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Social Skills and Executive Function Deficits in Children With the 22q11 Deletion Syndrome

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Abstract

The 22q11 Deletion Syndrome (22q11DS) is among the most frequent gene deletion disorders, occurring once in every 6,000 live births. Descriptive reports have suggested marked social differences in affected children. Empirical studies are needed to verify possible social skills deficits among children with 22q11DS, and also to examine possible associations between their frequently reported executive function deficits and social anomalies.

Fifty-two parents of affected children (n = 52) and participating control siblings (n = 26) completed the Social Skills Rating System (SSRS) and Behavior Inventory of Executive Function (BRIEF).

When compared with control siblings, children with 22q11DS had significantly lower SSRS ratings for Cooperation, Assertion, Responsibility, and Self-Control. Affected children had significantly higher BRIEF ratings for Initiation, Planning, Working Memory, and Monitoring. In affected children, global Social Skill was negatively correlated with BRIEF Global Composite scores. Initiation and Monitoring significantly predicted Social Skill. Children with 22q11DS have marked differences in social skill development which are associated with executive dysfunction.

Keywords

22q11 Deletion Syndrome; executive function; social skill; remediation; children; neuropsychology

The 22q11 Deletion Syndrome (22q11DS) is one of the most common known genetic disorders, estimated to occur in one of every 6,000 live births (Botto et al., 2003). The syndrome results in the loss of a 1.5 megabase region on the long arm of chromosome 22 at the 11.2 site. In the vast majority of cases, the deletion is not transmitted by either parent (*de novo*). Children with 22q11DS have an array of anomalies believed to be associated with the loss of genes in this region, including physical, neurocognitive, behavioral, and social differences. With regard to their physical phenotype, over 180 possible anomalies have been described (Ryan et al., 1997), the most common of which include structural differences of the head, ears, throat, and neck, possibly accompanied by early feeding difficulties (reflux), immunologic problems, heart defects of widely varying severity, and early hypotonia. Prior to the identification of a single underlying deletion, children with this syndrome were identified according to their primary physical problem, including DiGeorge Syndrome (primary immunological deficit), Conotruncal Anomaly Face Syndrome (primary heart defect with facial dysmorphologies), and

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Velo-Cardio-Facial Syndrome (primary dysmorphologies of the face and head with varying cardiac irregularities). Any given child, however, may have several or none of these physical problems (although most children have been noted to have at least minor structural facial differences). In this way, the physical phenotype associated with 22q11DS is quite broad, and may determine whether and how early a child's deletion is detected.

Although their physical phenotype varies greatly, the neurocognitive development of children with 22q11DS is proving to be far more consistent. Woodin et al. (2001) administered neuropsychological batteries to 50 children with 22q11DS who ranged in age from 6 to 17. Performances on a sequencing test (Trail-Making Test A) was within normal limits; however, children with 22q11DS had impaired performances (scores greater than 2 standard deviations below the mean) on an executive function test of visual scanning and working memory (Trail-Making Test B).

Sobin, Daniels, et al. (2005) administered the NEPSY and Stanford—Binet Intelligence Scale to 35 children with 22q11DS and 12 unaffected control siblings who ranged in age from 5 to 12 years old. No differences were found between the groups on tests of verbal or quantitative ability. However, children with 22q11DS had impaired performance on measures of visual attention, sensorimotor ability, and executive function.

Neurocognitive tests with specific links to underlying brain pathways have also been administered. The Attention Network Test was administered to 32 children with 22q11DS and 20 control siblings. The children ranged in age from 5 to 11.5 years old. Children with 22q11DS did not differ from control siblings on the Orienting or Alerting Attention measures; however, affected children's Executive Attention scores were in the impaired range (Sobin et al., 2004).

Sobin, Kiley-Brabeck, and Karayiorgou (2005) also administered a prepulse inhibition paradigm (PPI) to 25 children with 22q11DS and 23 control siblings who ranged in age from 6 to 13 years old. The paradigm was designed to assess pre-attentive processing, or "sensorimotor gating" in children. Significant group differences were found; PPI in children with 22q11DS was 20% less than in control siblings.

Thus, recent studies suggest that children with the 22q11DS have marked impairment in visual attention and executive function (Sobin, Daniels, et al., 2005; Sobin et al., 2004; Sobin, Kiley-Brabeck, & Karayiorgou, 2005; Woodin et al., 2001) and a range of secondary learning problems that appear to stem from these primary deficits. With regard to behavior, disinhibition, impulsivity, withdrawal, and shyness have been most consistently reported (McCandless, Scott, & Robin, 1998; McDonald-McGinn et al., 1997; Thomas & Graham, 1997). The impact that attention and executive function deficits have on daily behaviors, however, has never been studied in children with 22q11DS.

Social skill differences among children with 22q11DS have rarely been examined, and current reports are predominantly descriptive. Poor social competence, concrete thinking, and difficulties generalizing previous experience to novel situations, were among the first social traits to be observed among children with 22q11DS (Furst, Dool, & Rourke, 1995). Affected children were also described as having poor social interactions, impaired decision-making skills (Thomas & Graham, 1997), and general social immaturity (Shprintzen, 2000). The only empirical analysis of social behavior in children with 22q11DS (Woodin et al., 2001) was in a study reporting mean scores for eight subscales on the Achenbach Child Behavior Checklist (Achenbach, 1991). In this study, mean subscale scores in the clinically significant range (*T* score > 65) for 50 children with the 22q11DS ranging in age from 6 to 17 were found for only the Social Problems and Attention subscales.

Social functioning is a critical aspect of a child's life. Social skill deficits markedly impact a child's immediate quality of life as well as their long-term functioning, within the family as well as among peers. Prosocial behavior has been associated with academic success as suggested by a study showing that peer nominations of cooperative and helpful students were associated with greater academic competence (Wentzel, 1993). In this study, prosocial behavior was an independent positive predictor of both grade point average (GPA) and Stanford Test of Basic Skills scores (STBS) among 423 sixth and seventh graders, 52% of whom were male. Conversely, antisocial behavior (measured by peer nominations of children who start fights and break rules) was an independent negative predictor of GPA and STBS. (Controlling for confounding variables such as absenteeism, teacher preference, IQ, family environment, sex, and ethnicity did not change these associations.)

Social skills typically refer to specific prosocial abilities that increase the likelihood of positive evaluation and positive responses from others (Mash & Terdal, 1997); these skills include actions such as sharing, helping, initiating conversations, asking for help, and giving compliments (Elliot, Malecki, & Demaray, 2001). Prosocial behavior also includes more complex sets of behaviors such as adaptive social problem solving and the modulation of emotion (Mash & Terdal, 1997). Understanding of social skills has come in part from analysis of its developmental progression. Preschoolers' social behavior revolves around reciprocal, pretend play, and depends on the child's ability to attend to an activity, share roles, and display positive affect (Hymel & Rubin, 1985). During the grade school years, "best friendships" emerge and social interactions begin to include group games with complex rule sets. Successful grade-school social interactions require the ability to attend to, comprehend, maintain, and comply with group and social rule sets (Hartup, 1983). Norm-breaking behaviors in both elementary and secondary school often lead to peer rejection (Coie, Dodge, & Kupersmidt, 1990). In adolescence with the onset of puberty, teenagers become increasingly self-aware and self-conscious. More subtle types of reciprocity and communication become the foundations of successful adolescent social skills (Laursen, 1993).

Conceived in these ways, social competence requires a variety of cognitive abilities associated with executive function. Executive function refers to a core set of cognitive processes that are necessary to undertake and complete goal-directed behavior in novel problem-solving situations with multiple and perhaps conflicting response options (Welsh & Pennington, 1988). Many cognitive functions together are included in this process, including response inhibition, working memory, maintaining and shifting cognitive set, visualization and manipulation of information, strategizing, selecting a response from among competing choices, and maintaining task goals (Pennington & Ozonoff, 1996).

Moreover, the development of executive function overlaps that of social skill. The development of executive ability can be observed beginning around age 3 with the acquisition of response inhibition and impulse control (Diamond & Taylor, 1995). The most rapid period of measurable increase in executive function seems to occur between ages 5 and 8 (Anderson, Anderson, Northam, Jacobs, & Mikiewica, 2001; Klenberg, Korkman, & Latti-Nuuttia, 2001; Korkman, Kemp, & Kirk, 2001: Levin et al, 1991; Welsh, Pennington, & Grassier, 1991), although the subsets of abilities required for complex problem solving continue to develop through early and later adolescence (Korkman et al., 2001). Visual and auditory attention mature by age 10 (Klenberg et al., 2001); whereas executive fluency, described as the ability to plan, monitor, and evaluate performance (Klenberg et al., 2001), working memory (Klenberg et al., 2001; Korkman et al., 2001), and goal-setting abilities (Anderson et al., 2001), continue to develop throughout adolescence.

Thus, when considered in the context of social behavior, the functions that comprise "executive" ability are logically associated, and as Hartup (1985) suggested, social and

academic skills may be subserved by common self-regulatory abilities. Social interaction constitutes a constantly changing novel situation requiring the simultaneous evaluation of multiple types of information. It requires response delay, consistent visual and auditory attention, constant updating of information, and moment-to-moment evaluation of the course of the interaction. Flexibility is key, as is the comparison of past experience with present circumstances.

The possibility that behaviors associated with executive and attentional dysfunction may be associated with lowered social skill behavior is suggested by research in other pediatric populations. Most notably, marked deficits in social skill behavior have been repeatedly found in studies of children with Attention Deficit Hyperactivity Disorder (ADHD; American Psychological Association, 1994; see, e.g., Frankel & Feinberg, 2002; Hinshaw & Melnick, 1995; Hynd, Hern, & Voeller, 1991; Matthys, Cuperius, & Van England, 1999). However, specific associations between behaviors associated with attention and executive dysfunction, and social skill deficits, have only rarely been examined. One study that examined these associations used the Behavior Rating Inventory of Executive Function (BRIEF) and the Vineland Adaptive Behavior Scale (VBAS). The research was conducted with 35 children with autism, ranging in age from 6 to 17 years. The VBAS Socialization domain score was significantly correlated with BRIEF Initiate and Working Memory subscale scores (Gilotty, Kenworth, Sirian, Black, & Wagner, 2002).

Although children with the 22q11DS have been shown to have deficits on neurocognitive and neuropsychological tests that measure executive function and attention (Sobin, Daniels, et al., 2005; Sobin et al., 2004; Sobin, Kiley-Brabeck, & Karayiorgou, 2005; Woodin et al., 2001), whether the daily behavior of children with 22q11DS reflects these neurocognitive deficits has never been examined. Quantifying daily behaviors that may be associated with executive dysfunction is an important step in understanding the possible roots of social behavior differences.

This study had three aims. To expand the empirical literature on social skill behavior in children with the 22q11DS we compared empirical ratings of the social skill in affected children and their sibling controls. To determine whether deficits in executive function and attention would be evident in their daily behaviors, we compared ratings on a scale measuring daily behaviors associated with executive and attentional dysfunction in these same groups of children. To explore possible associations between behaviors associated with social skill and executive dysfunction, we examined correlations between behavioral measures of social skill and executive dysfunction. We hypothesized that, as compared with sibling controls, children with 22q11DS have lower scores on a measure of social skill competence, clinically significant (higher) ratings on a measure of daily behaviors associated with executive function deficits, and a correlation between global measures of social skill competence and daily behaviors associated with executive dysfunction.

METHOD

Participants

Data for this study are based on parent ratings of observable social skill behavior (Social Skills Rating System Scale, SSRS) and behaviors associated with executive dysfunction (BRIEF) for 78 children, including 52 confirmed positive (via florescence *in situ* hybridization) for 22q11DS (3.91–16.27 years old, $M = 8.41$, $SD = 2.88$) and 26 sibling control participants (3.58–13.08 years old, $M = 8.98$, $SD = 2.64$). The children and families are from highly diverse regions spanning an 880-mile radius around New York City. All siblings in the target age range who did not have a history of learning difficulties were invited to participate. There were no families

involved in the study that had more than one child available for any of the three age groupings (preschool, school-age, or secondary-level). For this study, all available participants were used.

The children are participants in an ongoing, federally funded longitudinal study at Rockefeller University in New York City. Families for this study were recruited via parent support groups, Web site postings, doctors' offices, and genetic counselors. Each family participated in an annual, 2-day assessment. Parent-report of social-emotional functioning, behaviors associated with executive function, social skills, temperament, and sensorimotor integration were obtained for all affected children and all control siblings. In addition, affected children were administered up to two neuropsychological batteries, two neurophysiologic tests, and computerized test batteries. The neuropsychological tests included measures of attention, executive function, visual—spatial processing, nonverbal reasoning, basic language and motor skills. Control siblings were administered neurophysiologic tests, computerized batteries, and tests of motor skill. Parental informed consent and child verbal assent was obtained on the first morning of testing.

An analysis was calculated to determine the power for the primary analyses including 43 affected children and 20 control siblings with equal variances. With effect size = 0.80, and alpha = 0.05, the projected power of the primary analyses was 0.95. Children with 22q11DS whose parents completed the SSRS and the BRIEF were offered feedback about their child's results. All other information pertaining to the affected child's neurocognitive performance was detailed in a formal report written on completion of the child's annual evaluation.

Genders were equally distributed between the groups with girls accounting for 48.1% (25/52) of the affected group and 51.3% (40/78) of the total sample ($df = 1$, $N = 78$, $\chi^2 = .64$, $p = .42$). The majority of participants were White or Non-Hispanic (81.2%, 69/85) with approximately 10.5% identified as Other or Mixed Heritage ($n = 9/85$), 3.5% African American (3/85), 2.4% Asian or Pacific Islander (2/85), and 2.4% Hispanic (2/85). Approximately 81% (80.7%) of participants were in elementary school (63/78). The remaining participants were in preschool (9.0%, 7/78) and secondary school (10.3%, 8/78). A chi-square analysis examined differences between affected and unaffected groups with regard to school level. The frequencies of school levels among affected and unaffected children did not differ ($df = 2$, $N = 78$, $\chi^2 = 1.68$, $p = .43$).

The Hollingshead Index of Social Position (ISP) was used to calculate the socioeconomic status of participating families. The mean ISP rating for the participating families was in the upper middle class ($M = 25.62$, $SD = 9.37$). Twelve percent of the participating families were rated as upper class, 64% were upper middle class, 22% were middle class, and 2% were lower middle class. None of the participating families were rated in the lower class bracket.

Instruments

Social Skills Rating System—The Social Skills Rating System—Parent Version (Gresham & Elliott, 1990) is a norm-referenced, parent-report behavior rating scale that was designed to screen and classify social skills of children in the preschool through high school grades. The SSRS computes a standardized Total Social Skills scaled score (TSSS), and four nominal behavior ratings for the social subskills of cooperation, responsibility, assertion, and self-control.

The SSRS was standardized on a national sample of 4,170 children from Grades 3 through 10 whose demographic characteristics were representative of children in the United States. A smaller, national, try-out sample of children ages 3 to 5 ($N = 200$) was utilized to develop the preschool norms. A recent review of six child-based social skills measures (Demaray, Ruffalo, & Carlson, 1995) recommended using the SSRS because of its sound reliability and validity,

multi-informant approach, and its direct links to interventions. Across all forms and levels of the SSRS, the median coefficient alpha reliability for the TSSS was .90 (Demaray et al., 1995). Parent test—retest reliability was .85 to .87 for Social Skills, .65 to .84 for Problem Behaviors, and .93 for Academic Competence (Gresham & Elliott, 1990).

The SSRS takes approximately 20 min to complete. Informants use a 3-point Likert-type scale that is intended to summarize the perceived frequency of particular types of behaviors. A rating of zero means the *behavior never happens*, a rating of 1 means the behavior *sometimes occurs*, and a rating of 2 means the behavior occurs *very often*. The SSRS ASSIST Computer Software program was used to score SSRS protocols for all participating children. Raw scores (0, 1, 2), subscale and TSSS were computed. Cutoff points of plus and minus 1 standard deviation established Behavior Level categories. The four subscale scores (cooperation, assertion, responsibility, and self-control) were reported as nominal behavior rating scores. A score of "1" indicated that the child had "fewer" of the given skills than same-age peers, a "2" indicated an "average" number of the skills, and a "3" indicated more of the rated skills. Computer derived standard scores were obtained for the TSSS (M = 100, SD = 15). After the data entry was completed, each protocol was examined for data entry accuracy and every fifth protocol was rechecked to ensure data accuracy.

Behavior Rating Inventory of Executive Functions—Parent Version—The BRIEF—Parent Version is an 86-item, parent-report rating scale of executive function behaviors. BRIEF items were selected to assess eight domains of everyday behavior most likely to occur among children with executive dysfunction, including inhibition (ability to not act on an impulse), shifting (ability to change freely from one situation, activity, or thought to another as the situation requires), emotional control (ability to regulate emotions), initiate (ability to self-start tasks or problem solve on one's own), working memory (hold information in mind to complete a task), plan and organize (plan and manage current and future task demands), organization of materials (ability to organize work, play space, etc.), and monitoring (ability to monitor own work or behavior). Clustered behavior domain scores provide indexes of Behavioral Regulation, including scores from inhibition, shifting, and emotional control subscales, and Metacognition, including scores from initiate, working memory, plan an organize, organization of materials, and monitor subscales. The Global Executive Composite provides a composite measure of all subscales. In addition, the BRIEF has two validity scales, one for consistency and one for negativity (Gioia, Isquith, Guy, & Kenworthy, 2000).

The BRIEF was standardized using children from rural, suburban, and inner-city areas throughout the state of Maryland. The number of students from underrepresented groups represented U.S. Censure figures at the time of the instrument's development, thereby ensuring the cross-cultural validity of the instrument. Parent-report forms were completed for 1,419 children ages 5 through 18. The internal consistency for the parent-report version was high, with a Cronbach alpha ranging from .80 to .98; test—retest reliability over a 2-week period was .81 (Gioia et al., 2000). The content validity of the BRIEF was supported by high interrater agreement. Convergent and divergent validity was also established comparing scores on the BRIEF with conceptually matched subscales from other behavior ratings scales, including the ADHD-Rating Scale—IV, Child Behavior Checklist (CBCL), and Conners' Rating Scale. Correlations of BRIEF scores with the ADHD-Rating Scale—IV ranged from .42 to .73, .44 to .72 with the CBCL, and .52 to .77 with the Conners' Rating Scale. Correlations between BRIEF scores and measures of emotional functioning were not correlated. (Gioia et al., 2000).

Parents answer each item using a 3-point Likert-type scale with 1 = *never*, 2= *sometimes*, and 3 = *often*. Raw scores are tallied for each of the eight subscales and the three composite scales. Based on the child's chronological age, raw scores are then computed into T scores (M = 50,

SD = 10). *T* scores ≥ 65 indicate clinically significant impairment (Gioia et al., 2000); *T* scores between 60 and 64 indicate borderline clinically significant scores.

RESULTS

Statistical analyses were conducted with StatView 5.0.1 (Standard Version). All summary and subscale scores from the SSRS and BRIEF were tested using the Kolmogorov-Smirnov Test (Daniel, 1991) to examine score distributions. No significant deviations from normality were found. An equality of variances *F* test revealed no significant differences in the variances of TSSS between the 22q11DS and control groups, $F(22, 39) = .56$, $p = .13$. Gender comparisons were run for all variables; no significant gender differences were found and genders were combined.

In this sample, we have observed marked developmental delays among affected preschool children (ages 3–5) characterized by emotional immaturity and a lack of age-appropriate social independence. For this reason, we confined our primary analyses to children ages 5.5 to 13, who were in Kindergarten through sixth grade (older children included had been held back in school). For comparison, secondary descriptive analyses were conducted with both the preschool and secondary school samples.

Social Skills Rating Scale

It was hypothesized that children with 22q11DS would score lower on ratings of social skills. Table 1 shows the mean total SSRS scaled scores of children with 22q11DS and sibling controls. The mean TSSS for children without the syndrome was within 1 standard deviation above the mean whereas the mean score of the affected children was greater than 1 standard deviation below the mean. The standard deviations of the two samples were close, although the affected children's range of scores was 17 points broader than those of unaffected children.

Descriptive statistics for the categorical behavior ratings associated with each SSRS subscale are shown in Table 2. Chi-square analyses suggested significant group differences with regard to the number of children with "fewer" skills in the areas of cooperation ($df = 2$, $N = 63$, $\chi^2 = 8.27$, $p = .02$), assertion ($df = 2$, $N = 63$, $\chi^2 = 16.86$, $p < .01$), and responsibility ($df = 2$, $N = 63$, $\chi^2 = .19.68$, $p < .01$). Groups did not differ with regard to the frequency of children with reduced skills in self-control ($df = 2$, $N = 63$, $\chi^2 = 5.38$, $p = .07$).

It was hypothesized that children with 22q11DS have lower mean TSSS as compared with control siblings. An unpaired *t* test suggested marked mean score differences between groups, *M diff* = 19.94, $df = 61$, $t = 6.32$, $p < .01$.

Behavior Rating Inventory of Executive Function

It was also hypothesized that children with 22q11DS as compared with control siblings have higher (worse) scores on a measure of daily behaviors associated with executive dysfunction. As Table 3 shows for four of the eight BRIEF subscales, mean scores of children with 22q11DS were more than 1 standard deviation above the mean (approaching "clinical significance," discussed earlier), with one of these (Working Memory) in the clinically significant range. An unpaired *t* test suggested group differences in the BRIEF Global Composite scale, *M diff* = 15.43, $df = 60$, $t = -5.80$, $p < .01$. Control participants' mean score was 1 standard deviation below the mean score of the affected children.

All of the BRIEF subscale scores were significantly correlated for children with 22q11DS ($p \leq .01$), indicating the use of a multivariate analysis of variance for analyses of group differences on individual BRIEF subscale scores (Tabachnick & Fidell, 2001). A Bonferroni correction was used to control Type I error rates ($p < .005$). As seen in Table 4, statistically significant

group differences were found on the four clinically significant subscales, including initiation, planning, working memory, and monitoring.

Associations Between Social Skills and Daily Behaviors Associated With Executive Dysfunction

It was hypothesized that TSSS would be associated with the BRIEF Global Composite Score (GCT), as well as clinically significant subscales. A simple regression found that the BRIEF GCT score accounted for 71.2% of the total variance in this sample of affected children's TSSS, $R = .71$, $F = 61.59$, $p < 01$.

Secondary analyses were used to examine which areas of executive dysfunction most strongly predicted TSSS. Among children with 22q11DS, Initiation ($r = -.59$, $p < .01$), Working Memory ($r = -.54$, $p < .01$), Planning ($r = -.46$, $p < .01$), and Monitoring ($r = -.61$, $p < .01$), were all negatively associated with TSSS. In contrast, TSSS was not associated with Initiation ($r = .07$, $p = .75$), Working Memory ($r = -.06$, $p = .80$), Planning ($r = -.19$, $p = .41$), or Monitoring ($r = -.06$, $p = .80$) among sibling controls.

A standard multiple regression analysis was then performed to predict affected children's TSSS from the subscales Initiation, Working Memory, Planning, and Monitoring. In this analysis, two predictors were identified: Monitoring ($t = -2.47$, $p < .05$) and Initiation ($t = -2.04$, $p < .05$). The four subscale variables together accounted for 69.3% of the total variance in affected children's TSSS, $R = .69$, $F = 8.10$, $p < .01$ (see Table 5).

Secondary Descriptive Analysis of Preschool and Secondary School-Age Children

Given the very small samples, only descriptive statistics were completed on the preschool and secondary school samples. In affected preschool children ($n = 6$), the mean TSSS score was in the impaired range ($M = 78.67$, $SD = 8.87$, range = 61–86). The only preschool-level control participant scored greater than 1 standard deviation above the mean.

Secondary school children (ages 13.5–16.2) with 22q11DS ($n = 6$) also were rated as having impaired social abilities ($M = 82.67$, $SD = 10.71$, range = 67–91); control participants ($n = 2$) were rated as having above average social skills ($M = 122.5$, $SD = 12.02$, range = 114–131).

Descriptive analyses were examined for BRIEF scores of children ages 13 and above (preschool children are too young for the BRIEF). The GCT for affected secondary school children was in the clinically significant range ($M = 66.50$, $SD = 12.60$). Secondary school children with the deletion also scored in the clinically significant range on Shifting ($M = 71.33$, $SD = 5.68$), Emotional Control ($M = 64.00$, $SD = 11.00$), Initiation ($M = 66.17$, $SD = 8.50$), Working Memory ($M = 62.17$, $SD = 13.38$), Planning ($M = 67.00$, $SD = 13.94$), Organization ($M = 60.33$, $SD = 8.09$), and Monitoring subscales ($M = 63.83$, $SD = 14.78$).

DISCUSSION

Previously, children with 22q11DS were described as having poor social interactions (Thomas & Graham, 1997), being socially immature (Shprintzen, 2000), and unable to control their behaviors and emotions in social settings (Golding-Kushner et al., 1985), with one study (Woodin et al., 2001) reporting a notably elevated Social Problems subscale mean score on the Achenbach CBCL. The findings reported here add to this literature by empirically quantifying specific types of social skill deficits in children with 22q11DS.

When compared with sibling control participants, children with 22q11DS had significantly lower global Social Skills that were approximately 1 standard deviation below the mean of control siblings. With regard to individual functioning domains, affected children were found

to have specific deficits in the areas of Cooperation, Assertion, and Responsibility. None of the affected children were rated as having "more" of these subskills, suggesting that deficits in these areas are experienced by most children with the deletion. Self-control skills of children with 22q11DS, however, did not differ from those of sibling controls. These findings begin to substantiate previous clinical impressions that children with the deletion have marked social difficulties, and help to specify the areas of greatest difficulty. Thus, because of the impact that social skills can have on academic achievement (discussed earlier), evaluating and addressing social difficulties must be included in any assessment or remediation program for children with 22q11DS. Intervention efforts should specifically help affected children improve their abilities to cooperate with others, initiate or assert social interactions, and be responsible in social and interpersonal situations.

Although several studies have shown marked deficits on neurocognitive measures of executive function and attention in children with 22q11DS, whether these deficits are apparent in the everyday behaviors of affected children was not previously considered. When ratings of everyday behaviors associated with executive function were examined (BRIEF), elementary-school-age children with 22q11DS differed significantly from control siblings. Group differences were found on the GCT, as well as for four of eight subscales on which affected children's scores approached the clinically significant range. These included Initiation, Working Memory, Planning, and Monitoring. The findings suggest that children with 22q11DS exhibit specific and observable behavioral difficulties associated with previously reported deficits in neurocognitively assessed attention and executive function, and these markedly impact their daily functioning. Although out of the scope of this report, in future studies it will be important to examine associations between specific neurocognitive measures and these behavioral domains.

Determining possible associations between social competence and behaviors associated with executive dysfunction may be critical for understanding how to intervene. When the BRIEF GCT was used to predict Total Social Skills ability in affected children, a significant association was found, with BRIEF scores accounting for approximately 71% of the variance of social skills ratings in this sample. The correlation between these measures may suggest that daily behaviors associated with executive dysfunction and social skills deficits stem from a common disruptive source characteristic of the 22q11DS. This suggestion is perhaps further strengthened by the lack of association between social skills and behaviors associated with executive function in control siblings. In fact, it has been proposed that social and academic deficits might result from common neurocognitive deficits with a brain-based origin (Kavale & Forness, 1996; Steinberg & Avenovoli, 2000; Waterhouse, 2002). This is an important area for further investigation.

In secondary analyses, specific correlations were found between Total Social Skills ability and domains from the Metacognition Index, namely the following: Initiation, Planning, Working Memory, and Monitoring. When considering only those subscale scores that were 1 standard deviation or more above the standardization sample mean, Initiation and Monitoring were the most significant predictors of overall social skills capability. Initiation is the ability to instigate activities, social interactions, homework, or daily responsibilities, whereas monitoring is the ability to observe, check, evaluate, and change as needed one's behavior. It is important to note that not all children with the deletion scored in a clinically significant range on these measures. Thus, identifying which children are at greatest risk for social problems may be importantly linked to their individual ability to initiate and monitor activities. These might also be key areas to target in programs of remediation. Interestingly, Working Memory did not emerge as a significant predictor of general social skill ability. Because this was the highest of the mean BRIEF scores among affected children, it is possible that the lack of correlation between BRIEF

Working Memory subscale and global social competence is attributable to a relatively small score range among affected children. This finding requires further exploration.

Limitations

Although the SSRS was one of the best social skills rating scales available, it measures social competence as opposed to social impairment. Although this ensures a range of scores among control participants, its capacity to adequately scale deficits is limited. Indeed, several parents commented that the SSRS failed to fully capture the variety and depth of their children's social difficulties. Future studies might include two scales, one that rates deficits and one that rates competence. The findings here reported were based exclusively on parent report of behavior. Although this can be a rich source of information regarding a child's behavior in the home environment, it is also subject to a variety of potential biases. An examination of the BRIEF negativity scale found that none of the parent reports suggested a negative bias toward the affected child. However, in the future, clinician-administered scales accompanied by a full assessment (including observations) of behavioral functioning would likely provide important additional data. This sample was predominantly White and the generalizability of these results to 22q11DS children from other racial and ethnic groups is unknown. The participating families were self-selected for participation which can result in a sample biased toward either more or less severely affected children. The applicability of these findings to the general population of children with 22q11DS remains speculative.

Summary

These data replicate previous research showing associations between social ability and behaviors associated with executive dysfunction (Gilotty et al., 2002). The replication may be especially informative because the previous research examined a very different clinical sample and used a different social skills outcome measure (both studies used the BRIEF). Importantly, however, the specific executive function deficits found to be associated with social skills differed between the two populations. For children with 22q11DS, initiation and monitoring were important predictors of social abilities; whereas for children with autism, Gilotty et al. (2002) found that initiation and working memory predicted VABS socialization scores (discussed earlier).

Previous studies have suggested that disturbances in fundamental cognitive processes (such as executive function) may be at the root of social disturbances, and, as such, should be the target of remediation services (Hughes, Dunn, & White, 1998; Kavale & Forness, 1996; Nigg, Quamma, Greenberg, & Kusche, 1999). More than a few researchers have found social skills training programs to be largely ineffective (Kavale & Forness, 1996), or, at best, to yield mixed results (Coie & Krehbiel, 1984). Named among the factors that may influence program efficacy is the perceived origin of social skill deficits (Kavale & Forness, 1996). Perhaps the failure of programs to effectively remediate social skills is a lack of attention to the cognitive factors, and in particular executive deficits, that perhaps underlie social deficits. If so, associations between social skill and daily behavior associated with executive function may have specific implications for social skill intervention. When a child has notable social skill deficits, screening for executive dysfunction may be warranted. Conversely, when executive dysfunction is identified, social skills must also be closely evaluated. Perhaps most importantly, these findings suggest that for children with a combination of social and executive deficits, intervention programs must target behaviors associated with executive functions, and in particular initiating and monitoring, to successfully remediate social competence.

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Table 1

Total Social Skills Scores Descriptive Statistics for 22q11DS and Control Participants

Age Level and Diagnostic Group	M	SD	Range
Preschool			
22q11DS[a]	78.67	8.87	61.0–86.0
Control participant[b]	122.00	n/a	n/a
Elementary school age (K—6)			
22q11DS[c]	84.98	13.15	62.0–114.0
Control participants[d]	104.91	9.83	88.0–123.00
Secondary school age			
22q11DS[a]	82.67	10.71	67.0 – 91.0
Control participants[e]	122.5	12.02	114.0

Note. $N = 78$. K—6 = Kindergarten through sixth grade.

[a] $n = 6$.

[b] $n = 1$.

[c] $n = 40$.

[d] $n = 23$.

[e] $n = 2$.

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Table 2

Elementary School-Age Frequency Percentages for SSRS Subscales: Category Scores by Participant Group

	22q11DS[a]			Controls[b]		
SSRS Subscale	**Fewer**	**Average**	**More**	**Fewer**	**Average**	**More**
Cooperation (%)	40	60	0	8.7	87.0	4.3*
Assertion (%)	50.0	50.0	0	4.3	82.6	13.0*
Responsibility (%)	60.0	40.0	0	4.3	91.3	4.3*
Self-Control (%)	30.0	70.0	0	13.0	78.3	8.7

Note. $N = 63$. SSRS = Social Skills Rating System; DS = Deletion Syndrome.

[a] $n = 40$.

[b] $n = 23$.

* Chi Square $p < .02$

Table 3

Elementary School-Age Mean BRIEF Subscale Scores by Participant Group

Subscale	M	SD	Range
Inhibition			
Control	48.32	7.63	38–69
22q11DS	58.50	9.76	38–82
Shift			
Control	44.36	6.57	37–60
22q11DS	58.23	11.28	41–83
Emotional Control			
Control	48.32	11.84	35–73
22q11DS	56.98	10.72	38–80
Initiate			
Control	44.68	6.53	35–63
22q11DS	60.13[c]	11.29	42–84
Working Memory			
Control	46.41	8.41	36–66
22q11DS	65.01[d]	10.41	36–84
Planning			
Control	47.36	9.91	37–69
22q11DS	61.03[c]	13.21	38–98
Organization			
Control	49.46	10.74	34–69
22q11DS	54.55	8.21	35–73
Monitoring			
Control	45.91	9.47	33–69
22q11DS	60.65[c]	9.59	36–80
Global Composite			
Control	46.32	9.07	36–67
22q11DS	61.75[c]	10.30	36–86

Note. $N = 62$; one child missing due to incomplete scoring by parent.

[a] $n = 22$.

[b] $n = 40$.

[c] Score approaching clinical significance.

[d] Clinically-significant score.

Table 4

Elementary School-Age MANOVA Results for Clinically-Significant BRIEF Subscale Scores by Participant Group

Subscale	df	F	h	p
Initiate	1	34.62	.59	<.01
Working Memory	1	51.68[a]	.66	<.01
Planning	1	17.92[a]	.46	<.01
Monitoring	1	33.85[a]	.59	<.01

Note. *N* = 62. MANOVA = multivariate analysis of variance; BRIEF = Behavior Inventory of Executive Function.

[a]Significant Bonferroni p < .005.

Table 5

Multiple Regression Analysis Predicting K—6, Affected Children's TSSS Scores from the Four Clinically-Significant BRIEF Subscale Scores

BRIEF Subscales	B	SE B	b	t
Monitoring	−.55	.22	−.40	−2.47*
Initiate	−.50	.25	−.43	−2.04*
Planning	−.30	.26	−.24	−1.17
Working Memory	.31	.23	.31	1.35

Note. $N = 40$. K—6 = Kindergarten through sixth grade; MANOVA = multivariate analysis of variance; BRIEF = Behavior Inventory of Executive Function; $R = .69$, $R^2 = .48$

*$p < .05$.