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Seizures and Pain Uncertainty Associated with Parenting Stress and Rett Syndrome

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Abstract

Data were collected parenting stress, adaptive behavior, pain, and health issues from the caregivers of 35 girls and women with Rett syndrome (mean age = 20.3). A majority (60%) of parents reported stress in the clinical range on at least one subscale of the Parenting Stress Index – Short Form. Seizures and uncertainty about their daughter's gastrointestinal pain experience were significantly associated with higher levels of parenting stress. No other child factors (adaptive behavior, age, residential status) were significantly related to parenting stress. Factors related to chronic health concerns (seizures, ambiguous pain presentation) may be important when considering family stress issues in relation to general outcomes for girls with Rett syndrome and related developmental disorders.

Keywords

Rett syndrome; Parenting stress; Pain; Seizures

Parents of children with chronic medical issues associated with neurodevelopmental disabilities are at increased risk for experiencing prolonged periods of stress,[1–5] the negative effects of which have been well documented. 6–8 [10,11] Parenting stress may also have indirect effects on the health of the child through decreased medical intervention adherence.[12–15] Therefore, identifying factors that may predict levels of parenting stress

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among families with children with neurodevelopmental disabilities with specialized health needs may be helpful in identifying parents who may need additional assistance in maintaining appropriate medical treatment regimens.

Caregivers of individuals Rett syndrome face stressors related to medical issues and developmental concerns. Rett syndrome is a severe neurodevelopmental disorder that affects primarily females. Following often normal peri-natal development, girls with Rett syndrome show a loss of communication and motor skills, and the development of stereotypical hand movements.[16] A number of health problems, including seizures, scoliosis, and apnea are common in this population.[17] Due to the pervasive nature of the effects of the syndrome, individuals with Rett syndrome typically require life-long assistance in all areas of their lives.

Few studies have examined the effects of caring for a child with Rett syndrome on parents' stress and health. In one previous study, the parents (N=29) reported higher levels of parenting stress than parents of typically developing children, although most parents still scored within the normal range.[18] None of the reported characteristics of the individuals with Rett syndrome (e.g., age, adaptive behavior) were found to be significantly related to parenting stress in their sample. A second study found that, although most child factors were unrelated to parenting stress, breathing abnormalities in the children were related to lower physical, but not mental health scores among parents, whereas bone fractures in the preceding two years and the presence of stereotyped facial movements were related to lower mental health scores only. Neither of the previous two studies examined parenting stress in relation to pain or other specific health issues common in Rett syndrome. Therefore, the goal of the current preliminary study was to revisit the issue of parenting stress and its relation to pain and health issues experienced by individuals with Rett syndrome.

Methods

Subjects

Forty-five survey packages were returned from a 1-time anonymous survey through regional Rett syndrome parent groups in the upper Midwest (20.3% response rate). Five families contacted the research team to indicate that they did not have a child with Rett syndrome. Of the returned surveys, 35 (78%) had completed the Parenting Stress Index, Short Form. The other measures (described below) were returned by all of the respondents. All of the respondents were parents or guardians for girls and women with clinical diagnoses of Rett syndrome. Individuals across the lifespan were represented in the sample, with ages ranging from 2 to 49 years (mean = 20.3). The majority (30, 85.7%) of the surveys were completed by the mother, three (8.6%) by the father, and two (5.7%) by foster parents. Twenty-six (74.3%) of the females with Rett syndrome lived with the caregiver who completed the survey, and the remaining nine (25.7%) lived in group homes or nursing facilities. All study procedures were approved by the University of Minnesota and Gillette Children's Specialty Healthcare human subjects review committees.

Measures

Parenting stress was measured using the Parenting Stress Index, Short Form[20]; a 36-item parent-report scale that provides a total score and three subscale scores (Parental Distress; Parent-Child Dysfunctional Interaction, and Difficult Child).

Adaptive behavior and maladaptive behavior were measured using the Inventory for Client and Agency Planning[21], a 16-page, self-administered structured questionnaire including four adaptive behavior subscales (Motor Skills; Social and Communication Skills; Personal Living Skills; and Community Living Skills), and a total adaptive behavior score.

Pain and health problems were measured by the Rett Syndrome Health Questionnaire, a questionnaire developed by the researchers to assess current and past health problems experienced by females with Rett syndrome. It consisted of (a) a list of health problems common to individuals with Rett syndrome (e.g., seizures, scoliosis, appetite problems), and (b) a list of sources of pain (e.g., gastrointestinal; therapy-related), and questions regarding the frequency and severity of the pain experienced by the individual with Rett syndrome (with items modified from the Non-Communicating Children's Pain Checklist – Revised).[21]

Data Analysis

Due to the relatively small sample size and non-normality of the variables examined, nonparametric statistical tests were used for all analyses. Missing data were excluded from the analyses. Analyses were conducted to determine whether the individuals who did not complete the Parenting Stress Index differed from the rest of the sample. Results suggested that there were no differences in the age, residential placement, adaptive behavior of the individual with Rett syndrome, or the relationship of the respondent to the individual.

Results

Parenting Stress

Scores above the 85th percentile of the normative sample on any subscale of the Parenting Stress Index are considered to be clinically significant. Of the 35 caregivers that completed the scale, 21 (60%) met criteria for clinically significant levels of parenting stress on at least one subscale. Fifteen (43%) fell into the clinically significant range for the total parenting stress score, and 20 (57%) had clinically significant scores on the difficult child subscale (see Table 1).

No significant relationships between parenting stress scores and the age, residential status (e.g., with parents vs. in nursing home), relationship of the respondent to the affected individual (e.g., mother vs. father vs. foster parent), or adaptive behavior scores were found.

Pain and Health Issues

As expected, a large proportion of the sample reported the presence of medical and related health issues including scoliosis (26; 59%), sleep problems (22; 50%), seizures (20; 47%), and appetite problems (15; 34%). Caregivers whose children had diagnosed seizure disorders had significantly higher scores on the parental distress (Mann-Whitney $U = 66$, p

= .007), parent-child dysfunctional interaction ($U = 83$, $p = .040$), difficult child ($U = .66$ $p = .007$), and total stress scales ($U = 59$, $p = .005$). No other health problems were significantly related to parenting stress scores.

A significant omnibus result showed a difference between groups (‘yes’, N = 22; ‘no’, N = 5; ‘unsure’, N = 8) reporting gastrointestinal pain on the Parent-Child Dysfunctional Interaction subscale. Post hoc analyses showed that there were no significant differences between the ‘yes’ and ‘no’ groups, or between the ‘unsure’ and ‘no’ groups. Those who responded ‘unsure’ had significantly higher scores on the Parent-Child Dysfunctional Interaction subscale than caregivers who answered ‘yes’ ($\chi^2_{(2)}= 7.257$; $p = .027$). Gastrointestinal pain was not related to any of the other parenting stress scores. Similarly, all other pain sources, pain frequency and pain severity were unrelated to parenting stress in this sample.

Discussion

In the present sample, the majority of caregivers scored in the clinical range for parenting stress on at least one subscale. General child factors (e.g., age, residential placement, adaptive behavior) were unrelated to parenting stress scores. These results suggest that the demands of caring for an individual with Rett syndrome put parents and caregivers at risk for elevated parenting stress across the lifespan, and that this stress is not necessarily reduced/relieved when the parents or guardians are no longer the day-to-day caregivers.

Among the numerous health issues reported, only seizures were significantly related to parenting stress in this sample. These results suggest that the presence of a seizure disorder may further add to the stress experienced by caregivers of Rett syndrome. Although preliminary, this finding emphasizes the importance of identifying and addressing comorbid health issues, which may have an impact on the quality of life of children and their families, beyond the effects of Rett syndrome more generally.

Two previous studies[1,2] linked epilepsy to elevated parenting stress among parents of otherwise typically-developing children. To our knowledge, however, no other studies have examined the link between seizures and parenting stress in individuals with neurodevelopmental disabilities. Because seizure management requires reliable administration of medication, and previous studies have shown a link between elevated parenting stress and decreased medical adherence, further studies are warranted explicitly examining whether parental stress among Rett syndrome families is related to parental adherence to prescribed medical treatments for their daughters.

With the exception of parent-reported gastrointestinal pain, the frequency, severity, and sources of pain were unrelated to parenting stress scores. Interestingly, it was not the parents who reported that their daughters experienced gastrointestinal pain that reported higher levels of stress, but those who reported that they were unsure of whether or not their child was in pain. This finding raises an important issue regarding the assessment of pain in individuals with Rett syndrome and more generally individuals with neurodevelopmental disabilities and significant communicative or motor deficits.

Several limitations of this small-scale study should be noted. First, the limited response rate, and the missing Parenting Stress Index forms may have biased the results in some way. Our survey return rate of a little over 20% is a fairly typical response rate for a 1-time anonymous survey with no follow up. Although low response rates do not guarantee less accurate results, (see research regarding survey sampling, response rates, and accuracy comparisons for small and large return rates[26]), it would be prudent to consider the results as preliminary and specific to this sample. In addition, all data were collected via caregiver report, making them susceptible to over- or under-estimation by the respondents. Given the topic of the study, however, parental perceptions of behavior and health may be more relevant to the caregivers' stress than more seemingly objective measures. In addition, the small sample size precluded more complex statistical analyses that may have revealed multifaceted relationships between factors.

Despite the limitations, the results suggest several variables that could be tested in future larger studies as potential risk factors for elevated parenting stress among caregivers of children with Rett syndrome and other neurodevelopmental disabilities. We think that further work investigating what is and is not related to parental stress – and why - among families with daughters with Rett syndrome has important implications for both affected individuals and their caregivers. Identifying specific predictive factors (like seizures) may be helpful in identifying families that require additional assistance in both managing their own physical and mental health status[27], and in adhering to the numerous pharmacological and behavioral therapies required to manage their child's various health issues.

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Table 1

Parenting stress scores as measured by the PSI-SF

Statistic	PD	DC	P-CDI	Total
Minimum	12	12	13	40
Maximum	58	53	47	148
Median	25	26	28	79
# clinically sig.	6	20	14	15
% clinically sig.	17%	57%	40%	43%

Note: PD = Parental Distress; DC = Difficult Child; P-CDI = Parent-Child Dysfunctional Interaction; Total = Total Parenting Stress. Scores are considered clinically significant if they are above the 85th percentile of the norming sample of the PSI-SF.