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Family Environment and Behavior Problems in Children, Adolescents, and Adults with Fragile X Syndrome

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Abstract

We examine how the family environment is associated with aspects of the Fragile X syndrome phenotype during childhood, adolescence, and adulthood. Mothers of children ($n = 48$), adolescents ($n = 85$), and adults ($n = 34$) with Fragile X syndrome participated in a multisite study. For children and adults with Fragile X syndrome, the presence of warmth and positivity and the absence of criticism were associated with fewer behavior problems. Although a higher level of criticism was significantly associated with greater behavior problems, there were only trend-level associations between levels of warmth and positivity and behavior problems during the adolescent years. The provision of family psychoeducation programs, which can reduce parental criticism, would likely benefit both the individual with Fragile X syndrome and the family.

Keywords

Fragile X syndrome; expressed emotion; family environment; life course

Individuals with Fragile X syndrome have cognitive and communication impairments and co-occurring conditions such as inattention/hyperactivity, anxiety, depression, and autism symptoms (Abbeduto, Brady, & Kover, 2007; Bailey, Raspa, Olmsted, & Holiday, 2008). There is some evidence that these impairments and conditions worsen during adolescence and early adulthood (Hatton et al., 2006). There is considerable heterogeneity in the course of Fragile X syndrome (Bailey, Raspa, Holiday, Bishop, & Olmsted, 2009; Bailey et al., 2008; Hartley et al., 2011), reflecting the influence of gender and intelligence as well as biomarkers of Fragile X, including mutation of the *Fragile X mental retardation 1 (FMR1)* gene. Less is known about how the family environment influences the course of Fragile X

syndrome. The present research is based on a multisite study investigating family adaptation to Fragile X syndrome and how the family context is associated with heterogeneity in life course patterns of development and functioning of individuals with Fragile X syndrome (Bailey, Hatton, Tassone, Skinner, & Taylor, 2001; Seltzer et al., in press; Warren, Brady, Sterling, Fleming, & Marquis, 2010). Given the centrality of the family in influencing lifelong development, in this study we examine how the family environment is associated with aspects of the Fragile X syndrome phenotype across three stages of the family life course: childhood, adolescence, and adulthood.

Family Environment and Child Development

The general literature on child development points to the bidirectional nature of parent–child relationships—how the child’s behaviors influence parents, and how parental behavior and the family environment shape the child’s social and emotional functioning (Pardini, 2008; Rutter, 2008). One characteristic of parenting behavior and the family’s environment that has received considerable research attention is *parental responsivity*, which refers to behavior characterized by warmth, nurturance, stability, predictability, and contingency on the child’s response (Spiker, Boyce, & Boyce, 2002). Responsivity has been shown to play an important role in enhancing child development (Osofsky & Thompson, 2000). Children whose mothers display more responsivity during the first several years of life achieve language milestones earlier (Landry, Smith, Swank, Assel, & Vellet, 2001), score higher on cognitive tests (Landry, Smith, Swank, & Miller-Loncar, 2000), develop better social skills (Calkins, Smith, Gill, & Johnson, 1998), and have fewer emotional and behavior problems (Goldberg, Corter, Lojkasek, & Minde, 1990). Maternal warmth, a component of responsivity, has long been shown to be predictive of higher levels of child adaptation and behavioral functioning (Caspi et al., 2004; Spera, 2005).

At the opposite end of the continuum from highly responsive and warm parenting is the display of high levels of parental criticism and negativity. Parental criticism may amplify emotional and behavioral dysregulation in children, resulting in the development of more serious behavior problems (Granic & Patterson, 2006; Snyder, Stoolmiller, Wilson, & Yamamoto, 2003). In addition, parental communications characterized by negativity, criticism, and low warmth have been shown to foster depressive cognitive styles in children, including self-criticism and low self-worth (Garber & Flynn, 2001; Radke-Yarrow, Belmont, Nottelmann, & Bottomly, 1990). Low maternal warmth and acceptance and high hostility and control have been linked to anxiety symptoms in children (see Degnan, Almas, & Fox, 2010), presumably because they heighten children’s perceptions of threat and reduce their feelings of mastery. The potential for parent negativity to foreshadow increased behavioral disturbance has led to the development of effective interventions aimed at reducing parents’ negativity and criticism toward their children and adolescents with behavior problems (e.g., Barkley, Edwards, & Robin, 1999).

Family Environment and Intellectual and Developmental Disabilities

There has been a strong tradition of research aiming to identify aspects of the family environment that can enhance the well-being of children with intellectual and developmental disabilities (IDD; see review by Hatton & Emerson, 2003). In one of the first studies, Mink, Nihira, and Meyers (1983) found that children in families that were more cohesive and expressive in their communication had higher levels of adaptive behavior, fewer behavior problems, and experienced less peer isolation. In a subsequent study of 218 children of families of children with IDD, Mink and Nihira (1987) showed that high levels of family cohesion and parental involvement predicted higher levels of community and social functioning. Hauser-Cram and colleagues (1999) studied families of children with

developmental delays and found that a more positive family environment when the child was age 3, as indexed by the cohesion and expressiveness subscales of the Family Environment Scale (Moos & Moos, 1986), predicted lower levels of both internalizing and externalizing behavior problems at age 5 (Mitchell & Hauser-Cram, 2009) and growth in social skills at age 10 (Hauser-Cram, Warfield, Shonkoff, & Krauss, 2001). Similarly, several studies have demonstrated a positive association between maternal responsivity and the development of children with IDD (see Warren & Brady, 2007, for a review), and other studies have shown that high levels of maternal criticism are associated with an increase in behavior problems of adolescents and adults with autism (e.g., Baker, Smith, Greenberg, Seltzer, & Taylor, 2011), confirming patterns in the general population.

Although there has been great interest in how the family environment affects the development of persons with a developmental disability (DD), there is a parallel body of research suggesting that the direction of effects are just as likely to flow from child to parent as from parent to child. The behavior problems and symptoms of children with autism spectrum disorder (ASD) are a significant source of stress for parents (Hastings & Brown, 2002; Lecavalier, Leone, & Wiltz, 2006; Lounds, Seltzer, Greenberg, & Shattuck, 2007). In a study of the bidirectional nature of effects between mothers and their adolescent and adult children with ASD, Smith, Greenberg, Seltzer, and Hong (2008) found that children's impairments in social reciprocity were associated with lower levels of maternal praise 18 months later. Orsmond, Seltzer, Greenberg, and Krauss (2006) similarly found that less severe maladaptive behaviors and fewer social impairments in children with ASD predicted higher quality parent-child relationships.

Less is known about the impact of the family environment on individuals with Fragile X syndrome. In one of the few studies exploring the effects of the family on young children with Fragile X syndrome, Warren and colleagues (2010) reported that early maternal responsivity, measured in 55 dyads of mother and child with Fragile X syndrome, predicted child language outcomes at 36 months of age, after controlling for child developmental level and autism symptoms. In another study, Wheeler, Hatton, Reichardt, and Bailey (2007) found that mothers of children with Fragile X syndrome exhibited high levels of warmth and displayed very low levels of negative behaviors such as punitiveness or corporal punishment as measured by the Maternal Rating Scale (Dewitt et al., 1997; Landry, Smith, Miller-Loncar, & Swank, 1997). Children scored higher on receptive language skills when their mothers presented the child with feedback or greater choices regarding an activity (i.e., maintaining behavior) and when the mothers made more verbal and nonverbal attempts to link activities, objects, or persons (i.e., scaffolding behaviors). Wheeler and colleagues (2010) also found evidence for bidirectional effects as the child's behavior was more strongly related to maternal responsivity than maternal traits such as depression and stress. Other research examined the effect of the quality of the family environment in families of 80 boys and 40 girls with Fragile X syndrome (age range = 6–17, $M = 10.8$); the quality of the home environment was found to be related to fewer autistic symptoms (Hessl et al., 2001), better cognitive outcomes (Dyer-Friedman et al., 2002), and better adaptive behavior (Glaser et al., 2003).

The present study is an investigation of the association between aspects of the family environment and behavior problems of individuals with Fragile X syndrome. We were guided in the design of this study by two gaps in past research. First, no previous Fragile X syndrome family study reflected a life course perspective, including those on families of children, adolescents, and adults. Since individuals with Fragile X syndrome, especially males, often live with their families well into adulthood, the influence of the family across the full family life course warrants investigation. Second, although past research examined the effects of different aspects of the family environment (e.g., responsivity, warmth,

punitiveness) on behavior problems, no previous study investigated how different dimensions of the family environment may have different effects depending on the age of the child. Therefore, in this study, we examine the relationship between different dimensions of the family environment and behavior problems, comparing families of children, adolescents, and adults with the full mutation of Fragile X syndrome.

Expressed Emotion

One strategy for characterizing the family environment builds on the concept of expressed emotion, which measures the emotional valence of the family (Hooley, 2007). A large number of studies have implicated expressed emotion in predicting symptom exacerbations across a broad range of mental health disorders and medical conditions, including schizophrenia, mood disorders, eating disorders, Alzheimer's disease, asthma, diabetes, and Parkinson's disease (see Hooley, 2007; Wearden, Tarrier, Barrowclough, Zastowny, & Rahill, 2000).

More recently, investigators have applied this concept in research on families of children with IDD. In one of the first such studies, high levels of criticism and emotional overinvolvement were found to be present only in a minority of families of adolescents and adults with ASD (18.1% for criticism and 10.1% for emotional overinvolvement; Greenberg, Seltzer, Hong, & Orsmond, 2006). Furthermore, it was found that lower maternal expressed emotion longitudinally predicted declining levels of behavior problems (Greenberg et al., 2006) and that changes in criticism in particular predicted behavior problems over an 8-year period (Baker et al., 2011). However, expressed emotion has not yet been evaluated as a measure of the family environment for individuals with Fragile X syndrome.

Present Study

In the present study, we examine two primary research questions: (a) Are there differences in the family environment of children with Fragile X syndrome at three stages of the family's life course: childhood, adolescence, and adulthood? and (b) Are the associations between aspects of the family environment and behavior problems similar across the three stages of the family's life course? In addition, in a follow-up analysis, we compare the family environment of adults with Fragile X syndrome to families of adults with ASD to determine whether there were diagnostic-group differences. The latter group of families was drawn from our longitudinal study of these families (Greenberg et al., 2006; Seltzer et al., 2011). When the Fragile X syndrome study began, the autism study was in its 10th year, and by that time, the sample included only a small number ($n = 15$) of adolescents with ASD and no children with ASD; thus, the analysis of diagnostic-group differences could not mirror the within-Fragile X syndrome analyses with age-group comparisons.

Regarding our first research question, based on our previous research on families of individuals with autism (Baker et al., 2011; Greenberg et al., 2006) and the research conducted by Wheeler and colleagues (2007) on mothers of young children with Fragile X syndrome, we expected to find relatively low rates of criticism (less than 25% of the sample) expressed by mothers of individuals with Fragile X syndrome across the life course. However, mothers of young children with Fragile X syndrome were expected to express significantly lower rates of criticism than mothers of adolescents or adults with Fragile X syndrome, reflecting the normative sanctions against criticizing young children who are perceived as having less developmental control over their behavior. We also hypothesized that mothers of adolescents and adults would express low rates of emotional overinvolvement, based on the existing body of research on families of children with developmental disabilities that has generally found that less than a quarter of study sample

members are characterized by high levels of emotional overinvolvement (Greenberg et al., 2006; Peris & Baker, 2000). However, we hypothesized that higher levels of emotional overinvolvement would be expressed by mothers of young children with Fragile X syndrome than by mothers of adolescents or adults because young children require significant protection and attention. Regarding positive dimensions of the family's environment, we expected moderate and high levels of warmth and positive remarks to be expressed by mothers at all stages of the life course, based on the findings of Wheeler and colleagues (2007).

Regarding our second research question, which examined the relationship between the family environment and behavior problems in the individual with Fragile X syndrome, we hypothesized that criticism would be positively related, and warmth and positive remarks would be negatively related, to behavior problems across all three age groups. We did not expect emotional overinvolvement to be predictive of behavior problems or symptoms for any of the three groups.

With respect to the comparison of families of adults with Fragile X syndrome to families of adults with ASD, we expected higher levels of criticism and lower levels of warmth and positive remarks to be expressed by mothers of adults with ASD because mothers of individuals with ASD must cope with a greater intensity of behavior problems than families of individuals with Fragile X syndrome (Abbeduto et al., 2004). However, based on the large literature finding similar relationships between measures of expressed emotion and behavior problems across diagnostic groups (Wearden et al., 2000), we did not expect to find differences in the pattern of correlations between different aspects of the family's environment and behavior problems across the two samples of families of adults with developmental disabilities.

Method

The data for this study comes from a larger, longitudinal multisite study of family adaptation to Fragile X syndrome. Although the larger study is longitudinal, only one wave of the Five Minute Speech Sample (Magaña et al., 1986), which is used to operationalize the different dimensions of expressed emotion, had been collected, and thus, this analysis is based on cross-sectional data on a total of 167 families of children ($n = 48$), adolescents ($n = 85$), and adults ($n = 34$) with Fragile X syndrome. Documentation from laboratory or medical records confirming that the son or daughter had the full mutation of the gene causing Fragile X syndrome was a requirement for participation. Mothers were recruited through service agencies, clinics, and foundations across the United States and from university-based research registries of families having a child with IDD. The sample was restricted to mothers with the premutation who provided complete data on the study measures.

Participants

Mothers of children with Fragile X syndrome ($n = 48$) participated in a longitudinal study of family adaptation to Fragile X syndrome (Warren & Brady, 2007). The Expressed Emotion protocol (Magaña et al., 1986) was introduced at the third wave of the study. Children's ages ranged from 10 to 40 months at the first data collection point (in 2003), and all lived in the parental home. In families with two or more children with Fragile X syndrome, the youngest child was the focal child for the present study. The median household income was between \$50,000 and \$80,000 in 2008, with incomes ranging from less than \$15,000 to \$100,000 or more. The majority of mothers were White (94%), currently married (75%), and had at least some college education (88%).

Mothers of adolescents ($n = 85$) and adults ($n = 34$) with Fragile X syndrome were selected from a companion longitudinal (3-wave) study of family adaptation to Fragile X syndrome (Seltzer et al., in press). The Expressed Emotion protocol was introduced at the first wave of the study. Median household income was between \$80,000 and \$89,000 in 2008, but a range in income was represented (less than \$9,999 to \$160,000 or more). The majority of mothers were White (95%), currently married (82%), and had at least some college education (83%), similar to the sample of families of children.

For the current analysis, age 22 and older was used as the marker for adulthood as children with disabilities may continue in high school until their 22nd birthday. For the current analysis, the sample of children ranged in age from 6 to 8 ($M = 7.2$), the sample of adolescents ranged in age from 12 to 21 ($M = 15.9$), and the sample of adults ranged in age from 22 to 43 ($M = 27.4$). Their mothers' ages were similarly diverse ($M = 38.5, 46.8, 55.8$ years, respectively). The majority of individuals with Fragile X syndrome were sons (83%, 84%, and 94% in the three age groups, respectively) and had ID (81%, 82%, and 82%, respectively). Neither the gender of the child nor the percent of those with ID differed across the three age cohorts.

Procedure

Mothers were interviewed (in their homes for those with school-aged children and by telephone for families of adolescents and adults), and they completed standardized self-administered measures. As part of the interviews, we administered the Five Minute Speech Sample (FMSS; Magaña et al., 1986) as the measure of the family environment. Past research has found that the FMSS, whether administered in person or over the telephone, produces equivalent ratings in studies of families of individuals with IDD (Beck, Daley, Hastings, & Stevenson, 2004).

For the FMSS, a family member is asked to speak for five minutes to describe his or her relationship with the person with the disability and his or her thoughts and feelings about the individual. The FMSS has been shown to produce valid and reliable ratings of expressed emotion (EE) in a variety of diagnostic groups (Van Humbeeck, Van Audenhove, De Hert, Pieters, & Storms, 2002), including families of individuals with IDD (Beck et al., 2004; Hastings, Daley, Burns, & Beck, 2006), and to correlate significantly with observed parent-child interactions among families in the general population (McCarty, Lau, Valeri, & Weisz, 2004).

The FMSS is coded with respect to both verbal content and vocal tone, and overall expressed emotion is calculated from the ratings of the criticism and emotional overinvolvement dimensions. In studies in which expressed emotion has been decomposed, the criticism dimension has been found to be a more robust predictor of symptom relapse than emotional overinvolvement, particularly in studies of individuals with IDD (Greenberg et al., 2006; Hastings et al., 2006; Peris & Baker, 2000). However, the appropriateness of various emotional overinvolvement codes has been questioned, especially for families of young children and when an individual has IDD. Emotional overinvolvement includes three codes: "excessive" positive remarks, self-sacrificing remarks, and remarks expressing parental overprotection. Wamboldt, O'Connor, Wamboldt, Gavin, and Klinnert (2000) suggested that positive remarks are similar to the dimension of warmth and raised questions about whether positive remarks should be included as an indicator of emotional overinvolvement. In our own research, positive remarks were associated with improved functioning in individuals with ASD (Smith et al., 2008), providing support for a different conceptualization than in the original paradigm of expressed emotion. Therefore, in the present study, we separate the self-sacrificing and overprotectiveness components of

emotional overinvolvement from the positive remarks component, as measured by the FMSS.

We further used a code for parental warmth, which has been shown to be related to lower levels of relapse in Mexican American individuals with schizophrenia (Lopez et al., 2004) and to reduced autism symptoms in a study of families of adolescents and adults with ASD (Smith et al., 2008). Warmth and positive remarks are overlapping constructs, but warmth reflects the parent's tone in describing the family member with the disability as well as expressions of sympathy, empathy, and interest in the family member, whereas positive remarks is a quantitative measure of the number of positive comments made by the parent during the FMSS.

A common protocol for transcribing the speech samples was used for families in all three life stages. FMSSs were rated for four measures of the family environment: criticism, emotional overinvolvement, positive remarks, and warmth. These ratings were performed by an independent rater with over 30 years of experience coding the FMSS for all aspects of expressed emotion. The same rater performed the coding for all three age groups of families of individuals with Fragile X syndrome as well as for the families of adults with ASD. We conducted a reliability study between this rater and another trained FMSS rater for 20 families of individuals with Fragile X syndrome. The percentage agreement was 95% ($\kappa = .91$) for the rating of criticism, and 90% for the rating of overinvolvement ($\kappa = .85$). The interrater reliability, as measured by the correlation between the two raters, for positive remarks was $r = .83$ ($p < .001$). In an earlier study (Smith et al., 2008), reliability was established for warmth in a study of 15 speech samples. The two raters had an interrater reliability of $r = .79$ ($p < .001$). In the Fragile X syndrome study, the reliability in the coding of warmth was replicated on a sample of 12 families; the correlation between the two raters was $r = .80$ ($p < .001$).

Measures

Criticism—Mothers were rated as *high* (2), *borderline* (1), or *low* (0) in criticism using procedures developed by Magaña and colleagues (1986). Respondents were rated high on criticism if they (a) made a negative opening remark, (b) described their relationship with their son or daughter in negative terms, or (c) made one or more criticisms about their son or daughter during the course of the speech sample. Mothers were rated borderline on criticism if they did not satisfy the above requirements but made one or more statements of dissatisfaction about their relationship with their son or daughter. Ratings of low criticism were given if no critical comments were present in the speech sample.

Emotional overinvolvement—Emotional overinvolvement was rated as high for this study if the family member either expressed excessive self-sacrificing or overprotective feelings toward the son or daughter or experienced an “emotional display” (e.g., crying) during the 5-min speech sample. A rating of borderline emotional overinvolvement was given if there was some evidence for the presence of self-sacrificing or overprotective feelings but the mother was below the threshold for being coded as excessive. Low emotional overinvolvement represents the absence of such indicators.

Positive remarks—As part of the protocol for coding the FMSS, raters coded the number of positive remarks the mother made about her son or daughter. Because few mothers made more than five positive remarks, scores of 5 or greater were top-coded to 5.

Warmth—Ratings of warmth were also coded from the FMSS. Guidelines from the Camberwell Family Interview (Vaugh & Leff, 1976) were utilized for coding warmth.

Warmth ratings were based on (a) tone of voice, (b) spontaneity of expression of sympathy, concern, and empathy, and (c) expression of interest in the child. Warmth was rated on a 5-point scale from 0 (*no warmth*) to 4 (*high warmth*).

Child behavior problems—Mothers completed the Child Behavior Checklist (CBCL; Achenbach & Rescorla, 2001) for sons/daughters who were 18.5 years of age or younger and the Adult Behavior Checklist (ABCL; Achenbach & Rescorla, 2003) for sons and daughters who were older than 18.5 years of age. Mothers indicated how true it was that their son or daughter exhibited each of 106 behaviors in the past 6 months. Each item was rated on 3-point scale: 0 (*not true*), 1 (*somewhat or sometimes true*), 2 (*very true or often true*). Scores for the three summary scales, Total Problems, Internalizing Problems, and Externalizing Problems, were computed. Reliability and validity for the CBCL and ABCL are well-established (Achenbach & Rescorla, 2001).

Analysis

Analysis of variance was used to examine differences between the three groups of families of adults with Fragile X syndrome and to examine differences between families of adults with Fragile X syndrome and families of adults with ASD with respect to aspects of the family environment and behavior problems. Eta square is reported as a measure of the effect size. Correlational analysis was used to examine whether associations between aspects of the family environment and behavior problems were similar across childhood, adolescents, and adulthood of persons with Fragile X syndrome, and whether the relationships between the family environment and behavior problems were similar for adults with Fragile X syndrome and adults with ASD.

Results

Our first research question investigated differences in the family environment in families of children, adolescents, and adults. In the preliminary analyses, we explored whether controlling for ID altered the results, but since it did not, ID was not included in the final analyses.

As shown in Table 1, levels of maternal criticism varied across the three age groups ($p < .05$), with follow-up tests indicating that mothers of children were rated as expressing higher levels of criticism than mothers of adolescents. There was a trend for levels of emotional overinvolvement to differ among the three groups, with mothers of adolescents showing higher levels of overinvolvement than mothers of children. The majority of mothers in all three groups had low levels of emotional overinvolvement (77%–91%). However, whereas approximately 10% of the mothers of children and mothers of adults were rated as high or borderline with respect to overinvolvement, 23% of mothers of adolescents were rated as high or borderline on overinvolvement. The three groups of mothers were very similar with respect to positive characteristics of the family environment. They made an average of between 3.0 and 3.3 positive remarks during the FMSS and showed a pattern of high or moderate levels of warmth (73%–88%). Thus, with respect to our first research question, negative dimensions of the family environment (i.e., criticism and overinvolvement) varied across the three groups whereas positive dimensions of the family environment (i.e., positive remarks and maternal warmth) were more stable across the three age cohorts.

As shown in Table 2, in regard to behavior problems in individuals with Fragile X syndrome, there were significant age-group differences with respect to internalizing problems ($F = 4.78, p < .01$) and total problems ($F = 3.84, p < .05$). Adolescents with Fragile X syndrome had a significantly higher frequency of internalizing problems than children with Fragile X syndrome, and children and adolescents had higher rates of total problems

than adults. There were no significant age-group differences in levels of externalizing problems.

Our second research question investigated whether similar associations are found between the indicators of the family environment and the son or daughter's behavior problems across the three life stages. As shown in Table 3, the associations between negative dimensions of the family environment and behavior problems were mainly similar across the three life stages. As hypothesized, maternal criticism was significantly related to externalizing symptoms for each of the three age groups, but it was unrelated to internalizing symptoms at any life stage. Criticism was also significantly associated with total problems for adolescents, but there was no significant association between criticism and total problems for children or adults (although the strength of the correlation was similar to those in the adolescent sample). Consistent with prior research, emotional overinvolvement was not related to internalizing, externalizing, or total problems scores for any of the three age groups.

In contrast, an age-related pattern was more evident in the relationship between positive dimensions of the family environment and behavior problems for children, adolescents, and adults. For both children and adults with Fragile X syndrome, higher numbers of positive remarks made by mothers during the FMSS were related to lower levels of externalizing and total problems (at the trend level for adults) in their sons and daughters with Fragile X syndrome. Positive remarks were significantly related to lower levels of internalizing behaviors in adults with Fragile X syndrome. However, for adolescents, there was a trend for positive remarks to be associated with lower levels of total problems. Among adolescents, positive remarks were unrelated to internalizing or externalizing problems. Similarly, higher levels of maternal warmth were significantly associated with fewer externalizing and total problems among children and adults with Fragile X syndrome; but among adolescents, only trend level associations were found. Maternal warmth was related to lower levels of internalizing behaviors at a trend level for adults with Fragile X syndrome but unrelated to internalizing problems for children or adolescents.

Diagnostic-Group Differences

As a follow-up analysis, we investigated the degree to which the family environment in adults with Fragile X syndrome differed from the family environment in families of adults with ASD, and whether behavior problems in adults with these two diagnoses were similarly or differentially associated with the family environment. For this analysis, we compared the families of 34 adults with Fragile X syndrome from the study reported above to 73 similarly aged adults with ASD, who are participating in our ongoing longitudinal study (Seltzer et al., 2011).

With respect to background characteristics, all adults in both diagnostic groups selected for the present analysis continued to live in the parental home at the time of the study. The sample of adults with ASD did not differ from the sample of adults with Fragile X syndrome with respect to child's age or mother's age, education, marital status, or ethnicity. However, the two samples were different with respect to household income (Fragile X syndrome, $M = \$86,000$, vs. ASD, $M = \$65,000$, $p < .01$), the percentage with ID (Fragile X syndrome, 82%, vs. ASD, 63%, $p < .05$), and the percentage of sons (Fragile X syndrome, 94%, vs. ASD, 74%, $p < .01$). Therefore, in the present follow-up analysis, family household income and the child's gender were controlled. The child's ID status was not controlled because gender and ID status were confounded in the Fragile X syndrome sample. Furthermore, since 18% of the adults with Fragile X syndrome also had a diagnosis of autism, we conducted a preliminary analysis comparing the adults with Fragile X syndrome and autism to the Fragile X syndrome-only group. There were no significant differences between the

two groups on any of the outcomes. Since ASD is part of the broader Fragile X syndrome phenotype, and to have adequate statistical power, we treated all adults with Fragile X syndrome as a single group.

In our comparison of mothers of adults with Fragile X syndrome versus mothers of adults with ASD, there were no significant diagnostic-group differences with respect to levels of criticism ($F = .29$, $\eta^2 = .003$) or emotional overinvolvement ($F = .44$, $\eta^2 = .004$) as expressed by the mothers. Of the mothers of adults with ASD, 63% were rated as borderline or high on criticism, not significantly different from the 50% of mothers of adults with Fragile X syndrome who were rated similarly. Although 21% of the mothers of adults with ASD but only 9% of the mothers of adults with Fragile X syndrome were classified as high or borderline with respect to emotional overinvolvement, this difference was not statistically significant.

Similarly, with respect to positive dimensions of the family environment, there were no group differences with respect to positive remarks ($F = 1.69$, $\eta^2 = .016$) or warmth ($F = .64$, $\eta^2 = .006$). Mothers of adults with Fragile X syndrome made an average 3.0 ($SD = 2.0$) positive remarks whereas mothers of adults with autism made an average of 2.4 ($SD = 1.8$) positive remarks about their child during the FMSS. Regarding warmth, the majority of the mothers of adults with Fragile X syndrome (88%) and ASD (70%) expressed moderate or high levels of warmth about the adult child during the FMSS. There were no diagnostic-group differences in levels of behavior problems (60.2 vs. 57.2 for internalizing behaviors; 56.4 vs. 55.7 for externalizing behaviors; and 59.6 vs. 56.6 for total problems for the ASD and Fragile X syndrome groups, respectively).

The associations of different dimensions of the family environment with the behavioral profile of the adult child also were remarkably similar across the two groups. As shown in Table 4, maternal criticism was positively and significantly related to levels of externalizing behaviors for adults with Fragile X syndrome and adults with ASD. Furthermore, for adults with ASD, levels of maternal criticism were significantly related to total behavior problems. Although the correlation between maternal criticism and total problems was not significant for adults with Fragile X syndrome, the magnitude of the correlation was similar to the correlation for adults with ASD. Maternal criticism was unrelated to internalizing behavior problems both for adults with Fragile X syndrome and for adults with ASD. Consistent with our examination of the associations between emotional overinvolvement and behavior problems at three points in the life course of individuals with Fragile X syndrome, we found no relationship between emotional overinvolvement and behavior problems in adults with ASD.

Positive dimensions of the family environment were significantly related to lower levels of behavior problems in both groups of adults with disabilities. For adults with Fragile X syndrome, positive remarks were significantly related to fewer internalizing problems and associated with fewer externalizing and total problems at a trend level. For adults with ASD, levels of positive remarks made by the mothers in the FMSS were significantly and inversely related to levels of externalizing and total problems, and at a trend level for internalizing problems. Warmth was associated with significantly lower levels of externalizing and total behavior problems for both groups of adults.

Discussion

In this study, we used a life course perspective to examine aspects of the family environment in families of individuals with Fragile X syndrome. Using cross-sectional methods, we compared families of school-aged children, adolescents, and adults with Fragile X

syndrome. For the most part, the family environment was similar across the life course, which may suggest that patterns of positivity and negativity are established early in parenthood and are persistent across time. The one exception to this pattern of similarity in family environment at different life stages was with respect to levels of maternal criticism, which were unexpectedly higher in the sample of children than in the samples of adolescents and adults. There also was a trend for mothers of adolescents to be more emotionally overinvolved than mothers with young children with Fragile X syndrome. These patterns, based on cross-sectional data, suggest resiliency in mothers of individuals with Fragile X syndrome at various stages of the life course because most showed consistently high levels of warmth and positive remarks. Longitudinal data are needed to confirm these cross-sectional patterns.

Consistent with the much larger body of research on expressed emotion in the general population (Christiansen, Oades, Psychogiou, Hauffa, & Sonuga-Barke, 2010; Tompson et al., 2010), high levels of criticism were related to high levels of externalizing problems in children, adolescents, and adults with Fragile X syndrome. Also, our finding that emotional overinvolvement was unrelated to behavior problems at any of the three life stages in families of individuals with Fragile X syndrome is consistent with prior research on families of individuals with developmental disabilities (Peris & Baker, 2000). In this study, we operationalized emotional overinvolvement in terms of high levels of self-sacrificing behaviors and/or high levels of overprotectiveness. We conducted a follow-up analysis and found that neither of these two dimensions was related to any of the indicators of behavior problems. Since most boys and many girls with Fragile X syndrome are diagnosed in the early childhood period and experience developmental delays, their parents are thrust early on into the role of being protective of their child and needing to making personal sacrifices to accommodate to their child's condition. Thus, for individuals with Fragile X syndrome and other developmental disabilities, high levels of parental involvement and protectiveness may be normative. In contrast, for individuals with mental illness, high levels of parental emotional overinvolvement have shown to be problematic (Hooley, 2007). Future research is needed to examine whether a much higher threshold of emotional overinvolvement must be reached before it has a detrimental effect on the lives of individuals with IDD.

With respect to positive dimensions of the family environment, for families of children and adults with Fragile X syndrome there was a general pattern for high levels of positive remarks and warmth to be related to lower levels of behavior problems. These findings are consistent with the large body of research on the relationship between positive aspects of the family environment and behavior problems in children (Caspi et al., 2004; Eisenberg et al., 2001; Smith et al., 2008). However, this pattern was not as evident in families of adolescents with Fragile X syndrome. The period of adolescence for individuals with Fragile X syndrome is fraught with transitions and uncertainty. Adolescents with Fragile X syndrome experience the same biological changes experienced by individuals without IDD during this stage of life but may have far fewer coping resources available to negotiate those life changes. Parents may become increasingly anxiously knowing that exiting high school is on the horizon, marking the end of many entitlement services that they have come to depend on. The heightened stress for families in the general population during adolescence and the even greater level of stress for families of adolescents with Fragile X syndrome, may serve to attenuate relationships between positive dimensions of the family's environment and the behavior of the adolescent with Fragile X syndrome during this life stage. In addition, the general literature on adolescence indicates that, during this period, parents and adolescents rate their level of closeness and interdependence lower (Laursen & Williams, 1997) and spend less time together (Larson, Richards, Moneta, Holmbeck, & Duckett, 1996) than during childhood.

In our follow-up analysis comparing families of adults with Fragile X syndrome and families of adults with ASD, there were few diagnostic-group differences in the nature of the family environment or how different aspects of the family environment were related to behavior problems. Most surprising, the strength of the associations between characteristics of the family environment and behavior problems in the adults with both conditions were virtually identical.

There have been a great number of studies finding support for the relationship between dimensions of expressed emotion and psychiatric symptoms and behavior problems in persons with disabilities (cf. Hooley, 2007; Wearden et al., 2000). This study provides further evidence that these associations are similar in families of individuals with Fragile X syndrome. It suggests that interventions, in particular psychoeducational interventions, which have been shown to lower criticism in other populations, could be adapted for families with Fragile X syndrome. However, since our data are correlational and do not permit inference about causality, intervention studies are a necessary next step to determine whether family treatments designed to lower expressed emotion and promote positivity in families of persons with psychiatric disabilities have a similar beneficial effect on adolescents and adults with Fragile X syndrome.

Although there appeared to be some elevation in criticism among families of young children with Fragile X syndrome, overall the profile of the three groups of families was one of high levels of warmth and positive feelings toward their children with an appropriate level of emotional involvement, consistent with Wheeler and colleagues (2007). Thus, the interventions described above should be targeted to the small subgroup of families with high criticism and low positivity. Even in the face of their child's challenging behaviors and the fact that many of the mothers themselves were at risk for emotional problems because of their genetic status (i.e., they were carriers of the premutation of the *FMR1* gene), these mothers maintained a positive family environment. These data indicate a pattern of family strength and resiliency, a pattern that is being increasingly documented by researchers studying the long-term impacts of parenting children with developmental disabilities (Blacher & Baker, 2007).

Several limitations of the study must be acknowledged. First, we did not have a measure of IQ on all sample members. As a proxy for IQ, we initially controlled for ID status but found it was not significant and did not affect our results and, therefore, dropped it from the final analysis. Our inability to adequately control for IQ status of the children is an important limitation of the study. Second, our sample of mothers of children, adolescents, and adults with Fragile X syndrome consisted of volunteers who were predominantly White and higher in socioeconomic status. Consequently, the representativeness of the sample and the generalizability of the findings to more diverse populations are not known. Third, the data are cross-sectional. The correlations between the family environment and child behaviors likely reflect bidirectional transactional processes that unfold over many years. Longitudinal research is needed to examine the temporal nature of these relationships. Fourth, we measured the family environment through the mothers' levels of expressed emotion and positivity; future research should include measures obtained from fathers and siblings. Finally, different modes of administering the FMSS were used in this study. Whereas the FMSS was administered during a face-to-face interview for families of children with Fragile X syndrome and adults with ASD, the FMSS was administered over the telephone for families of adolescents and adults with Fragile X syndrome. Yet, Beck and colleagues (2004) found good agreement on expressed emotion ratings applied to telephone and face-to-face interviews in mothers of individuals with ID.

In conclusion, this is the first study to examine life course patterns of the family environment in families of individuals with Fragile X syndrome. Overall, the data show a predominant and mostly stable pattern of positivity in the families, and that the presence of warmth and positivity and the absence of criticism are associated with lower levels of behavior problems in children and adults with Fragile X syndrome. Adolescents with Fragile X syndrome were similarly affected by levels of criticism in the family, but their behavior was not significantly associated with levels of warmth and positivity. Past research that has suggested a wear-and-tear effect of family caregiving (e.g., Clark, Bond, & Hecker, 2007) would predict that there would be age-associated increases in negativity and decreases in positivity among families providing care over a period of decades. In contrast, the present data suggest a pattern of resiliency in families of individuals with Fragile X syndrome, as the majority of mothers maintain high levels of positivity over time and continue to provide a family environment that fosters behavioral regulation in their sons and daughters. However, the small number of families who are rated high in criticism and low in warmth have sons and daughters at risk for externalizing behaviors. These families are in need of interventions and supports to assist them to increase the positivity of the family environment, with likely benefits not only for the son or daughter with Fragile X syndrome, but for the mother and the family as a whole.

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References

- Abbeduto L, Brady N, Kover ST. Language development and Fragile X syndrome: Profiles, syndrome-specificity and within-syndrome differences. *Mental Retardation and Developmental Disabilities Research Reviews*. 2007; 13:36–46. [PubMed: 17326110]
- Abbeduto L, Seltzer MM, Shattuck P, Krauss MW, Orsmond G, Murphy M. Psychological well-being and coping in mothers of youths with autism, Down syndrome, or Fragile X syndrome. *American Journal on Mental Retardation*. 2004; 109:237–254. [PubMed: 15072518]
- Achenbach, T.; Rescorla, LA. *Manual for the ASEBA School-Age Forms & Profile: An integrated system of multi-informant assessments*. Burlington, VT: University of Vermont, Research Center for Children, Youth, and Families; 2001.
- Achenbach, TM.; Rescorla, LA. *Manual for ASEBA Adult Forms & Profiles*. Burlington, VT: University of Vermont, Research Center for Children, Youth, and Families; 2003.
- Bailey DB, Hatton DD, Tassone F, Skinner M, Taylor AK. Variability in FMRP and early development in males with Fragile X syndrome. *American Journal on Mental Retardation*. 2001; 106:16–27. [PubMed: 11246709]
- Bailey D, Raspa M, Holiday D, Bishop E, Olmsted M. Functional skills of individuals with Fragile X syndrome: A lifespan cross-sectional analysis. *American Journal on Intellectual & Developmental Disabilities*. 2009; 114:289–303.10.1352/1944-7558-114.4. 289-303 [PubMed: 19642710]
- Bailey D, Raspa M, Olmsted M, Holiday D. Co-occurring conditions associated with FMR1 gene variations: Findings from a national parent survey. *American Journal of Medical Genetics, Part A*. 2008; 146A:2060–2069. [PubMed: 18570292]

- Baker JK, Smith LE, Greenberg JS, Seltzer MM, Taylor JL. Change in maternal criticism and behavior problems in adolescents and adults with autism across a 7-year period. *Journal of Abnormal Psychology*. 2011; 120(2):465–475.10.1037/a0021900 [PubMed: 21319925]
- Barkley, RA.; Edwards, GH.; Robin, AL. *Defiant teens: A clinician's manual for assessment and family intervention*. New York, NY: Guilford; 1999.
- Beck A, Daley D, Hastings RP, Stevenson J. Mother's expressed emotion towards children with and without intellectual disabilities. *Journal of Intellectual Disability Research*. 2004; 48:628–638. [PubMed: 15357682]
- Blacher J, Baker B. Positive impact of intellectual disability on families. *American Journal of Mental Retardation*. 2007; 112:330–348. [PubMed: 17676958]
- Calkins SD, Smith CL, Gill KL, Johnson MC. Maternal interactive style across contexts: Relations to emotional, behavioral, and physiological regulation during toddlerhood. *Social Development*. 1998; 7:350–369.
- Caspi A, Moffitt TE, Morgan J, Rutter M, Taylor A, Arseneault L, Polo-Tomas M. Maternal expressed emotion predicts children's antisocial behavior problems: Using monozygotic-twin differences to identify environmental effects on behavioral development. *Developmental Psychology*. 2004; 40:149–161. [PubMed: 14979757]
- Christiansen H, Oades RB, Psychogiou L, Hauffa BP, Sonuga-Barke EJ. Does the cortisol response to stress mediate the link between expressed emotion and oppositional behavior in Attention-Deficit/Hyperactivity-Disorder (ADHD)? *Behavioral & Brain Functions*. 2010; 6:1–11. [PubMed: 20047681]
- Clark MS, Bond MJ, Hecker JR. Environmental stress, psychological stress, and allostatic load. *Psychology, Health, & Medicine*. 2007; 12:18–30.
- Degnan K, Almas A, Fox N. Temperament and the environment in the etiology of childhood anxiety. *Journal of Child Psychology & Psychiatry*. 2010; 51:497–517. [PubMed: 20158575]
- Dewitt SJ, Sparks JW, Swank PR, Smith K, Denson SE, Landry SH. Physical growth of low birth weight infants in the first year of life: Impact of maternal behaviors. *Early Human Development*. 1997; 47:19–34. [PubMed: 9118826]
- Dyer-Friedman J, Glaser B, Hessel D, Johnston C, Huffman L, Taylor A, Reiss AL. Genetic and environmental influences on the cognitive outcomes of children with Fragile X syndrome. *Journal of the American Academy of Child & Adolescent Psychiatry*. 2002; 41:237–244. [PubMed: 11886017]
- Eisenberg N, Gershoff ET, Fabes RA, Shepard SA, Cumberland AJ, Losoya SH, Murphy BC. Mothers' emotional expressivity and children's behavior problems and social competence: Mediation through children's regulation. *Developmental Psychology*. 2001; 37:475–490. [PubMed: 11444484]
- Garber J, Flynn C. Predictors of depressive cognitions in young adolescents. *Cognitive Therapy & Research*. 2001; 25:353–376.
- Glaser B, Hessel D, Dyer-Friedman J, Johnston C, Wisbeck J, Taylor A, Reiss A. Biological and environmental contributions to adaptive behavior in Fragile X syndrome. *American Journal of Medical Genetics*. 2003; 117A:21–29. [PubMed: 12548736]
- Goldberg S, Corter C, Lojkasek M, Minde K. Prediction of behavior problems in four-year-olds born prematurely. *Development and Psychopathology*. 1990; 2:15–30.
- Granic I, Patterson GR. Toward a comprehensive model of antisocial development: A dynamic systems approach. *Psychological Review*. 2006; 113:101–131.10.1037/0033-295X.113.1.101 [PubMed: 16478303]
- Greenberg JS, Seltzer MM, Hong J, Orsmond GI. Bidirectional effects of expressed emotion and behavior problems and symptoms in adolescents and adults with autism. *American Journal on Mental Retardation*. 2006; 111:229–249. [PubMed: 16792426]
- Hartley SL, Seltzer MM, Raspa M, Olmsted MG, Bishop EE, Bailey DB. Exploring the adult life of men and women with Fragile X syndrome: Results from a national survey. *American Journal of Intellectual & Developmental Disabilities*. 2011; 116(1):16–35.
- Hastings RP, Brown T. Behavior problems of children with autism, parental self-efficacy, and mental health. *American Journal of Mental Retardation*. 2002; 107:222–232. [PubMed: 11966335]

- Hastings RP, Daley D, Burns C, Beck A. Maternal distress and expressed emotion: Cross sectional and longitudinal relationships with behavior problems of children with intellectual disabilities. *American Journal of Mental Retardation*. 2006; 111:48–61. [PubMed: 16332156]
- Hatton C, Emerson E. Families with a person with intellectual disabilities: Stress and impact. *Current Opinion in Psychiatry*. 2003; 16:497–501.
- Hatton DD, Sideris J, Skinner M, Mankowski J, Bailey DB, Roberts J, Mirrett P. Autistic behavior in children with Fragile X syndrome: Prevalence, stability, and the impact of the FMRP. *American Journal of Medical Genetics Part A*. 2006; 140:1804–1813. [PubMed: 16700053]
- Hauser-Cram P, Warfield M, Shonkoff J, Krauss M. Children with disabilities: A longitudinal study of child development and parent well-being. *Monographs of the Society for Research in Child Development*. 2001; 66:1–131.10.1111/1540-5834.00151
- Hauser-Cram P, Warfield M, Shonkoff J, Krauss M, Upshur C, Sayer A. Family influences on adaptive development in young children with Down syndrome. *Child Development*. 1999; 70:979–989.10.1111/1467-8624.00071 [PubMed: 10446730]
- Hessl D, Dyer-Friedman J, Glaser B, Wisbeck J, Barajas RG, Taylor A, Reiss AL. The influence of environmental and genetic factors on behavior problems and autistic symptoms in boys and girls with Fragile X syndrome. *Pediatrics*. 2001; 108:E88. [PubMed: 11694672]
- Hooley JM. Expressed emotion and relapse of psychopathology. *Annual Review of Clinical Psychology*. 2007; 3:329–352.
- Landry SH, Smith KE, Miller-Loncar CL, Swank PR. Predicting cognitive-linguistic and social growth curves from early maternal behaviors in children at varying degrees of biological risk. *Developmental Psychology*. 1997; 33:1040–1054. [PubMed: 9383626]
- Landry SH, Smith KE, Swank PR, Assel MA, Vellet S. Does early responsive parenting have a special importance for children's development or is consistency across early childhood necessary? *Developmental Psychology*. 2001; 37:387–403. [PubMed: 11370914]
- Landry SH, Smith KE, Swank PR, Miller-Loncar CL. Early maternal and child influences on children's later independent cognitive and social functioning. *Child Development*. 2000; 71:358–375. [PubMed: 10834470]
- Larson RW, Richards MH, Giovanni M, Holmbeck G, Duckett E. Changes in adolescents' daily interactions with their families from ages 10 to 18: Disengagement and transformation. *Developmental Psychology*. 1996; 32:744–754.
- Laursen, B.; Williams, VA. Perceptions of interdependence and closeness in family and peer relationships among adolescents with and without romantic partners. In: Hulman, S.; Collins, AW., editors. *Romantic relationships in adolescence: Developmental perspectives*. San Francisco, CA: Jossey-Bass; 1997. p. 3-20.
- Lecavalier L, Leone S, Wiltz J. The impact of behavior problems on caregiver stress in young people with autism spectrum disorders. *Journal of Intellectual Disabilities Research*. 2006; 50:172–183.
- Lounds J, Seltzer MM, Greenberg JS, Shattuck PT. Transition and change in adolescents and young adults with autism: Longitudinal effects on maternal well-being. *American Journal of Mental Retardation*. 2007; 112:401–417. [PubMed: 17963433]
- Lopez SR, Hipke KN, Polo AJ, Jenkins JH, Karno M, Vaughn C, Snyder KS. Ethnicity, expressed emotion, attributions, and course of schizophrenia: Family warmth matters. *Journal of Abnormal Psychology*. 2004; 113:428–439. [PubMed: 15311988]
- Magaña AB, Goldstein MJ, Karno M, Miklowitz DJ, Jenkins J, Falloon IRH. A brief method for assessing expressed emotion in relatives of psychiatric patients. *Psychiatry Research*. 1986; 17:203–212. [PubMed: 3704028]
- McCarty C, Lau A, Valeri S, Weisz J. Parent-child interactions in relation to critical and emotionally overinvolved expressed emotion (EE): Is EE a proxy for behavior? *Journal of Abnormal Child Psychology*. 2004; 32:83–93.10.1023/BLJACP.0000007582.61879.6f [PubMed: 14998113]
- Mink IT, Nihira K. Direction of effects: Family life styles and behavior of TMR children. *American Journal on Mental Retardation*. 1987; 92:57–64.
- Mink IT, Nihira K, Meyers C. Taxonomy of family life styles: I. Homes with TMR children. *American Journal of Mental Deficiency*. 1983; 87:484–497. [PubMed: 6220609]

- Mitchell DB, Hauser-Cram P. Early predictors of behavior problems: Two years after early intervention. *Journal of Early Intervention*. 2009; 32:3–16.
- Moos, RH.; Moos, BS. Family Environment Scale manual. 2. Palo Alto, CA: Consulting Psychologists Press; 1986.
- Orsmond GI, Seltzer MM, Greenberg JS, Krauss MW. Mother-child relationship quality among adolescents and adults with autism. *American Journal of Mental Retardation*. 2006; 111:121–137. [PubMed: 16466284]
- Osofsy, JD.; Thompson, MD. Adaptive and maladaptive parenting: Perspectives on risk and protective factors. In: Shonkoff, JP.; Meisels, SJ., editors. *Handbook of early childhood intervention*. 2. New York, NY: Cambridge University Press; 2000. p. 54-75.
- Pardini D. Novel insights into longstanding theories of bidirectional parent-child influences: Introduction to the special section. *Journal of Abnormal Child Psychology*. 2008; 36:627–631. [PubMed: 18437550]
- Peris T, Baker B. Applications of the expressed emotion construct to young children with externalizing behavior: Stability and prediction over time. *Journal of Child Psychology and Psychiatry*. 2000; 41:457–462. [PubMed: 10836675]
- Radke-Yarrow, M.; Belmont, B.; Nottelmann, E.; Bottomly, L. Young children's self-conceptions: Origins in the natural discourse of depressed mothers and their children. In: Cicchetti, D.; Beeghly, M., editors. *The self in transition: Infancy to childhood*. Chicago, IL: University of Chicago Press; 1990. p. 345-361.
- Rutter, M. *Rutter's child and adolescent psychiatry*. 5. Malden, MA: Blackwell; 2008.
- Seltzer MM, Barker ET, Greenberg JS, Hong J, Coe C, Almeida D. Differential sensitivity to life stress in *FMR1* premutation carrier mothers of children with Fragile X syndrome. *Health Psychology*. (in press).
- Seltzer, MM.; Greenberg, JS.; Taylor, JL.; Smith, LE.; Orsmond, GI.; Esbensen, A.; Hong, J. Adolescents and adults with autism spectrum disorder. In: Amaral, DG.; Dawson, G.; Geschwind, D., editors. *Autism spectrum disorders*. New York, NY: Oxford University Press; 2011.
- Smith LE, Greenberg JS, Seltzer MM, Hong J. Symptoms and behavior problems of adolescents and adults with autism: Effects of mother-child relationship quality, warmth, and praise. *American Journal on Mental Retardation*. 2008; 113:387–402.10.1352/2008.113:387–402 [PubMed: 18702558]
- Snyder J, Stoolmiller M, Wilson M, Yamamoto M. Child anger regulation, parental responses to children's anger displays, and early child antisocial behavior. *Social Development*. 2003; 12:335–360.
- Spera C. A review of the relationship among parenting practices, parenting styles, and adolescent school achievement. *Educational Psychology Review*. 2005; 17:125–146.
- Spiker, D.; Boyce, GC.; Boyce, LK. Parent-child interactions when young children have disabilities. In: Glidden, LM., editor. *International review of research in mental retardation*. Vol. 25. San Diego, CA: Academic Press; 2002. p. 35-70.
- Tompson MC, Pierre CB, Boger KD, McKowen JW, Chan PT, Freed RD. Maternal depression, maternal expressed emotion, and youth psychopathology. *Journal of Abnormal Child Psychology*. 2010; 38:105–117. [PubMed: 19693663]
- Van Humbeeck G, Van Audenhove C, De Hert M, Pieters G, Storms G. Expressed emotion: A review of assessment instruments. *Clinical Psychology Review*. 2002; 22:321–341.
- Vaugh CE, Leff JP. The measurement of expressed emotion in the families of psychiatric patients. *British Journal of Social & Clinical Psychology*. 1976; 15:157–165. [PubMed: 938822]
- Wamboldt FS, O'Connor SL, Wamboldt MZ, Gavin LA, Klinnert MD. The five minute speech sample in children with asthma: Deconstructing the construct of expressed emotion. *Journal of Child Psychology & Psychiatry*. 2000; 41:887–898. [PubMed: 11079431]
- Warren SF, Brady NC. The role of maternal responsivity in the development of children with intellectual disabilities. *Mental Retardation & Developmental Disabilities Research Reviews*. 2007; 13:330–338. [PubMed: 17979201]

- Warren SF, Brady N, Sterling A, Fleming K, Marquis J. Maternal responsivity predicts language development in young children with Fragile X syndrome. *American Journal on Intellectual & Developmental Disabilities*. 2010; 115(1):54–75. [PubMed: 20025359]
- Wearden AJ, Tarrier N, Barrowclough C, Zastowny TR, Rahill AA. A review of expressed emotion in health care. *Clinical Psychology Review*. 2000; 20:633–666. [PubMed: 10860170]
- Wheeler A, Hatton D, Reichardt A, Bailey D. Correlates of maternal behaviours in mothers of children with Fragile X syndrome. *Journal of Intellectual Disability Research*. 2007; 51:447–462. [PubMed: 17493028]
- Wheeler AC, Hatton DD, Holloway VT, Sideris JJ, Neebe EC, Roberts JE, Reznick JS. Maternal responses to child frustration and requests for help in dyads with Fragile X syndrome. *Journal of Intellectual Disability Research*. 2010; 54:501–515. [PubMed: 20426796]

Table 1

Family Environment: By Child's Age Categories

Measures of family environment	Children with FXS (<i>n</i> = 48)	Adolescents with FXS (<i>n</i> = 85)	Adults with FXS (<i>n</i> = 34)	<i>F</i> value/ η^2 <i>df</i> = (2,164)
Criticism ^a				
High	31%	16%	29%	<i>F</i> = 3.67 [*]
Borderline	40%	31%	21%	η^2 = .043
Low	29%	53%	50%	
EOI ^b				
High	0%	7%	6%	<i>F</i> = 2.78 ⁺
Borderline	10%	16%	3%	η^2 = .033
Low	90%	77%	91%	
Positive Remarks				
< 5 positive remarks	71%	61%	59%	<i>F</i> = .60,
5+ positive remarks	29%	39%	41%	η^2 = .007
Mean positive remarks	3.0 (1.1)	3.3 (1.7)	3.0 (2.0)	
Warmth				
None/very little	2%	5%	3%	<i>F</i> = .79
Some	25%	14%	9%	η^2 = .010
Moderate	56%	61%	73%	
High	17%	20%	15%	
Mean warmth	3.3 (1.1)	3.5 (1.1)	3.4 (0.9)	

Note. FXS = Fragile X syndrome; EOI = emotional overinvolvement.

^aPost hoc follow-up tests showed that families of children had higher levels of criticism than families of adolescents: *M* difference = .38 (*SE* = .14), *p* < .05.

^bPost hoc follow-up tests showed that families of adolescents had higher levels of overinvolvement than families of children; *M* difference = .20 (*SE* = .09), *p* < .08.

⁺*p* < .07.

^{*}*p* < .05.

Table 2

Behavior Problems: By Child's Age Categories

Measures of behavior problems	Children with FXS (<i>n</i> = 48)	Adolescents with FXS (<i>n</i> = 85)	Adults with FXS (<i>n</i> = 34)	<i>F</i> value/ η^2 <i>df</i> = (2,164)
ABCL/CBCL				
Internalizing ^a	54.6 (8.9)	59.4 (8.8)	57.2 (8.2)	<i>F</i> = 4.78 ** η^2 = .055
Externalizing	56.0 (8.6)	53.6 (8.6)	55.7 (7.3)	<i>F</i> = 1.53 η^2 = .018
Total problems ^b	60.6 (7.9)	60.6 (7.3)	56.6 (6.5)	<i>F</i> = 3.84 * η^2 = .045

Note. FXS = Fragile X syndrome; ABCL = Adult Behavior Checklist; CBCL = Child Behavior Checklist.

^aPost hoc follow-up tests showed families of children had fewer internalizing problems than families of adolescents ($p < .01$).

^bPost hoc follow-up tests showed that families of adults had fewer total behavior problems than families of children ($p < .05$) and families of adolescents ($p < .05$).

*
 $p < .05$.

**
 $p < .01$.

Table 3

Correlations between Family Environment Measures and Adult Behavior Checklist and Child Behavior Checklist Variables

Measures of family environment	Children with FXS (<i>n</i> = 48)	Adolescents with FXS (<i>n</i> = 85)	Adults with FXS (<i>n</i> = 34)
Criticism			
Internalizing	.11	.06	.10
Externalizing	.29 [*]	.26 [*]	.34 [*]
Total problems	.21	.28 [*]	.22
EOI			
Internalizing	.17	-.18	.13
Externalizing	-.08	-.01	.06
Total problems	-.02	-.11	.05
Positive remarks			
Internalizing	-.21	-.15	-.35 [*]
Externalizing	-.35 [*]	-.17	-.31 ⁺
Total problems	-.31 ^{**}	-.18 ⁺	-.33 ⁺
Warmth			
Internalizing	-.13	-.15	-.29 ⁺
Externalizing	-.38 ^{**}	-.19 ⁺	-.37 [*]
Total problems	-.30 [*]	-.21 ⁺	-.38 [*]

Note. FXS = Fragile X syndrome; EOI = emotional overinvolvement.

⁺ $p < .10$.

^{*} $p < .05$.

^{**} $p < .01$.

Table 4

Adults with Fragile X Syndrome Versus Autism Spectrum Disorder: Correlations between Family Environment Measures and ABCL/CBCL Variables Controlling for Gender and Family Income

Measures of family environment	Adults with FXS (<i>n</i> = 34)	Adults with autism (<i>n</i> = 73)
Criticism		
Internalizing	.14	.16
Externalizing	.37 *	.33 **
Total problems	.27	.29 *
EOI		
Internalizing	.19	.11
Externalizing	.08	-.07
Total problems	.08	.03
Positive remarks		
Internalizing	-.40 *	-.20 ⁺
Externalizing	-.31 ⁺	-.25 *
Total problems	-.32 ⁺	-.31 **
Warmth		
Internalizing	-.28	-.16
Externalizing	-.36 *	-.44 ***
Total problems	-.36 *	-.40 ***

Note. ABCL = Adult Behavior Checklist; CBCL = Child Behavior Checklist; FXS = Fragile X syndrome; EOI = emotional overinvolvement.

⁺ $p < .10$.

* $p < .05$.

** $p < .01$.

*** $p < .001$.