Author Manuscript

Author Manuscript

Author Manuscript

Author Manuscript

HHS Public Access

Author manuscript

Am J Intellect Dev Disabil. Author manuscript; available in PMC 2020 August 31.

Published in final edited form as:

Am J Intellect Dev Disabil. 2017 September ; 122(5): 359–373. doi:10.1352/1944-7558-122.5.359.

DIFFERENTIAL RELATIONSHIPS OF ANXIETY AND AUTISM SYMPTOMS ON SOCIAL SKILLS IN YOUNG BOYS WITH FRAGILE X SYNDROME

Debra L. Reisinger[1], **Jane E. Roberts**[1]

[1]Department of Psychology, University of South Carolina

Abstract

Social skills are critical for academic, social, and psychological success of children with both typical and atypical development. Boys with fragile X syndrome (FXS) are at high risk for social skill impairments given intellectual impairments and secondary conditions. The present study examines the impact of adaptive behavior, autism symptoms and anxiety symptoms to social skills at the composite and subdomain level in boys with FXS across age. This cross-sectional study included boys with FXS (3 – 14 years) contrasted to age matched typical control boys. Results revealed that social skills are generally within developmental expectations with adaptive behavior as the primary predictor. Anxiety and autism symptoms emerged as additive risk factors, particularly in the areas of responsibility and self-control.

Keywords

fragile X syndrome; autism; anxiety; social skills

Social skills are learned, socially accepted behaviors involving initiations and responses across a variety of interactive and specific situations (Frey, Elliot, & Gresham, 2011; Merrell & Gimpel, 2014). Although social skills fall under the broad social behavior umbrella, they are considered a specific class of social behaviors exhibited by an individual to successfully complete a social task (Gresham, 2016). Social skills allow children to develop meaningful social relationships and have been shown to play a key role in children's academic, social, and psychological outcomes (Frey, Elliot, & Gresham, 2011; Gillis, Callahan, & Romanczyk, 2011). They rely on the interplay of a variety of psychological constructs and basic human traits including: personality, intelligence, language, perception, appraisal, attitude, and behavior-environment interactions (Merrell & Gimpel, 2014). In early childhood, core social skills include: listening to others, following rules, taking turns, asking for help, cooperating with peers, and controlling emotions at times of conflict (Elliot, Roach, & Beddow, 2008). However, social skills vary and evolve across development with the acquisition of specific social skills typically emerging and improving across early childhood, middle childhood, and adolescence (Merrell & Gimpel, 2014).

Corresponding Author: Correspondence concerning this article should be addressed to Debra L. Reisinger, University of South Carolina, Department of Psychology, 1512 Pendleton St., Columbia, SC 29208 (reisingd@email.sc.edu).

Author Manuscript Author Manuscript Author Manuscript Author Manuscript

Social skill deficits are present across the spectrum of impairment including those without a formal diagnosis to those with one or more specific diagnoses or conditions. In individuals with intellectual disabilities (ID), limitations in social skills are a central characteristic with negative effects on their overall adaptive behavior (De Bildt et al, 2005). Difficulties have also been shown in children with learning disabilities, language impairment, anxiety disorders, and autism spectrum disorders (Griswold & Townsend, 2015; Merrell & Gimpel, 2014; Schalock et al., 2010). Specific social patterns have been identified as characteristic of certain disorders within the literature. For example, individuals with non-specific ID tend to display significant deficits or limitations in their social skills across all ages (Merrell & Gimpel, 2014), whereas individuals with Down syndrome show relative strengths in their socialization skills despite their intellectual functioning (Fidler, Hepburn, & Rogers, 2006). Similarly, research in other disorders such as Williams syndrome, who are found to be hypersocial, and chromosome 22q11.2 deletion syndrome, who have significant social deficits, has suggested that social functioning is not and should not be solely related to cognitive functioning (Doyle, Bellugi, Korenberg, & Graham, 2004; Shashi et al., 2012). A common assumption is to assume social deficits are related to cognitive deficits, or diagnostic overshadowing (Feinstein & Singh, 2007). In individuals with high anxiety, both clinical and non-clinical samples have shown poorer social skills (Coplan, Prakash, O'Neil, & Armer, 2004; Schneider, 2009; Spinrad et al., 2004) with longitudinal studies suggesting anxiety predicts future social skill deficits (Thorell, Bohlin, & Rydell, 2004). For individuals with autism spectrum disorder (ASD), social skill deficits are one of the core features represented by impairments in social communication present across the range of cognitive and language abilities (Carter, Davis, Klin, & Volkmar, 2005).

Fragile X Syndrome

Fragile X syndrome (FXS) is caused by a trinucleotide (CCG) repeat expansion on the fragile X mental retardation 1 (FMR1) gene associated with reductions in FMR1 protein resulting in physical, cognitive, and behavioral deficits (Hagerman & Hagerman, 2002; Hall et al., 2009). FXS is the leading known heritable cause of ID occurring in 1 in 4,000 males and is associated with a variety of co-occurring conditions including social impairments, autism and anxiety (Cordeiro et al., 2011; Hall, Lightbody, Huffman, Lazzeroni, & Reiss, 2009; Kau et al., 2004). One of the most frequent and disabling behavioral abnormalities is social avoidance in young boys with FXS that is associated with social anxiety and hyperarousal (Kau et al, 2004; Kaufmann et al, 2004; Roberts et al., 2009; Roberts, Weisenfield, Hatton, Heath, Kaufmann, 2007). Reports of adaptive socialization, friendships and interpersonal skills indicate deficits across all these domains with the degree to which males with FXS are able to interact with others cited as one of the most critical factors predicting adult outcomes (Bailey, Raspa, Holiday, Bishop, & Olmsted, 2008; Hartley et al., 2011; Klaiman et al., 2014).

One of the most prominent features associated with impaired socialization in FXS is the presence of autism symptoms. ASD is one of the most highly recognized behavioral abnormalities occurring in males with FXS with 60-70% of males with FXS meeting diagnostic criteria for ASD in comparison to 30-60% of all children with FXS meeting diagnostic criteria for ASD (Klusek et al., 2014; Talisa, Boyle, Crafa, & Kaufmann, 2014;

Author Manuscript Author Manuscript Author Manuscript Author Manuscript

Thurman et al., 2014). There is controversy in the literature about the overlap between ASD and FXS. Some purport that FXS is a subtype of the spectrum of ASD, whereas others believe they represent two distinct conditions with fundamental differences including more mild social and communication impairments in FXS coupled with more impaired general cognitive abilities (Abbeduto, McDuffie, & Thurman, 2014; Bailey, Hatton, Mesibov, Ament, & Skinner, 2000; Kau et al., 2004; Kaufmann et al., 2004). Overall, males with FXS who meet diagnostic criteria for ASD are at risk for markedly poorer outcomes compared to those with only FXS (Abbeduto, McDfufie & Thurman, 2014; Hartley et al., 2011).

The presence of anxiety disorders has also been listed as one of the primary predictors of impaired socialization in FXS with approximately 70-83% of individuals with FXS diagnosed with one or more anxiety disorders (Cordeiro et al., 2011; Kaufmann et al., 2004; Tonnsen et al., 2013). Anxiety symptoms within FXS have been associated with a range of behavioral problems including: poor eye contact, gaze aversion, excessive shyness, hand flapping, hand biting, aggression, and autism symptoms (Boyle & Kaufmann, 2010; Tranfaglia, 2011). Research has shown that poor social behaviors are related to social anxiety within ASD and FXS (Hall et al., 2009).

To date, research has primarily focused on examining broad socialization indices in FXS rather than focus on important social skills that contribute to social success and are central for later academic and social outcomes. Despite the importance of social skills to outcomes in both typical and atypical conditions, no study has reported on age effects or variables that relate to social skills in FXS. This work is complex given the overlapping features across ID, ASD and anxiety and the measurement challenges associated with limited insight and impaired communication problems in FXS (Cordeiro et al., 2011). Although challenging, the present study aims to develop a better understanding of the potential relationships of anxiety and ASD symptoms on social skills within young boys with FXS to add to the literature on the social behavioral phenotype of FXS. The current study examines social skills at the composite and subdomain level in young boys with FXS in contrast to typically developing (TD) boys and in relation to age, adaptive behavior, anxiety symptoms and autism symptomology. The research questions and hypotheses are as follows:

1. What is the relationship between social skills and age in young boys with FXS compared to chronological age-matched TD boys controlling for adaptive behavior? We hypothesized that boys with FXS would display significant deficits in social skills compared to TD boys across chronological age with strong adaptive skills as a significant predictor of greater social skills.
2. How are features of autism and anxiety in boys with FXS related to their social skill development controlling for adaptive behavior? We hypothesized that elevated features of anxiety and autism spectrum disorders would be associated with decreased social skill competency.

Author Manuscript Author Manuscript Author Manuscript Author Manuscript

Method

Participants

Data were drawn from a series of three longitudinal studies examining development and achievement in males with FXS. Each of the longitudinal studies were complementary; however, they each had independent specific aims and methodology as evidenced by some currently published literature (Baker et al., 2011; Hooper et al., 2008; Klusek et al., 2015). Participants were recruited across the United States through FXS parent list serves, FXS parent support groups, genetic clinics, developmental evaluation centers, early intervention programs, and ongoing research studies. Control participants were recruited locally through flyers and word of mouth. All males with FXS had the FMR1 full mutation based on standard DNA testing. Participants were excluded from the study if they had any preexisting sensory conditions (e.g., hearing or vision impairment). The typically developing boys had no reported or suspected developmental concerns and fell within the average range (e.g., between 80 and 120) of functioning on developmental measures completed through the longitudinal studies. Developmental measures varied across the three longitudinal studies with some participants being administered the Mullen Scales of Early Learning (MSEL; Mullen, 1995) or the Leiter International Performance Scale-Revised (Leiter-R; Roid & Miller, 1997).

To answer the first research question comparing social skills between FXS and TD boys, 64 males with FXS (ages 3-7 years, 6 months, mean age = 4.38 years) and 65 TD males (ages 3-6 years, 6 months, mean age = 4.53 years) from the longitudinal studies were included. Although several participants had multiple data points available, we selected each participant's first data point and matched all scores from the same data point for both sets of data to identify emerging relationships on social skills. Groups were matched on chronological age (t= 0.692, p=0.50) according to the criteria outlined by Mervis and Robinson (2003). First, we compared the TD and FXS boys matched on chronological age for the full sample independent of adaptive behavior or developmental level. Due to the series of longitudinal studies measuring developmental level with different measures (MSEL or Leiter-R), we utilized the Vineland Adaptive Behavior Scales (VABS; Sparrow, Balla, & Cicchetti, 1984) to obtain a measure of current developmental functioning across all participants to maximize the power of the analyses. Next, we compared a subset of 21 TD (ages 3-4 years, 2 months, mean age = 3.47) and 44 FXS (ages 3-4 years, 7 months, mean age = 3.55) boys matched on chronological age (t=0.68, p=0.50) utilizing the same above mentioned criteria that had complete social skills data and adaptive behavior data, which was used as a proxy for developmental level. Due to the potential confound with adaptive behavior and social skills (e.g., socialization is one of the domains on the adaptive measure), we supplemented analyses utilizing a similar subsample (three of the participants within the larger sample had either adaptive behavior or developmental level, but not both) of 21 TD (ages 3-4 years, 2 months, mean age = 3.47) and 45 FXS (ages 3-4 years, 7 months, mean age = 3.54) boys and included developmental level in the model using the Early Learning Composite (ELC) from the MSEL to verify the results found utilizing adaptive behavior.

Author Manuscript Author Manuscript Author Manuscript Author Manuscript

To answer the second research question examining predictors of social skills within the group of males with FXS, we expanded our dataset to 102 males with FXS (ages 3-14 years, 3 months, mean age = 6.34 years) who had social skills data available. This FXS dataset was able to be expanded given we did not have to match to the typical controls. All of the participants had either autism severity data or anxiety data available; however, 4 boys with FXS did not have CARS data and 13 FXS boys did not have anxiety data available. Table 1 and Table 2 summarize the descriptive statistics for the samples and subsamples.

Measures

Social Skills.—Parent report of children's social skills was obtained using the preschool version (for ages 3 years to 4 years, 11 months) and elementary version (for grades K-6) of the Social Skills Rating System (SSRS; Gresham & Elliot, 1990; Frey, Elliot, & Gresham, 2011). The SSRS is a standardized questionnaire that measures the perceived frequency with which social skills are displayed in the home and the community, problem behaviors that might interfere with the acquisition of these important social skills, and academic competence. The SSRS is a norm referenced 3-point rating scale (never, often, very often) including 55 questions. The SSRS is a well-established measure of social skills with young, school age children with an internal reliability ranging from 0.73 – 0.93 across forms and subscales (Matson & Wilkins, 2009) and strong internal consistencies with high alpha coefficients for the parent version of 0.89 (S. Van der Oord et al., 2005). Data from our study also indicate high internal consistency with a Cronbach's alpha of 0.92 for both measures. Total social skills standard scores were used in our analyses.

There are four core subdomains of social skills in the SSRS: cooperation, assertion, responsibility and self-control (Gresham & Elliot, 1990). Cooperation includes behaviors such as helping others, sharing materials, and complying with rules and directions. Assertion includes initiating behaviors, asking for information, introducing oneself, and responding to the action of others, such as peer pressure or insults. Responsibility behaviors include demonstrating the ability to communicate with adults properly, such as questioning household rules and asking for permission. Self-control behaviors emerge in situations of conflict, including responding appropriately to teasing, and in non-conflict situations such as taking turns and compromising. The SSRS does not provide scaled scores for the prosocial domains; however, for these analyses, participant's raw scores were used for the prosocial subscales. Raw scores represent the rating of how often a child is displaying a certain behavior of a rating from 0 ("never"), 1 ("sometimes"), and 2 ("often"). The range of raw scores for each subscale is 0-20 across both the preschool and early elementary form.

Adaptive Behavior.—The Vineland Adaptive Behavior Scales, Survey Form (VABS) was used to assess the development of adaptive functioning (Sparrow, Balla, & Cicchetti, 1984). The VABS is a semi-structured interview that was administered to the participant's parents to obtain their overall adaptive functioning from four subdomains (communication, daily living skills, socialization, and motor skills). The VABS provides an adaptive behavior composite score (mean of 100 and standard deviation of 15) that was used in the current study to control for adaptive functioning across groups. The VABS was used due to inconsistent measures of cognitive functioning across participants as a consistent measure of

Author Manuscript Author Manuscript Author Manuscript Author Manuscript

development/intelligence was not available across the participants. The VABS has been reported to have good test-retest reliability with ranges from 0.76 to 0.93 (Rosenbaum, Saigal, Szatmari, & Hoult, 1995). Additionally, the VABS has shown adequate concurrent validity ranging from 0.40-0.70 (Harrison, 1987).

Developmental Level.—The MSEL is a standardized assessment of cognitive and motor abilities yielding a norm-referenced full-scale cognitive functioning in infants and young children. Four (visual reception, receptive language, expressive language, and fine motor) domains on the MSEL comprise the ELC. The ELC standard score was utilized in the present study for a subset of participants due to the potential confound of socialization being measured in the VABS and to verify that any found results remain after controlling for developmental level. Adequate reliability and concurrent validity for the MSEL have been reported with the Bailey Scales of Infant Development (Mullen, 1995).

Anxiety Symptomology.—Anxiety symptoms were measured through parent report using the Child Behavior Checklist (CBCL/ 1.5-5, 6-18; Achenbach, 1991; Achenbach & Rescorla, 2001). The CBCL has two separate forms for children, ages 1.5-5 and 6-18 years, containing syndrome specific scales, broad composite scales, and DSM IV-based symptoms scales. Raters score the items on a scale of 0 (not true), 1 (somewhat or sometimes true), and 2 (very true or often true), based on their child's behavioral, emotional, and social functioning. The current study uses the DSM IV-Anxiety subscale T -score, consisting of six of the eight CBCL anxiety/depression syndrome scale items, which have been shown to be consistent with the DSM-IV anxiety disorder criteria and are present on both forms (Achenbach, Dumenci, & Rescorla, 2003). A T-score 70 or above is considered clinically significant with 17% (N=15) of the present study's data meeting this criteria. For the DSM-IV anxiety scale, previous research has shown adequate reliability with a Cronbach's alpha of 0.77 and adequate concurrent validity of 0.59 (Nakamura, Ebesutani, Bernstein, & Chorpita, 2009).

Autism Symptomology.—Autism symptoms were assessed with the Childhood Autism Rating Scale (CARS; Schopler et al., 1988). The CARS is an examiner rating scale of behavioral symptoms and characteristics of autism symptom severity in children that has been used in several laboratories (Brock & Hatton, 2010; Roberts et al., 2007). The CARS consists of 15 areas rated on a scale of 1 (typical development) to 4 (severely abnormal) that cover a range of behaviors including body use, activity level, and communication (verbal and nonverbal). A total score of 30 or above is considered to have mild to severe autism symptoms, whereas a score of less than 30 is considered to have minimal to no symptoms. In the present study, the total score was utilized with 32% of the FXS sample meeting criteria, which is consistent with previous reports (Bailey et al., 1998; Hatton et al., 2006). Examiners were trained to complete the CARS using videotaped training materials, review of cases and achievement of at least 80% reliability. Examiners rated the CARS through consensus scoring based on direct observation, parent interviews, and parent rating scales. The CARS has been shown to be a reliable measure with ratings of 0.73-0.90 (Magyar & Pandolfi, 2007). Data from our study indicates a high level of internal consistency with a Cronbach's alpha of 0.96. Several studies have attested to the CARS validity and its ability

Author Manuscript Author Manuscript Author Manuscript Author Manuscript

to discriminate between autism and non-autism samples (Bailey et al., 2000; Hatton et al., 2006) and across other developmental disorders (Chlebowski, Green, Barton, & Fein, 2010; Rellini et al., 2004).

Procedures

Participants were primarily assessed in their home with trained examiners administering the MSEL and the participant's parent completing the SSRS and CBCL within a 2-week window of the direct assessment. A trained examiner administered the VABS to the participant's parent. Trained examiners completed the CARS upon the completion of each assessment based on their observation of the child during the assessment. Some of the typically developing sample did not have VABS or MSEL scores due to it not being administered across all studies for which data were pulled. Some of the FXS participants did not have CARS or CBCL data due to data collection errors or participant non-compliance.

Data Analysis

Preliminary analyses were conducted to examine outliers, nonnormality, linearity, and homogeneity of residuals. A Pearson correlation analysis revealed that there was little relationship between autism symptomology (CARS) and anxiety symptoms (CBCL), $r(98) = 0.10$, $p < .05$, two-tailed. All data were converted to z-scores to standardize measures on the same scale. Regression analyses were used to examine the relationship between social skills and chronological age while including adaptive behavior and developmental level in the models, respectively, between FXS and TD boys. Next, regression analyses were completed to examine the relationship of autism symptomology and chronological age in predicting social skills in boys with FXS. Finally, regression analyses were utilized to examine the relationship of anxiety severity and chronological age on social skills in boys with FXS. The final two models included adaptive behavior to examine the relationships of anxiety and autism symptom severity on social skills above and beyond the effects of developmental level in boy with FXS.

Results

Development of Overall Social Skills in FXS and TD Boys

Chronological Age.—A regression model analyzed the effects of chronological age between groups (i.e., TD and FXS) on total social skills and the prosocial subscales with an interaction indicated (B=0.30, SE=0.17, t=2.36) on total social skills, $F(3, 125)=57.12$, $p<0.001$, $R^2=0.58$ (Table 3). As shown in Figure 4, both TD and FXS boys show an incline in total social skills with older age; however, boys with FXS show lower total social skills compared to their TD peers. When examining the prosocial subscales, a significant interaction of chronological age and group was observed on the assertion subscale (B=0.36, SE=0.14, t=2.54; $F(3, 125)=37.99$, $p<0.001$, $R^2=0.47$) and responsibility subscale (B=0.37, SE=0.11, t=3.36; $F(3, 125)=89.93$, $p<0.001$, $R^2=0.68$). Conversely, no significant interaction of chronological age and group was observed on the cooperation (B=0.20, SE=0.15, t=1.32; $F(3, 125)=29.57$, $p<0.001$, $R^2=0.41$) and self-control (B=0.07, SE=0.15, t=0.50; $F(3, 125)=25.19$, $p<0.001$, $R^2=0.37$) subscales.

Author Manuscript Author Manuscript Author Manuscript Author Manuscript

Adaptive Behavior.—A regression model examined the effects of chronological age and group on total social skills and the prosocial subscales while including adaptive behavior in the model to determine the effect of adaptive behavior on social skill development. No significant interaction or main effects were found while controlling for adaptive behavior, $F(4,60)=18.43$, $p<0.001$, $R^2=0.55$ (Table 3). Within the prosocial subscales, no significant interactions or main effects were observed for the cooperation subscale, $F(4, 60)=8.97$, $p<0.001$, $R^2=0.37$. For assertion, a significant main effect was found for adaptive behavior ($B=0.53$, $SE=0.19$, $t=2.82$), $F(4, 60)=11.35$, $p<0.001$, $R^2=0.43$. Within the self-control subscale, the main effect of group was approaching significance ($B=0.91$, $SE=0.46$, $t=1.97$), $F(4, 64)=6.96$, $p<0.001$, $R^2=0.26$. Lastly, for responsibility, a significant main effect was found for group ($B=0.83$, $SE=0.32$, $t=2.62$) and adaptive behavior ($B=0.48$, $SE=0.15$, $t=3.20$), $F(4, 60)=29.85$, $p<0.001$, $R^2=0.67$. Overall, these results suggest that adaptive behavior, not chronological age, largely accounts for the group differences in social skills and the prosocial subscales in boys with FXS compared to TD boys (Figure 1).

Developmental Level.: Due to our measure of adaptive behavior including a socialization component, supplemental regression models were used to verify that these results remain when including developmental level in the model for the subset of participants who were administered the MSEL. A regression model was used to examine the effects of chronological age and group on total social skills and the prosocial subscales while including developmental level in the model on a similar subset of participants who had such data available. No interaction or main effects were found when including developmental level, $F(4, 61)=15.49$, $p<0.001$, $R^2=0.50$ (Table 3). Within the prosocial subscales, no significant interactions or main effects were observed for the cooperation ($F(4, 61)=7.61$, $p<0.001$, $R^2=0.33$) or self-control ($F(4, 61)=7.23$, $p<0.001$, $R^2=0.32$) subscales. For the assertion subscale, developmental level was approaching significance ($B=0.02$, $SE=0.01$, $t=1.87$), $F(4, 61)=9.85$, $p<0.001$, $R^2=0.39$. For responsibility, group was approaching significance ($B=-1.29$, $SE=0.65$, $t=-1.97$), $F(4, 64)=18.91$, $p<0.001$, $R^2=0.54$. These results support our earlier findings suggesting that adaptive behavior and/or developmental level, not chronological age, accounts for the group differences in social skills across age in boys with FXS compared to TD boys.

Autism Symptomology in FXS on Social Skills

Regression analyses were used to analyze the hypothesis that increased autism symptomology is associated with decreased social skills in boys with FXS on chronological age. As shown in Table 4, a significant interaction of chronological age ($B=0.56$, $SE=0.17$, $t=3.31$) and autism symptomology ($B=0.11$, $SE=0.53$, $t=0.83$) on total social skills ($F(4,93)=15.65$, $p<0.001$, $R^2=0.38$) was found while including adaptive behavior in the model. When examining the effect of autism symptomology and chronological age on the prosocial subscales, an interaction was only found for the responsibility ($B=-0.21$, $SE=0.81$, $t=-2.55$) subscale ($F(4, 93)=31.41$, $p<0.001$, $R^2=0.57$). For the cooperation, assertion, and self-control subscales, no interactions were observed ($ps>0.05$). To probe the interactions, CARS scores were used to create three equal groups (e.g., low [$N=32$, *range*=18-25], medium [$N=33$, *range*=25-29], and high [$N=33$, *range*=30-48]) and graphed. As shown in Figure 2, boys with FXS who have low autism symptomology show a significant increase in

Author Manuscript Author Manuscript Author Manuscript Author Manuscript

their total social skills as they get older compared to the other two groups. Those with mid-level autism symptomology have higher total social skills at younger ages with minimal increases in their social skills as they get older; however, those with high autism symptomology have very low total social skills at young ages with minimal increases in social skills at older ages. For responsibility, all three groups show an increase in their responsibility skills as they get older; however, the medium and high autism symptomology groups do not show similar increases in their social skills as the low autism symptomology group. Overall, the high autism symptomology group shows minimal increases in their responsibility skills as they get older.

Anxiety Symptoms in FXS on Social Skills

Regression analyses were used to analyze the hypothesis that anxiety symptoms will be a predictor of social skill deficits in boys with FXS. As shown in Table 5, an interaction of age (B=0.76, SE=0.26, t=2.96) and anxiety symptoms (B=0.38, SE=0.36, t=1.04) on social skills (F(4,84)= 8.75, p<0.001, R^2=0.26) was found controlling for adaptive behavior. Results suggest that the effects of anxiety symptoms on social skills varies with age in boys with FXS. To probe this interaction, anxiety scores were used to create three equal groups (e.g., low, medium, and high) and graphed. As shown in Figure 2, both the low and medium groups of anxiety symptoms show a significant increase of social skills with age. However, the group with high scores of anxiety symptoms shows very little progression in social skills as they get older. When examining the prosocial subscales, a significant interaction of chronological age and anxiety (B=−0.21, SE=0.10, t=−2.16) was observed only for the self-control subscale (F(4, 84)=7.83, p<0.001, R^2=0.27). Similar to the above, anxiety scores were used to create three groups (e.g., low, medium, and high) and graphed. Children with FXS with high anxiety showed a decrease in their self-control skills as they get older. The medium and low anxiety groups show an increase in their social skills as they get older. No significant interactions were found for the cooperation, assertion, or responsibility subscales.

Discussion

Social skills are socially acceptable learned behaviors that rely on a range of cognitive and affective processes that support positive social interactions across a variety of settings (Frey, Elliot, & Gresham, 2011; Gresham & Elliot, 1990). Specific social skills that emerge and develop in early childhood include: getting along with others, listening to others, following rules, taking turns, maintaining eye contact, and controlling emotions at times of conflict (Elliot, Roach, & Beddow, 2008). Understanding social skills and the factors that predict and support effective social skills is critical to optimize academic, social, and psychological outcomes and for guiding diagnostic and treatment efforts. The developmental trajectory of social skills typically reflects the emergence and refinement of social skills with increasing age across childhood and into adolescence and adulthood (Merrell & Gimpel, 2014). Cognitive skills impact social skills with individuals diagnosed with ID typically presenting with social skill deficits (Merrell & Gimpel, 2014). However, cognitive deficits do not solely account for social skill deficits as shown in several genetic disorders including Down syndrome and Williams syndrome in which socialization was a strength above reported cognitive ability (Doyle et al., 2004; Fidler, Hepburn, & Rogers, 2006) and in 22q11.2 in

Author Manuscript Author Manuscript Author Manuscript Author Manuscript

which no relationship of cognitive ability and social skills are reported (Shashi et al., 2012). Given the importance of social skills for functional outcomes across a number of domains, the present study examined early social skills and its relationship with autism symptomology and anxiety severity within boys with FXS, a genetic disorder associated with ID and at high risk for poor social skill development. To date, this is the only study to examine specific social skills in FXS, and we do so by contrasting their profile to TD boys and by examining the relationship of adaptive behavior, anxiety and autism symptoms on social skills.

Overall, our results suggest that boys with FXS showed significant social skill deficits in global and across prosocial subdomains due to a complex set of predictors and interactions between adaptive behavior, anxiety severity, autism symptoms, and chronological age. In support of our first hypothesis, boys with FXS displayed social skill impairments across both global and subdomain levels in comparison to same-aged typically developing peers. These deficits, however, were largely accounted for by lower adaptive behavior and developmental level indicating that better social skills of young boys with FXS are associated with older age and are within developmental expectations. They are not an area of relative weakness overall. Given the etiological variability in social skills across genetic disorders with cognitive impairments indicating a lack of correspondence between developmental level and social skills (Doyle et al., 2004; Fidler, Hepburn, & Rogers, 2006; Shashi et al., 2012), our findings indicate that developmental level does play a role in the social skill development of males with FXS with lower developmental skills associated with lower social skills. However, two domains of social skills, Responsibility and Self-Control, appear to represent significant relative weaknesses as these domains appear more significantly impaired even when developmental level is considered.

To identify relationships and potential mechanistic factors, we examined the relationship of autism symptom severity and anxiety symptomology on early social skills in boys with FXS. Our results suggest that elevated autism features were, in fact, associated in a complex nuanced manner. Specifically, more severe autism symptoms were associated with lower social skills at older ages. This divergence in social skills appears to emerge around 5 years of age at which time boys with FXS who have low autism symptom severity display better social skills with older age. In contrast, those with a moderate and high degree of autism symptom severity display little to no change in social skills as they get older and start off with significantly lower social skills around the age of three. Overall, these findings suggest that boys with FXS who have relatively moderate levels of ASD display deficits in their social skills and could potentially benefit from intervention.

When examining the core prosocial subscales and autism symptom severity, only the Responsibility subscale was significantly affected by age and autism symptom severity. Similar to their overall social skills, boys with FXS responsibility skills were significantly hindered, with moderate and high levels of autism symptomology playing a role in comparison to boys with FXS and low autism symptoms . Items on the Responsibility subscale appear to focus on the child’s independence and communication skills (e.g., questioning household rules, politely refusing requests, asking for permission). Thus, given that one of the core features of ASD is impaired social communication, there is coherence regarding elevated ASD features and impaired social skills in the Responsibility domain.

Author Manuscript Author Manuscript Author Manuscript Author Manuscript

These findings suggest that ASD comorbidity contributes to social skill deficits, specifically their responsibility skills, in boys with FXS, but does not account for these deficits entirely. These results are consistent with previous literature suggesting that lower socialization skills are associated with higher autism risk and that individuals with high autism symptoms are at risk for markedly poorer outcomes compared to those with FXS and low autism symptoms, but not across all domains of social skills (Brock & Hatton, 2010; Kau et al., 2004; Kaufmann et al., 2004). Overall, social skill deficits are an area of concern for boys with FXS and even more so for boys with FXS with high autism symptom severity.

Given the association of elevated anxiety to reduced social competence in the general population (Costello et al., 2003) and the high co-morbidity of anxiety disorders in FXS (Cordeiro et al., 2011; Tonnsen et al., 2013), we examined these associations within our sample. Results indicated a somewhat complex effect on early social skills and older age, but only for those with low to mid-level anxiety severity, whereas those with a high degree of anxiety symptoms demonstrated a flat profile of social skills as they get older similar to those with high autism symptomology. Even though the overall sample had low anxiety symptoms (17% were above the clinical cutoff), effects were still found for those with high anxiety symptoms. When teasing apart the prosocial subscales, only the Self-Control subscale was affected by age and anxiety, which differed compared to those with high autism symptomology. Items on the Self-Control subscale include skills reflecting following instructions, avoiding problematic situations, controlling emotions, and exercising self-regulation. Previous research has reported that individuals with FXS have poor self-regulation skills which can increase the risk for the development of anxiety disorders or exacerbate self-control impairments in the presence of elevated anxiety symptoms (Roberts et al., 2014; Tonnsen et al., 2013). Our results are consistent with the previous literature suggesting that poor social behaviors are related to elevated anxiety severity within FXS (Boyle & Kaufmann, 2010; Hall et al., 2009; Tonnsen et al., 2013).

Limitations.

The results of the present study should be considered with caution due to limitations. First, the sample size for the TD group with adaptive behavior scores was relatively small, but still powerful enough to find effects. Next, anxiety and autism symptom ratings relied on parent or examiner report, which could be underestimated in comparison to other physiological, experimental, or observational measures of anxiety and ASD. Additionally, the present study utilized adaptive behavior as a proxy for developmental level. Finally, the analyses were limited to using a cross-sectional design.

Summary and Future Directions.

The present study aimed to expand our understanding of the FXS social behavioral phenotype by increased knowledge of early social skills and factors related to social skill competence. Our results indicate a general pattern of social skill impairments that are below chronological age expectations, however, in line with developmental expectations. Boys with FXS who have high and moderate autism symptomology displayed significantly worse social skills, especially in the area of responsibility. Furthermore, boys with FXS who presented with high anxiety showed significant deficits in the area of self-control (emotion

Author Manuscript Author Manuscript Author Manuscript Author Manuscript

control, self-regulation). Thus, these two areas might represent targets for the development of interventions for boys with FXS who display either elevated ASD or anxiety features. Furthermore, in contrast to the social profiles of other genetic disorders, social deficits in FXS appear to be related to their developmental functioning. In summary, these results point to developmental levels in FXS as playing a significant role in young boys' social skill deficits, with high levels of anxiety and autism symptomology as additive risk factors. Reduced social skills in the Responsibility domain are largely driven by elevated autism symptoms while impaired Self-Control social skills are closely aligned with more severe anxiety symptoms. Thus, discrete social skill impairments emerged in association with well-documented aspects of the FXS behavioral phenotype. Therefore, boys with FXS displayed a unique social profile distinct from those with FXS with comorbid diagnoses; however, those with comorbid ASD or anxiety symptoms also demonstrate a specific social profile that is distinct from boys with only FXS. These distinct social profiles further differentiate ASD and anxiety within FXS.

Future directions could include obtaining a larger sample across a variety of ages to examine how social skill deficits change over time to add strength to the current findings. Also, using biobehavioral markers, experimental and observational measures to obtain a more accurate representation of anxiety, autism symptoms, and social skills within FXS rather than relying on examiner and parental report is recommended. In addition, further examining how ASD and anxiety play different roles in their effects on social skills within boys with FXS is important. Future research could also examine social skill deficits in females with FXS to see if they are similar or different compared to boys with FXS. Lastly, results from this study could be utilized to design interventions for these individuals to increase their potential adult outcomes and success within their environment.

Acknowledgements:

This article is based on Debra Reisinger's thesis submitted to the University of South Carolina in partial fulfillment of the requirements for the master's degree. This study was supported by the National Institute of Child Health and Human Development (P30-HD003110-35S1), the National Institute of Mental Health (R01MH0901194-01A1), and the office of Special Education Programs, U.S. Department of Education (H023C53334 and H324C010007). The present study was presented as a poster at the International Meeting for Autism Research in Atlanta, GA (May, 2014).

References

Abbeduto L, McDuffie A, & Thurman AJ (2014). The fragile X syndrome–autism comorbidity: what do we really know?. Frontiers in genetics, 5.

Achenbach TM, Rescorla LA. Manual for the ASEBA school-age forms & profiles. Burlington: University of Vermont, Research Center for Children, Youth, & Families; 2001.

Achenbach TM. Manual for the child behavior checklist/4–18 and 1991 profile. Burlington: University of Vermont Department of Psychiatry; 1991.

Achenbach TM, Dumenci L, & Rescorla LA (2003). DSM-oriented and empirically based approaches to constructing scales from the same item pools. Journal of clinical child and adolescent psychology, 32(3), 328–340. [PubMed: 12881022]

American Psychiatric Association. (2013). Diagnostic and statistical manual of mental disorders (5th ed.). Washington, DC: Author.

Author Manuscript Author Manuscript Author Manuscript Author Manuscript

Bailey DB Jr, Hatton DD, Mesibov G, Ament N, & Skinner M (2000). Early development, temperament, and functional impairment in autism and fragile X syndrome. Journal of Autism and Developmental Disorders, 30(1), 49–59. [PubMed: 10819120]

Bailey DB Jr, Raspa M, Holiday D, Bishop E, & Olmsted M (2009). Functional skills of individuals with fragile X syndrome: A lifespan cross-sectional analysis. Journal Information, 114(4).

Bailey DB, Raspa M, Olmsted M, & Holiday DB (2008). Co-occurring conditions associated with FMR1 gene variations: Findings from a national parent survey. American journal of medical genetics part A, 146(16), 2060–2069.

Baker S, Hooper S, Skinner M, Hatton D, Schaaf J, Ornstein P, & Bailey D (2011). Working memory subsystems and task complexity in young boys with Fragile X syndrome. Journal of Intellectual Disability Research : JIDR,55(1), 19–29. [PubMed: 21121991]

Baranek GT, Roberts JE, David FJ, Sideris J, Mirrett PL, Hatton DD, & Bailey DB (2008). Developmental trajectories and correlates of sensory processing in young boys with fragile X syndrome. Physical & occupational therapy in pediatrics, 28(1), 79–98. [PubMed: 18399048]

Boyle L, & Kaufmann WE (2010, 11). The behavioral phenotype of FMR1 mutations *In* American Journal of Medical Genetics Part C: Seminars in Medical Genetics (Vol. 154, No. 4, pp. 469–476). Wiley Subscription Services, Inc., A Wiley Company.

Brock M, & Hatton D (2010). Distinguishing features of autism in boys with fragile X syndrome. Journal of Intellectual Disability Research, 54(10), 894–905. [PubMed: 20704635]

Bryson SE, Zwaigenbaum L, Brian J, Roberts W, Szatmari P, Rombough V, & McDermott C (2007). A prospective case series of high-risk infants who developed autism. Journal of autism and developmental disorders, 37(1), 12–24. [PubMed: 17211728]

Carter AS, Davis NO, Klin A, & Volkmar FR (2005). Social development in autism Handbook of Autism and Pervasive Developmental Disorders, Volume 1, Third Edition, 312–334.

Chang YC, Quan J, & Wood JJ (2012). Effects of anxiety disorder severity on social functioning in children with autism spectrum disorders. Journal of Developmental and Physical Disabilities, 24(3), 235–245.

Chlebowski C, Green JA, Barton ML, & Fein D (2010). Using the childhood autism rating scale to diagnose autism spectrum disorders. Journal of autism and developmental disorders, 40(7), 787–799. [PubMed: 20054630]

Coplan RJ, Prakash K, O'Neil K, & Armer M (2004). Do you" want" to play? Distinguishing between conflicted shyness and social disinterest in early childhood. Developmental psychology, 40(2), 244. [PubMed: 14979764]

Cordeiro L, Ballinger E, Hagerman R, & Hessl D (2011). Clinical assessment of DSM-IV anxiety disorders in fragile X syndrome: prevalence and characterization. Journal of neurodevelopmental disorders, 3(1), 57–67. [PubMed: 21475730]

Costello EJ, Mustillo S, Erkanli A, Keeler G, & Angold A (2003). Prevalence and development of psychiatric disorders in childhood and adolescence. Archives of general psychiatry, 60(8), 837–844. [PubMed: 12912767]

De Bildt A, Serra M, Luteijn E, Kraijer D, Sytema S, & Minderaa R (2005). Social skills in children with intellectual disabilities with and without autism. Journal of Intellectual Disability Research, 49(5), 317–328. [PubMed: 15817049]

Doyle TF, Bellugi U, Korenberg JR, & Graham J (2004). “Everybody in the world is my friend” hypersociability in young children with Williams syndrome. American Journal of Medical Genetics Part A, 124(3), 263–273.

Elliott SN, Roach AT, & Beddow P (2008). Best practices in preschool social skills training In Thomas A & Grimes J (Eds.), Best practices in school psychology IV (pp. 1531–1546). Washington, DC: National Association of School Psychology.

Feinstein C, & Singh S (2007). Social phenotypes in neurogenetic syndromes. Child and Adolescent Psychiatric Clinics of North America, 16(3), 631–647. [PubMed: 17562583]

Fidler D, Hepburn S, & Rogers S (2006). Early learning and adaptive behaviour in toddlers with Down syndrome: evidence for an emerging behavioural phenotype?. Down Syndrome Research and Practice, 9(3), 37–44.

Author Manuscript Author Manuscript Author Manuscript Author Manuscript

Frey JR, Elliott SN, & Gresham FM (2011). Preschoolers' Social Skills: Advances in Assessment for Intervention Using Social Behavior Ratings. School Mental Health, 3(4), 179–190.

Garber KB, Visootsak J, & Warren ST (2008). Fragile X syndrome. European Journal of Human Genetics, 16(6), 666–672. [PubMed: 18398441]

Gillis JM, Callahan EH, & Romanczyk RG (2011). Assessment of social behavior in children with autism: The development of the Behavioral Assessment of Social Interactions in Young Children. Research in Autism Spectrum Disorders, 5(1), 351–360.

Gresham FM (2016). Social skills assessment and intervention for children and youth. Cambridge Journal of Education, 46(3), 319–332.

Gresham FM, & Elliott SN (1990). Social Skills Rating System: Preschool, Elementary Level. American Guidance Service.

Hagerman RJ & Hagerman PJ (2002). Fragile X syndrome Outcomes in neurodevelopmental and genetic disorders. (198–219). Cambridge: Cambridge University Press.

Hall SS, Lightbody AA, Hirt M, Rezvani A, & Reiss AL (2010). Autism in fragile X syndrome: a category mistake?. Journal of the American Academy of Child & Adolescent Psychiatry, 49(9), 921–933. [PubMed: 20732628]

Hall SS, Lightbody AA, Huffman LC, Lazzeroni LC, & Reiss AL (2009). Physiological correlates of social avoidance behavior in children and adolescents with fragile X syndrome. Journal of the American Academy of Child & Adolescent Psychiatry, 48(3), 320–329. [PubMed: 19182690]

Harrison PL (1987). Research with adaptive behavior scales. The Journal of Special Education, 21(1), 37–68.

Hatton DD, Sideris J, Skinner M, Mankowski J, Bailey DB, Roberts J, & Mirrett P (2006). Autistic behavior in children with fragile X syndrome: prevalence, stability, and the impact of FMRP. American journal of medical genetics Part A, 140(17), 1804–1813.

Heilman KJ, Harden ER, Zageris DM, Berry-Kravis E, Porges SW. Autonomic regulation in fragile X syndrome. Developmental Psychobiology. 2011;53(8):785–795. doi: 10.1002/dev.20551. [PubMed: 21547900]

Hooper SR, Hatton D, Sideris J, Sullivan K, Hammer J, Schaaf J, … & Bailey DB Jr (2008). Executive functions in young males with fragile X syndrome in comparison to mental age-matched controls: Baseline findings from a longitudinal study. Neuropsychology, 22(1), 36. [PubMed: 18211154]

Kau AS, Tierney E, Bukelis I, Stump MH, Kates WR, Trescher WH, & Kaufmann WE (2004). Social behavior profile in young males with fragile X syndrome: Characteristics and specificity. American Journal of Medical Genetics Part A, 126(1), 9–17.

Kaufmann WE, Cortell R, Kau AS, Bukelis I, Tierney E, Gray RM, … & Stanard P (2004). Autism spectrum disorder in fragile X syndrome: communication, social interaction, and specific behaviors. American Journal of Medical Genetics Part A, 129(3), 225–234.

Klaiman C, Quintin EM, Jo B, Lightbody AA, Hazlett HC, Piven J, … & Reiss AL (2014). Longitudinal profiles of adaptive behavior in fragile X syndrome. Pediatrics, 134(2), 315–324. [PubMed: 25070318]

Klusek J, Hunt AW, Mirrett PL, Hatton DD, Hooper SR, Roberts JE, & Bailey DB (2015). Reading and Phonological Skills in Boys with Fragile X Syndrome. Journal of Autism and Developmental Disorders, 45(6), 1699–1711. [PubMed: 25448919]

Klusek J, Martin GE, & Losh M (2014). Consistency between research and clinical diagnoses of autism among boys and girls with fragile X syndrome. Journal of Intellectual Disability Research, 58(10), 940–952. [PubMed: 24528851]

Lord C, Rutter M, DiLavore PC, Risi S, Gotham K, & Bishop S (2012). Autism Diagnostic Observation Schedule, 2nd Edition (ADOS-2). Torrance: Western Psychological Services.

Magyar CI, & Pandolfi V (2007). Factor structure evaluation of the childhood autism rating scale. Journal of Autism and Developmental Disorders, 37(9), 1787–1794. [PubMed: 17437070]

Matson JL, & Wilkins J (2009). Psychometric testing methods for children's social skills. Research in Developmental Disabilities, 30(2), 249–274. [PubMed: 18486441]

Merrell KW, & Gimpel G (2014). Social skills of children and adolescents: Conceptualization, assessment, treatment. Psychology Press.

Author Manuscript Author Manuscript Author Manuscript Author Manuscript

Mervis CB, & Robinson BF (2003). Methodological issues in cross-group comparisons of language and/or cognitive development In Levy Y & Schaeffer J (Eds.), Language competence across populations: Toward a definition of specific language impairment (pp. 233–258). Mahwah, NJ: Lawrence Erlbaum.

Moskowitz LJ & Jones EA, (2015). Uncovering the evidence for behavioral interventions with individuals with fragile X syndrome: A systematic review. Research in Developmental Disabilities, 38, 223–241. [PubMed: 25575286]

Mullen EM (1995). Mullen scales of early learning (pp. 58–64). Circle Pines, MN: AGS.

Nakamura BJ, Ebesutani C, Bernstein A, & Chorpita BF (2009). A psychometric analysis of the child behavior checklist DSM-oriented scales. Journal of Psychopathology and Behavioral Assessment, 31(3), 178–189.

Rellini E, Tortolani D, Trillo S, Carbone S, & Montecchi F (2004). Childhood Autism Rating Scale (CARS) and Autism Behavior Checklist (ABC) correspondence and conflicts with DSM-IV criteria in diagnosis of autism. Journal of autism and developmental disorders, 34(6), 703–708. [PubMed: 15679189]

Roberts JE, Clarke MA, Alcorn K, Carter JC, Long AC, & Kaufmann WE (2009). Autistic behavior in boys with fragile X syndrome: social approach and HPA-axis dysfunction. Journal of neurodevelopmental disorders, 1(4), 283–291. [PubMed: 21547720]

Roberts JE, Tonnsen BL, Robinson M, McQuillin SD, & Hatton DD (2014). emperament factor structure in fragile X syndrome: The Children's Behavior Questionnaire. Research in developmental disabilities,35(2), 563–571. [PubMed: 24380785]

Roberts JE, Weisenfeld LAH, Hatton DD, Heath M, & Kaufmann WE (2007). Social approach and autistic behavior in children with fragile X syndrome. Journal of autism and developmental disorders, 37(9), 1748–1760. [PubMed: 17180715]

Roid GH, & Miller LJ (1997). Leiter-R Performance Scale, Revised. Wood Dale, IL: Stoelting Co.

Rosenbaum P, Saiga S, Szatmari P, & Hoult L (1995). Vineland adaptive behavior scales as a summary of functional outcome of extremely low-birthweight children. Developmental Medicine & Child Neurology, 37(7), 577–586. [PubMed: 7542210]

Schalock RL, Borthwick-Duffy S, Bradley V,Buntinx WHE, Coulter DL, Craig EM,… Yeager MH (2010). Intellectual disability: Definition, classification, and system of supports (11th ed.). Washington, DC: American Association on Intellectual and Developmental Disabilities. 444 North Capitol Street NW Suite 846, Washington, DC 20001.

Schneider BH (2009). An observational study of the interactions of socially withdrawn/anxious early adolescents and their friends. Journal of Child Psychology and Psychiatry, 50(7), 799–806. [PubMed: 19298472]

Schopler E, Reichler RJ, & Renner BR (1988). The childhood autism rating scale (CARS). Los Angeles: Western Psychological Services.

Shashi V, Veerapandiyan A, Schoch K, Kwapil T, Keshavan M, Ip E, & Hooper S (2012). Social skills and associated psychopathology in children with chromosome 22q11. 2 deletion syndrome: implications for interventions. Journal of Intellectual Disability Research, 56(9), 865–878. [PubMed: 21883601]

Sparrow SS, Balla DA, & Cicchetti DV (1984). Vineland adaptive behavior scales: Interview edition, survey form manual. Circle Pines, MN: American Guidance Service.

Spinrad TL, Eisenberg N, Harris E, Hanish L, Fabes RA, Kupanoff K, … & Holmes J (2004). The relation of children's everyday nonsocial peer play behavior to their emotionality, regulation, and social functioning. Developmental Psychology, 40(1), 67. [PubMed: 14700465]

Thorell LB, Bohlin G, & Rydell AM (2004). Two types of inhibitory control: Predictive relations to social functioning. International Journal of Behavioral Development, 28(3), 193–203.

Thurman AJ, McDuffie A, Hagerman R, & Abbeduto L (2014). Psychiatric symptoms in boys with fragile X syndrome: a comparison with nonsyndromic autism spectrum disorder. Research in developmental disabilities, 35(5), 1072–1086. [PubMed: 24629733]

Tonnsen BL, Malone PS, Hatton DD, & Roberts JE (2013). Early Negative Affect Predicts Anxiety, not Autism, in Preschool Boys with Fragile X Syndrome. Journal of abnormal child psychology, 41(2), 267–280. [PubMed: 23011214]

Author Manuscript Author Manuscript Author Manuscript Author Manuscript

Tonnsen BL, Shinkareva SV, Deal SC, Hatton DD, & Roberts JE (2013). Biobehavioral Indicators of Social Fear in Young Children With Fragile X Syndrome. American Journal on Intellectual and Developmental Disabilities, 118(6), 447–459. [PubMed: 24432858]

Tranfaglia MR (2011). The psychiatric presentation of fragile x: evolution of the diagnosis and treatment of the psychiatric comorbidities of fragile X syndrome. Developmental neuroscience, 33(5), 337–348. [PubMed: 21893938]

Author Manuscript Author Manuscript Author Manuscript Author Manuscript

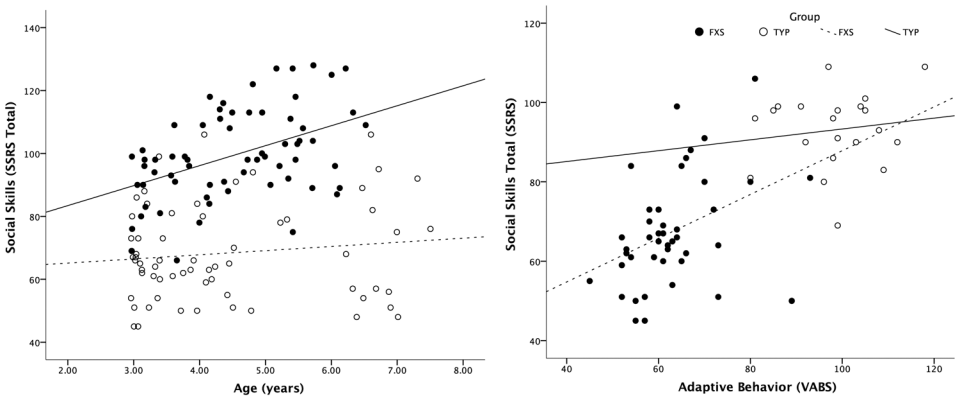

Figure 1.

In the left pane, social skills are graphed against chronological age for boys with FXS and TD controls. In the right pane, social skills are graphed against adaptive behavior as measured by the VABS for a subsample of boys with FXS and TD controls.

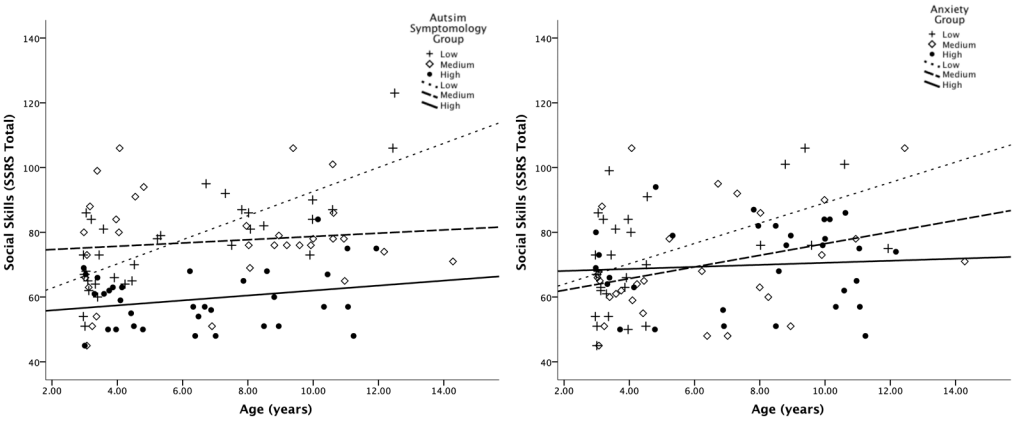

Figure 2.

In the left pane, varying levels of autism symptomology are graphed on social skills and chronological age. In the right pane, varying levels of anxiety severity are graphed on social skills and chronological age.

Author Manuscript Author Manuscript Author Manuscript Author Manuscript

Author Manuscript Author Manuscript Author Manuscript Author Manuscript

Am J Intellect Dev Disabil. Author manuscript; available in PMC 2020 August 31.

Table 1

Descriptive Statistics of FXS and TD Variables

Variable	FXS			TD			FXS Subsample			TD Subsample		
	n	**Mean**	**SD**	***n***	**Mean**	**SD**	***n***	**Mean**	**SD**	***n***	**Mean**	**SD**
Chronological Age (years)	64	4.38	1.44	65	4.53	1.01	44	3.55	0.52	21	3.47	0.38
SSRS Total standard score	64	68.28	15.30	65	99.51	14.36	44	67.30	13.76	21	93.10	9.61
SSRS Cooperation raw score*	64	6.84	4.01	65	12.15	2.80	44	6.57	3.91	21	11.48	2.18
SSRS Assertion raw score*	64	8.84	3.73	65	14.60	2.97	44	8.84	3.19	21	13.24	2.66
SSRS Responsibility raw score*	64	2.63	3.19	65	10.14	3.66	44	1.86	2.55	21	7.90	2.81
SSRS Self-control raw score*	64	7.89	3.86	65	13.12	2.89	44	7.89	3.67	21	12.52	2.23
VABS Adaptive Behavior Composite							44	62.73	9.75	21	98.33	9.98
MSEL Early Learning Composite							45	51.97	5.29	21	106.38	13.05

Note. FXS is fragile X syndrome; TD is typically developing controls; VABS is the Vineland Adaptive Behavior Scales; MSEL is the Mullen Scale of Early Learning; SSRS is the Social Skills Rating System

*standard scores are not available

Author Manuscript

Author Manuscript

Author Manuscript

Author Manuscript

Table 2

Descriptive Statistics for Within FXS Analyses

Variable	FXS		
	n	Mean	SD
Chronological Age (years)	102	6.59	3.12
VABS Adaptive Behavior Composite	102	51.50	14.53
SSRS Total standard score	102	71.06	15.72
SSRS Cooperation raw score	102	7.83	4.00
SSRS Assertion raw score	102	8.91	3.65
SSRS Responsibility raw score	102	4.18	3.91
SSRS Self-control raw score	102	8.55	3.83
CARS Total score	98	28.19	6.36
CBCL DSM Anxiety T-score	89	57.56	8.84

Note. FXS is fragile X syndrome; VABS is the Vineland Adaptive Behavior Scales; SSRS is the social skills rating system; CARS is the Childhood Autism Rating Scale; CBCL is the Child Behavior Checklist

Author Manuscript

Author Manuscript

Author Manuscript

Author Manuscript

Table 3

Summary of Regression Analyses for Age, Group, and Adaptive Behavior to Predicting Total Social Skills

Variables	*df*	*B*	SE_B	*t*	*p*
Intercept	125	2.04	0.17	11.54	<0.001*
Chronological Age	125	0.08	0.07	1.07	0.29
Group	125	−1.40	0.11	−12.44	<0.001*
Chronological Age*Group	125	0.30	0.13	2.36	0.02*
Including Adaptive Behavior					
Intercept	65	−0.23	0.14	−1.63	0.10
Adaptive Behavior	65	0.47	0.18	2.69	0.09
Chronological Age	65	0.15	0.12	1.26	0.90
Group	65	0.70	0.37	1.89	0.06
Chronological Age*Group	65	0.27	0.27	1.01	0.31
Including Developmental Level					
Intercept	66	1.23	1.82	0.67	0.50
Developmental Level	66	0.01	0.01	0.62	0.54
Chronological Age	66	0.11	0.22	0.50	0.62
Group	66	−1.12	0.74	−1.52	0.14
Chronological Age*Group	66	0.30	0.51	0.58	0.57

Author Manuscript

Author Manuscript

Author Manuscript

Author Manuscript

Table 4

Summary of Regression Analyses for Autism Symptomology on Predicting Social Skills in Boys with FXS

Variables	***df***	***B***	SE_B	***t***	***p***
Intercept	93	38.01	18.09	2.10	0.04*
Adaptive Behavior	93	0.28	0.14	2.07	0.04*
Chronological Age	93	0.56	0.17	3.31	0.001*
Autism Symptomology	93	0.11	0.53	0.21	0.83
Autism Symptomology*Age	93	−0.01	0.01	−2.08	0.04*

Author Manuscript

Author Manuscript

Author Manuscript

Author Manuscript

Table 5

Summary of Regression Analyses for Anxiety on Predicting Social Skills in Boys with FXS

Variables	*df*	*B*	SE_B	*t*	*p*
Intercept	84	−2.30	23.75	−0.10	0.92
Adaptive Behavior	84	0.60	0.13	4.66*	<0.001*
Chronological Age	84	0.76	0.26	2.96*	0.004*
Anxiety	84	0.38	0.36	1.04	0.30
Chronological Age*Anxiety	84	−0.001	0.01	−2.00*	0.049*