burlington, VT: University of Vermont, Department of Psychiatry; 1991a.

Achenbach, TM. Integrative Guide for the 1991 CBCL 4-18, YSR, and TRF profiles. Burlington, VT: University of Vermont, Department of Psychiatry; 1991b.

Briegel W, Schneider M, Otfried Schwab K. 22q11.2 deletion syndrome: Behaviour problems of children and adolescents and parental stress. Child: Care, Health and Development. 2007; 34:319–324.

Cai X, Kaiser AP, Hancock TB. Parent and teacher agreement on child behavior checklist items in a sample of preschoolers from low-income and predominantly african american families. Journal of Clinical Child and Adolescent Psychology. 2004; 33:303–312. [PubMed: 15136195]

De Smedt B, Devriendt K, Fryns JP, Vogels A, Gewillig M, Swillen A. Intellectual abilities in a large sample of children with velo-cardio-facial syndrome: An update. Journal of Intellectual Disabilities Research. 2007; 51:666–670.

Dennis M, Francis DJ, Cirino PT, Schachar R, Barnes MA, Fletcher JM. Why IQ is not a covariate in cognitive studies of neurodevelopmental disorders. Journal of the International Neuropsychological Society. 2009; 15:331–343. [PubMed: 19402919]

Fagan J, Fantuzzo J. Multirater Congruence on the Social Skills Rating System: Mother, Father, and Teacher Assessments of Urban Head Start Children's Social Competencies. Early Childhood Research Quarterly. 1999:229–242.

Feinsten C, Eliez S, Blasey C, Reiss A. Psychiatric disorders and behavioral problems in children with velocardiofacial syndrome: Usefulness as phenotypic indicators of schizophrenia risk. Biological Psychiatry. 2002:312–318.

Gothelf D, Feinstein C, Thompson T, Gu E, Penniman L, Van Stone E, Kwon H, Eliez S, Reiss A. Risk factors for the emergence of psychotic disorders in adolescents with 22q11.2 deletion syndrome. American Journal of Psychiatry. 2007; 164:663–669. [PubMed: 17403981]

Gothelf D, Lombroso P. Genetics of childhood disorders: XXV. Velocardiofacial syndrome. Journal of the American Academy of Child and Adolescent Psychiatry. 2001; 40

NIH-PA Author Manuscript

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NIH-PA Author Manuscript

Jansen P, Dujiff S, Beemer F, Vorstman J, Klaassen P, Morcus M, Heineman-de Boer J. Behavioral problems in relation to intelligence in children with 22q11.2 deletion syndrome: A matched control study. American Journal of Medical Genetics. 2007; 143:574–580. [PubMed: 17318841]

Knoff, HM. Best practices in personality assessment. In: Thomas, A.; Grimes, J., editors. Best practices in school psychology IV. Bethesda, MD: National Association of School Psychologists; 2002. p. 1281-1302.

Lajiness-O'Neill R, Beaulieu I, Asamoah A, Titus J, Bawle E, Ahmad S, Kirk J, Pollack R. The neuropsychological phenotype of velocardiofacial syndrome (VCFS): Relationship to psychopathology. Archives of Clinical Neuropsychology. 2006; 21:175–184. [PubMed: 16307864]

Lewandowski KE, Shashi V, Berry PM, Kwapil T. Schizophrenic-like neurocognitive deficits in children and adolescents with chromosome 22q11.2 deletion syndrome. American Journal of Medical Genetics Part B: Neuropsychiatric Genetics. 2007; 144:27–36.

McDonald-McGinn DM, Kirschner R, Goldmuntz E, Sullivan K, Eicher P, Gerdes M, Zackai EH. The Philadelphia story: The 22q11.2 deletion: Report on 250 patients. Genetic Counseling. 1999; 10:11–24. [PubMed: 10191425]

McDonald-McGinn DM, LaRossa D, Goldmuntz E, Sullivan K, Eicher P, Gerdes M, Zackai EH. The 22q11.2 deletion: Screening, diagnostic workup, and outcome of results; Report on 181 patients. Genetic Testing. 1997; 1:99–108. [PubMed: 10464633]

Moss E, Batshaw M, Solot C, Gerdes M, McDonald-McGinn D, Driscoll D, Emanual B, Zackai E, Wang P. Psychoeducational profile of the 22q11.2 microdeletion: A complex pattern. Journal of Pediatrics. 1999; 134

Murphy KC, Jones LA, Owen MJ. High rates of schizophrenia in adults with velo-cardiofacial syndrome. Archives of General Psychiatry. 1999; 56:940–5. [PubMed: 10530637]

Niklasson L, Rasmussen P, Óskarsdóttir S, Gillberg C. Neuropsychiatric disorders in the 22q11 deletion syndrome. Genetics in Medicine. 2001; 3:79–84. [PubMed: 11339385]

Niklasson L, Rasmussen P, Óskarsdóttir S, Gillberg C. Chromosome 22q11 deletion syndrome (CATCH 22): Neuropsychiatric and neuropsychological aspects. Developmental Medicine & Child Neurology. 2002; 44:44–50. [PubMed: 11811651]

Papolos DF, Faedda GL, Veit S, Goldberg R, Morrow B, Kucherlapati R, Shprintzen RJ. Bipolar spectrum disorders in patients diagnosed with velo-cardiofacial syndrome: Does a hemizygous deletion of chromosome 22q11 result in bipolar affective disorder? American Journal of Psychiatry. 1996; 153:1541–7. [PubMed: 8942449]

Rutter, M.; Graham, P.; Yule, W. Clinics in Developmental Medicine Nos 35-36. London: Spastics International Medical Publications/Heinemann Medical Books; 1970. A neuropsychiatric study in childhood.

Sattler, JM.; Hoge, RD. Assessment of Children: Behavioral, social, and clinical foundations. 5th. San Diego, CA: Jerome M. Sattler, Publisher, Inc.; 2006.

Shashi V, Veerapandiyan A, Schoch K, Kwapil T, Keshavan M, Ip E, Hooper S. Social skills and associated psychopathology in children with chromosome 22q11.2 deletion syndrome: Implications for interventions. Journal of Intellectual Disabilities Research. 2011; 56(3):1–14.10.1111/j.1365-2788.2011.01477.x

Schoch K, Harrell W, Hooper SR, Ip EI, Saldana S, Kwapil TR, Shashi V. Applicability of the Nonverbal Learning Disability Model for Children with 22q11.2 Deletion Syndrome. Journal of Learning Disabilities. in press.

Shprintzen RJ, Goldberg R, Golding-Kushner KJ, Marion RW. Late-onset psychosis in the velo-cardiofacial syndrome. American Journal of Medical Genetics. 1992; 42:141–2. [PubMed: 1308357]

Sobin C, Kiley-Brabeck K, Hadley Monk S, Khuri J, Karayiorgou M. Sex differences in the behavior of children with the 22q11 deletion syndrome. Psychiatry Research. 2009; 166:24–34. [PubMed: 19217670]

Swillen A, Devriendt K, Legius E, Eyskens B, Dumoulin M, Gewillig M, Fryns J. Intelligence and psychosocial adjustment in velocardiofacial syndrome: A study of 37 children and adolescents with VCFS. Journal of Medical Genetics. 1997; 34:453–458. [PubMed: 9192263]

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Tassé M, Lecavalier L. Comparing parent and teacher ratings of social competence and problem behaviors. American Journal on Mental Retardation. 2000; 105:252–259. [PubMed: 10934567]

Tezenas Du Montcel S, Mendizabai H, Ayme S, Levy A, Phillip N. Prevalence of 22q11 microdeletion. Journal of Medical Genetics. 1996; 33:719. [PubMed: 8863171]

Verhulst FC, Akkerhuis GW. Agreement between parents' and teachers' ratings of behavioral/emotional problems of children aged 4-12. Journal of Child Psychology and Psychiatry. 1989; 30:123–136. [PubMed: 2925818]

Woodin M, Wang P, Aleman D, McDonald-McGinn D, Zackai E, Moss E. Neuropsychological profile of children and adolescents with the 22q11.2 microdeletion. Genetics in Medicine. 2001; 3:34–39. [PubMed: 11339375]

Young AS, Shashi V, Schoch K, Kwapil T, Hooper SR. Discordance in diagnoses and treatment of psychiatric disorders in children and adolescents with 22q11.2 deletion syndrome. Asian Journal of Psychiatry. 2011; 4:119–24. [PubMed: 21743818]

Youngstrom E, Loeber R, Stouthamer-Loeber M. Patterns and correlates of agreement between parent, teacher, and male adolescent ratings of externalizing and internalizing problems. Journal of consulting and clinical psychology. 2000; 68:1038–1050. [PubMed: 11142538]

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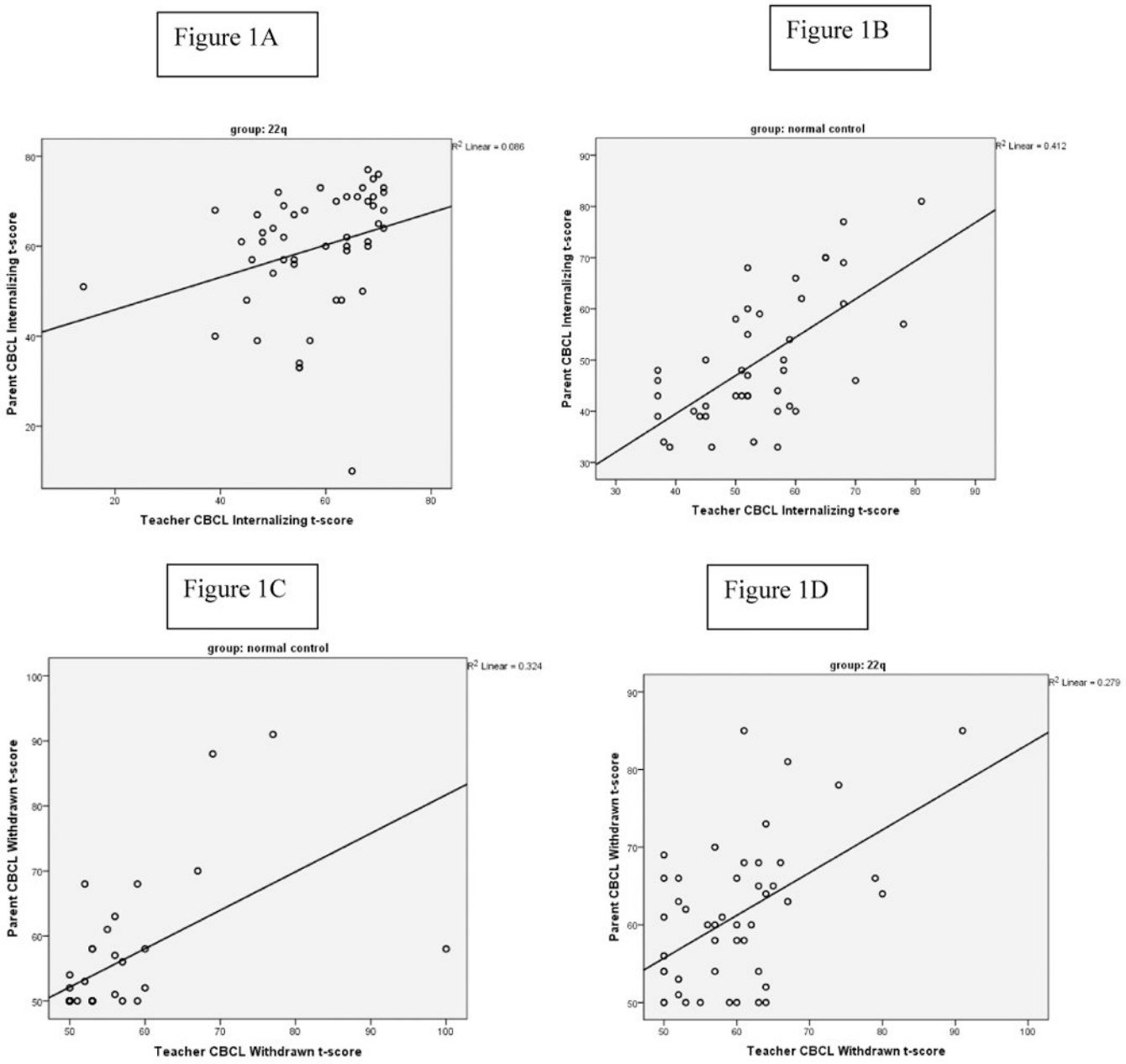

Figure 1A-D.

Bland-Altman plots of agreement between parent and teacher reports on behavioral and emotional problems in children with 22q11Ds, compared to control subjects. R^2 indicates the percentage of agreement between the parents and the teachers for each measure

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Table 1

Participant Sociodemographic Information

	22q11DS Mean (SD)	**Control Mean (SD)**	**t-value**
Overall SES	31.68 (13.33)	34.37 (13.89)	-1.03
Age at initial assessment	10.47 (2.57)	11.16 (2.22)	-1.60
WISC-4 FSIQ	73.28 (12.38)	97.68 (11.91)	-11.12***
Gender	49% Male	53% Male (Fisher's Exact Test p=.725	
Ethnicity	86% Caucasian	64% Caucasian (X^2=18.5, p<.01)3	

Notes. WISC-4 = Wechsler Intelligence Scale for Children – 4th Edition.

FSIQ= Full-scale IQ

*** p<0.001,

** p<0.01,

* p<0.05

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Table 2

Group Comparisons on the Parent CBCL Rating Scale Controlling for Ethnicity

CBCL Scale	22qDS11 (n = 67)	Control (n = 59)	F-Value	p-Value	Partial Eta Square
		Summary Scales			
Internalizing	59.81 (12.86)	49.69 (12.74)	19.10	.002	**.134**
Externalizing	52.91 (9.24)	49.66 (11.5)	2.88	.09	.023
Total Problem Score	61.18 (9.66)	50.76 (12.25)	27.78	.000	**.184**
		Clinical Scales			
Anxious/Depressed	59.66 (9.18)	54.75 (8.15)	9.64	.002	.073
Withdrawn	61.00 (8.89)	55.12 (8.65)	14.05	.000	**.103**
Somatic	62.18 (9.08)	55.39 (7.24)	20.52	.000	**.143**
Social Problems	64.21 (8.64)	54.29 (7.83)	44.78	.000	**.267**
Thought Problems	61.67 (9.77)	54.80 (7.83)	18.10	.000	**.128**
Attention	64.78 (9.85)	58.76 (8.74)	13.18	.000	.097
Delinquent	54.57 (4.90)	54.76 (6.34)	0.05	.824	.000
Aggressive	56.13 (6.77)	54.59 (6.95)	1.43	.234	.012

Notes. CBCL = Child Behavior Checklist. Medium and large effect sizes are in bold

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Table 3

Group Comparisons on the Teacher TRF Rating Scale

TRF Scale	22qDS11 (n = 54)	Control (n = 44)	F-Value	p-Value	Partial Eta Square
		Summary Scales			
Internalizing	57.96 (11.02)	53.73 (10.7)	3.7	.06	.03
Externalizing	52.39 (9.76)	53.23 (9.50)	.14	.7	.001
Total Problem Score	57.7 (10.29)	55.86 (8.91)	.88	.35	.009
		Clinical Scales			
Anxious/Depressed	58.91 (8.27)	57.09 (8.02	1.20	.28	.01
Withdrawn	59.65 (8.64)	55.09 (9.03)	6.48	.01	.063
Somatic	55.85 (7.24)	54.16 (6.04)	1.53	.22	.016
Social Problems	62.89 (8.27)	55.16 (7.07)	24.07	.000	**.200**
Thought Problems	56.57 (8.68)	53.73 (5.95)	3.42	.07	.034
Attention	59.61 (8.15)	58.07 (6.92)	0.;99	.32	.010
Delinquent	53.59 (5.05)	54.39 (6.76)	0.44	.507	.005
Aggressive	55.74 (5.77)	55.82 (8.62)	0.003	.96	.000

Notes. TRF = Teacher Report Form. Medium and large effect sizes are in bold

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Table 4

Parent-teacher Correlations of Summary Scale Reports in Children with 22q11DS and Controls

	TEACHER RATINGS					
PARENT RATINGS	**Internalizing**		**Externalizing**		**Total Problems**	
	22q11	**Controls**	**22q11**	**Controls**	**22q11**	**Controls**
Internalizing	0.14	(.63)**	0.02	.42*	0.06	.54**
Externalizing	0.04	0.17	0.18	0.22	0.04	0.15
Total Problems	0.14	.50**	0.15	0.28	0.1	.41*

Notes. Correlations were calculated controlling for Type of Teacher and Medication Status. CBCL = Child Behavior Checklist.

*** $p<0.001$,

** $p<0.01$,

* $p<0.05$.

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Table 5

Pearson Partial Correlations of Parent-teacher Ratings of Clinical scales Children with 22q11DS and Control Children

	TEACHER RATINGS															
	Anx/Dep		Withdrawn		Somatic		Soc Probs		Thought Probs		Attent Probs		Rule Break		Aggression	
PARENT RATINGS	**22q**	**Con**	**22q**	**Con**	**22q**	**Con**	**22q**	**Con**	**22q**	**Con**	**22q**	**Con**	**22q**	**Con**	**22q**	**Con**
Anxious/Depressed	.34*	.67***	0.17	.49*	-0.26	.42*	0.12	.85***	0.25	0.24	0.12	0.29	-0.01	0.22	0.04	0.31
Withdrawn	0.09	.61**	.48**	.56**	-0.11	0.32	0.19	.76***	0.13	0.2	-0.02	0.28	0	0.31	-0.02	0.23
Somatic	0.09	.46*	-0.07	0.11	0.14	0.15	0.07	.45*	0.25	0.2	-0.02	0.39	0.09	0.1	0.2	0.32
Social Problems	0.04	.60**	0.17	0.35	-0.05	0.32	0.09	.77***	0.1	0.07	-0.06	0.14	-0.02	0.1	0.1	0.24
Thought Problems	0.02	.46*	-0.01	0.33	-0.28	0.21	0.02	.69***	.43**	0.27	-0.12	0.2	-0.1	-0.08	-0.02	0.08
Attention Problems	-.33*	0.14	-0.13	0.28	0.07	0.23	-0.13	0.28	0.07	0.13	0.01	.44*	-0.05	0.14	0.01	0.02
Rule Break /Delinq	0.09	0.12	0	0.17	0.07	0.27	0.11	.52**	.42**	0.03	0.17	0.11	0.14	0.2	0.16	0.14
Aggression	0.25	0.34	0.05	0.29	-0.17	0.31	0.14	.63**	.35*	0.05	0.04	-0	0.02	0.16	0.1	0.13

Notes. Correlations were calculated controlling for Type of Teacher and Medication Status. CBCL = Child Behavior Checklist. Con = Control Group.

*** $p<0.001$,

** $p<0.01$,

* $p<0.05$.