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Association of the family environment with behavioural and cognitive outcomes in children with chromosome 22q11.2 deletion syndrome

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Abstract

Background—Children with 22q11.2 deletion syndrome (22q11DS) are at risk for social-behavioural and neurocognitive sequelae throughout development. The current study examined the impact of family environmental characteristics on social-behavioural and cognitive outcomes in this pediatric population.

Method—Guardians of children with 22q11DS were recruited through two medical genetics clinics. Con senting guardians were asked to complete several questionnaires regarding their child's social, emotional and behavioural functioning, as well as family social environment and parenting styles. Children with 22q11DS were asked to undergo a cognitive assessment, including IQ and achievement testing, and measures of attention, executive function and memory.

Results—Modest associations were found between aspects of the family social environment and parenting styles with social-behavioural and cognitive/academic outcomes. Regression models indicated that physical punishment, socioeconomic status, parental control and family organisation significantly predicted social-behavioural and cognitive outcomes in children with 22q11DS.

Conclusion—Characteristics of the family social environment and parenting approaches appear to be associated with functional outcomes of children with 22q11DS. Understanding the impact of environmental variables on developmental outcomes can be useful in determining more effective targets for intervention. This will be important in order to improve the quality of life of individuals affected by 22q11DS.

Keywords

chromosome 22q11.2 deletion syndrome; DiGeorge syndrome; family environment; parenting; social-behavioural functioning; velocardiofacial syndrome

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Introduction

Deletion of chromosome 22 at band q11.2 (22q11DS) is a common genetic condition – also known as DiGeorge syndrome or velocardiofacial syndrome – with an incidence of 1/2000 to 1/1600 (Shprintzen 2008). In addition to a range of medical complications, it is well documented that children with 22q11DS have associated cognitive impairment and academic learning problems.

Research has indicated that 80–100% of these children exhibit cognitive deficits, with an average IQ score of 75 (Swillen *et al*. 1997; Gerdes *et al*. 1999; De Smedt *et al*. 2007; Niklasson & Gillberg 2010). A closer examination of specific cognitive abilities reveals deficits in a number of domains, many of which are independent of IQ, including visualspatial processing, executive functioning, attention, verbal learning and working memory (Sobin *et al*. 2005; Lewandowski *et al*. 2007a). Decrements in cognitive ability can lead to poor academic performance in these children, and indeed, research has found notable deficits in arithmetic among children with 22q11DS, whereas reading and spelling skills are relatively stronger (Moss *et al*. 1999; Simon 2008; De Smedt *et al*. 2009).

In addition to significant neurocognitive and academic impairment, children with 22q11DS are at risk for emotional and behavioural problems. Research has indicated that upwards of 66% of youth with 22q11DS meet criteria for at least one psychiatric diagnosis (Young *et al*. 2011), which is significantly greater than the diagnostic rate observed in the general paediatric population [approximately 11% of children ages 8–15 (Merikangas *et al*. 2010)]. More specifically, these children exhibit a wide range of psychopathology including anxiety disorders, depression, attentiondeficit/hyperactivity disorder (ADHD) and oppositional defiant disorder (Swillen *et al*. 1999; Baker & Skuse 2005; Kiley-Brabeck & Sobin 2006a; Jolin *et al*. 2009; Shashi *et al*. 2012). Consistent with high rates of diagnosed psychiatric disorders, parents of children with 22q11DS also report elevated rates of internalising and externalising symptoms in their children (Young *et al*. 2011). Psychiatric and behavioural problems early in development are thought to represent a major risk factor for more severe psychopathology later in life (Gothelf *et al*. 2007; Lewandowski *et al*. 2007b). This is a very relevant consideration for children with 22q11DS as it has been shown that these children are 25 times more likely to develop a serious mental illness later in adolescence through early adulthood, and approximately one-third of this population will develop schizophrenia spectrum disorders (Shprintzen *et al*. 1992; Murphy *et al*. 1999; Gothelf *et al*. 2007).

Given the rates of cognitive and psychiatric disability in children with 22q11DS, it is not surprising that there is also a high risk for poor social functioning in this population. A recent study by Shashi and colleagues (2012) compared social functioning in children with 22q11DS with typically developing peers, and reported remarkable deficits in the 22q11DS group. Specifically, deficits emerged on parent-reported questionnaires and also on more objective measures of social skills, such as facial emotion recognition tasks. These findings are consistent with previous research, which has similarly reported social skill deficits among children with 22q11DS (Kiley-Brabeck & Sobin 2006b). Importantly, these behavioural problems have not been shown to be associated with intellectual impairment in

this population, which suggests that they may be specifically related to a behavioural phenotype of 22q11DS rather than a general cognitive disability (Jansen *et al*. 2007; Shashi *et al*. 2012).

While significant research has documented the presence of cognitive, psychological and social impairments in children with 22q11DS, less work has examined the contextual risk factors that may be contributing to this neurobehavioural phenotype. Although it is reasonable to expect that the microdeletion of multiple genes would confer cognitive and/or social-psychological disability upon these children, it is not an all-encompassing explanation of the processes at hand. Rather, it is likely that there are complex interactions between genetic and environmental factors that influence neurocognitive and social-psychological outcomes. In particular, it has been widely established in the general population that the home environment provides a direct context for the child's development, and thus has an important impact on cognitive and psychosocial outcomes in children (Farah *et al*. 2006; Farah 2010). However, very limited research has specifically examined if, and how, components of the family environment relate to functional outcomes in children with 22q11DS.

To date, only two groups have examined the relationship between child outcomes (i.e. cognitive, psychiatric and/or behavioural functioning) and aspects of the family environment. Prinzie *et al*. (2004) assessed the relationship between parenting styles and personality characteristics of children with 22q11DS. They reported that parental warmth was associated with conscientiousness and emotional stability in children, while parental control was positively associated with degree of child dependence and negatively correlated with conscientiousness. Differences in parental and family context were not related to the child's IQ, gender or cardiac defects. Recently, Shashi *et al*. (2010b, 2012) examined the relationship between parental socioeconomic status (SES) and neuropsychological and behavioural functioning in children with 22q11DS compared with typically developing peers. In general, their results indicated significantly worse functioning across cognitive, academic and behavioural domains in children with 22q11DS compared with the agematched control group. Looking specifically at the family environment, among children with 22q11DS, lower SES was correlated with poorer social skills and greater levels of maladaptive behaviour, and a significant positive relationship between SES and IQ/achievement variables was found. Taken together, these studies suggest that family environmental factors, such as SES, have an important statistical and clinically meaningful relationship with behavioural outcomes among children with 22q11DS.

Collectively, the research to date affords some insight into the lives of families affected by 22q11DS, in that higher SES and more positive parenting responses are associated with better emotional functioning. Better understanding of additional contextual variables of the family environment, such as social climate, may help clinicians identify useful targets for intervention.

To further examine potential environmental factors, the present study examined family social climate variables, including cohesion, expressiveness, values, organisation, and level

of structure and control in addition to dimensions of parenting in relation to neurobehavioural outcomes of children with 22q11DS.

In typically developing children, it has been shown that the family social and parenting environment influences both cognitive and behavioural functioning (Baumrind 1966, 1967; Jonson-Reid *et al*. 2010; Hewage *et al*. 2011; Whitley *et al*. 2011). There are currently no published data that have examined the family social and parenting environment and its relationship to cognitive, academic and behavioural outcomes in children with 22q11DS, however. Based on the empirical data in typically developing children, and previous data showing the effects of other family environment variables (e.g. SES and parenting style) on children with 22q11DS, the following hypotheses were made in the context of the measured variables in this study: (1) more adaptive social-behavioural functioning in children would be associated with family social environments high in expressiveness and active-recreational orientation, and low in child independence and conflict; (2) adaptive social-behavioural functioning would be associated with specific parenting traits including a high degree of nurturance and control. Parental discipline styles, including inconsistent punishment and physical punishment were each hypothesised to have a negative relationship with adaptive social-behavioural functioning, whereas following through on discipline was predicted to have a positive relationship; and (3) familial achievement orientation and control, in addition to parental organisation, would have a positive association with overall functioning and cognitive and academic outcomes, whereas passive punishment strategies would have a negative relationship with these outcomes. A final goal of the present study was to examine the family social environment and parenting style variables as potential predictors of social-behavioural and cognitive outcomes in children with 22q11DS.

Methods

Participants

Participants included 48 children and adolescents diagnosed with 22q11DS, confirmed by fluorescence *in situ* hybridisation (Vysis, Gaithersburg, MD, USA) or microarray analyses (http://www.affymetrix.com). Participants were recruited from genetics clinics at two medical centres located in the south-eastern USA. Both sites were used for assessments and the institutional review boards of both medical centres approved the study. All caregivers completed written informed consent, and children provided assent when applicable, in accordance with procedures of the Institutional Review Board. The study was offered to 63 eligible families, 48 of whom participated.

The child and adolescent participants ranged in age from 9 to 18 years, with an average age at study enrolment of 12.46 years (SD = 2.07). The sample was 54.2% male and largely white (83.3%), with African-Americans (4.2%), Hispanics (6.3%) and multiracial (6.3%) racial groups also being represented in the sample. The majority of adults completing the parent portion of the evaluation were mothers (39; 81.3%), in addition to eight fathers (16.7%) and one grandfather (2.1%). The Hollingshead Two-Factor Index of Social Position (Hollingshead 1957; Cirino *et al*. 2002) placed the sample within the middle socioeconomic stratum (M = 29.3, SD = 13.45).

Neurocognitive and behavioural measures

The participants underwent neurocognitive and achievement testing over one to two sessions to ascertain academic achievement, and *a priori* determined aspects of neurocognition. Parental interviews and questionnaires were also administered to ascertain demographic information, and behavioural/social, and emotional functioning. Individual outcome variables will be described in detail below.

Overall social and psychological functioning of child participants was measured using the Children's Global Assessment Scale [CGAS (Shaffer *et al*. 1983)], a clinician-rated subjective index of global functioning of the child based on levels of impairment in the home, at school and with peers. Potential ratings span between 1 and 100, with higher ratings indicating better overall functioning. Overall SES was measured for each family using the Hollingshead Two-Factor Index of Social Position (Hollingshead 1957), with lower numbers indicating higher SES. SES was calculated for each parent, and the lower of the two numbers (e.g. the highest SES level) was used as a measure of family SES.

Psycho-educational estimates consisted of a full scale IQ using the Wechsler Intelligence Scale for Children – IV (Wechsler 2003) and broad reading ability using the Wechsler Individual Achievement Test – III [WIAT-III (Wechsler 2009)]. The IQ and achievement results are part of prior reports from our group, with highly significant differences found between children with 22q11DS and age-matched control subjects on all the measures (Shashi *et al*. 2010a,b). Executive functioning was measured by the computerised Wisconsin Card Sorting Test Version 4 – Research Edition (Chelune & Baer 1986; Chase-Carmichael *et al*. 1999), assessing the executive functions of set-shifting, planning and problem-solving, organisation and impulsivity. The per cent of perseverative errors on this task was used as an index of inhibitory control.

Behavioural and psychological functioning was assessed using the Child Behaviour Checklist [CBCL (Achenbach 1991)], the Diagnostic Interview Schedule for Children [DISC-IV (Shaffer *et al*. 2000)] and Social Skills Rating System [SSRS (Gresham & Elliott 1990)]. All three of these measures were completed by parents. The CBCL is a widely used measure of emotional and behavioural functioning of children ages 6–18. Parents respond to questions regarding their child's social, academic, behavioural and emotional functioning across home and school domains. Total Competency scales are calculated based on children's performance and involvement in school, social and academic domains, and clinical scales are computed for several areas of problem functioning. In the present study, scales for Total Problem Behaviours and Total Competence were used as outcome variables. The DISC-IV is a computerised, fully structured diagnostic instrument that assesses 34 common Diagnostic and Statistical Manual – IV (DSM-IV) psychiatric diagnoses of children and adolescents, by ascertaining the presence or absence of symptoms. In the present study, DSM-IV axis I disorders were evaluated. Social functioning was assessed using the SSRS, a 30-item questionnaire that assesses social competence, prosocial skills and problem behaviours in children grades K-12. The SSRS Total Social Skills and Problem Behaviour composite scores served as outcomes in the present study.

Environmental characteristics

Information pertaining to the family social environment and parenting style was measured through parent-completed questionnaires. Specifically, the Family Environment Scale – Real Form [FES-R (Moos & Moos 1986)] and Parenting Dimensions Inventory Short Form [PDI-S (Power 2002)] were used to examine general aspects of the family social environment and specific parenting approaches respectively.

The FES-R is a 90-item, true-false measure that assesses people's perceptions of their current family social environment. The FES-R is composed of 10 sub-scales: Cohesion (alpha coefficient $\alpha = 0.78$), Expressiveness ($\alpha = 0.69$), Conflict ($\alpha = 0.75$), Independence ($\alpha = 0.61$), Achievement orientation ($\alpha = 0.64$), Intellectual/Cultural orientation ($\alpha = 0.78$), Active-Recreational orientation ($\alpha = 0.67$), Moral-Religious emphasis ($\alpha = 0.78$), Organisation ($\alpha = 0.76$) and Control ($\alpha = 0.67$). This scale has been broadly used in clinical and research settings as an evaluative measure of a family's dynamic social environment, including research on congenital abnormalities such as cystic fibrosis (Breslau 1983), intellectual disabilities (Blacher *et al*. 1987; Howell *et al*. 2007), and other disorders such as Down's syndrome (Pueschelo & Myers 1994) and fragile-X syndrome (Kuo *et al*. 2002).

The PDI-S is a 27-item measure of parenting style that assesses six dimensions of parenting: Nurturance (alpha coefficien $\alpha = 0.72$), Following Through on Discipline ($\alpha = 0.66$), Type of Control (alpha not reported), Amount of Control ($\alpha = 0.71$), Inconsistency ($\alpha = 0.82$) and Organisation ($\alpha = 0.80$). The items for Nurturance, Inconsistency, Following Through on Discipline and Family Organisation consist of a series of descriptive elements on six-point scales. The items assessing Amount of Control consist of a series of opposing statements for which parents must choose the statement that they agree with most. Finally, five disciplinary situations are presented where parents indicate on Likert scales how likely it is that they would use different types of discipline. For the current study, two particular discipline strategies in the Type of Control dimension were examined: Letting the Situation Go (a = 0.83) and Physical Punishment (a = 0.85). These particular strategies were examined because they can be conceptualised as proxies for polar parenting approaches – permissive (e.g. poorly controlled and lax discipline) and authoritarian styles (e.g. tightly controlled and rigid) respectively – as identified in Baumrind's (1966 in Baumrind's (1967) seminal parenting literature. The literature has shown that permissive styles are linked to maladaptive behaviours (e.g. substance use, school misconduct) in adolescence, while authoritarian strategies can negatively impact self-esteem and self-concept, and in turn social-emotional functioning (Lamborn *et al*. 1991). Given the impact that these parenting styles might have on latter functioning in children, these particular variables from the PDI-S seemed to warrant consideration here. The PDI-S has been widely used in a variety of research settings as an evaluative and predictive tool for populations including children with cerebral palsy (Cunningham *et al*. 2009), and those in special education (Redden *et al*. 2003).

Statistical methods

Statistical tests were performed using the SPSS statistical software version 19.0. Data were initially examined for normality and, as some variables were not normally distributed, a

conservative approach utilising non-parametric tests was taken. Pearson correlations were computed to determine associations among neurocognitive, achievement, social/ behavioural, family social environment and parenting variables. Wherever indicated, Bonferroni correction for multiple comparisons was applied, to account for the number of correlations that were examined. In order to examine whether family social environment/ parenting dimensions predicted neurocognitive and/or social/behavioural characteristics of children with 22q11DS, a series of linear regression analyses were conducted. Variables from the FES-R and PDI-S that were either significantly correlated with the cognitive, academic and social/ behavioural outcomes, or theoretically relevant, were used in the regression analyses. Means, standard deviations and correlations among the outcome variables and the family and parenting variables that were significantly associated with outcomes are shown in Tables 1 and 2.

Results

Descriptives for the FES-R/PDI-S

Given that the focus of this study was to determine the familial correlates of social-behavioural, cognitive and academic functioning in these children, only brief descriptions will be given here regarding group outcomes on the FES-R and PDI-S. On the FES-R, families in our study had scores high on Cohesion, Expressiveness, Intellectual orientation, Recreational orientation, Moral-Religious orientation, Organisation and Control and lower in Conflict, when compared with norms (see Table 1). They were lower than typical families on Achievement Orientation and Independence. On the PDI-S, parents of children with 22q11DS had scores high on Nurturance, Organisation, Following Through on Discipline and Control. They were low on Physical Punishment and Inconsistency.

> Hypothesis 1. Relationship between social-behavioural functioning and family social environment (FES-R).

In order to examine this relationship, correlational analyses between the CBCL (Problems Score and Competency), SSRS (Social skills and Problem Behaviours) and the 10 FES-R sub-scales were conducted. Contrary to our expectations, no significant relationships were found between children's social-behavioural functioning and these FES-R variables. See Table 2 for specific correlation values.

> Hypothesis 2. Relationship between social-behavioural functioning and parenting style (PDI-S)

A second set of correlational analyses for social-behavioural functioning included the parenting traits measured by the PDI-S. The use of physical punishment was negatively associated with CBCL Total Competency ($r = -0.42$; $P < 0.01$;Table 3) and SSRS Total Social Skills ($r = -0.38$, $P < 0.05$), and positively associated with CBCL Problem Behaviours ($r = 0.31$; $P < 0.05$; Table 3). Thus greater use of physical punishment was associated with lower levels of social and academic competence in our sample of children, and higher levels of problem behaviours. There were no other significant associations.

Hypothesis 3. Relationship between child neurocognitive and overall functioning outcomes and family social environment characteristics, parenting traits and disciplinary strategies

To test this hypothesis, correlational analyses were performed between the 10 FES-R variables, the six PDI-S variables, overall functioning (CGAS), executive functions and academic reading outcomes. As expected, there was a significant association between children's overall functioning and the family's SES ($r = -0.34$; $P < 0.05$). There were also significant associations between overall functioning and the FES-R Achievement orientation ($r = 0.38$; $P < 0.05$; Table 2), and the PDI-S Organisation ($r = 0.35$; $P < 0.05$; Table 3) variables. Thus, higher overall functioning was associated with more affluent SES (lower SES score), greater family emphasis on achievement and greater parental organisation. SES, in turn, was significantly associated with the FES-R Intellectual-Cultural orientation ($r = -0.36$, $P < 0.05$) and the PDI-S Physical Punishment ($r = 0.32$, $P < 0.05$). More affluent SES (lower SES scores) was significantly associated with greater levels of family interest in political, intellectual and other cultural activities, and lower levels of physical punishment.

Regarding neurocognitive and achievement performance, children's executive functioning [Wisconsin Card Sorting Task (WCST) per cent perseverative errors] was positively associated with the PDI-S Organisation ($r = 0.37$, $P < 0.05$; Table 3). Academic achievement in reading was also significantly associated with PDI-S Organisation ($r = 0.38$, $P < 0.05$; Table 3), in addition to FES-R Control ($r = 0.40$, $P < 0.05$; Table 2). Lax disciplinary strategies (i.e. ‘Letting the Situation Go’ per the PDI-S) was negatively associated with achievement in reading in this population, as hypothesised ($r = -0.43$, $P < 0.01$; Table 3). Contrary to hypotheses, there was no association between familial achievement orientation and academic reading outcomes of children.

Because of the number of overall correlations, application of the Bonferroni correction for the total number of correlations would not allow for the retention of the significance of all our correlational results. However, it is to be noted that many of the correlations were of medium effect size (Tables 2 and 3) and thus it is likely that a larger sample size would enable results that would survive correction for multiple comparisons.

Family and parenting predictors of functional outcomes in children with 22q11DS

In order to examine possible family environment and parenting predictors of social-behavioural functioning in children with 22q11DS, four hierarchical regression analyses were conducted. Because children with 22q11DS tend to have high rates of diagnosis of anxiety disorders and ADHD, and are particularly prevalent diagnoses in our sample, we first evaluated the relation between Anxiety disorders and ADHD and social-behavioural functioning with *t*-tests examining differences in CBCL Competency and Total Problems and the SSRS social skills and problem behaviours in those with and without an Anxiety disorder and with and without ADHD. We found that children with an Anxiety disorder had significantly higher CBCL Total Problems t-score compared with those who did not ($t = 3.31$, $P < 0.01$), as well as lower social skills ($t = 6.06$, $P < 0.001$) and more problem behaviours ($t = 2.32$, $P < 0.05$) on the SSRS. Children with ADHD had lower CBCL Total Competency scores, compared with those without this diagnosis ($t = 2.05$, $P < 0.05$), but

were not significantly different on CBCL Total Problems or the SSRS variables. To account for these significant differences, it was determined to use the anxiety and ADHD diagnosis as a covariate in the relevant regression analysis.

For each regression model, demographic and psychiatric diagnoses (anxiety disorders or ADHD), were entered in the first step of each model as controls. In the second step, the FES-R and PDI-S variables were entered into the model to allow for a direct comparison of coefficients. If theoretically relevant, an interaction was added in the third step to examine the interactive effect beyond that of the individual variables. Each continuous variable was standardised and centred to reduce multicollinearity among the variables and allow for more meaningful interpretations of the regression coefficients. Results of the three hierarchical regressions that included significant FES-R or PDI-S variables are presented in Table 4.

CBCL Total Problem Behaviours

The hierarchical regression of CBCL Total Problem Behaviours was significant [$\beta = 0.47$, $F_{5,35} = 4.6$, $P < 0.01$]. The presence of an anxiety disorder, as previously discussed, was entered into the first step of the model, and was indeed a significant predictor [$\beta = 0.47$, $t(39) = 3.31$, $P < 0.01$]. FES-R Conflict, PDI-S Physical Punishment and Nurturance were included in the second block, and an interaction term between PDI-S Physical Punishment and FES-R Conflict was entered into the third step of this model. Physical Punishment was the only unique predictor of CBCL Total Problems in the second step [$\beta = 0.29$, $t(37)$, $P < 0.05$]. Although the final step of the model was significant, the interaction of physical punishment and conflict did not uniquely contribute to the prediction of CBCL Total Problems. The final model accounted for an unadjusted 35.2% of the variance in CBCL Total Problems.

CBCL Total Competency

The hierarchical regression of CBCL Total Competency was also significant [$F_{4,34} = 3.04$, $P < 0.05$). Having an ADHD diagnosis was entered in as a covariate in the first step as previously discussed. Results indicated that this significantly predicted lower total competency [$\beta = -0.32$, $t(37) = -2.05$, $P < 0.05$]. The PDI-S Physical Punishment and Organisation dimensions as well as their interaction were included in the model in the second and third steps respectively. Again, only Physical Punishment uniquely contributed to the prediction of CBCL Total Competency with higher scores on the physical punishment dimension predicting lower competency [$\beta = -0.36$, $t(35) = -2.49$, $P < 0.05$]. The final model accounted for an unadjusted 26.3% of the variance in CBCL Total Competency.

SSRS Total Social Skills

The hierarchical regression of parent-rated SSRS Total (Social) Skills was also significant [$F_{5,36} = 14.5$, $P < 0.001$). The presence of an anxiety disorder diagnosis and the child's gender were entered as covariates in the first and second steps respectively. Having an anxiety disorder [$\beta = -0.65$, $t(40) = -5.64$, $P < 0.001$] and being female [$\beta = -0.31$, $t(39) = -2.77$, $P < 0.01$] significantly predicted lower parent-rated total social skills. PDI-S Physical Punishment and FES-R Independence were included in the third block of this model, as well as their interaction as the fourth step. In this model, Physical Punishment scores uniquely

contributed to the prediction of SSRS Total Skills, such that greater endorsement of physical punishment by parents predicted lower competency [$\beta = -0.39$, $t(38) = -4.07$, $P < 0.001$]. The final model accounted for an unadjusted 66.8% of the variance in SSRS Total Skills.

SSRS Problem Behaviours

In the hierarchical regression of SSRS Problem Behaviours, having an anxiety disorder diagnosis was entered as a covariate in the first step. This significantly predicted more parent-rated behavioural problems in children with 22q11DS [$\beta = 0.35$, $t(39) = 2.34$, $P < 0.05$]. PDI-S Physical Punishment, FES-R Active-Recreational orientation and Religious/Moral were added to the model in the second step. None of the family and parenting variables significantly predicted problem behaviours and hence no interaction was included. The final model accounted for an unadjusted 28.7% of the variance in SSRS Problem Behaviours.

Neurocognitive and overall functioning

Contrary to hypotheses, regression models of cognitive (i.e. executive functioning, measured by the WCST perseverative errors), academic (i.e. Broad Reading from the WIAT) and global functioning (per the CGAS) outcomes were non-significant. Neither family social environment (i.e. intellectual-cultural orientations, degree of control and organisation of the home) nor parenting variables (i.e. parenting traits and parental disciplinary strategies) were significant predictors here.

Discussion

The current study examined the impact of family social environmental factors on the social-emotional, behavioural and cognitive functioning in children with 22q11DS. Results from the FES-R and PDI-S indicated that, in general, families of children with 22q11DS exhibited many positive qualities, such as high organisation, cohesion, expressiveness and low levels of conflict. They tended to have lower scores on academic achievement orientation and independence, perhaps a reflection of lower parental expectations because of the cognitive impairments experienced by children with the condition. Parents in our sample reported, in general, low rates of physical punishment, and high rates of nurturance, organisation and control.

The aim of the current study was to utilise the FES-R and PDI-S scales to examine whether elements of the family social environment (e.g. level of support, conflict, control and value orientations) and approaches to parenting (e.g. punishment style, approach to problem-solving) predict functional outcomes in children with 22q11DS. At present, there is very little empirical literature regarding the role of the family social environment on behavioural outcomes in this population. Only two prior studies have examined the family environment in general in the 22q11DS population; both finding significant relationships (Prinzie *et al*. 2004; Shashi *et al*. 2012). Overall, the findings in the present study suggest that the family social environment may indeed be associated with functional outcomes in children with 22q11DS.

The most striking result involved parental use of physical punishment, which was significantly associated with multiple behavioural outcomes. There was a relationship between the use of physical punishment and social (SSRS) and behavioural problems (CBCL) such that greater use of physical punishment was associated with lower levels of overall academic and social competency, and higher levels of behaviour problems. The potentially maladaptive effect of physical discipline was further supported in our regression models in which it uniquely predicted the presence of behavioural problems, low overall competency (across social, emotional and school domains) and poor social skills. These findings were significant despite the fact that the levels of use of physical punishment in our sample were relatively low compared with the general population.

There is much controversy in the field of child development regarding corporal punishment strategies in the home. While a thorough discussion of this is outside the scope of this paper, briefly, a meta-analysis of corporal punishment of children by their parents indicated that this disciplinary strategy results in undesired consequences, affecting social, behavioural and psychological health (Gershoff 2002). Physical punishment strategies reflect an authoritarian approach to discipline, which may be ineffective and perhaps contraindicated for children with special needs. Physical punishment, as opposed to authoritative approaches (e.g. having clearly defined consequences and discussions regarding behaviour), may prevent children with 22q11DS from understanding *why* certain behaviours are maladaptive. In addition, these youth are at risk for modelling aggressive or generally maladaptive behaviour consistent with physical discipline. Ultimately, using physical discipline may make it more difficult for children with 22q11DS to internalise social rules about appropriate and inappropriate behaviour. This may explain some of the behavioural and social impairments associated with physical punishment in our sample. Indeed, this theoretical process, by which functional outcomes are associated with punishment styles, has also been delineated for typically developing populations (Gershoff).

The present study also observed relationships between dimensions of the family environment, overall levels of child functioning and neurocognitive abilities. There was a significant association between family SES and children's global functioning, such that higher affluence (lower SES number) was correlated with a higher CGAS rating. This finding is consistent with previous research (including the cohort in this study;(Shashi *et al*. 2010b,2012), which indicated that SES was a significant predictor of social skills, and IQ, but not higher neurocognitive functioning or reading achievement in children with 22q11DS. It is possible that families with a higher SES have increased access to high-quality resources (e.g. educational opportunities for children, clinical care providers) that help promote healthy functioning in children, which is supported by the fact that in our study higher SES was also related to greater interest in political, intellectual and cultural activities. In addition, a higher SES may attenuate some of the stress in families (e.g. by affording families more resources), thereby enabling parents more time to shape and promote positive outcomes across functional domains such as school, and in social relationships. We also noted in our findings that SES was related to the use of physical punishment, which we previously discussed as having an impact on several social-behavioural outcomes.

Several relationships were also observed between child neurocognitive outcomes and parent organisation. Specifically, the results of this study indicated that the level of parent organisation on the PDI-S was positively related to children's overall functioning (per the CGAS as well as better executive functioning, and achievement in reading. FES-R Control was also associated with reading ability in that a greater number of family rules and procedures were related to higher reading achievement scores. Although these family-level characteristics were not significant predictors of functioning in our regression models, the observed relationships are nonetheless important to consider. The impact of family-level characteristics – in this instance, organisation and parental control – may be working to facilitate behavioural outcomes of children with 22q11DS through direct and indirect mechanisms. On a direct level, families high in organisation and parental control may be shaping adaptive behaviour in their children by providing them with concrete rules and structure. These rules can be internalised, and ultimately provide a rubric that can be applied and adapted by children in varying contexts. Thus, a child whose parents set consistent rules and expectations regarding school may be more likely to comply with homework, resulting in stronger potentially academic and cognitive skills. This same child may also exhibit more compliant behaviour in a school setting because he or she internalised rules about behaviour and respect for adults through the structure at home.

While this would likely be the case for any child, in the context of 22q11DS, children may require more externally imposed structure, support and control, given the risk for cognitive or social handicaps associated with the genetic deletion (Feinstein *et al*. 2002; Lewandowski *et al*. 2007a). The importance of organisation and control for these children is further supported by our finding that a more permissive disciplinary strategy (i.e. Letting the Situation Go) was negatively associated with reading ability. It is possible that insufficient structure or lack of consequences can impede these children from reaching their maximal potential, particularly with respect to early developmental functions. In this regard, the family environment may become a crucial player in the promotion of children's resilience.

Environmental influences can also impact children's behaviour through indirect means. In particular, having a child with chronic medical needs can create a level of stress that may deregulate the family environment. Indeed, research has suggested that families with a child with 22q11DS have particularly high levels of stress and marital discord in the home environment (Prinzie *et al*. 2004) and that poor psychosocial adjustment is exacerbated among homes with familial cases of 22q11DS (Gothelf *et al*. 2007). In light of this, it is possible that increased organisation and control can help compensate for some of the challenges that may otherwise be present among families with a child with 22q11DS. For instance, having an organised environment, in which parents maintain control, may mitigate the stress from anticipated or unanticipated medical, psychological or social needs of their child with 22q11DS. By reducing the level of stress in the home, children are provided with a more adaptive environment in which to develop. Indeed, previous research has shown that children who are resistant to ecological stressors have more adaptive behaviour and better overall functioning across multiple contexts (Luthar 1991; Holmes *et al*. 1999). This may be the case here, and could account for the adaptive role played by familial organisation and parental control.

The current study provides modest to moderate evidence for the association of family social environmental factors in the outcomes of children with 22q11DS. It was initially anticipated that family environment traits would be stronger, more robust predictors of outcomes in these children. However, results did not indicate an association between the majority of our cognitive outcomes with theoretically relevant environmental factors (e.g. intellectual orientation and achievement motivation of a family). Ultimately, this is likely due to the fact that cognitive deficits in children with 22q11DS are biologically mediated because of the deletion (Simon *et al*. 2005; DeBoer *et al*. 2007). In contrast, our data revealed more significant associations between family social factors and achievement outcomes, which are more susceptible to environmental influences.

There were several limitations to the current study. First, the current study did not employ a comparison group, which limits the interpretation of results. Notably, we were unable to make comparisons about differences in the family social environment between children with 22q11DS and typically developing peers or other comparison groups, although some of the observed relationships between the home environment and child outcomes are similar to typically developing populations.

The use of the Total Competency score on the CBCL may have led to findings that do not directly reflect the children's functioning, as some of the items may be considered to be parent-mediated, such as providing the child with opportunities to play sports and to socialise. With a cross-sectional study as ours, causal inferences are hard to make, as the direction of the correlations we found between the family's social environment and social-behavioural functioning could be in either direction. The results also did not retain significance after correction for multiple comparisons; however, the effect sizes for the significant correlations between the family social environment and the children's social-behavioural functioning indicates that with a larger sample size, the results would be more robust.

In addition, we were limited by the fact that the majority of our measures were completed by parents. Although we had objective cognitive and academic data, information regarding the home environment, social, emotional and behavioural functioning (with the exception of a CGAS rating) was based on parental report. There has been significant discussion in the paediatric literature regarding the ideal informant, because of recognised discrepancies between parents, teachers, children and healthcare provider reports (Sawyer *et al*. 1999; Wake *et al*. 2000). This study would have been strengthened by collecting data from multiple informants, specifically from children and teachers, in addition to gathering more objective data of social or behavioural functioning (e.g. facial expression recognition, problem-solving of social vignettes, parent–child interactions).

Finally, our study was limited by a relatively small sample size. With only 48 families, our analytical approach was restricted to correlational and regression models. We could not explore models of mediation or moderation, which would be particularly interesting because it could help us identify specific targets for intervention. Further, while there were trends in our data, we failed to reach statistical significance for many of the models examined in our analyses. A larger sample size could have strengthened our findings and helped us better

understand which environmental characteristics are particularly meaningful for this population.

Thus, future research needs to continue examining the influence of environmental variables on the functional outcomes of children with 22q11DS. There is a dearth of data in this field regarding environmental influences, which limits our understanding of those factors involved with the development of these children. The current study provided a novel examination of family-level social factors specifically. While more research should continue to parse out family-level influences, future studies should also expand this lens to include other factors in the child's ecological system. This may include an examination of school-based factors or community-level factors that collectively influence child development. Children with 22q11DS are at a high risk for a number of social, emotional and psychiatric sequelae, and the ability to identify environmental risk or resistance factors will aid in the development of effective interventions.

In summary, the current study contributed to the very small body of literature regarding the role of the family social environment in the functional outcomes of children with 22q11DS. The current results indicated a maladaptive effect of physical punishment, and suggested some protective role for high parental control and familial organisation. While these specific findings need to be explored further in future studies, more generally, this study provides evidence that the environment may play an important role in the outcomes of children with 22q11DS.

References

Achenbach, TM. Manual for the CBCL. University of Vermont; Burlington, VT: 1991.

Baker KD, Skuse DH. Adolescents and young adults with 22q11 deletion syndrome: psychopathology in an at-risk group. British Journal of Psychiatry. 2005; 186:115–20. [PubMed: 15684233]

Baumrind D. Effects of authoritative parental control on child behavior. Child Development. 1966; 37:887–907.

Baumrind D. Child care practices anteceding three patterns of preschool behavior. Genetic Psychology Monographs. 1967; 75:43–88. [PubMed: 6032134]

Blacher J, Nihira K, Meyers CE. Characteristics of home environment of families with mentally retarded children: comparison across levels of retardation. American Journal of Mental Deficiency. 1987; 91:313–20. [PubMed: 3812601]

Breslau, N. Family care of disabled children: effects on siblings and mothers. In: Rubin, L.; Thompson, G.; Bilenker, R., editors. Comprehensive Management of Cerebral Palsy. Grune & Stratton; NewYork: 1983. p. 299-309.

Chase-Carmichael CA, Ris MD, Weber AM, Schefft BK. Neurologic validity of the Wisconsin Card Sorting Test with a pediatric population. Clinical Neuropsycholog y. 1999; 13:405–13.

Chelune GJ, Baer RA. Developmental norms for the Wisconsin Card Sorting test. J Clinical and Experimental Neuropsycholog y. 1986; 8:219–28.

Cirino PT, Chin CE, Sevcik RA, Wolf M, Lovett M, Morris RD. Measuring socioeconomic status: reliability and preliminary validity for different approaches. Assessment. 2002; 9:145–55. [PubMed: 12066829]

Cunningham SD, Warschausky S, Thomas PD. Parenting and social functioning of children with and without cerebral palsy. Rehabilitation Psychology. 2009; 54:109–15. [PubMed: 19618710]

De Smedt B, Devriendt K, Fryns JP, Vogels A, Gewil-lig M, Swillen A. Intellectual abilities in a large sample of children with Velo-Cardio-Facial Syndrome: an update. Journal of Intellectual Disability Research. 2007; 51:666–70. [PubMed: 17845235]

De Smedt B, Swillen A, Verschaffel L, Ghesquiere P. Mathematical learning disabilities in children with 22q11.2 deletion syndrome: a review. Developmental Disability Research Review. 2009; 15:4–10.

DeBoer T, Zhongle W, Lee A, Simon TJ. Hippocampal volume reduction in children with chromosome 22q11.2 deletion syndrome is associated with cognitive impairment. Behavioral and Brain Functions. 2007; 3:54–63. [PubMed: 17956622]

Farah, MJ. Mind, Brain, and Education in Socioeconomic Context. Springer; New York: 2010.

Farah MJ, Shera DM, Savage JH. Childhood poverty: specific associations with neurocognitive development. Brain Research. 2006; 1110:166–74. [PubMed: 16879809]

Feinstein C, Eliez S, Blasey C, Reiss AL. Psychiatric disorders and behavioral problems in children with velocardiofacial syndrome: usefulness as phenotypic indicators of schizophrenia risk. Biological Psychiatry. 2002; 51:312–18. [PubMed: 11958782]

Gerdes M, Solot C, Wang PP, Moss E, LaRossa D, Randall P, et al. Cognitive and behavior profile of preschool children with chromosome 22q11.2 deletion. American Journal of Medical Genetics. 1999; 85:127–33. [PubMed: 10406665]

Gershoff ET. Corporal punishment by parents and associated child behaviors and experiences: a meta-analytic and theoretical review. Psychological Bulletin. 2002; 128:539–79. [PubMed: 12081081]

Gothelf D, Feinstein C, Thompson T, Gu E, Penniman L, Van SE. Risk factors for the emergence of psychotic disorders in adolescents with 22q11.2 deletion syndrome. American Journal of Psychiatry. 2007; 164:663–9. [PubMed: 17403981]

Gresham, FM.; Elliott, SN. Social Skills Rating System Manual. American Guidance Services; Circle Pines, MN: 1990.

Hewage C, Bohlin G, Wijewardena K, Lindmark G. Executive functions and child problem behaviors are sensitive to family disruption: a study of children of mothers working overseas. Developmental Science. 2011; 14:18–25. [PubMed: 21159084]

Hollingshead, AB. Two Factor Index of Social Position. Yale University; New Haven, CT: 1957.

Holmes CS, Yu Z, Frentz J. Chronic and discrete stress as predictors of children's adjustment. Journal of Consulting and Clinical Psychology. 1999; 67:411–19. [PubMed: 10369062]

Howell A, Hauser-Cram P, Kersh JE. Setting the stage: early child and family characteristics as predictors of later loneliness in children with developmental disabilities. American Journal on Mental Retardation. 2007; 112:18–30. [PubMed: 17181387]

Jansen PW, Duijff SN, Beemer FA, Vorstman JA, Klaassen PW, Morcus ME, et al. Behavioral problems in relation to intelligence in children with 22q11.2 deletion syndrome: a matched control study. American Journal of Medical Genetics. 2007; 143:574–80. [PubMed: 17318841]

Jolin EM, Weller RA, Jessani NR, Zackai EH, McDonald-McGinn DM, Weller EB. Affective disorders and other psychiatric diagnoses in children and adolescents with 22q11.2 Deletion Syndrome. Journal of Affective Disorders. 2009; 119:177–80. [PubMed: 19269692]

Jonson-Reid M, Presnall N, Drake B, Fox L, Bierut L, Reich W, et al. Effects of child maltreatment and inherited liability on antisocial development: an official records study. Journal of the American Academy of Child and Adolescent Psychiatry. 2010; 49:321–32. quiz 431. [PubMed: 20410725]

Kiley-Brabeck K, Sobin C. Social skills and executive function deficits in children with the 22q11 Deletion Syndrome. Applied Neuropsychology. 2006a; 13:258–68. [PubMed: 17362146]

Kiley-Brabeck K, Sobin C. Social skills and executive function deficits in children with the 22q11 deletion syndrome. Applied Neuropsychology. 2006b; 13:258–68. [PubMed: 17362146]

Kuo AY, Reiss AL, Freund LS, Huffman LC. Family environment and cognitive abilities in girls with fragile-X syndrome. Journal of Intellectual Disability Research. 2002; 46:328–39. [PubMed: 12000584]

Lamborn SD, Mounts NS, Steinberg L, Sanford M. Patterns of competence and adjustment among adolescents from authoritative, authoritarian, indulgent, and neglectful families. Child Development. 1991; 62:1049–65. [PubMed: 1756655]

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

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

Luthar SS. Vulnerability and resilience: a study of high-risk adolescents. Child Development. 1991; 62:600–16. [PubMed: 1914628]

Merikangas KR, He JP, Brody D, Fisher PW, Bourdon K, Koretz DS. Prevalence and treatment of mental disorders among U.S. children in the 2001–2004 NHANES. Pediatrics. 2010; 125:75–81. [PubMed: 20008426]

Moos, RH.; Moos, BM. Family Environment Scale: Manual. Consulting Psychologists Press; Palo Alto, CA: 1986.

Moos, RH.; Moos, BM. Family Environment Scale Manual and Sampler Test: Development, Applications and Research. 4th. Mind Garden, Inc.; Palo Alto, CA: 2009.

Moss EM, Batshaw ML, Solot CB, Gerdes M, Donald-McGinn DM, Driscoll DA, et al. Psychoeducational profile of the 22q11.2 microdeletion: a complex pattern. Journal of Pediatrics. 1999; 134:193–8. [PubMed: 9931529]

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

Niklasson L, Gillberg C. The neuropsychology of 22q11 deletion syndrome. A neuropsychiatric study of 100 individuals. Research in Developmental Disabilities. 2010; 31:185–94. [PubMed: 19815377]

Power, TG. Parenting Dimensions Inventory – Short Version (PDI-S): A Research Manual. Washington State University; Pullman, WA: 2002.

Prinzie P, Swillen A, Maes B, Onghena P, Vogels A, Van Hooste A, et al. Parenting, family contexts, and personality characteristics in youngsters with VCFS. Genetic Counseling. 2004; 15:141–57. [PubMed: 15287414]

Pueschelo SM, Myers BA. Environmental and temperament assessments of children with Down's syndrome. Journal of Intellectual Disability Research. 1994; 38:195–202. [PubMed: 8193455]

Redden SC, Ramey SL, Ramey CT, Forness SR, Brezausek CM. Special education placements among former head start children in kindergarten: a descriptive multi-site study. Education & Treatment of Children. 2003; 26:128–49.

Sawyer M, Antoniou G, Toogood I, Rice M. A comparison of parent and adolescent reports describing the health-related quality of life of adolescents treated for cancer. International Journal of Cancer Supplement. 1999; 12:39–45.

Shaffer D, Gould MS, Brasic J, Ambrosini P, Fisher P, Bird H, et al. A Children's Global Assessment Scale (CGAS). Archives of General Psychiatry. 1983; 40:1228–31. [PubMed: 6639293]

Shaffer D, Fisher P, Lucas CP, Dulcan MK, Schwab-Stone ME. NIMH Diagnostic Interview Schedule for Children Version IV (NIMH DISC-IV): description, differences from previous versions, and reliability of some common diagnoses. Journal of the American Academy of Child and Adolescent Psychiatry. 2000; 39:28–38. [PubMed: 10638065]

Shashi V, Howard TD, Keshavan MS, Kaczorowski J, Berry MN, Schoch K, et al. COMT and anxiety and cognition in children with chromosome 22q11.2 deletion syndrome. Psychiatry Research. 2010a; 178:433–6. [PubMed: 20488547]

Shashi V, Keshavan M, Kaczorowski J, Schoch K, Lewandowski KE, McConkie-Rosell A, et al. Socioeconomic status and psychological function in children with chromosome 22q11.2 deletion syndrome: implications for genetic counseling. Journal of Genetic Counseling. 2010b; 19:535–44. [PubMed: 20680421]

Shashi V, Veerapandiyan A, Schoch K, Kwapil TR, Keshavan MS, Ip EH, et al. Social skills and associated psychopathology in children with chromosome 22q11.2 deletion syndrome: implications for interventions. Journal of Intellectual Disability Research. 2012; 56:865–78. [PubMed: 21883601]

Shprintzen RJ. Velo-cardio-facial syndrome: 30 years of study. Developmental Disabilities Research Reviews. 2008; 14:3–10. [PubMed: 18636631]

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

Simon TJ. A new account of the neurocognitive foundations of impairments in space, time and number processing in children with chromosome 22q11.2 deletion syndrome. Developmental Disability Research Reviews. 2008; 14:52–8.

Simon TJ, Bish JP, Bearden CE, Ding L, Ferrante S, Nguyen V, et al. A multilevel analysis of cognitive dysfunction and psychopathology associated with chromosome 22q11.2 deletion syndrome in children. Development and Psychopathology. 2005; 17:753–84. [PubMed: 16262991]

Sobin C, Kiley-Brabeck K, Karayiorgou M. Associations between prepulse inhibition and executive visual attention in children with the 22q11 deletion syndrome. Molecular Psychiatry. 2005; 10:553–62. [PubMed: 15520831]

Swillen A, Devriendt K, Legius E, Eyskens B, Dumoulin M, Gewillig M, et al. Intelligence and psychosocial adjustment in velocardiofacial syndrome: a study of 37 children and adolescents with VCFS. Journal of Medical Genetics. 1997; 34:453–8. [PubMed: 9192263]

Swillen A, Devriendt K, Legius E, Prinzie P, Vogels A, Ghesquiere P, et al. The behavioural phenotype in velo-cardio-facial syndrome (VCFS): from infancy to adolescence. Genetic Counseling. 1999; 10:79–88. [PubMed: 10191433]

Wake M, Hesketh K, Cameron F. The Child Health Questionnaire in children with diabetes: cross-sectional survey of parent and adolescent-reported functional health status. Diabetes Medicine. 2000; 17:700–7.

Wechsler, D. Wechsler Intelligence Scale for Children. 4th. The Psychological Corporation; San Antonio, TX: 2003.

Wechsler, D. Wechsler Individual Achievement Test. 3rd. The Psychological Corporation; San Antonio, TX: 2009.

Whitley E, Gale CR, Deary IJ, Kivimaki M, Batty GD. Association of maternal and paternal IQ with offspring conduct, emotional, and attention problem scores. Transgenerational evidence from the 1958 British Birth Cohort Study. Archives of General Psychiatry. 2011; 68:1032–8. [PubMed: 21969461]

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]

Table I

Means and standard deviations by variable

Measure	22ql IDS Mean (SD)	Measure	22qllDS Mean (SD)	Normal families Mean (SD)*	Distressed families Mean (SD)*
CBCL Total Problem Behaviour	64.0 (8.86)	FES Cohesion	7.22 (2.25)	6.73 (1.47)	5.25 (2.13)
CBCL Total Competency†	36.8 (8.02)	FES Expressiveness	5.79 (1.97)	5.54 (1.61)	4.71 (1.78)
CBCL Oppositional Behaviour	58.4 (6.95)	FES Conflict	2.14 (1.97)	3.18 (1.91)	4.02 (2.07)
SSRS Total Social Skills‡	85.9 (15.97)	FES Independence	5.91 (1.64)	6.66 (1.26)	6.03 (1.35)
SSRS Problems‡	107.8 (15.95)	FES Achievement orientation	4.66 (2.10)	5.47 (1.62)	5.33 (1.58)
WIAT Broad Reading‡	85.2 (18.18)	FES Intellectual orientation	6.20 (2.28)	5.56 (1.82)	4.62 (1.98)
WCST	88.3 (1 1.37)	FES Active-Recreational	5.33 (2.35)	5.33 (1.96)	4.15 (1.96)
CGAS	64.7 (7.37)	FES Moral-Religious	6.27 (2.37)	4.75 (2.03)	4.51 (1.96)
SES§	29.3 (13.45)	FES Organisation	5.68 (2.61)	5.47 (1.90)	5.07 (1.97)
PDI Physical Punishment	0.5 (0.66)	FES Control	5.22 (2.02)	4.26 (1.84)	4.61 (1.89)
PDI Organisation	3.9 (1.2)				
PDI Letting the Situation Go	0.1 (0.27)				
PDI Nurturance	4.83 (1.2)				
PDI Inconsistency	2.31 (.86)				
PDI Following Through on Discipline	3.91 (1.18)				
PDI Amount of Control	3.91 (1.21)				

*Cited in Moos & Moos (2009). Distressed norms derived from families with depressed individuals, children in crisis, psychiatric patients and alcohol use problems. Distressed families are lower on cohesion, expressiveness, independence, intellectual and recreational orientation and higher on conflict.

†*T*-scores, where M = 50, SD = 10.

‡Standard scores, M = 100, SD = 15.

§SES is determined from Hollingshead Index; higher numbers indicate lower SES.

22qn11DS, 22qn11.2 deletion syndrome; SD, standard deviation; CBCL, Child Behaviour Checklist; SSRS, Social Skills Rating Scale; WIAT, Wechsler Individual Achievement Test - III; WCST, Wisconsin Card Sorting Task; CGAS, Children's Global Assessment Scale; PDI, Parenting Dimension Inventory.

Physical Punishment: range 0–3, with higher scores indicating more physical punishment; Organisation: range 1–6, higher scores indicating more organisation; Letting the Situation Go: 0–3, with higher responses indicating parent more likely to let situation go; Nurturance: 1–6, higher responses indicate more nurturance; Inconsistency: 1–6, higher scores indicate a higher likelihood of inconsistency; Following Through on Discipline: 1–6, higher scores indicate more following through on discipline; Amount of Control: 0–5, higher scores representing greater parental control.

FES: Family Environment Scale; raw scores, possible scores: 0–9.

FES Control - higher scores indicate more structured procedures in the family; Achievement orientation - higher scores indicate greater family emphasis on achievement and competition.

SES: socioeconomic status; lower values indicate higher SES.

Table 2

Correlation matrix of study outcome variables with the Family Environment Scale - Real Form (FES-R)

	Cohesion	Expressiveness	Conflict	Independence	Achievement orientation	Intellectual-Cultural orientation	Active-Recreation orientation	Moral-Religious emphasis	Organisation	Control
Social-behavioural measures										
CBCL Competency Total	0.26	−0.25	−0.06	−0.28	0.21	0.14	0.29	0.14	0.17	0.18
CBCL Behaviour Problems Total	−0.03	−0.06	0.16	0.00	−0.1 1	0.12	−0.01	0.12	−0.17	0.01
SSRS Total Social Skills	0.09	−0.06	0.02	−0.29	0.28	0.04	0.20	0.06	0.14	−0.15
SSRS Problem Behaviour Total	0.06	0.03	0.25	0.05	−0.07	0.21	0.13	0.20	−0.11	0.19
Neurocognitive and overall functioning										
CGAS	0.01	−0.12	0.02	−0.18	**0.376***	0.12	0.19	0.03	0.22	0.24
Hollingshead SES	−0.06	−0.07	−0.07	0.04	0.01	**−0.364***	−0.22	0.18	0.02	0.05
WIAT - Reading	0.14	0.08	0.06	−0.24	0.17	−0.04	0.06	0.12	0.26	**0.400***
WCST - Perseverative Errors	0.02	0.12	0.29	−0.03	0.003	0.12	0.05	−0.03	0.03	0.28

*Correlation is significant at the 0.05 level;

**correlation is significant at the 0.01 level.

Medium and large effect sizes are in bold.

CBCL, Child Behaviour Checklist; SSRS, Social Skills Rating Scale; WIAT, Wechsler Individual Achievement Test - III; WCST, Wisconsin Card Sorting Task; CGAS, Children's Global Assessment Scale; FES, FamilyEnvironment Scale; SES, socioeconomic status.

Table 3

Correlation matrix of study outcome variables with the Parenting Dimensions Inventory Short Form (PDI-S)

	Nurturance	Inconsistency	Following Through on Discipline	Organisation	Amount of Control	Let Situation Go	Physical Punishment
Social-behavioural measures							
CBCL Competency Total	0.25	−0.24	0.23	0.30	0.05	0.12	**−0.421****
CBCL Behaviour Problems Total	0.08	0.01	−0.10	**−0.1**	0.03	0.01	**0.311***
SSRS Total Social Skills	0.18	−0.10	0.01	0.09	−0.03	−0.13	**−0.380***
SSRS Problem Behaviours	0.03	0.10	−0.00	0.06	0.12	0.08	0.28
Neurocognitive and overall functioning measures							
CGAS	0.12	−0.07	0.07	**0.347***	0.09	−0.25	−0.01
Hollingshead SES of family	**−0.14**	0.14	0.13	−0.21	0.14	0.12	**0.315***
WIAT - Reading	0.16	0.00	−0.03	**0.376***	0.08	**−0.430****	0.00
WCST - Perseverative Errors	−0.04	0.28	−0.18	**0.372***	0.04	−0.06	−0.13

* Correlation is significant at the 0.05 level;

** correlation is significant at the 0.01 level.

Medium and large effect sizes are in bold.

CBCL, Child Behaviour Checklist; SSRS, Social Skills Rating Scale; WIAT, Wechsler Individual Achievement Test - III; WCST, Wisconsin Card Sorting Task; CGAS, Children's Global Assessment Scale; FES, Family Environment Scale; SES, socioeconomic status.

Table 4

Hierarchical regression analyses to differentially predict social-emotional functioning in children with 22qn11.2 deletion syndrome (22qn11DS)

	Anxiety dignosis			**FES conflict**			**PDI Physical Punishment**			**PDI Nurturance**			**Physical Punishment × Conflict**		
Criterion	***β***	***R²***	***f²***	***β***	***R² change***	***f²***	***β***	***R² change***	***f²***	***β***	***R² change***	***f²***	***β***	***R² change***	***f²***
CBCL Total Problem Behaviours	0.47**	0.22	**0.28**	0.14	0.02	0.04	0.29*	0.08	0.12	0.05	0.002	0.004	1.2	0.02	0.03
	ADHD diagnosis			**PDI Organisition**			**PDI Physical Punishment**			**Physical Punishment × Organisation**					
Criterion	*β*	*R²*	*f²*	*β*	*R²* Change	*f²*	*β*	*R²* Change	*f²*	*β*		*R²* change		*f²*	
CBCL Total Competency	−0.32*	0.10	**0.11**	0.18	0.03	0.03	−0.36*	0.13	**0.17**	−0.12		0.00		0.00	
	Anxiety diagnosis			**Child Gender**			**PDl Physical Punishment**			**FES Independence**			**Physical Punishment × Independence**		
Criterion	*β*	*R²*	*f²*	*β*	*R²* change	*f²*	*β*	*R² change*	*f²*	*β*	*ΔR²*	*f²*	*β*	*R²* change	*f²*
SSRS Total Social Skills	−0.65**	0.42	**10.8**	−0.30**	0.95	**0.20**	−0.39**	0.146	**0.43.**	0.05	0.003	0.009	0.08	0.00	0.00

*$P < 0.05$.

**$P < 0.01$. Medium (0.15) and large (0.35) effect sizes (f^2 are in bold.

CBCL, Child Behaviour Checklist; SSRS, Social Skills Rating Scale; PDI, Parenting Dimension Inventory; FES, Family Environment Scale; SES, socioeconomic status.