Parental Distress, Family Functioning, and Social Support in Families with and without a Child with Neurofibromatosis 1*

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Objective To compare parental adjustment, social support, and family functioning between families of children with neurofibromatosis 1 (NF1) and a group of demographically similar comparison families, and to examine the impact of disease severity. Methods Questionnaires were completed at home by parents of 54 children with NF1 (54 mothers and 42 fathers) and 51 comparison children (49 mothers and 32 fathers).

Results Few differences between groups were identified for parental distress, social support, or family environment. Greater neurological impairment in children with NF1 was associated with greater distress, more family conflict, less positive mealtime interactions, and less social support from the perspectives of mothers. Conclusions Overall, parents of children with NF1 appear similar to parents of comparison children. Mothers who have children with NF1 characterized by greater neurological impairment may be at risk for more difficulties. Future work exploring long-term adjustment for these mothers as well as interventions to ameliorate any potential difficulties may be appropriate.

Key words disease severity; family functioning; neurofibromatosis; parents.

Neurofibromatosis (NF1), an autosomal dominant genetic disorder, has a prevalence rate of 1/2,000 to 1/5,000 (Rasmussen & Friedman, 2000). Clinical features include café-au-lait spots, neurofibromas, optic nerve tumors, and osseous dysplasias. NF1 is a progressive disorder, with additional complications such as vascular abnormalities, cosmetic disfigurement, headaches, and motor coordination difficulties likely to develop (Riccardi & Eichner, 1986).

Pediatric chronic conditions such as NF1 have the potential to adversely affect the adjustment of parents and the family. Mothers of children with chronic illness may be especially at risk as they often bear the primary responsibility for their child’s daily care (Quittner et al., 1998). Concern about the child’s well-being, medical challenges, and fear for the future can put strain on the psychosocial and financial resources of a family. Parents of children with NF1 must cope with possible life-threatening complications for their child, cosmetic deformities, visual impairment, orthopedic complications that may require repeated surgeries, learning problems, and difficulties with attention. Physical challenges and cognitive risk (Hyman, Shores, & North, 2005) may result in an uncertain future for children with NF1. As adults, individuals with NF1 have been described as experiencing numerous psychiatric problems, including depressive and anxiety problems (Samuelsson & Riccardi, 1989). In addition, Zoller and Rembeck (1999) reported that a high proportion of adults with NF1 were diagnosed with psychiatric or affective disorders, especially dysthymia.

Although no studies were identified that specifically examined parental adjustment or family functioning in response to having a child with NF1, considerable research has focused on the functioning of parents of children with chronic conditions. Results have been mixed. Some work has suggested minimal increased risk for parental distress.
(Daltroy et al., 1992; Greenberg, Kazak, & Meadows, 1989; Noll et al., 1994); parents in other studies have reported symptoms of depression, anxiety, and posttraumatic stress (Barakat et al., 1997; Canning, Harris, & Kelleher, 1996; Kazak et al., 1997; Noll et al., 1995). Several studies have suggested that though the majority of parents report minimal difficulties, a subset may experience clinically significant levels of distress (Gerhardt et al., 2003; Manne et al., 1996; Quittner et al., 1998; Shore, Austin, & Dunn, 2004).

When examining family functioning, several studies have indicated few differences between families of children with chronic conditions and comparison families (Gerhardt et al., 2003; Greenberg, et al., 1989; Kazak et al., 1997; Noll et al., 1994, 1995). In contrast, other research has found that parents of children with chronic conditions describe their families as rigid and low in flexibility relative to comparison families (Kazak & Meadows, 1989; Madan-Swain et al., 1994).

A number of factors have been explored that may account for variability in the adaptation of parents and families with a child with a chronic illness, including social support and child disease severity. For instance, difficulties may be exacerbated when there is a lack of social support available to the family (Manne, Duhamel, & Redd, 2000). In particular, some work has indicated an impact of social support on psychosocial functioning only for mothers (Barakat et al., 1997) or only for fathers (Wijnberg-Williams, Kamps, Klip, & Hoekstra-Weebers, 2006). Although research regarding the impact of child disease severity or functional impairment has been mixed (Gerhardt et al., 2003, Lustig, Ireys, Sills, & Walsh, 1996; Lutz, Barakat, Smith-Whitley, & Ohene-Frempong, 2004), chronic conditions that affect neurological functioning may be particularly challenging for parents as these conditions have been associated with increased behavioral difficulties in children (Vannatta, Gerhardt, Wells, & Noll, 2007). Thus, it is possible that families of children with NF1 who experience greater neurological involvement may be especially at risk for increased parental distress and family difficulties.

The literature on the psychosocial functioning of parents and families of children with chronic illness is limited by a number of methodological problems that have made findings difficult to interpret. Such problems have included a failure to report recruitment rates, lack of comparison groups, minimal data from fathers, and a lack of standardized measures. In addition, a reliance on data obtained during clinic visits, in hospital settings, or using disease-specific questionnaires may create a focusing illusion or misallocation of attention (Kahneman, Krueger, Schkade, Schwarz, & Stone, 2006; Schkade & Kahneman, 1998) on disease status and impairment (Smith, Schwarz, Roberts, & Ubel, 2006).

This study examined parental distress, social support, and family functioning (both general and at mealtimes) in families of children with NF1 as well as the impact of disease severity on these variables. We sought to evaluate general family functioning as well as family functioning at mealtimes, since previous work by family systems clinicians (Ackerman, 1958) had suggested that home visits to observe mealtime interactions are a sensitive metric of general family functioning (Satter, 1999). A group of families without a child with chronic illness who were demographically similar to the families affected by NF1 was included as the comparison point to clarify the impact of NF1 on the outcomes of interest. Data from both groups of families were obtained at the family’s home using psychometrically sound measures. In addition, some researchers have cautioned that the unique perspectives of fathers are rarely represented in the pediatric psychology literature, or are obscured by failing to examine maternal and paternal effects separately (Phares, Lopez, Fields, Kamboukos & Duhig, 2005). Thus, we attempted to include both mothers and fathers. We expected that: (a) parents of children with NF1 would report greater distress, more family difficulties, and less social support relative to comparison families; (b) greater social support would be associated with less parental distress, and this association would be stronger for parents of children with NF1; and (c) disease severity would be positively associated with parental distress and family difficulties. Finally, exploratory analyses examined whether family functioning mediated the association between disease severity and parental distress.

Method
This study was part of a larger project examining the adjustment of children with NF1 and their families. The first phase included data collection on peer relationships in the classroom, followed by home-based data collection with families of children with NF1 and comparison peers. This study focused on parent adjustment and family environment data from the home-based assessment and was approved by the local institutional review board.

Participants
Children with NF1, ages 7–15 years, who received care at a NF clinic in a Midwestern children’s hospital were identified via medical records. Those in full-time special
education were excluded, resulting in 65 eligible children. Children receiving services for learning disabilities, including those in part-time special education services, were eligible. Of these 65 children, two families and four schools declined the school-based assessment, and five additional families declined the home visit, leaving 54 (83%) families. Data were obtained from 54 mothers and 42 fathers of children with NF1.

The family of the classmate who was of the same gender and race and who was closest in birth date to the child with NF1 was contacted to participate. If they declined, the family of the child whose birthday was next closest was called. Forty-three (84%) families were first choice comparison families. All were screened to ensure that none had a child with a severe chronic illness. Data were collected from 51 comparison families (49 mothers and 32 fathers). All female caregivers (e.g., biological mothers, stepmothers, grandmothers, aunts, etc.) in the current study are referred to as “mothers” and all male caregivers (e.g., biological fathers, stepfathers, grandfathers, and uncles) are called “fathers.”

Procedure
Following written informed consent, each parent completed questionnaires independently in the home with staff assistance. All families were compensated for their time.

Measures

Demographic Background Questionnaire
This instrument assessed background characteristics of the family (e.g., parental age, education, marital status, income, and number of children in the home) (Noll et al., 1996). Socioeconomic status (SES) was determined using the Revised Duncan (Nakao & Treas, 1992), an occupation-based measure of SES. Use of occupation-based measures of SES have been recommended because these measures have been found to provide a stable indicator of SES that is congruent with current practice at the Bureau of the Census and this information can be obtained quickly from families (Entwisle & Astone, 1994; Hauser, 1994).

Symptom Checklist 90-Revised (SCL-90-R)
This 90-item self-report measure assessed frequency of psychological symptoms over the past 2 weeks (Derogatis, 1983). Nine dimensions of psychological distress and three global scales [i.e., Positive Symptom Distress, Positive Symptom Total, and Global Severity Index (GSI)] are derived. The GSI, considered the best summary measure of distress, includes the number and intensity of symptoms experienced. This measure has adequate reliability (Derogatis, 1983), and has been used to assess distress in studies of parents of children with chronic illnesses (Gerhardt et al., 2003; Levers, Brown, Lambert, Hsu, & Eckman, 1998; Noll et al., 1995; Wijnberg-Williams et al., 2006).

Family Environment Scale (FES)
Parents completed this 90-item measure assessing the social environment of the family. The FES measures 10 dimensions of family functioning, each of which has shown adequate psychometric properties (Moos & Moos, 1994). However, four higher-order scales [e.g., Family Relationship Index (FRI), Supportive, Conflicted, and Controlling] have been shown to be useful with families of children with chronic illness and have demonstrated good validity (Kronenberger & Thompson, 1990; Moos & Moos, 1994). This instrument has been widely used in studies of families of children with chronic illness (Gerhardt et al., 2003; Noll et al., 1994, 1995; Phipps, Dunavant, Lensing & Rai, 2005; Thompson et al., 2003).

Norbeck Social Support Interview (NSSI)
Parents were interviewed separately about the number of significant individuals in their lives (network size) and their satisfaction with support from each (perceived functional support) using six structured questions (Norbeck, Lindsey, & Carieri, 1981). This measure has satisfactory reliability and validity (Gigliotti, 2002; Norbeck & Anderson, 1989; Norbeck, Lindsey, & Carieri, 1983), and has been used previously with parents of children with chronic illness (Florian & Krulik, 1991; Gerhardt et al., 2003; Noll et al., 1995).

About Your Child’s Eating-Revised (AYCE-R)
This measure assesses parent’s concern and satisfaction with their child’s nutrition, as well as perceptions of family mealtime interactions (Davies, Ackerman, Davies, Vannatta, & Noll, 2007). Three factor-derived scales include: (a) Child Resistance to Eating, (b) Positive
Mealtime Environment, and (c) Parent Aversion to Mealtime. Adequate convergent validity and internal consistency for these scales have been demonstrated ($\alpha = .72–.89$) (Davies et al., 2007; Davies, Noll, Davies, & Bukowski, 1993). This measure has been found to be significantly associated with several indices of family functioning (Davies et al., 2007) and was included in the current study to describe family functioning specifically during mealtimes.

Medical Chart Review
A clinical geneticist rated children with NF1 on three severity scales (Table I) during a medical chart review of the clinic visit closest to the home-based data collection. Six children were not rated, because there was no clinic visit within 1 year of the data collection, and one child’s medical chart was unavailable. First, a general medical severity scale (Riccardi, 1982) was used to assess overall disease severity. Second, an appearance scale was used to rate the visibility features of the disease including neurofibromas, café-au-lait spots, and facial or skeletal abnormalities (adapted from Ablon, 1996). Finally, a neurological impairment scale was based on neurological examination; presence of headaches, seizures, or vision impairment; learning difficulties, behavior problems, and brain tumors. Chart review included records of formal testing by schools or psychologists for learning disabilities, attention deficit hyperactivity disorder (ADHD), and cognitive function for many, but not all, of the children. Scales ranged from “1” minimal to “4” severe.

These scales were previously validated for reliability independently by scoring patients from chart records. Specifically, a total of 22 charts (not participants in the current study) were reviewed by two physicians familiar with NF1; patients were independently scored on each of the severity scales (Medical, Appearance, and Neurological). Scales were clarified somewhat from the initial version to make them less subjective, with the final version giving 84% agreement for scores on the three severity scales (Schorry et al., unpublished data). In no case did the score for one of the severity scales differ by more than one point between the two physicians. A later version of this scale was published, where the neurological impairment scale was replaced by separate developmental and behavioral scales, with 87% agreement between the same two physicians for the four scales (Sebold, Lovell, Hopkin, Noll, & Schorry, 2004).

For medical severity, 17 (36%) children had minimal disease, 19 (40%) had mild disease, 10 (21%) had moderate disease, and 1 (2%) had severe disease. For the appearance scale, 30 (64%) children had a minimal rating, 13 (28%) had a mild rating, 1 (2%) had a moderate

<table>
<thead>
<tr>
<th>General*a</th>
<th>Appearance*b</th>
<th>Neurological</th>
</tr>
</thead>
<tbody>
<tr>
<td>Minimal</td>
<td>Few NF1 features, café-au-lait and freckling only, with no compromise of health</td>
<td>No visible tumors outside of clothing, normal gait and posture</td>
</tr>
<tr>
<td>Mild</td>
<td>Mild hypertension, asymptomatic tumors (plexiforms or optic glioma)</td>
<td>Many café-au-lait spots, less than 25 cutaneous neurofibromas, normal gait, and posture</td>
</tr>
<tr>
<td>Moderate</td>
<td>Orthopedic complications requiring bracing or surgery, large or symptomatic plexiforms, and moderate pain</td>
<td>25–75 cutaneous neurofibromas, many visible. Visible skeletal feature with moderate impact</td>
</tr>
<tr>
<td>Severe</td>
<td>Intractable seizures, severe chronic pain, visual impairment, inoperable tumors, and malignancies</td>
<td>More than 100 cutaneous neurofibromas, including face, facial or orbital plexiforms, severe scoliosis, tibial dysplasia requiring bracing, or associated with limp</td>
</tr>
</tbody>
</table>

*aBased on Riccardi, 1982.
*bBased on Ablon, 1996.

While we have used the term neurological impairment to describe our scale here, we have used neurological severity to describe this scale in previously published work (Noll et al., 2007). The actual scales are identical.
rating, and 3 (6%) had a severe rating. Finally, for the neurological impairment scale, 7 (15%) children had minimal impairment, 20 (43%) had mild impairment, 19 (40%) had moderate impairment, and 1 (2%) had severe impairment.

Analyses

Parental adjustment, family environment, mealtime climate, and social support were compared between groups (NF1 and comparison) using two-tailed, independent t-tests (α = .05). Pearson correlations were used to examine associations between disease severity and parental and family functioning. Hierarchical multiple regressions examined the impact of social support on parental distress (GSI). Group status (NF1 vs. comparison) was entered in the first step followed by either network size or perceived functional support in step 2. For step 3, the interaction of group with support was entered. All predictor variables were centered, and the interaction term was formed by multiplying the centered variables (Holmbeck, 2002).

Hierarchical multiple regression analyses were also used to examine whether family functioning mediated the association between disease severity and parental distress (Holmbeck, 2002). Family functioning variables were considered potential mediators if they demonstrated a significant correlation with the independent variable (e.g., disease severity) and the dependent variable (e.g., GSI). The significance of the indirect effect was evaluated with a bootstrapping procedure, using a macro for SPSS downloaded from http://www.comm.ohio-state.edu/ahayes (Preacher & Hayes, 2004).

Power analyses using GPOWER (Faul & Erdfelder, 1992) indicated that the sample of 103 mothers (α = .05) produced 71% power and the sample of 74 fathers (α = .05) produced 56% power to detect a medium effect size (d = 0.5) for t-tests. For correlations involving disease severity measures, the sample of 47 mothers and 37–38 fathers provided 46–56% power to detect a medium effect size (r = 0.3). The number of participants varies slightly between measures because not all individuals completed all items from all questionnaires.

Results

Demographic and Background Characteristics

Families of children with NF1 and comparison families did not significantly differ on a number of demographic variables (Table II). Eighty percent of mothers of children with NF1 and 86% of comparison mothers were married, χ² (1, n = 104) = 0.74, NS.

<table>
<thead>
<tr>
<th>Variable</th>
<th>NF1</th>
<th>Comparison</th>
</tr>
</thead>
<tbody>
<tr>
<td>Family SES³</td>
<td>52.20</td>
<td>50.24</td>
</tr>
<tr>
<td>Number of children in home</td>
<td>2.50</td>
<td>2.40</td>
</tr>
<tr>
<td>Father education⁴</td>
<td>13.98</td>
<td>13.34</td>
</tr>
<tr>
<td>Mother education</td>
<td>13.70</td>
<td>13.48</td>
</tr>
<tr>
<td>Father’s age (years)⁵</td>
<td>43.70</td>
<td>42.47</td>
</tr>
<tr>
<td>Mother’s age (years)⁵</td>
<td>39.45</td>
<td>38.28</td>
</tr>
<tr>
<td>Child’s age (years)⁴</td>
<td>10.98</td>
<td>10.98</td>
</tr>
<tr>
<td>Child’s estimated IQ⁶</td>
<td>95.15</td>
<td>98.41</td>
</tr>
</tbody>
</table>

³The number of participants varies between measures because not all individuals completed all items from all questionnaires.
⁴All t-values are nonsignificant, two-tailed tests.
⁵SES, Socioeconomic status. Higher scores indicate greater occupational status.
⁶nNF1 = 43; nCOMP = 38.
⁷IQ estimates are based on the Vocabulary and Block Design subtests of the WISC-R.

Parental Distress

No significant differences were identified between groups (NF1 vs. comparison) for mothers or fathers (Table III) on the SCL-90-R. The number of mothers and fathers exceeding the clinical cutoff for the SCL-90-R (i.e., a T-score > 63 for the GSI or at least two subscales) was also examined. Twenty-eight percent (n = 15) of mothers of children with NF1 and 24% (n = 12) of comparison mothers had clinically significant distress, χ² (1, n = 103) = 0.10, NS. Similarly, 34% (n = 14) of fathers of children with NF1 and 47% (n = 15) of comparison fathers exceeded the clinical cutoff, χ² (1, n = 73) = 1.04, NS.

Family Environment and Mealtime Climate

Family environment and mealtime climate were similar between groups with one exception; mothers of children with NF1 reported greater child resistance to eating relative to comparison mothers (Table IV). Pearson correlations between mother and father report of corresponding family functioning variables ranged from low to moderate in families of children with NF1 (r = 0.32–0.69) and in comparison families (r = 0.20–0.57).

Social Support

There were no group differences in mother or father report of social support (Table IV). Hierarchical multiple regressions were completed to examine the impact of social support on parental distress. According to mothers, greater perceptions of functional support were associated with less maternal distress.
(Standardized $B = -0.23$, $p < .05$). No other main effects for group or interactions were identified.

### Child Age

Additional exploratory analyses were conducted to examine whether parent and family outcomes varied as a function of child age for families with a child with NF1 and comparison families. Analyses of variance, using a $2 \times 2$ design, were completed to examine the interaction of group (NF1 and comparison) with child age (based on a median split of 10.3 years). One significant interaction was identified for mother report of child resistance to eating on the AYCE-R, $F (1, 99) = 4.62, p < .05$; however, the interaction was not significant after appropriate consideration for multiple comparisons.

### Parental NF1

Twenty-five percent of children with NF1 had a parent affected with NF1 (nine mothers and six fathers). Exploratory nonparametric tests indicated no significant differences between mothers with NF1 and mothers without NF1 on our outcomes of interest.
Table V. Associations of Disease Severity Ratings with Parental Distress, Family Environment, Mealtime Climate, and Social Support for Mothers (n = 47) and Fathers (n = 37–38) of Children with NF1

<table>
<thead>
<tr>
<th></th>
<th>General disease severity</th>
<th>Appearance</th>
<th>Neurological impairment</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Mother Report</strong></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Parental distress (GSI)</td>
<td>0.13</td>
<td>0.04</td>
<td>0.34*</td>
</tr>
<tr>
<td><strong>Family environment (FES)</strong></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Family relationship index</td>
<td>−0.14</td>
<td>−0.12</td>
<td>−0.37*</td>
</tr>
<tr>
<td>Supportive</td>
<td>0.08</td>
<td>−0.01</td>
<td>−0.35*</td>
</tr>
<tr>
<td>Conflict</td>
<td>0.18</td>
<td>0.06</td>
<td>0.44**</td>
</tr>
<tr>
<td>Controlling</td>
<td>−0.17</td>
<td>−0.02</td>
<td>−0.21</td>
</tr>
<tr>
<td><strong>Mealtime climate (AYCE-R)</strong></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Resistance to eating</td>
<td>0.03</td>
<td>0.04</td>
<td>0.14</td>
</tr>
<tr>
<td>Positive mealtime interactions</td>
<td>−0.37*</td>
<td>−0.15</td>
<td>−0.50***</td>
</tr>
<tr>
<td>Parent aversion to mealtime</td>
<td>0.09</td>
<td>0.11</td>
<td>0.17</td>
</tr>
<tr>
<td><strong>Social support (NSSI)</strong></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Network size</td>
<td>0.03</td>
<td>−0.16</td>
<td>−0.19</td>
</tr>
<tr>
<td>Functional support</td>
<td>0.02</td>
<td>−0.12</td>
<td>−0.34*</td>
</tr>
<tr>
<td><strong>Father report</strong></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Parental distress (GSI)</td>
<td>0.33*</td>
<td>0.06</td>
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<td>0.08</td>
<td>0.06</td>
<td>−0.03</td>
</tr>
</tbody>
</table>

*p < .05, **p < .01, ***p < .001, two tailed tests.

However, fathers with NF1 reported greater social support network size (M = 8.40, SD = 1.14) than fathers without NF1 (M = 5.92, SD = 2.36), U = 29.50, p < .05.

**Disease Severity**

Correlations examined the associations of NF1 disease severity (e.g., general, appearance, or neurological) with parental distress, family functioning, mealtime climate, and social support (Table V). Few significant correlations were identified for fathers. However, according to mothers, greater neurological impairment for children was associated with greater distress, more family conflict, less positive mealtime interactions, and less social support. In addition, more mothers of children with NF1 who had moderate to severe neurological impairment (N = 10) were in the clinical range on the SCL-90-R relative to mothers of children with minimal to mild severity ratings (n = 5), χ² = (n = 47) 5.24, p < .05.

**Mediational Analyses**

Hierarchical multiple regressions examined whether family functioning mediated the association between disease severity and parental distress (GSI). For mothers, only neurological impairment was significantly associated with distress and family functioning (FRI and Positive Mealtime Interactions; Table V). In the regression analysis, neurological impairment was entered in step 1 (change in R² = 0.11, p < .05), followed by mother-reported FRI and Positive Mealtime Interactions in step 2 (change in R² = 0.15, p < .05; Fig. 1). The unstandardized β-weight for the neurological impairment scores became nonsignificant after the mediators were added to the model. Post hoc analyses using the bootstrap method for multiple mediators indicated that as a set, the family functioning variables were significant mediators (95% confidence interval (CI) = 0.08–0.97). However, examining the indirect effects of each variable separately indicated that neither FRI (estimated unstandardized β = 0.07; 95% CI around β = −0.13 to 0.26) nor Positive Mealtime Interactions (estimated unstandardized β = 0.23; 95% CI around β = −0.02 to 0.93) were significant mediators.

For fathers, only general disease severity score was significantly associated with parental distress (Table IV). General disease severity was also significantly associated

*Because of high correlations between FRI scores and the Conflict (r = −0.91) and Supportive scores (r = 0.77), only FRI scores were used in regression models.
Figure 2 Meditational model in which mother-reported Positive Mealtime Interactions mediate the association between general disease severity ratings of children with NF1 and paternal distress. Path values not in parentheses are zero-order correlations; those in parentheses are standardized β-coefficients from the regression equation with the other predictors included. *p < .05, two-tailed tests.

Discussion

This study examined the effect of childhood NF1 on parent and family functioning. Strengths included using demographically similar comparison families; psychometrically sound measures; both mother and father report of multiple domains of functioning; and multiple measures of disease severity. Contrary to our hypothesis, few differences between groups were identified for parental adjustment, social support, family environment, or mealtime climate, indicating substantial resilience among families of children with NF1. These findings are consistent with other studies of children with various chronic conditions that have used a comparison group of children without a chronic illness (Gerhardt et al., 2003; Greenberg et al., 1989).

Children with NF1 were perceived by mothers as exhibiting greater resistance to eating on the AYCE-R compared to comparison children. Because NF1 is not typically associated with eating/feeding problems, the AYCE-R was used in the present study as a measure of family functioning at mealtimes. Thus, maternal perceptions of children’s greater resistance to eating may be indicative of maternal perceptions of family conflict at mealtime. Items on the AYCE-R include “I feel that it is a struggle or fight to get my child to eat” and “there are arguments between me and my child over eating.” However, we did not find similar reports of family conflict on the FES according to parents of children with NF1, consistent with other studies using the FES among families of children with chronic conditions and comparison families (Gerhardt et al., 2003; Greenberg et al., 1989; Noll et al., 1995). In contrast, in several studies examining the functioning of families of pediatric cancer survivors, mothers reported their families to be rigid and inflexible relative to comparison mothers (Kazak & Meadows, 1989; Madan-Swain et al., 1994). Both of these studies included small samples of children, a number of whom had learning difficulties. In particular, Kazak and Meadows (1989) found that mothers of children who were receiving learning assistance perceived their families as less adaptable than mothers of other survivors. Similarly, fathers of children receiving learning assistance perceived their families as less cohesive and less adaptable than fathers of other survivors.

As expected, greater maternal perceptions of social support were associated with less distress, although this association was not stronger for mothers of children with NF1. While several studies of families of pediatric cancer survivors indicated that less social support was associated with greater posttraumatic stress symptoms in mothers (Manne et al., 2000) and greater psychological distress in fathers (Wijnberg-Williams et al., 2006), these studies did not include a group of comparison families. The results from our study are consistent with previous work suggesting few differences between groups of parents in level of social support (Gerhardt et al., 2003; Kazak et al., 1997; Noll et al., 1995) or its effect on parental distress (Gerhardt et al., 2003; Levers et al., 1998).

Despite the lack of between-group differences, we found difficulties for a subgroup of mothers of children with NF1. Consistent with our hypothesis, greater neurological impairment was associated with maternal reports of greater distress, more family conflict, fewer positive mealtime interactions, and less perceived social support. In addition, significantly more mothers of children with NF1 who had moderate to severe neurological impairment ratings exceeded clinical cutoffs for distress than mothers with children with minimal to mild ratings. Because of the genetic nature of NF1, it is possible that some of these mothers were also coping...
with the effects of the disease on themselves, which in combination with the increased demands of caring for a child with more severe disease, might be expected to contribute to greater distress. However, only three of the mothers with children with more severe disease had NF1 themselves.

In contrast to our findings involving the neurological impairment scale, few significant associations were found between psychosocial measures and ratings of general disease severity and appearance. This is concordant with a broader literature examining disease severity or functional limitations in children with other chronic illnesses. In these studies, few significant associations with parent and family functioning have been found (Canning et al., 1996; Gerhardt et al., 2003; Lutz et al., 2004) unless ratings were made by the same reporter, usually the mother (Lustig et al., 1996; Silver, Bauman, & Ireys 1995). Previous work with the current sample (Noll et al., 2007) has also indicated that greater general disease and appearance severity were unrelated to most child outcomes, but greater neurological impairment was associated with increased social, emotional, and behavioral difficulties in children with NF1. Our current findings appear to further demonstrate the influence of disease severity on parental functioning as well.

Few significant findings were identified for father report in the current study. Several studies also have indicated few significant differences between fathers of children with other chronic conditions and fathers of children without a chronic illness, both in their own adjustment, as well as their perception of family functioning (Gerhardt et al., 2003; Kazak & Meadows, 1989; Quitnner et al., 1998). It is noteworthy that our lack of findings using the SCL-90-R for fathers does not appear to be a result of under-reporting of symptoms. Fathers in both groups reported many symptoms of distress. Our previous work with the SCL-90-R and fathers of school-aged children has shown a similar pattern (Gerhardt et al., 2003; Noll et al., 1995) and highlight the critical role of an appropriate comparison group. These results may suggest that fathers of school-aged children, regardless of whether or not they have a child with a chronic illness, experience significant distress as a group, or alternatively, that the nonpatient norms for this measure may not be appropriate (Gerhardt et al., 2003). Although efforts are made to obtain norms that are both representative and contemporary, reliance on comparisons to normative data has been criticized because of potential problems with regional and cohort effects (Achenbach & Howell, 1993; Sandberg, Meyer-Bahlburg, & Yager, 1991). It is clear from these results that continued attention to the perspective of fathers is important in pediatric psychology.

Finally, it was expected that among families of children with NF1, family difficulties would mediate the association between disease severity and parental distress. Taken together, variables indicating poorer family functioning (i.e., lower FRI scores and less positive mealtime interactions) were significant mediators of the association between a child’s neurological impairment and maternal distress, but post hoc analyses indicated that neither variable was a significant mediator when examined separately. Interestingly, mother report of positive mealtime interactions was a potential mediator between general disease severity in children and paternal distress. However, post hoc analyses indicated no significant mediation. Additional work is needed here to better understand the factors that contribute to greater paternal distress in these families. While these models were tested with family functioning as the potential mediator between disease severity and parental distress, directionality of the associations between these variables cannot be determined without longitudinal research. It is possible that parental distress acts as the mediator between a child’s disease severity and family functioning.5

This study had several limitations. First, the lack of significant differences between groups may be the result of reduced power, particularly for analyses involving father report. However, effect sizes in most cases were small, suggesting that differences between groups were not clinically significant. Second, generalizability was limited by having few parents of children with more severe disease, who appear to be at greatest risk for difficulties. We were also unable to fully explore the impact of parental NF1 on parent and family functioning. Although about 50% of children with NF1 would be expected to have inherited the condition, only 23% of our sample had a parent with NF1 (nine mothers and six fathers) despite our rigorous recruitment strategies. While exploratory analyses revealed few differences between parents with NF1 and those without NF1, future research is needed to address the effect on family functioning when both parent and child are affected.

Third, our measures focused on general parental adjustment (e.g., SCL-90-R) and family functioning (e.g.,

5Exploratory analyses were run with maternal distress as the mediator between neurological impairment and FRI scores or Positive Mealtime Interactions, as well as with paternal distress as the mediator between general disease severity and mother report of Positive Mealtime Interactions. While the β-weights were suggestive of mediation in all cases, post hoc analyses indicated that maternal and paternal distress were not significant mediators.
FES). Measures that focus directly on parenting stress in general or stress specific to pediatric chronic illness might provide additional insight to the challenges faced by parents of a child with NF1. However, it is important to note that contextual variables have been suggested to play a pivotal role when obtaining information regarding subjective functioning (Kahneman et al., 2006). For instance, using disease-specific measures that ask questions about disease status and functional impairments or obtaining data in clinic or hospital settings may create a focusing illusion or misallocation of attention (Schkade & Kahneman, 1998) on health and disease status when conducting assessments of emotional well-being. Thus, responses to questions about day-to-day well-being may be altered, when cognition has been primed or focused on disability and medical difficulty (Smith et al., 2006).

Fourth, our scales of medical involvement lack robust psychometric data. With regard to our measure of neurological impairment, we did not obtain additional neurocognitive data or detailed information about learning disabilities on all the children. Many of the children had formal neurocognitive testing in their medical charts, but they all were not tested using the same protocol. Comparison children, however, were not screened for learning disabilities or cognitive impairment. It is certainly possible that some of the comparison children had learning disabilities. We might have obtained larger between group effects had we compared families with children with NF1 with a group of comparison families, who were screened for learning disabilities or cognitive impairment.

Finally, no correction for multiple comparisons was made for analyses involving disease severity. Though several significant associations were found between pediatric neurological impairment and parent/family functioning, spurious effects must be considered when interpreting results and findings need to be replicated. Future research should: (a) include multiple sites to improve generalizability and sample size, (b) address adjustment over time as children with NF1 become adolescents and young adults, and (c) include multiple perspectives such as child report and objective observations of family functioning.

Although most parents and families in the current study appear to be functioning similarly to comparison parents and families, many parents (especially fathers) of school-age children reported numerous symptoms of distress. While this may not be worse for mothers or fathers of children with NF1, our data remain a concern. Additionally, mothers of children with NF1 who have greater neurological impairment may be at risk for more difficulties. Our previous work with this sample demonstrated that greater neurological impairment was also significantly associated with mother report of children’s acting out behaviors (Noll et al., 2007). Numerous studies have indicated that behavioral problems in children with chronic illness are associated with greater parental distress and family conflict (levers et al., 1998; Kronenberger & Thompson, 1990; Manne et al., 1996; Thompson et al., 2003). When children with NF1 have significant neurological involvement, our data suggest these children and their families may be at the highest risk for problems and in need of assistance. Finally, several studies have suggested that parents who perceive their child’s disease as intrusive or who experience greater caregiver burden or parenting stress are more likely to report distress and family difficulties (Canning et al., 1996; Lustig et al., 1996; Streisand, Kazak, & Tercyak, 2003). Future work is needed to explore these factors as well, especially for families with a child with more neurologically severe NF1.

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