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Family functioning mediates adaptation in caregivers of individuals with Rett syndrome

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ABSTRACT

Objective: The objective of this study was to investigate factors related to family functioning and adaptation in caregivers of individuals with Rett syndrome (RS).
Methods: A cross-sectional quantitative survey explored the relationships between demographics, parental self-efficacy, coping methods, family functioning and adaptation. A forward-backward, stepwise model selection procedure was used to evaluate variables associated with both family functioning and adaptation. Analyses also explored family functioning as a mediator of the relationship between other variables and adaptation.
Results: Bivariate analyses ($N = 400$) revealed that greater parental self-efficacy, a greater proportion of problem-focused coping, and a lesser proportion of emotion-focused coping were associated with more effective family functioning. In addition, these key variables were significantly associated with greater adaptation, as was family functioning, while controlling for confounders. Finally, regression analyses suggest family functioning as a mediator of the relationships between three variables (parental self-efficacy, problem-focused coping, and emotion-focused coping) with adaptation.
Conclusion: This study demonstrates the potentially predictive roles of expectations and coping methods and the mediator role of family functioning in adaptation among caregivers of individuals with RS, a chronic developmental disorder.
Practice implications: A potential target for intervention is strengthening of caregiver competence in the parenting role to enhance caregiver adaptation.

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1. Introduction

Rett syndrome (RS) is a panethnic neurodevelopmental disorder, and is one of the major causes of severe intellectual disability in females, affecting approximately one in 10,000 [1–3]. RS is characterized by the loss of intellectual functioning, as well as fine and gross motor skills (e.g., purposeful hand use, communicative abilities) after a period of seemingly normal development. Individuals with RS can also experience a deceleration of head growth, stereotypic hand movements, seizures, respiratory dysfunction, and ataxic or apraxic gait [2,4]. During the rapid destructive stage of RS, which is usually initiated between one and four years of age, children can experience disturbing behavioral problems that can be very trying for their families including, for example, episodes of screaming, panic attacks, and self-injurious behavior [2,5]. Accordingly, caregivers of individuals with RS can face an array of psychosocial challenges.

It has been shown that parents of girls with RS experience more stress [6] and less satisfaction with their marital relationships [4] than controls. While one study reported normative levels of depression in mothers of girls with RS [6], most evidence suggests an elevated risk for depression and reduced health-related quality of life [7]. Another study showed that better mental health in mothers of girls with RS was associated with better family functioning [8]. Family functioning is a dynamic phenomenon that has been described as a family's ability to achieve goals integral to the lives of its members [9]. While there have been no studies of

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the relationships between parental self-efficacy, coping strategies, and adaptation in families of girls with RS, some of these associations have been established in studies looking at parents of children with other genetic disorders [10,11].

Thompson's Transactional Stress and Coping model [10,12–14], provides a useful framework for understanding how an individual adapts to a stressor within the family. The cognitive processes of appraisals and expectations, methods of coping, and family functioning are hypothesized to mediate adaptation to being a caregiver of an individual with a chronic condition in Thompson's model. There is not sufficient evidence for claiming path relationships among these constructs, but this model suggests a transactional rather than a unidirectional relationship in which each factor impacts the others over time. The objectives of the current study were to (1) investigate potential relationships among parental self-efficacy, coping methods, family functioning, and caregiver adaptation, and (2) evaluate family functioning as a mediator of the relationships between potential predictor variables and caregiver adaptation.

Per the model depicted in Fig. 1 [15], we specified the following hypotheses regarding relationships of other predictor variables and family functioning: higher levels of parental self-efficacy, a greater proportion of problem-focused coping, and a lesser proportion of emotion-focused coping will be associated with more effective family functioning (relationship *A*). Regarding relationships of predictor variables with adaptation, we made the following hypotheses: higher levels of parental self-efficacy, a greater proportion of problem-focused coping, and a lesser proportion of emotion-focused coping will be associated with greater caregiver adaptation (relationship *C*), as will more effective family functioning (relationship *B*). Finally, we hypothesized that family functioning would be a mediator of the relationships between adaptation and both parental self-efficacy and coping methods (relationship *C* vs. relationship *D*).

2. Methods

2.1. Sample

The study population consisted of men and women who self-reported as the primary caregivers of a child with RS, and who were able to read and write in English. Cohabitation with the child with RS was an additional inclusion criterion. To attain a diverse representation of the population, participants were recruited from four clinics across the eastern and southeastern United States, the 25th Annual Education & Awareness Conference for Rett Syndrome, and various RS organizations and websites. All interested individuals were invited to complete the paper surveys that were mailed to their homes or to visit the online version available on a SurveyMonkey® platform. Participants were compensated with a modest gift card. Informed consent was obtained from all participants, and the study was approved by ethics committees from all involved institutions.[1]

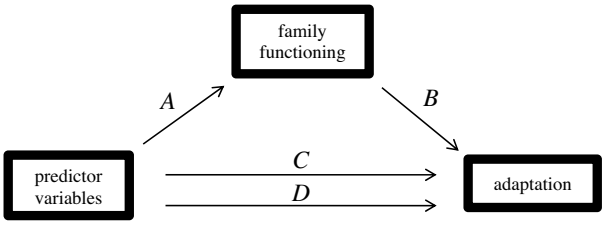

Fig. 1. This mediation model was adapted from Gelfand and colleagues [16]. Predictor variables include parental self-efficacy, problem-focused coping, and emotion-focused coping. *C* denotes the relationships between predictor variables and adaptation without family functioning included in the model, whereas *D* denotes the relationship including family functioning in the model.

2.2. Measures

This study used a cross-sectional research design with a quantitative survey that was available in both paper and electronic versions. The survey was pilot tested to determine the clarity and comprehensibility of questions, as well as to gain feedback on content. In addition to demographics questions, the survey instrument was composed of validated measures of parental self-efficacy, coping methods, family functioning, and adaptation.

Demographic Information collected included age, gender, race, ethnicity, marital status, education, income, perceived finance burden (extent of agreement with "Healthcare costs for RS are a financial burden to me and my family"), workload ("How much of the work do you share with other members in your family with regard to caring for you child with RS?" contrasting responses indicating all/most of the work with responses indicating some degree of sharing), health of caregiver (5-point item ranging from "poor" to "excellent"), amount of caregiver sleep per night, relationship to child with RS, number of children with RS, presence of children requiring special services, number and type of adults living in the household, frequency of information-seeking behavior (4-point item ranging from "never" to "regularly"), and use of support programs. The current age of the individual with RS and his/her age at diagnosis were also recorded. In addition, two 5-point items were used to assess the caregiver's subjective impression of the severity of their child's condition compared to children of the same age with RS ("not at all severe" to "very severe") and without RS ("doing a lot worse" to "doing a lot better"). This information was used to evaluate these variables' potential as confounders in linear regression analyses, as well as to assess external validity.

Parental Self-Efficacy was assessed using the Efficacy component of the Parenting Sense of Competence Scale (PSOC) [16], which indicates the degree to which the parent feels competent, capable of problem solving, and familiar with parenting. Each of the seven items allowed for responses on a 6-point scale ranging from "strongly disagree" to "strongly agree." This subscale has previously been applied as an independent measure, with a reported internal consistency of 0.71 [17]. Summed scores could range from 7 to 42, with higher scores indicating greater parental self-efficacy.

Coping Methods were assessed using the Ways of Coping Checklist-Revised (WCC-R) [18]. Respondents were asked to estimate how often they use each of the 42 coping strategies, classified under the following five domains: problem-focused, wishful thinking, seeks social support, blamed self, and avoidance. This scale has been shown to be valid and reliable with Cronbach's alphas of the subscales ranging from 0.73–0.88 [18]. As proposed by Zakowski and colleagues [19], the blamed self, wishful thinking, and avoidance subscales were combined into one emotion-focused coping score. Finally, the two proportions were calculated by dividing the score sums of problem- and emotion-focused coping separately into the total sum of all coping strategies used.

Family Functioning was assessed using the Family Assessment Measure III (FAM-III). The FAM-III is based on the Process Model of

[1] This study was approved by the National Human Genome Research Institute (NHGRI) Institutional Review Board (IRB; Protocol # T-HG-0021), the Johns Hopkins Bloomberg School of Public Health IRB (Protocol #: IRB00001946), the University of Alabama at Birmingham IRB (Protocol #X090417004), the Self Regional Healthcare IRB (IRB associated with Greenwood Genetic Center), and the Baylor College of Medicine (Protocol #H-14884). Approval was also obtained from the research compliance reviewer at the Kennedy Krieger Institute.

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