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Behavior Problems, Maternal Internalizing Symptoms, and Family Relations in Families of Adolescents and Adults with Fragile X Syndrome

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Abstract

Background—Studies have linked the behavior problems of children with fragile X syndrome (FXS) to maternal well-being, but less is known about how behavior problems relate to important family factors such as marital satisfaction and family cohesion.

Method—Married mothers of 115 adolescents and adults with FXS completed questionnaires and interviews, and maternal CGG repeat length was obtained by medical/laboratory records or by blood analysis.

Results—Indirect effects were present between behavior problems and family variables in that behavior problems were positively related to maternal internalizing symptoms which were, in turn, negatively associated with both family cohesion and marital satisfaction. Direct associations between behavior problems and family relationship variables were not significant.

Conclusions—Findings suggest the importance of intervening with behavior problems in individuals with FXS and identify maternal mental health as a potentially powerful conduit for the effects of child behavior on relationships within these families. Implications for targeted interventions are discussed.

Keywords

fragile X syndrome; family; behavior problems; cohesion; marital satisfaction

Fragile X Syndrome (FXS) is the leading known cause of inherited intellectual disability and results from a mutation of the *FMR1* gene on the long arm of the X chromosome (Hagerman & Hagerman, 2002). In addition to cognitive delay, the behavioral phenotype of FXS often includes features of autism (e.g., poor eye contact, perseverative behavior) and attention-deficit/hyperactivity disorder (e.g., inattention, hyperactivity), as well as symptoms of social

anxiety (Bailey et al., 2008; Hatton et al., 2002). Mothers of individuals with FXS report high levels of distress as compared to both the general population and to mothers of children with certain other disabilities (e.g., Down syndrome; Abbeduto et al., 2004; Sarimski, 1997; Wheeler et al., 2007); however, considerable variability in well-being exists (Poelmann et al., 2003; Lewis et al., 2006).

As is the case for mothers of children with other developmental disabilities (e.g., Davis & Carter, 2008; McIntyre et al., 2002), variability in well-being among mothers of individuals with FXS appears to be tied in part to behavioral characteristics of the child. Behavior problems in individuals with FXS have been associated with maternal stress and distress (e.g., depression, anxiety) in studies focusing on childhood, adolescence, and young adulthood (Abbeduto et al., 2004; Hall et al., 2007; Johnston et al., 2003; Wheeler, et al., 2007). Although the link between children's behavior problems and poorer well-being in mothers of children with FXS is relatively well established, the degree to which behavior problems and poor maternal mental health might affect broader aspects of family functioning has been investigated less frequently. It is likely that maternal distress brought on and/or amplified by difficult child behavior might spillover beyond the parent-child dyad, challenging more distal dyadic relationships and the broader family climate (Bailey et al., 2007). The current study examined associations between behavior problems in adolescents and adults with FXS, maternal internalizing symptoms, and two key family outcomes: family cohesion and mothers' marital satisfaction.

One of the most studied aspects of interpersonal functioning within families of individuals with FXS has been family-level cohesion, or the closeness and supportiveness of the family unit. Johnston and colleagues (2003) found that behavior problems in a sample of children and adolescents with FXS were positively associated with aspects of maternal stress, which were, in turn, linked to lower family cohesion. Interestingly, behavior problems in this study were associated directly with family cohesion at the level of a trend only. Hall et al. (2007), found associations between children's behavior problems and maternal internalizing symptoms (anxiety and depression), but did not find significant links between behavior problems and broad family functioning; however, cohesion was not examined specifically and the association between internalizing symptoms and the family environment was not a focus. The few findings to date suggest that either the behavior problems of individuals with FXS are unrelated to family cohesion or that the effects may be indirect, through compromised maternal well-being.

To our knowledge, no study has examined the marital relationships of parents of individuals with FXS. Findings involving families of children with other developmental disabilities are mixed, suggesting that the presence of a child with a disability may or may not impart risk for poor marital functioning (see Hartley, Seltzer, Barker, et al., in press, for a review). Examinations of the correlates of marital functioning *within* families of children with developmental problems have produced somewhat mixed results. Baker and colleagues (2002) found that the behavior problems of preschoolers with developmental delays related to parents' reports of a negative impact on their marriages. Kersh et al. (2006) reported an association between the behavior problems of 10-year-old children with developmental disabilities and marital quality for mothers but not for fathers. Depressive symptoms in this study were related to both child behavior, and to marital quality — again raising the possibility of indirect effects.

Research and theory related to indirect effects in genetic syndromes has tended to focus on children's behavior as a mechanism for associations between syndrome status and parental stress or coping (Hodapp, 1997; Hodapp & Dykens, 2009). The current study expands upon

investigations of indirect effects between genotype and family response to examine potential pathways among specific areas of functioning *within* families of individuals with FXS.

In addition to experiences shared with other families of children with developmental disabilities, families of individuals with FXS may also encounter unique challenges. Unlike many commonly studied developmental disorders, FXS has a known genetic inheritance linked to particular family members who themselves may exhibit certain vulnerabilities as a function of their carrier status (Bailey et al., 2008; Franke et al., 1998). Family interaction and communication around issues related to FXS are likely to be challenging for these families (Bailey, 2010), potentially complicating family dynamics and marital relations.

The majority of studies on FXS and family process have focused on children and adolescents or very young adults with FXS (e.g., Abbeduto et al., 2004; Hall et al., 2007; Johnston et al., 2003; Lewis et al., 2006; Wheeler et al., 2007). However, changes in the FXS phenotype over time (Dykens et al., 2000) and in parenting challenges related to caring for an adult child with a disability (Seltzer et al., 2001), make consideration of family process in FXS important across the continuum of the family life cycle. Although considerable variability exists, many adults with FXS continue to require significant care (Bailey et al., 2009), and recent findings from our laboratory have linked the behavior of adolescents and adults with FXS to maternal daily stress, using daily diaries and cortisol measurement (MASKED). Understanding the pathways by which behavior problems might affect mental health and relationships in families of adolescents and adults with FXS could identify the most beneficial avenues for interventions aimed at improving the lives of these families.

The Current Study

We hypothesized that behavior problems in the individuals with FXS would be positively associated with maternal internalizing (i.e., depressive and anxiety) symptoms, which would, in turn, relate negatively to both marital satisfaction and family cohesion. Furthermore, we predicted significant indirect effects between behavior problems and the family relationship variables through maternal internalizing symptoms.

The possibility of maternal internalizing symptoms as a pivotal mechanism for the influence of children's behavior problems on families of individuals with FXS is particularly intriguing given the suggestion that maternal carrier status (i.e., the premutation of the *FMR1* gene) may lead to increased risk for internalizing problems, above and beyond the effects of caring for a child with a disability (Bailey et al., 2008; Franke et al., 1998). The current study therefore accounted for variations in the genotype of the mothers (i.e., the number of CGG repeats) and focused only on mothers with the *FMR1* premutation (CGG = 55 to 200), due to the qualitatively different phenotype of individuals with the full mutation (CGG > 200; Bailey et al., 2008; Hagerman & Hagerman, 2002). We also considered additional factors found or suspected to relate to our variables of interest (Abbeduto et al., 2004; Johnston et al., 2003; Seltzer et al., 2009).

Method

Participants

Families were drawn from a larger study examining the caregiving of adolescents and adults with FXS (MASKED). Inclusion required that the mother was the biological parent of a son or daughter with the *FMR1* full mutation. The sons and daughters with FXS were required to be 12 years of age or older, and to live in the parental home or to have at least weekly contact with their mothers. Documentation from a health care professional confirming that

the son or daughter had the full mutation of the gene causing FXS was also required. Recruitment occurred through service agencies, clinics, and foundations across the United States, and from a university-based research registry. If a mother had multiple children with FXS, she was asked to report on the child who was living with her. If more than one child with FXS was co-residing, the mother reported on the child who she believed was the most severely affected.

The larger study included 147 families; however, 25 of the mothers were unmarried or separated (19 divorced, 3 widowed, 2 never married, and 1 separated), leaving 122 married mothers. Six of these mothers exhibited either the full mutation ($n = 3$) or were mosaic ($n = 3$), based upon the number of CGG repeats identified. One mother tested within the normal range, with 30 repeats. The final sample therefore included 115 premutation mothers and their adolescent and adult children with FXS. Our analytic sample did not differ significantly from the group excluded on any variable of interest.

Individuals with FXS in the current sample ranged in age from 12 to 41 years ($M = 20.46$, $SD = 6.60$) and 86% were male. Eighty four percent held a diagnosis of intellectual disability (ID) as per mother report. Mothers reported that 33 individuals with FXS (29%) had been professionally diagnosed with an autism spectrum disorder (ASD), and record review supported the presence of ASD in all but 4 of these reports (see MASKED for procedure). Average maternal age was 50.45 years ($SD = 7.15$) and 60% of mothers held a 4-year college degree (all graduated high school). The majority of mothers (97%) identified themselves as Caucasian non-Hispanic and 81% of the mothers were married to the target child's biological father. Mean household income fell between \$80,000 and \$100,000, and mothers reported an average of between 1 and 2 children with a disability ($M = 1.75$, $SD = .74$, range 1 – 4; for FXS specifically: $M = 1.50$, $SD = .68$, range 1 – 4). Nearly all (90%) of the sons and daughters with FXS were living in the parental home.

Procedures and Measures

All procedures were conducted in accordance with our Institutional Review Board. Mothers provided data through questionnaires and telephone interviews that typically lasted one hour. Mothers also supplied medical or laboratory records of their CGG repeat length ($n = 30$) or provided a blood sample to Kimball Genetics, Inc. ($n = 85$) from which their repeat length was measured.

Child behavior problems—Behavior problems in the individuals with FXS were measured by the Total Problems scale from the age-appropriate version of either the Child or Adult Behavior Checklist (C/ABCL; Achenbach & Rescorla, 2001, 2003). Mothers reported on the degree to which their children exhibited certain behaviors over the past 6 months on a scale ranging from 0 (*not true*) to 2 (*very true*). The CBCL and ABCL are widely used and have demonstrated good reliability and validity in the general population (Achenbach & Rescorla, 2001, 2003), and for individuals with disabilities, including FXS (Hall et al., 2007; Hatton et al., 2002; Johnston et al., 2003; Tenneij & Koot, 2007; Wheeler et al., 2007). The CBCL and ABCL each produce standardized *T*-scores derived from age-based norms; thus scores from each form were comparable in that they reflected how the individuals with FXS functioned in relation to others in their age group (Achenbach & Rescorla, 2003). *T*-Scores of less than 60 fell within the *normal* range; those between 60 and 63 were considered *borderline*, and scores 64 and above were considered *clinically significant* (Achenbach & Rescorla, 2001).

Maternal internalizing symptoms—Mothers' depressive symptomatology was measured with the Center for Epidemiologic Studies Depression Scale (CES-D; Radloff,

1997). The CES-D asked participants to rate how often each of 20 statements applied to them within the last week, from 0 (*rarely or none of the time, less than 1 day*) to 3 (*most or all of the time, 5 – 7 days*). CES-D scores above 15 indicate clinically-significant depressive symptoms. The CES-D is reliable and valid, and has demonstrated convergence with other measures of depression. The 9-item tension-anxiety scale from the Profile of Mood States (POMS; McNair et al., 1981) was used to index maternal anxiety symptoms. Mood states of tension/anxiety were described and mothers rated how they had felt that week on scales ranging from 1 (*not at all*) to 5 (*extremely*). The POMS tension-anxiety scale has demonstrated high associations with other measures of anxiety including the State-Trait Anxiety Inventory (STAI; Spielberger et al., 1970). To our knowledge, the POMS anxiety scale does not have a formal clinical cut-off, but based on a standardization study by Nyenhuis and colleagues (1999), scores above 15.75 are thought to represent clinical significance for individuals over 55 years of age, with a higher threshold of 17.20 for younger groups. Maternal symptom data were normalized through square-root transformation prior to analysis due to positive skew. Maternal depression and anxiety symptoms were examined separately for initial analyses; however, these scores were combined (standardized and averaged together) for use in models due to the high comorbidity and shared clinical features of these problems (e.g., Klenk, Strauman, & Higgins, 2011; van Lang, Ferdinand, Ormel, & Verhulst, 2006), and the likelihood of a strong association between them (e.g., $r = .71$ has been reported for mothers of young children with FXS; Wheeler et al., 2007).

Marital satisfaction—Marital satisfaction was measured with 6 items from the 20-item Marital Satisfaction Scale of the Marital Satisfaction Questionnaire for Older Persons (Haynes et al., 1992). Mothers rated their satisfaction with particular areas of their marriage on a 6-point scale ranging from 1 = *very dissatisfied* to 6 = *very satisfied*. Only six items were chosen for the study in order to reduce respondent burden and because these six items demonstrated some of the highest factor loadings on the original scale (all $> .78$; Haynes et al., 1992). Within the current sample, principal component analysis revealed a single factor that explained 77% of the variance and all items demonstrated high loadings. The six items included mothers' satisfaction with the consideration shown by their spouse (loading = .91), the way disagreements were settled (.90), how decisions were made (.89), day-to-day support and encouragement (.86), spouses overall personality (.88), and how well the spouse listened to them (.83). Internal consistency for the 6-item scale in the current sample was $\alpha = .94$. The original Satisfaction scale has demonstrated good content validity, internal reliability, test-retest reliability (including on the item level), criterion validity, and construct validity (Haynes et al., 1992). Of note, the original scale has demonstrated moderate to high association with independent observations of marital behavior (Haynes et al., 1992).

Family cohesion—Family cohesion was measured with the Cohesion scale of the Family Environment Scale (FES; Moos & Moos, 1986). Mothers were asked to rate whether they felt that each of nine statements were *true* ("1") or not true (i.e., *false*, "0") of their family. Items included statements such as, "Family members really help and support one another," and "There is very little group spirit in our family." Items reflecting low cohesion were reverse coded and added to remaining items such that potential scores could range from 0 (low cohesion) to 9 (high cohesion). The FES-Cohesion scale has demonstrated validity in the general population (Moos, 1990) and has been used with families of children with disabilities (e.g., Floyd et al., 2004; Kelly et al., 2008), including FXS (Hall et al., 2007; Lewis et al., 2006). Internal consistency in the current sample was $\alpha = .77$.

Child intellectual disability, adaptive behavior, and autism symptoms—Mothers reported on whether or not their child had been diagnosed with intellectual disability, and

adaptive behavior of the individuals with FXS was measured for consideration in analyses. Mothers' completed the W-ADL (Maenner et al., under review), a 17-item measure of daily living skills (current $\alpha = .87$) that exhibits good psychometric properties, including strong associations with established measures of adaptive functioning in populations with ASD, and moderate relations with intelligence scores in children with ID and Down syndrome (Maenner et al., under review; Seltzer & Krauss, 1989). Scores on this measure converged well with mothers' reports of the children's ID diagnoses, with those who had ID having lower W-ADL scores than those who did not have ID $t = 2.81, p < .01, d = .53$. The adaptive behavior of males ($M = 22.76, SD = 5.63$) and females ($M = 25.44, SD = 4.69$) did not differ. Autism symptoms in the individuals with FXS were indexed using the Social Communication Questionnaire- current symptom version (Rutter et al., 2001), an autism screening measure that inquired about behavior related to the diagnostic criteria for ASD. Maternal responses for each item were scored dichotomously, 0 (*child does not display this behavior*) or 1 (*exhibits this behavior*) and the items were summed for the overall score. The current sample exhibited a mean on the SCQ of 19.71 ($SD = 7.18$), and scores corresponded strongly to supported maternal reports of ASD diagnoses, $t = 5.74, p < .001, d = 1.29$.

Data Analytic Plan

Correlations were examined first in order to understand potential direct associations between variables. A just-identified path model was then tested using Mplus v.5 (Muthén & Muthén, 2009) in which the behavior problems of the individuals with FXS predicted maternal internalizing symptoms, which, in turn, predicted marital satisfaction and family cohesion. Direct relations between behavior problems and the family relationship variables were also modeled. Finally, a pathway from marital satisfaction to family cohesion was included. An alternative model was also tested in order to assess the relative support for the hypothesized model in comparison to other possible structures. This model examined the indirect effects of behavior problems on maternal internalizing symptoms through the family relationship variables. Adequate fit for trimmed and/or alternative (i.e., not just-identified) models was concluded if the χ^2 divided by the degrees of freedom was under 2, the CFI was over .90, and the RMSEA was under .08.

Results

Descriptive Data

Descriptive data for the study variables of interest appear in Table 1. The average behavior problem score for individuals with FXS fell within the normal range ($T = 58.54$), slightly lower than, but remarkably consistent with, the average score reported for young children with FXS (61; Wheeler et al., 2007) and boys with FXS (60; Hatton et al., 2002). However, fewer than half (49%) of the individuals in our sample scored within the normal range of behavior problems, with a quarter (26%) scoring within the full clinical range.

Approximately one quarter (24%) of mothers scored within the clinical range for depressive symptoms, roughly consistent with previous reports based on parents of young children with FXS (Wheeler et al., 2007). Although the mean depressive symptom score for our sample was similar to that reported for the mothers of adolescents and young adults with FXS (11; Abbeduto et al., 2004), the variance of depression scores in the current sample was larger, resulting in a higher percentage of mothers scoring within the clinical range. Using the relevant age-based cut-offs for the POMS, approximately 12% of mothers appeared to have been exhibiting clinically-concerning symptoms of anxiety — somewhat lower than that reported for parents of young children with FXS (Wheeler et al., 2007).

Mothers in our sample reported rates of cohesion ($M = 7.22$) similar to previous studies of mothers of youths with FXS (Lewis et al., 2006), and higher cohesion than has been reported for families of children and adolescents in several psychometric evaluations of the FES scale (e.g., $M = 6.61$, Moos & Moos, 1986), families of children with disabilities ($M = 5.40$, Bourke-Taylor et al., 2009) and families of children with autism ($M = 6.5$, Kelly et al., 2008). Our use of an abbreviated scale precluded comparison to published marital satisfaction scores.

Mothers' CGG repeat lengths ranged from 67 to 180 ($M = 96.38$, $SD = 20.72$) and did not exhibit significant linear or quadratic associations with any variable of interest. As seen in Table 2, no demographic variable other than household income was related to the variables of interest in a manner that could confound findings. Additional demographics not included in Table 2 (e.g., whether father was the child's biological father) also were not significantly associated with the variables of interest. As predicted, maternal anxiety and depressive symptoms exhibited a high association, $r = .69$, so these variables were combined to form a single internalizing symptom variable. This composite was strongly correlated with anxiety and depressive symptoms ($r = .92$ and $.94$, respectively).

Correlations among Variables of Interest

Bivariate correlations demonstrated significant positive associations between behavior problems in the individuals with FXS and both maternal anxiety and depressive symptoms (and the internalizing composite; Table 2). Maternal internalizing symptoms were, in turn, negatively associated with both marital satisfaction and family cohesion. No direct association was found between behavior problems in the sons and daughters and either family relationship variable (marital satisfaction or family cohesion). None of the primary associations of interest differed as a function of residential status (see Table 2). The full sample was therefore used in model testing so as to maintain an appropriate participant-to-parameter ratio (Bentler & Chou, 1987).

Models and Indirect Paths

The hypothesized model was tested first, controlling for household income. Child age was also retained as a covariate given the large age range of the children. Mothers' CGG repeat length was not associated with any variable in preliminary models, so we did not include it in the final model. As seen in Figure 1, significant pathways were present from child behavior problems to maternal internalizing symptoms, and from internalizing symptoms to each family relationship variable. Indeed, indirect effects were present between children's behavior problems and both marital satisfaction, indirect effect = $-.09$, $z = -2.34$, $p < .05$ ($CI = -.160$ to $-.014$), and family cohesion, indirect effect = $-.08$, $z = -2.28$, $p < .05$ ($CI = -.150$ to $-.011$). Consistent with bivariate correlations, no direct effects were present between behavior problems and either of the family relationship variables. Examination of the hypothesized model with the direct pathways between behavior problems and family relationships omitted exhibited good fit, $\chi^2(2) = .15$, $CFI = 1.00$, $RMSEA = .00$. Of note, replacement of the directional path from marital satisfaction to family cohesion with a bidirectional path (i.e., correlating the error terms) did not significantly alter model findings, with the exception of a stronger pathway between maternal internalizing symptoms and family cohesion, $\beta = -.38$, $p < .001$, and a stronger indirect effect for family cohesion, $-.112$, $z = -2.66$, $p < .01$ ($CI = -.194$ to $-.029$).

The alternative model was then tested. This model included pathways between child behavior problems and each of the family relationship variables, which, in turn, predicted maternal internalizing symptoms. This model was not supported in that, consistent with the initial correlations, no direct associations between behavior problems and the family

relationship variables were significant (β s $< .11$, *ns*). Furthermore, marital satisfaction did not predict internalizing symptoms in this model ($\beta = -.12$, *ns*) and the direct association between behavior problems and maternal internalizing symptoms remained significant, $\beta = .22$, $p < .01$. This model with the direct effect trimmed exhibited poor fit, $\chi^2(1) = 7.00$, $p < .01$, CFI = .91, RMSEA = .23.

Discussion

Mothers of individuals with fragile X syndrome (FXS) are at increased risk for stress and poor psychological functioning (Abbeduto et al., 2004; Sarimski, 1997; Hartley, Seltzer, Hong, et al., in press; Seltzer et al., 2011; Wheeler et al., 2007). A critical task is to identify the contributors to distress among these mothers as well as to understand the potential systemic consequences of poor maternal mental health on affected families. Findings from the current study supported a model in which the behavior problems of adolescents and adults with FXS related to maternal internalizing symptoms, which, in turn, were associated with maternal report of both marital satisfaction and family cohesion.

Our findings suggest that established links between the behavior problems of individuals with FXS and maternal distress extend beyond childhood, and also suggest that indirect effects from behavior problems to family relationships may be present. These findings argue for a twofold approach to intervention, focused on reducing behavior problems in the individuals with FXS and on improving maternal mental health. Pharmacological interventions are the most common treatment for individuals with FXS, with some evidence of benefit; however, additional double-blind trials are necessary and gains to date have been modest (see Hall, 2009, for a review). As noted by Hall (2009) few studies (and primarily case reports) have focused on behavioral treatments aimed at improving functioning in individuals with FXS, despite the clear evidence for the efficacy of such treatments for children with similar disabilities (e.g., autism; Lovaas, 1987; Howlin et al., 2009). Recent behaviorally-oriented interventions are showing promise, but data on the degree to which these treatments may reduce behavior problems are not yet available (Hall, 2009).

Little is known regarding the effectiveness of psychopharmacological and/or psychosocial treatments for internalizing symptoms in mothers with the *FMR1* premutation who are caring for individuals with FXS. Extrapolation from research on the general population or even research on stressful caregiving may not be appropriate given evidence in the literature and from our laboratory demonstrating complex linkages between FXS-related genetic characteristics and maternal internalizing symptoms (MASKED, Bailey et al., 2008; Franke et al., 1998). Findings from the current study suggest that behavior problems of individuals with FXS may directly affect maternal mental health, but that these problems may not directly influence other aspects of family functioning. The indirect nature of these effects, through maternal well being, suggests that improving the manner with which mothers of individuals with FXS are supported, may be critical to buffering negative family response to child behavior problems.

A lack of direct association between behavior problems and family variables suggests potential resilience in these families. Indeed, comparisons to published values on the cohesion measure suggested that the families of adolescents and adults with FXS are no less cohesive than families of children and adolescents with other disabilities or with no developmental problems. This observation is even more striking given that mothers in the current sample exhibited high rates of internalizing symptoms and that these symptoms were associated with lower rates of cohesion. Taken together, these findings suggest that very high levels of cohesion may exist for intact families of individuals with FXS whose mothers remain low in internalizing symptoms. Indeed, mothers in the current sample who scored in

the lowest quartile of internalizing symptoms reported very high cohesion ($n = 27$, $M = 7.85$ on the 9-point scale, $SD = 1.83$). It is possible, however, that the co-residence or frequent-contact criteria for the current study may have reduced the involvement of less cohesive families; thus this finding of high cohesion may not generalize to all families of adults with FXS. Future studies would benefit from using the full Marital Satisfaction Questionnaire for Older Persons, so as to have the ability to compare the marital satisfaction of mothers of adolescents and adults with FXS with other populations. Findings from the current study do suggest, however, that poor maternal mental health may challenge the marriages of parents of individuals with FXS.

Although demographic variables were not the focus of the current investigation, it is notable that family income related to virtually every family variable of interest, and the associations were not explained by maternal education. Causality cannot be addressed in the current study; however, these findings suggest that financial resources may be closely tied to various dimensions of family functioning in this population as in the general population, and underscore the need to consider the social context in future research on, and in providing services to, families of individuals with FXS.

Limitations exist in the current study. Most significantly, the primary variables were based on maternal report, thus report bias is a concern; however, each of the family relationship measures has demonstrated agreement in past research with observational assessments (Haynes et al., 1992; Sanford et al., 1999), and the low correlations between mothers' reports of child behavior problems and their reports of other family factors argue against the presence of a global report bias. Nonetheless, observational and/or multi-informant report of marital quality and family cohesion would allow for stronger conclusions. Second, a longitudinal design would allow for clearer causal inferences. Indeed, bi-directional relations are possible, and a full reversal of the proposed model would result in an identical fit to the data. For theoretical, empirical, and practical reasons, we chose to combine maternal anxiety and depression, so future studies might consider examining these constructs separately. Similarly, we examined both internalizing and externalizing problems in the individuals with FXS. Although each of these types of problems is no doubt a source of stress for mothers, separate examinations may provide additional information. Many of our report measures asked about different periods of time (e.g., one week vs. 6 months), and increasing the consistency of the periods inquired about might produce more precise measurement. The lifespan orientation of the current study required consideration of the continuity between two versions of the behavior checklist forms, maintaining the developmental appropriateness of the measures but perhaps adding some degree of error. Although we examined correlational data separately for families with co-residing children versus the entire sample and found few differences, our final model depended upon the full sample for proper estimation. Finally, many of the effect sizes were modest, albeit in line with many of those found in examinations of family relations.

The current study suggests that established links between the behavior problems of individuals with FXS and maternal mental health persist into adolescence and adulthood. Results also indicate that maternal internalizing symptoms may play a pivotal role in links between child and family functioning, suggesting that reductions in behavior problems in individuals with FXS and decreases in maternal internalizing symptoms may produce cascading effects, promoting positive adaptation in these families more broadly.

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Figure 1.

Path model for the relations among behavior problems in the adolescents and adults with fragile X syndrome, maternal internalizing symptoms, and family factors, controlling for child age and household income. $+p < .10$, $*p < .05$, $**p < .01$, $***p < .001$

Table 1

Descriptive Data for Study Variables of Interest

Variable	Mean	SD	Range
Child Behavior Problems	58.54	7.15	44 to 76
Maternal Depressive Symptoms	10.69	9.41	0 to 49
Maternal Anxiety Symptoms	9.25	6.93	0 to 36
Maternal Internalizing Symptoms	0.00	0.89	−1.10 to 3.67
Family Cohesion	7.22	2.08	0 to 9
Marital Satisfaction	27.84	6.07	10 to 36

Table 2

Correlations among Variables of Interest and Demographics, for the Full Sample and for the Co-residing Sample Only

	1	2	3	4	5	6	7	8	9	10	11	12	13	14
1. Adapt. Behavior	--	.11	-.47***	.30**	n/a	-.17 [†]	.14	-.01	-.13	.00	-.07	.03	.01	-.11
2. Daughter ^a	.17 [†]	--	-.16 [†]	-.18 [†]	n/a	.07	-.01	-.04	.17 [†]	.07	.12	.01	-.04	.32**
3. Autism Sx	-.46***	-.19*	--	-.09	n/a	.12	-.09	-.13	.15	.02	.10	-.06	-.01	.21*
4. Child Age	.34***	-.08	-.08	--	n/a	.01	.06	-.06	-.03	-.07	-.04	.02	.05	-.26**
5. Not co-resident ^a	.23*	.21**	-.10	.38***	--	n/a	n/a	n/a	n/a	n/a	n/a	n/a	n/a	n/a
6. Child w/ Dis.	-.19*	-.01	.15	.00	-.06	--	-.10	-.14	.13	.12	.13	.17 [†]	.00	.06
7. Income	.13	-.05	-.10	.05	.04	-.08	--	.36***	-.19 [†]	-.36***	-.31**	.23*	.20*	-.04
8. Maternal Ed.	.06	-.03	-.15	.00	.21*	-.15	.34***	--	-.11	-.26**	-.22*	.06	.04	.11
9. Mom Anxiety	-.11	.13	.14	-.01	.00	.12	-.21*	-.09	--	.68***	.92***	-.24*	-.32**	.28**
10. Mom Depress.	.03	.02	.03	.02	.00	.10	-.38***	-.24*	.69***	--	.90***	-.42***	-.44***	.17 [†]
11. Mom Int. Sx.	-.05	.07	.10	.01	.00	.12	-.33**	-.20*	.92***	.90***	--	-.35***	-.42***	.22*
12. Marital Satis.	-.01	-.05	-.09	.02	.02	.18 [†]	.27**	.02	-.22*	-.39***	-.33***	--	.49***	-.09
13. Fam. Cohesion	-.02	-.03	-.05	-.01	-.02	-.04	.20*	.05	-.31**	-.41***	-.38***	.46***	--	-.09
14. Behavior Prob.	-.12	.19*	.25**	-.23*	-.18 [†]	.06	-.07	.06	.27**	.21*	.26**	-.12	-.10	--

Note: Correlations below the diagonal are for the entire sample ($n = 115$); those above the diagonal are for the coresiding group only ($n = 104$).^a1 = Yes.[†] $p < .10$.* $p < .05$.** $p < .01$.*** $p < .001$.

Adapt. = Adaptive, Sx = Symptoms, Child w/ Dis. = Number of children in the family with a disability, Ed. = Education, Depress. = Depression, Int. Sx = Internalizing Symptoms, Fam. = Family, Prob. = Problems.