Research in Developmental Disabilities 35 (2014) 2477–2486

Contents lists available at ScienceDirect

Research in Developmental Disabilities

Maternal well-being and child behavior in families with fragile X syndrome

Claire T. Hauser, Sara T. Kover, Leonard Abbeduto *

Waisman Center, University of Wisconsin-Madison, United States

ARTICLE INFO

Article history:
Received 23 January 2014
Received in revised form 8 June 2014
Accepted 12 June 2014
Available online 29 June 2014

Keywords:
Fragile X syndrome
Mental health
Maternal outcomes
Child behavior

ABSTRACT

The purpose of this study was to examine the bidirectional relationships relationship between maternal mental health status, maternal stress, family environment and behavioral functioning of children with fragile X syndrome (FXS), the leading cause of inherited intellectual disability. Children with FXS commonly demonstrate challenging behavior related to anxiety, attention, and aggression, whereas mothers of children with FXS have been identified as susceptible to mental health challenges due to their status as genetic carriers of the FXS premutation, as well as the environmental stressors of raising children with special needs. The longitudinal design of this study builds upon prior work that established a concurrent relationship among these factors in families of children with other intellectual disorders. Findings indicated that maternal mental health status was not significantly related to changes in levels of child challenging behavior, heightened child challenging behavior was related to improvements in maternal depression over time, and heightened levels of child challenging behavior was related to increased feelings of maternal closeness toward the child over time. The unexpected nature of the results regarding maternal depression and closeness provides new and more complex hypotheses about how mothers of special needs children demonstrate adaptation and resilience. The findings have implications for maternal and familial mental health treatment as well as future research.

1. Introduction

Fragile X syndrome (FXS) is the leading cause of inherited intellectual disability (Hagerman, 2008). The syndrome results from an expansion of a trinucleotide (CGG) sequence in the *FMR1* gene on the X chromosome, which leads to a deficit of FMRP, a protein that is essential for normal neural functioning (Bassell & Warren, 2008). In the full mutation case, which produces FXS, the CGG sequence is expanded to more than 200 repetitions compared to the healthy allele range of 15 to 54 repetitions (Brown, 2002). In the premutation case, the expansion is between 55 and 200 repetitions Brown, 2002). The premutation can result in both reduced FMRP levels and elevated levels of *FMR1* messenger RNA and possible RNA toxicity (Tassone et al., 2000). Although the premutation does not produce FXS, the premutation is associated with adverse phenotypic consequences, including comorbid conditions, such as the neurodegenerative disorder FXTAS (Cornish et al., 2008). In virtually all cases, FXS is inherited from the mother, who will be a carrier of either the *FMR1* premutation or full mutation (Nolin et al., 1996). Thus, FXS is a multigenerational disorder and the functioning of each family member is likely to

* Corresponding author. Present address: University of California, Davis MIND Institute, 2825 50th Street, Sacramento, CA 95817, United States. Tel.: +1 916 703 0234.
E-mail address: leonard.abbeduto@ucdmc.ucdavis.edu (L. Abbeduto).

http://dx.doi.org/10.1016/j.ridd.2014.06.012
0891-4222/

be affected by, and affect, the functioning of other family members (Seltzer et al., 2009). The aim of the present study was to characterize the dynamic bidirectional relationships that exist among child, mother, and family context over time in families affected by FXS.

1.1. Phenotype of children and youth with FXS

Most males with FXS have a moderate intellectual disability, and although females are less impaired on average, up to half also meet criteria for an intellectual disability (Hagerman & Hagerman, 2004). In addition to intellectual impairments, FXS is associated with an elevated rate of challenging behaviors relative to conditions such as Down syndrome, although there is considerable within-syndrome variability in this regard (Kau et al., 2004). Behavioral problems associated with FXS include social anxiety, hyperactivity, hypersensitivity to sensory stimuli, increased aggression, self-injurious behaviors, and attention problems (Kau et al., 2004; Kauffman et al., 2004). In addition, individuals with FXS commonly have impaired social and communicative skills (Abbeduto et al., 2004; Lewis et al., 2006) and other behaviors characteristic of autism (Bailey, Sideris, Roberts, & Hatton, 2008; McDuffie, Kover, Abbeduto, Lewis, & Brown, 2012). In fact, 25–33% of people with FXS meet criteria for a co-morbid diagnosis of autistic disorder, with the remainder displaying at least some autistic-like behaviors (Bailey et al., 2004; Brown et al., 1982; Demark, Feldman, & Holden, 2003; Lewis et al., 2006; Rogers, Wehner, & Hagerman, 2001). For males with FXS, a co-morbid autism diagnosis or more severe autism symptoms are associated with more problem and aberrant behavior (especially social avoidance and repetitive behavior), lower levels of adaptive behavior, more severe language impairments, and lower IQ scores relative to boys with FXS alone (Kau et al., 2004; McDuffie et al., 2010, 2012).

There is considerable evidence from studies of children with intellectual disabilities of various origins that child challenging behavior is a powerful predictor of maternal stress and poor mental health (Abbeduto et al., 2004; Roberts et al., 2009; Wheeler et al., 2010). At the same time, there is evidence that the child's challenging behaviors can be exacerbated by increased maternal stress and mental health problems, such as depression (Jouriles, Murphy, & O'Leary, 1989; NICHD, 1999; Orsmond et al., 2003; Osofsky & Thompson, 2000). In the present study, we were interested in the ways in which the challenging behavior of the son or daughter with FXS affects, and is affected by, the mental health of the mother as well as the family climate.

1.2. Maternal mental health

Biological mothers of individuals with FXS are at elevated risk for mental health concerns. In a sample of these mothers who had clinically significant levels of stress according to self-report, 63% also exceeded the clinical threshold on at least one other measure of maladaptive mental health (Bailey et al., 2008). Relative to women in the general population, biological mothers of children with FXS also display higher rates of social phobia, personality disorders (especially schizotypal personality disorder), major depressive disorder, panic disorder, and agoraphobia (Franke et al., 1998; Roberts et al., 2009). Depression and anxiety, however, are the most frequently diagnosed psychiatric disorders for women with the *FMR1* premutation (Franke et al., 1996). Rates of depression for women with the premutation have been identified as ranging from 16% to 40% (Bailey et al., 2008; Franke et al., 1996). Lifetime rates of depression have been cited at 56%, which is far higher than the 10–12% of women who experience depression in the general population (Wheeler, Hatton, Reichardt, & Bailey, 2007). The rate of current diagnoses of anxiety disorders in female carriers of the *FMR1* premutation has been found to be 17% (Bailey et al., 2008), and mothers of children with FXS have a frequency of anxiety disorders that is three times higher than that of mothers of children with autism and children who are typically developing (Franke et al., 1996).

It has been suggested that biological mothers of individuals with FXS are more susceptible to mental health problems in part because of their own genetic status as carriers of either the *FMR1* full mutation or premutation (Roberts et al., 2009). The evidence on this point, however, is equivocal. Thompson, Rogeness, McClure, Clayton, and Johnson (1996) found that mothers of individuals with FXS had a higher rate of depression (78%) than mothers of children with Down syndrome or spina bifida (37%) who, they argued, cope with similar environmental stressors. In fact, the rates of most types of challenging behaviors, including those reflective of externalizing problems, are higher in FXS than in Down syndrome, spina bifida, and many other syndromes (Dykens et al., 2000); thus, the Thompson et al. data are ambiguous as to the cause of maternal differences in mental health. In addition, Roberts et al. (2009) found that the age-of-onset for psychiatric diagnoses in mothers who were *FMR1* expansion carriers occurred much earlier than did their child's diagnosis, which led these investigators to suggest that the high prevalence of affective disorders among the mothers could not be attributed solely to the stress of raising a child with a developmental disability. However, Bailey, Raspa, Bishop, and Holiday (2009) found that the developmental problems of children with FXS are manifested and recognized by parents years in advance of the FXS diagnosis, which raises the possibility that child behavior and delays prior to diagnosis might still be contributing to maternal mental health problems. Nevertheless, relatives of mothers of children with FXS also have a higher frequency of affective disorders (20%) compared to the relatives of mothers of children with autism (11.7%) and relatives of mothers from the general population (3.3%; Franke et al., 1996), supporting the claim that being a premutation carrier confers risk for mental health problems in and of itself. It is likely, however, that it is the dual action of a genetic predisposition and the experiences associated with parenting a son or daughter with severe behavioral challenges that leads to less positive maternal mental health outcomes.