

2015

Anxiety and Autism Symptomology On Social Skills in Young Boys With Fragile X Syndrome

Debra L. Reisinger
University of South Carolina

Follow this and additional works at: <http://scholarcommons.sc.edu/etd>



Part of the [School Psychology Commons](#)

Recommended Citation

Reisinger, D. L. (2015). *Anxiety and Autism Symptomology On Social Skills in Young Boys With Fragile X Syndrome*. (Master's thesis). Retrieved from <http://scholarcommons.sc.edu/etd/3682>

This Open Access Thesis is brought to you for free and open access by Scholar Commons. It has been accepted for inclusion in Theses and Dissertations by an authorized administrator of Scholar Commons. For more information, please contact SCHOLARC@mailbox.sc.edu.

ANXIETY AND AUTISM SYMPTOMOLOGY ON SOCIAL SKILLS IN YOUNG BOYS WITH
FRAGILE X SYNDROME

by

Debra L. Reisinger

Bachelor of Science
College of Mount St. Joseph, 2012

Submitted in Partial Fulfillment of the Requirements

For the Degree of Master of Arts in

School Psychology

College of Arts and Sciences

University of South Carolina

2015

Accepted By:

Jane Roberts, Director of Thesis

Richard Nagle, Reader

Lacy Ford, Senior Vice Provost and Dean of Graduate Students

©Copyright by Debra L. Reisinger, 2015
All Rights Reserved.

ABSTRACT

Children with fragile X syndrome (FXS) are at high risk for developing several comorbid disorders, including autism spectrum disorder (ASD) and anxiety, that can substantially impair their social skills. However, to date, there are no studies that examine complex social skills within boys with FXS and the effects of emerging autism and anxiety symptoms on the development of their social skills. 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 study consisted of boys with FXS (ages 36 – 171 months) compared to chronologically aged matched typically developing peers. A combination of MANCOVA's and regression analyses revealed that developmental level in FXS is a main contributor to social skill deficits with high levels of anxiety and autism symptoms as additive risk factors independently, particularly in the areas of responsibility and self-control.

TABLE OF CONTENTS

ABSTRACT	iii
LIST OF TABLES	v
CHAPTER 1: INTRODUCTION	1
1.1 FRAGILE X SYNDROME	2
1.2 AUTISM SPECTRUM DISORDERS IN FXS.....	3
1.3 ANXIETY IN FXS	4
1.4 THE CURRENT STUDY	5
CHAPTER 2: METHOD	7
2.1 PARTICIPANTS	7
2.2 MEASURES	8
2.3 PROCEDURES.....	12
2.4 DATA ANALYSIS	12
CHAPTER 3: RESULTS	15
3.1 DEVELOPMENT OF OVERALL SOCIAL SKILLS IN FXS AND TD BOYS	15
3.2 AUTISM SYMPTOMOLOGY IN FXS ON SOCIAL SKILLS.....	17
3.3 ANXIETY SYMPTOMS IN FXS ON SOCIAL SKILLS	19
CHAPTER 4: DISCUSSION	23
4.1 LIMITATIONS	26
4.2 CONCLUSIONS AND FUTURE DIRECTIONS	26
REFERENCES	28

LIST OF TABLES

Table 2.1 Descriptive Statistics of FXS and TD Variables	13
Table 2.2 Descriptive Statistics of Supplemental FXS and TD Variables	13
Table 2.3 Descriptive Statistics for Within FXS Analyses	14
Table 3.1 Summary of Regression Analyses for Age, Group, and Adaptive Behavior to Predicting Total Social Skills	21
Table 3.2 Summary of Regression Analyses for Autism Symptomology on Predicting Social Skills in Boys with FXS	22
Table 3.3 Summary of Regression Analyses for Anxiety on Predicting Social Skills in Boys with FXS.....	22

CHAPTER 1

INTRODUCTION

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). Social skills 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, necessary social skills may include: listening to others, following rules, taking turns, asking for help, cooperating with peers, and controlling emotions at times of conflict. However, social skills vary and evolve across development with age significantly affecting the acquisition of specific social skills across early childhood, middle childhood, and adolescence (Merrell & Gimpel, 2014). In individuals with intellectual disabilities (ID) and autism spectrum disorders (ASD), limitations in social skills are a central characteristic, playing a major role in their overall adaptive behavior (De Bildt et al, 2005).

According to Hartup (1988), “the single best childhood predictor of adult adaptation is not school grades, and not classroom behavior, but rather, the adequacy with which the child gets along with other children” (p. 1). 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). Social skill deficits are not limited to one specific

diagnosis or condition. Difficulties have also been shown in children with ID, 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 ID across all ages tend to display significant deficits or limitations in their social skills (Merrell & Gimpel, 2014), whereas individuals with Down syndrome show strengths in their socialization skills (Fidler, Hepburn, & Rogers, 2006). In individuals with high anxiety, both clinical and non-clinical populations 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 ASD, social skill deficits have been identified as core underlying feature regardless of cognitive or language abilities (Carter, Davis, Klin, & Volkmar, 2005).

1.1 Fragile X Syndrome

Fragile X syndrome (FXS) is the leading heritable cause of ID, occurring in as many as 1 in 4,000 males, and is associated with a variety of co-occurring conditions, including autism and anxiety (Hall, Lightbody, Huffman, Lazzeroni, & Reiss, 2009; Kau et al., 2004). FXS is caused by a mutation on the trinucleotide (CCG) repeat expansion on the fragile X mental retardation 1 (FMR1) gene, which is located on the long arm of the X chromosome (Hagerman & Hagerman, 2002; Hall et al., 2009). When FMRP is reduced there is an increase in manifestations of the syndrome, including physical, cognitive, and behavioral deficits (Hagerman & Hagerman, 2002; Hall et al., 2009). FXS is characterized by mild to severe ID and other deficits including anxiety, social deficits,

and abnormalities in communication, gaze aversion, inattention, impulsivity, aggression and hyperactivity (Cordeiro et al., 2011). One of the most frequent and disabling behavioral abnormalities is social avoidance, or social withdraw in young boys with FXS (Kau et al, 2004; Kaufmann et al, 2004; Roberts, Weisenfield, Hatton, Heath, Kaufmann, 2007). In examining socialization skills, previous literature has identified specific deficits in overall socialization through the use of adaptive behavior measures within FXS (Bailey, Raspa, Holiday, Bishop, & Olmsted, 2008; Klaiman et al., 2014). Individuals with FXS have been shown to be interested in social interactions but, shyness and social anxiety may interfere with proper social behaviors resulting in social withdrawal. To date, work has primarily focused on examining these social approach-withdraw behaviors and broad socialization in FXS rather than focus on important social skills that contribute to their social success and are central for later academic and social outcomes.

1.2 Autism Spectrum Disorders in FXS.

Autism is a neurodevelopmental disorder that is represented on a continuum as autism spectrum disorders (ASD). ASD is characterized by social abnormalities, communicative abnormalities, stereotyped and repetitive behaviors, and restricted interests (American Psychiatric Association, 2013). ASD is one of the most severe and most recognized behavioral abnormalities occurring in males with FXS, making them at higher risk for ASD compared to typically developing individuals (Kau et al., 2004; Kaufmann et al., 2004). Approximately 30-60% of children with FXS meet diagnostic criteria for ASD (Talisa, Boyle, Crafa, & Kaufmann, 2014). Approximately 90% of males with FXS display at least one autistic behavior (Brock & Hatton, 2010; Roberts et

al., 2007). Several features of the ASD behavioral phenotype are typically displayed in individuals with FXS (Garber, Visootsak, & Warren, 2008).

It has been argued that boys with FXS have been shown to be socially shy, playful, and conversational, in contrast to boys with autism being shown to be socially oblivious, not playful, and exhibiting little in the way of conversational skills (Bailey et al., 2000). 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 feel they are two distinct conditions because they have been shown to differ fundamentally in many ways, including their social and communication skills (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 (Brock & Hatton, 2010). With ASD and FXS overlapping in the area of social behaviors, it is unclear whether such deficits within overall social skills and subdomains (e.g. cooperation, assertion, responsibility, and self-control) are characteristic of autism symptomology within the context of FXS rather than a characteristic of FXS itself.

1.3 Anxiety in FXS.

Anxiety disorders in the general population are one of the most common psychiatric disorders, occurring in 2.4 – 10.7% of children (Costello, Mustillo, Erkanli, Keeler, & Angold, 2003). FXS has been shown to be highly comorbid with anxiety disorders with anxiety being cited as one of the most frequent and impairing conditions associated with FXS, with approximately 70-83% of individuals with FXS being diagnosed (Cordeiro et al., 2011; Kaufmann et al., 2004; Tonnsen et al., 2013). It has been argued that anxiety

symptoms within FXS are a driving cause for many behavioral problems including: poor eye contact, gaze aversion, excessive shyness, hand flapping, hand biting, aggression, and autistic symptoms (Boyle & Kaufmann, 2010; Tranfaglia, 2011).

Anxiety is also highly comorbid in individuals with ASD with ranges of those having at least one comorbid anxiety disorder being between 29% up to 84% of the ASD population (Chang, Quan & Wood, 2012). Identifying anxiety disorders in those with ID, including those with FXS and ASD, can be rather difficult due to a potential lacking of verbal expression and personal insight (Cordeiro et al., 2011). Current literature has shown that poor social behaviors are related to social anxiety within ASD and FXS (Hall et al., 2009). The present study aims to develop a better understanding of the potential effects of anxiety symptoms on social skills within young boys with FXS to add the current literature on the social behavioral phenotype of FXS.

1.4 The Current Study

As previously mentioned, to the author's knowledge, no research has looked at these specific social skill deficits within children with FXS in addition to their relationship with age, adaptive behavior, autism and anxiety symptoms. A review study investigating intervention studies completed in individuals with FXS suggested that minimal interventions have been examined looking specifically at increasing social skills in FXS (Moskowitz & Jones, 2015). Developing a better understanding of social skill development in FXS can help guide diagnostic, treatment and intervention efforts within this population. 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 are as follows:

1. What is the development of social skills across age in young boys with FXS compared to chronological age matched TD boys controlling for adaptive behavior?
 - a. It is hypothesized that boys with FXS will display significant deficits in social skills compared to TD boys across chronological age.
 - b. It is hypothesized that controlling for adaptive behavior will significantly impact social skill levels within the FXS group compared to the TD group, potentially minimizing the differences between groups.
2. How do features of autism and anxiety in boys with FXS affect their social skill development controlling for adaptive behavior?
 - a. It is hypothesized that elevated features of autism (at both a categorical and continuous level) will be associated with decreased social skill competency.
 - b. It is hypothesized that elevated features of anxiety (at both a categorical and continuous level) will be associated with decreased social skill competency.

CHAPTER 2

METHOD

2.1 Participants

Data were drawn from a series of longitudinal studies through the Carolina Fragile X Project at the University of North Carolina. Participants were recruited across the United States through FXS parent list serves, FXS parent support groups, 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 conditions (e.g., hearing or vision impairment) that may have impacted the results of this study. The typically developing boys had no reported or suspected developmental concerns and fell within the average range of functioning on developmental measures completed through the study.

To answer the first research question comparing social skills between FXS and TD boys, 63 males with FXS (ages 36-90 months, mean age = 52.86 months) and 70 TD males (ages 36-90 months, mean age = 56.53 months) from the longitudinal studies were included. Groups were matched on chronological age with no difference in chronological age ($t = -1.35, p > 0.05$). First, we compared the TD and FXS boys matched on chronological age for the full sample independent of adaptive behavior or mental age. Next, we compared a subset of TD ($N = 21$) and FXS ($N = 48$) boys matched on chronological age ($t = -1.12, p > 0.05$) while controlling for adaptive behavior to represent

developmental level. Due to the potential confound with adaptive behavior and social skills being highly correlated, with this same subset, we completed supplemental analyses controlling for mental age to verify if the results found with adaptive behavior remain.

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 36-171 months, mean age = 79.06 months) who had data for the dependent variable. Although several participants had multiple data points available, we selected each participant's earliest data point and matched all scores from the same data point for both sets of data in an effort to identify emerging features of social skills. Table 2.1, Table 2.2, and Table 2.3 summarize the descriptive statistics.

2.2 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 with parent, teacher, and student (self) versions. 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). Additionally, the SSRS has shown strong

internal consistencies with high alpha coefficients for the parent version of 0.89 (S. Van der Oord et al., 2005). The current study focuses on the parent form for both the preschool and elementary forms. For the purpose of this study, total social skills standard scores were used to minimize differences between ages.

Currently, there are four core subdomains of social skills that are essential for effective social functioning for children: 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 nonconflict situations such as taking turns and compromising. For these analyses, raw scores were used for the prosocial subscales due to the SSRS not providing norms to convert the raw scores to standard scores. 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. 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 Mullen Scales of Early Learning (MSEL; Mullen, 1995) is a standardized assessment of cognitive and motor abilities yielding a norm-referenced full-scale IQ in infants and young children. Four (visual reception, receptive language, expressive language, and fine motor) out of five of the domains on the MSEL comprise the Early Learning Composite (ELC). The ELC 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 studies 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 studies data, the total score was utilized and based on this score 32% of the sample met criteria for mild to severe autism symptoms. 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 internal reliability ranging from 0.73-0.90 (Magyar & Pandolfi, 2007). Several studies have attested to the CARS validity and its ability to discriminate between autism and non-autism samples (Magyar & Pandolfi, 2007).

2.3 Procedures

Participants were seen over various time points within the lab and in their home where the participant's parent completed the SSRS and CBCL. A trained examiner administered the VABS to the participant's parent. Trained examiners completed the CARS upon the completion of each 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 (n=49). Some of the FXS participants did not have CARS or CBCL data due to data collection errors (n=4; n=13).

2.4 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 make sure all of the measures are standardized on the same scale. Regression analyses and a MANOVA were used to analyze the development of social skills across ages, examining chronological age and controlling for adaptive behavior, between FXS and TD boys. Next, regression analyses and a MANOVA were completed to examine the relationship of autism symptomology and chronological age in predicting social skills in boys with FXS while controlling for adaptive behavior. Finally, regression analyses and a MANOVA were utilized to examine the relationship of anxiety severity and chronological age on social skills in boys with FXS while controlling for adaptive behavior.

Table 2.1 Descriptive Statistics of FXS and TD Variables

Variable	FXS			TD		
	<i>n</i>	Mean	SD	<i>n</i>	Mean	SD
Chronological Age (months)	63	52.86	17.32	70	56.53	14.07
SSRS Total standard score	63	68.51	15.31	70	100.87	15.25
SSRS Cooperation raw score*	63	6.89	4.02	70	12.39	2.90
SSRS Assertion raw score*	63	8.90	3.73	70	14.77	3.08
SSRS Responsibility raw score*	63	2.67	3.19	70	10.47	3.79
SSRS Self-control raw score*	63	7.95	3.86	70	13.36	2.97

Note. FXS is fragile X syndrome; TD is typically developing controls; SSRS is the Social Skills Rating System; *standard scores are not available

Table 2.2 Descriptive Statistics of Supplemental FXS and TD Variables

Variable	FXS			TD		
	<i>n</i>	Mean	SD	<i>n</i>	Mean	SD
Chronological Age (months)	48	43.54	7.44	21	41.58	4.61
VABS Adaptive Behavior Composite	48	61.75	10.01	21	98.33	9.98
MSEL Early Learning Composite	48	52.04	5.32	21	106.38	13.05
SSRS Total standard score	48	67.46	14.19	21	93.10	9.61
SSRS Cooperation raw score*	48	6.52	3.85	21	11.48	2.18
SSRS Assertion raw score*	48	8.77	3.47	21	13.24	2.66
SSRS Responsibility raw score*	48	1.98	2.65	21	7.90	2.81
SSRS Self-control raw score*	48	7.96	3.69	21	12.52	2.23

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

Table 2.3 Descriptive Statistics for Within FXS Analyses

Variable	FXS		
	<i>n</i>	Mean	SD
Chronological Age (months)	102	79.06	37.42
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

CHAPTER 3

RESULTS

3.1 Development of Overall Social Skills in FXS and TD Boys

Chronological Age. A regression model was used to analyze the effects of chronological age between groups (i.e., TD and FXS) on total social skills. A significant interaction of chronological age and group ($B=0.16$, $SE=0.06$, $t=2.87$) on total social skills ($F(3, 130)=66.43$, $p<0.001$, $R^2=0.60$; Table 3.1) was found. As shown in Figure 3.1, we see that both TD and FXS boys show an incline in total social skills as they age however, boys with FXS are progressing at a lower rate compared to their chronologically same-aged TD peers. When examining the prosocial subscales, a significant interaction of chronological age and group was observed on the assertion subscale ($B=-0.33$, $SE=0.13$, $t=-2.59$) and responsibility subscale ($B=-0.33$, $SE=0.10$, $t=-3.41$). Conversely, no significant interaction of chronological age and group was observed on the cooperation ($B=-0.22$, $SE=0.13$, $t=-1.65$) and self-control ($B=-0.17$, $SE=0.14$, $t=-1.25$) subscales.

To further understand these social skill deficits within FXS as compared to TD boys, follow up analyses were conducted using the prosocial subscales of the SSRS. A MANCOVA indicted a significant effect of group on all the prosocial subscales: Cooperation ($F(2,131)= 82.07$, $p<0.001$) Assertion ($F(2,131)= 97.70$, $p<0.001$), Responsibility ($F(2,131)= 198.38$, $p<0.001$), and Self-Control ($F(2,131)= 81.18$, $p<0.001$). Bonferroni-corrected pairwise comparisons showed that the boys with FXS

had significantly less competent social skills than TD boys across all the prosocial subscales ($ps < 0.001$).

Adaptive Behavior. A regression model was utilized to examine the effects of chronological age and group on total social skills while controlling for adaptive behavior to see if the above found effects remain. No significant interaction or main effects were found while controlling for adaptive behavior ($F(4,64)=17.30, p < 0.001, R^2=0.49$; Table 3.1). Similar results were found when examining the prosocial scales individually. When controlling for adaptive behavior, no significant interactions or main effects were observed for the cooperation, assertion, self-control, and responsibility subscales. This suggests that adaptive behavior, not chronological age, accounts for the group differences change in social skills as they age in boys with FXS compared to TD boys (Figure 3.1).

Although there were no effects found within the overall model for the SSRS total score, follow up analyses were conducted to see if there were any group differences found within the prosocial subscales. A MANCOVA indicted a significant effect of group on two of the four prosocial subscales: Responsibility ($F(3,65)= 4.79, p=0.03$) and Self-Control ($F(3,65)= 5.08, p=0.03$). Bonferroni-corrected pairwise comparisons showed that the boys with FXS had significantly less competent social skills than TD boys in the areas of Responsibility and Self-Control when controlling for adaptive behavior ($ps=0.03$).

Developmental Level. To verify the above results with adaptive behavior remain, a supplemental regression model was used to examine the effects of chronological age and group on total social skills while controlling for developmental level. No significant interaction or main effects were found while controlling for developmental level

($F(4,64)=14.72, p<0.001, R^2=0.45$; Table 3.1). Similar results were found when examining the prosocial scales individually. When controlling for developmental level, no significant interactions or main effects were observed for the cooperation, assertion, self-control, and responsibility subscales. These results support our results found with adaptive behavior, suggesting that adaptive behavior or developmental level, not chronological age, accounts for the group differences change in social skills as they age in boys with FXS compared to TD boys.

Although there were no effects found within the overall model for the SSRS total score, follow up analyses were conducted to see if there were any group differences found within the prosocial subscales while controlling for developmental level. A MANCOVA indicted a significant effect of group only in the area of Responsibility ($F(3,65)= 4.65, p=0.04$). Similar to the results found with adaptive behavior, Bonferroni-corrected pairwise comparisons showed that the boys with FXS had significantly less competent social skills than TD boys in the area of Responsibility when controlling developmental level ($p=0.04$). Conversely, the area of Self-control is not significantly different when controlling for developmental level.

3.2 Autism Symptomology in FXS on Social Skills

Autism as a continuous score. Regression analyses were used to analyze the hypothesis that increased autism symptomology is associated with decreased social skills in boys with FXS across age. As shown in Table 3.2, 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 controlling for adaptive behavior. Results suggest that autism symptomology on

social skills varies with age in boys with FXS. When examining the effect of autism symptomology and chronological age on the prosocial subscales, a significant interaction was only found for the responsibility ($B=-0.21$, $SE=0.81$, $t=-2.55$) subscale. For the cooperation, assertion, and self-control subscales, no significant interactions were observed ($ps<0.05$).

To further understand the interaction between autism symptomology and age on total social skills, CARS scores were used to create three equal groups (e.g., low, medium, and high) and graphed. As shown in Figure 3.2, we see that boys with FXS who have low autism symptomology show a significant increase in their social skills as they get older compared to the other two groups. Those with mid-level autism symptomology have higher social skills at younger ages with minimal increases as they get older; however those with high autism symptomology have very low social skills at young ages with minimal increases in social skills at older ages.

Autistic symptomology as a categorical variable. The CARS cut-off score of 30 was utilized to separate the FXS sample into those with ASD (FXS+ASD; 30+, $n=22$) and those without ASD (<30, $n=37$) to examine whether the FXS only group has a distinct behavioral profile compared to those with FXS+ASD and TD boys ($n=21$), while controlling for adaptive behavior.

A MANCOVA indicted a significant effect of group on the SSRS total standard score ($F(2,76)= 8.15$, $p=0.001$) and on all the prosocial subscales: Cooperation ($F(2,76)= 5.45$, $p=0.006$) Assertion ($F(2,76)= 8.85$, $p<0.001$), Responsibility ($F(2,76)= 4.84$, $p=0.01$), and Self-Control ($F(2,76)= 5.09$, $p=0.008$). There were no significant effects of adaptive behavior on the SSRS total score or any of its subscales ($ps>0.05$).

Bonferroni-corrected pairwise comparisons showed that the boys with FXS+ASD had significantly less competent social skills than those with only FXS across the SSRS total score ($p<0.001$) and all of the prosocial subscales: Cooperation ($p=0.001$), Assertion ($p<0.001$), Responsibility ($p=0.04$) and Self-Control ($p=0.004$). Both groups with FXS, regardless of ASD status, displayed less competent social skills than their TD peers ($p<0.01$) across the SSRS total score and the Responsibility subscale score. The boys with only FXS did not perform significantly different than their TD peers on the subscales of Self-Control, Assertion, and Cooperation ($ps>0.05$) while controlling for adaptive behavior. The boys with FXS+ASD performed significantly worse than both groups across the SSRS total standard score and all of the prosocial subscales ($ps<0.04$).

3.3 Anxiety Symptoms in FXS on Social Skills

Anxiety as a continuous score. 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 3.3, a significant 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 while controlling for adaptive behavior. Results suggest that the effects of anxiety symptoms on social skills varies with age in boys with FXS. 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. No significant interactions were found for the cooperation, assertion, or responsibility subscales.

To further understand this interaction between anxiety symptoms and age on total social skills, anxiety was split into three equal groups (e.g., low, medium, and high) and

graphed. As shown in Figure 3.2, 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.

Anxiety as a categorical variable. To further understand the effects of anxiety symptoms on social skills within boys with FXS, anxiety was split into a categorical variable and follow up analyses were conducted with the SSRS total score and the prosocial subscales. The CBCL DSM Anxiety clinical significance score of 70 or greater was utilized to separate the FXS sample into those with high anxiety (n=15) and those with little to no anxiety (n=74).

A MANCOVA indicated trends toward significant effects of group for the SSRS total score ($F(3,85)= 3.85, p=0.053$), Assertion subscale ($F(3,85)= 3.57, p=0.062$), and Responsibility subscale ($F(3,85)= 2.97, p=0.088$). The subscales of Cooperation and Self-control were not found to differ significantly between groups ($p>0.05$).

Table 3.1 Summary of Regression Analyses for Age, Group, and Adaptive Behavior to Predicting Total Social Skills

Variables	<i>df</i>	<i>B</i>	<i>SE_B</i>	<i>t</i>	<i>p</i>
Intercept	130	-0.02	0.06	-0.36	0.72
Chronological Age	130	0.25	0.06	4.34	<0.001*
Group	130	0.70	0.06	12.65	<0.001*
Chronological Age*Group	130	0.16	0.06	2.87	0.005*
Controlling for Adaptive Behavior					
Intercept	64	-0.20	0.14	-1.45	0.15
Adaptive Behavior	64	0.44	0.17	2.60	0.12
Chronological Age	64	0.12	0.09	1.22	0.23
Group	64	0.71	0.37	1.93	0.06
Chronological Age*Group	64	0.17	0.26	0.64	0.52
Controlling for Developmental Level					
Intercept	64	1.44	1.30	1.11	0.27
Developmental Level	64	0.17	0.24	0.71	0.48
Chronological Age	64	0.60	0.95	0.64	0.53
Group	64	-1.01	0.73	-1.38	0.17
Chronological Age*Group	64	-0.21	0.50	-0.43	0.67

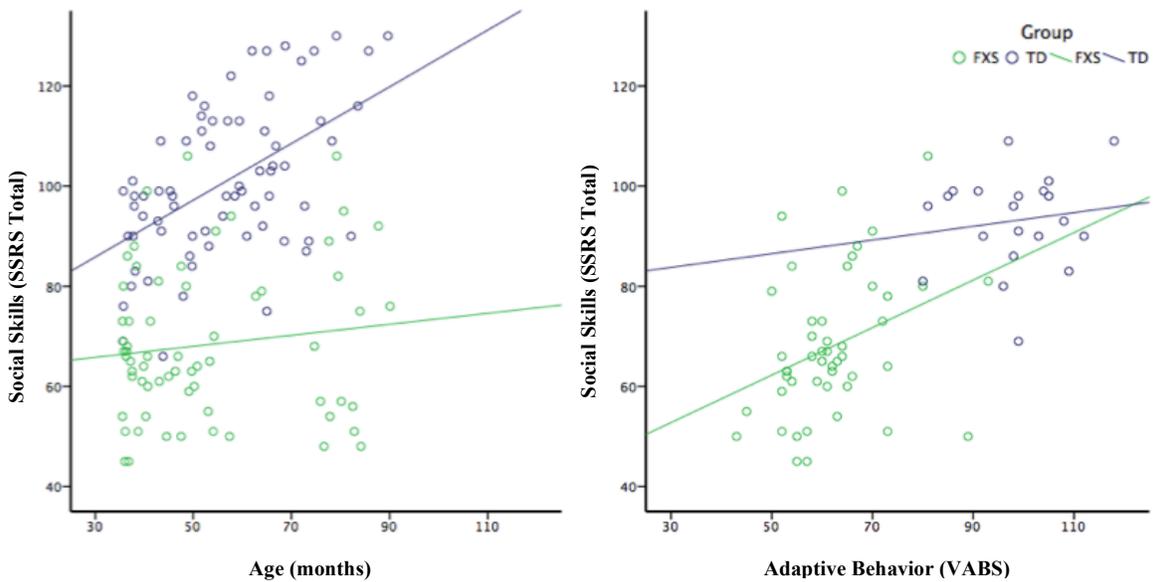


Figure 3.1 In the left pane, social skills are graphed against chronological age for boys with FXS and TD controls. In the right pane, social skills are graphed against adaptive behavior as measured by the VABS for a subsample of boys with FXS and TD controls.

Table 3.2 Summary of Regression Analyses for Autism Symptomology on Predicting Social Skills in Boys with FXS

Variables	<i>df</i>	<i>B</i>	<i>SE_B</i>	<i>t</i>	<i>p</i>
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*

Table 3.3 Summary of Regression Analyses for Anxiety on Predicting Social Skills in Boys with FXS

Variables	<i>df</i>	<i>B</i>	<i>SE_B</i>	<i>t</i>	<i>p</i>
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*

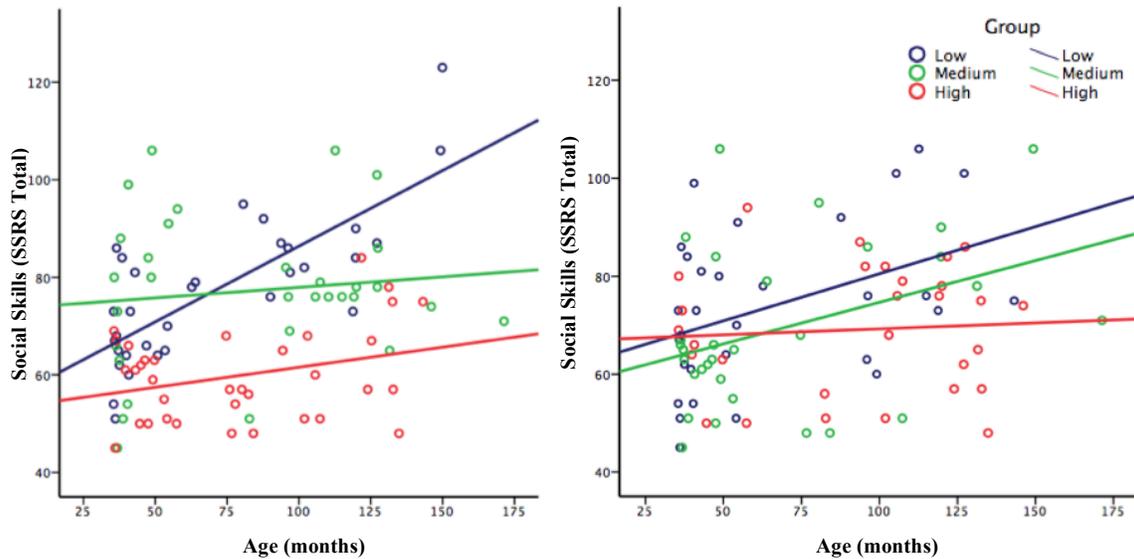


Figure 3.2 In the left pane, varying levels of autism symptomology are graphed on social skills and chronological age. In the right pane, varying levels of anxiety severity are graphed on social skills and chronological age.

CHAPTER 4

DISCUSSION

Social skills are socially acceptable learned behaviors that rely on a variety of psychological constructs and allow us to positively interact with peers across a variety of settings (Frey, Elliot, & Gresham, 2011; Gresham & Elliot, 1990). Specific social skills that we begin to 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 (Council for Exceptional Children, 2003; Gresham & Elliot, 1990). Understanding social skills and the factors that affect social skills is critical for academic, social, and psychological outcomes and for guiding diagnostic and treatment efforts. Social skill competence is predicted by age with increasing social skills developing across childhood into adolescence (Merrell & Gimpel, 2014). Cognitive skills also impact social skills, with individuals diagnosed with ID across all age ranges typically presenting with social skill deficits (Merrell & Gimpel, 2014). Given the importance of social skills and prevalence of deficits in those with intellectual disabilities, the present study examined social skill development and its association with autism symptomology and anxiety severity within boys with FXS, a genetic disorder at risk for poor social skill development. To date, this is the only study that has looked at specific social skills within FXS in comparison to TD boys and the relationship of anxiety and autism symptoms on those 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. The present study hypothesized that boys with FXS would display significant social skill deficits compared to TD boys across chronological age with adaptive behavior potentially minimizing group differences. In support of our first hypothesis, overall social skill impairments were largely accounted for by lower adaptive behavior and developmental level in the global and majority of the prosocial domains. Specifically, social skill deficits exist within boys with FXS in relationship to TD comparisons but when controlling for developmental level they show improvements across age. However, the Responsibility and Self-control subscales remained significantly impaired. This suggests that in the areas of self-control and responsibility, specific characteristics related to the social behavioral phenotype of FXS are playing a key role in these group differences outside of their developmental level.

In addition, the present study hypothesized that elevated features of autism would be associated with decreased social skill competency. Probing for a more comprehensive understanding of social skill development within boys with FXS, we found that elevated autism features were in fact associated in a complex nuanced manner. Specifically, increased autism symptoms were associated with decreased social skills only in older boys with FXS. Boys with FXS with a low degree of autism symptoms displayed elevated social skills at older ages. However, when examining the core prosocial subscale, only the area of responsibility was significantly affected by age and autism. These relationships held considering autism features at both the continuous and

categorical levels. When separating the FXS group into those with FXS only and those with FXS and ASD, the boys with FXS and ASD showed significantly less developed social skills compared to those with only FXS. Furthermore, both groups of FXS, regardless of ASD status, showed significantly delayed overall social skills and in the area of responsibility compared to their TD peers. These findings suggest that ASD comorbidity contributes to social skills deficits 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 only FXS, 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 traits.

Given the association of elevated anxiety to reduced social competence in the general population (Costello et al., 2003) and in those with anxiety disorders (Cordeiro et al., 2011; Tonnsen et al., 2013), we examined these associations within our FXS sample. Results indicated a somewhat complex effect with social skill development increasing with age only for those with low to mid-level anxiety severity whereas those with a high degree of anxiety symptoms demonstrated a flat profile with age similar to those with high autism symptomology. Even though overall the present studies sample had low anxiety symptoms, significant effects were still found within the small sample of those with high anxiety symptoms on social skills in these young boys with FXS. In addition, when teasing apart the prosocial subscales, only the area of self-control for boys with FXS was significantly affected by age and anxiety, which was different compared to

those with high autism symptomology. The results are consistent with the previous literature suggesting that poor social behaviors are related to anxiety severity within FXS (Boyle & Kaufmann, 2010; Hall et al., 2009; Tonnsen et al., 2013).

4.1 Limitations

The results of the present study should be considered with caution due to some 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 present study's sample was pulled from extant data and analyses were limited to using a cross-sectional design. Despite these limitations, collectively, the present study suggests that boys with FXS exhibit social skill deficits and that these deficits are related to autism symptomology, anxiety severity, and changes in age.

4.2 Conclusions and Future Directions

The present study aimed to expand our understanding of the FXS social behavioral phenotype. Clearly, social skills should be targeted for intervention, including early intervention, to help boys with FXS with their potential academic, social, and psychological outcomes. Boys with FXS who have high autism symptomology display significantly worse social skills, especially in the area of responsibility (e.g., introducing oneself, politely questioning unfair rules, asking permission before taking things, reporting accidents) suggesting a targeted area for intervention. Furthermore, boys with

FXS who report high anxiety severity show significant deficits in the area of self-control (e.g., appropriate tone of voice, avoid troublesome situations, controlling temper, cooperating with peers). Collectively, the present study's results point to developmental levels in FXS being a main contributor to social skill deficits with high levels of anxiety and autism symptomology as additive risk factors independently.

Future directions could include obtaining a larger sample across a variety of ages to examine how the 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. In addition, further examining how ASD and anxiety play different roles in their effects on social skills within boys with FXS. Also, future research could examine social skill deficits in females with FXS to see if they are similar or different compared to boys with FXS. Furthermore, examining these data utilizing longitudinal methods would build on the current study's findings to see how social skill deficits change over time. Lastly, results from this study should be utilized to design interventions for these individuals to increase their potential adult outcomes and success within their environment.

REFERENCES

- Abbeduto, L., McDuffie, A., & Thurman, A. J. (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, T. M., Dumenci, L., & Rescorla, L. A. (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.
- American Psychiatric Association. (2013). *Diagnostic and statistical manual of mental disorders* (5th ed.). Washington, DC: Author.
- Bailey Jr, D. B., Hatton, D. D., 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.
- Bailey Jr, D. B., 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, D. B., Raspa, M., Olmsted, M., & Holiday, D. B. (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.
- Baranek, G. T., Roberts, J. E., David, F. J., Sideris, J., Mirrett, P. L., Hatton, D. D., & Bailey, D. B. (2008). Developmental trajectories and correlates of sensory processing in young boys with fragile X syndrome. *Physical & occupational therapy in pediatrics*, 28(1), 79-98.
- Boyle, L., & Kaufmann, W. E. (2010, November). 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.
- Carter, A. S., Davis, N. O., Klin, A., & Volkmar, F. R. (2005). Social development in autism. *Handbook of Autism and Pervasive Developmental Disorders, Volume 1, Third Edition*, 312-334.
- Chang, Y. C., Quan, J., & Wood, J. J. (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.
- Coplan, R. J., 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.

- Cordeiro, L., Ballinger, E., Hagerman, R., & Hessel, D. (2011). Clinical assessment of DSM-IV anxiety disorders in fragile X syndrome: prevalence and characterization. *Journal of neurodevelopmental disorders*, 3(1), 57-67.
- Costello, E. J., 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.
- Council for Exceptional Children, Division for Learning Disabilities and Division for Research. (2003, Fall). Social skills instruction for students with learning disabilities. *Current Practice Alerts*, Issue 9, pp. 1-4.
- 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.
- 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.
- Frey, J. R., Elliott, S. N., & Gresham, F. M. (2011). Preschoolers' Social Skills: Advances in Assessment for Intervention Using Social Behavior Ratings. *School Mental Health*, 3(4), 179-190.
- Garber, K. B., Visootsak, J., & Warren, S. T. (2008). Fragile X syndrome. *European Journal of Human Genetics*, 16(6), 666-672.
- Gillis, J. M., Callahan, E. H., & Romanczyk, R. G. (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, F. M., & Elliott, S. N. (1990). *Social Skills Rating System: Preschool, Elementary Level*. American Guidance Service.
- Hagerman, R. J. & Hagerman, P. J. (2002). Fragile X syndrome. *Outcomes in neurodevelopmental and genetic disorders*. (198-219). Cambridge: Cambridge University Press.
- Hall, S. S., Lightbody, A. A., Hirt, M., Rezvani, A., & Reiss, A. L. (2010). Autism in fragile X syndrome: a category mistake?. *Journal of the American Academy of Child & Adolescent Psychiatry*, 49(9), 921-933.
- Hall, S. S., Lightbody, A. A., Huffman, L. C., Lazzeroni, L. C., & Reiss, A. L. (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.
- Harrison, P. L. (1987). Research with adaptive behavior scales. *The Journal of Special Education*, 21(1), 37-68.
- Kau, A. S., Tierney, E., Bukelis, I., Stump, M. H., Kates, W. R., Trescher, W. H., & Kaufmann, W. E. (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, W. E., Cortell, R., Kau, A. S., Bukelis, I., Tierney, E., Gray, R. M., ... & 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, E. M., Jo, B., Lightbody, A. A., Hazlett, H. C., Piven, J., ... & Reiss, A. L. (2014). Longitudinal profiles of adaptive behavior in fragile X syndrome. *Pediatrics*, 134(2), 315-324.
- Lord, C., Rutter, M., DiLavore, P. C., Risi, S., Gotham, K., & Bishop, S. (2012). *Autism Diagnostic Observation Schedule, 2nd Edition (ADOS-2)*. Torrance: Western Psychological Services.
- Magyar, C. I., & Pandolfi, V. (2007). Factor structure evaluation of the childhood autism rating scale. *Journal of Autism and Developmental Disorders*, 37(9), 1787-1794.
- Matson, J. L., & Wilkins, J. (2009). Psychometric testing methods for children's social skills. *Research in Developmental Disabilities*, 30(2), 249-274.
- Merrell, K. W., & Gimpel, G. (2014). *Social skills of children and adolescents: Conceptualization, assessment, treatment*. Psychology Press.
- Moskowitz, L. J. & Jones, E. A., (2015). Uncovering the evidence for behavioral interventions with individuals with fragile X syndrome: A systematic review. *Research in Developmental Disabilities*, 38, 223-241.
- Mullen, E. M. (1995). *Mullen scales of early learning* (pp. 58-64). Circle Pines, MN: AGS.
- Nakamura, B. J., Ebesutani, C., Bernstein, A., & Chorpita, B. F. (2009). A psychometric analysis of the child behavior checklist DSM-oriented scales. *Journal of Psychopathology and Behavioral Assessment*, 31(3), 178-189.

- Roberts, J. E., Weisenfeld, L. A. H., Hatton, D. D., Heath, M., & Kaufmann, W. E. (2007). Social approach and autistic behavior in children with fragile X syndrome. *Journal of autism and developmental disorders*, 37(9), 1748-1760.
- 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.
- Schalock, R. L., Borthwick-Duffy, S., Bradley, V., Buntinx, W. H. E., Coulter, D. L., Craig, E. M.,... Yeager, M. H. (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, B. H. (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.
- Schopler, E., Reichler, R. J., & Renner, B. R. (1988). *The childhood autism rating scale (CARS)*. Los Angeles: Western Psychological Services.
- Sparrow, S. S., Balla, D. A., & Cicchetti, D. V. (1984). *Vineland adaptive behavior scales: Interview edition, survey form manual*. Circle Pines, MN: American Guidance Service.
- Spinrad, T. L., Eisenberg, N., Harris, E., Hanish, L., Fabes, R. A., 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.

- Thorell, L. B., Bohlin, G., & Rydell, A. M. (2004). Two types of inhibitory control: Predictive relations to social functioning. *International Journal of Behavioral Development, 28*(3), 193-203.
- Tonnsen, B. L., Malone, P. S., Hatton, D. D., & Roberts, J. E. (2013). Early Negative Affect Predicts Anxiety, not Autism, in Preschool Boys with Fragile X Syndrome. *Journal of abnormal child psychology, 41*(2), 267-280.
- Tonnsen, B. L., Shinkareva, S. V., Deal, S. C., Hatton, D. D., & Roberts, J. E. (2013). Biobehavioral Indicators of Social Fear in Young Children With Fragile X Syndrome. *American Journal on Intellectual and Developmental Disabilities, 118*(6), 447-459.
- Tranfaglia, M. R. (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.