Implications of Resolving the Diagnosis of PKU for Parents and Children

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Objective To examine resolution of the diagnosis among parents of children with phenylketonuria (PKU) as a mechanism of adjustment for parents and children. Methods Reaction to diagnosis interviews were conducted with 52 mothers and 47 fathers of 55 children with PKU aged 2–12 years. The parents also completed questionnaires assessing their personal adjustment (stress symptoms), their child’s adjustment (behavior problems), and coping variables (personal hopefulness and coping strategies). Results Most mothers (69%) and fathers (77%) were resolved to their child’s diagnosis. Lower levels of parent stress were explained by higher personal hopefulness (14% of the variance for mothers and 21% for fathers) and resolution of the diagnosis (15% of the variance for mothers and 6% for fathers) after taking account of demographic variables and severity of the child’s PKU. Parent resolution, however, did not contribute independently to the variance explained in child behavior problems after taking account of coping variables and severity of PKU. Conclusions Resolution of the diagnosis of PKU is a strong indicator of parent adjustment, and assessment of parent reactions should be considered an integral component of clinical care. Further research is warranted in relation to the implications of parent resolution for the child’s response to PKU through different development stages and the effectiveness of interventions in aiding parent resolution.

Key words adjustment; child; coping; PKU; parent; resolution of the diagnosis.

The Impact of PKU

Parents play a pivotal role in the management of childhood phenylketonuria (PKU), but few studies have examined parent reactions and their adjustment to this condition. PKU is a rare inborn error of metabolism, detected by newborn screening (Waisbren, 1999). Children with PKU have an enzyme deficiency that affects the metabolism of phenylalanine, a specific amino acid contained in normal diets. If untreated, PKU causes brain damage and developmental delay. However, the detrimental effects of PKU are largely controllable through a protein-restricted diet, which is crucial during early childhood and recommended for the whole of life. Most children with PKU require blood testing, at least monthly, to monitor phenylalanine levels. In developed countries, children with PKU are usually managed by major pediatric centers, and most achieve normal physical and intellectual development. For a small minority, poor adherence to the diet leads to learning and behavioral difficulties (Sullivan & Chang, 1999). A central issue for parents is how they deal with the emotional impact of the diagnosis, which occurs abruptly soon after the birth and represents a serious threat to the child’s development. Moreover, the child’s diet is highly restrictive and exacting. Thus, PKU tends to have an intrusive impact on family life (Eiser, 1985), and both mothers and fathers tend to worry about their child’s development (Lord, Wastell, & Ungerer, 2005).

Resolving the Impact of the Diagnosis

While there have been few studies of parent reactions to PKU, common reactions to other serious childhood conditions have been well described. Numerous studies have highlighted the intensity of parental grief that is often associated with the diagnosis, as well as individual differences in reactions and coping styles (Beresford, 1994; Shapiro, 1983).
A grief response is usually triggered by the painful realization that the child has a condition that changes expectations of normal healthy development (Trout, 1983; Waisbren, 1980). Although the intensity of grief varies, initial reactions usually involve strong feelings of disappointment, sadness, or anger (Ormerod & Huebner, 1988; Thernlund, Dahlquist, Ivarsson, & Ludvigsson, 1996) and may include fear, numbness, and difficulties in retaining information (Alonzo, 2000; Jedlicka-Kohler, Gotz, & Eichler, 1996). Persistent trauma effects have been recognized among parents of children surviving life-threatening conditions such as cancer (Barakat et al., 1997), and similar reactions have recently been found in relation to PKU (Lord et al., 2005).

How parents process the emotional impact of the diagnosis has been a debated issue (Marvin & Pianta, 1996). An emphasis on pathological responses in the early literature (Shapiro, 1983) was followed by models depicting “stages” through which parents typically move toward adaptation (Clubb, 1991; Drotar, Baskiewicz, Irvin, Kennell, & Klaus, 1975; Trout, 1983), with the common theme of progression over time towards eventual adjustment (Blacher, 1984). While most parents appear to move through an initial crisis to a state of relative equilibrium, the notion of stages has been criticized as too linear and prescriptive (Allen & Affleck, 1985). The “stages” approach also implies the normal pathway is toward completion of grief, which has not accorded well with many parents whose experience has been described as chronic sorrow (Teel, 1991) or episodic grief (Hewson, 1997).

An important advance in this area has been the development of the concept of “resolution” of the child’s diagnosis, based on attachment theory, in a way that recognizes individual differences in coping styles and adjustment outcomes (Marvin & Pianta, 1996; Pianta, Marvin, Britner, & Borowitz, 1996). Marvin and Pianta (1996) define resolution as the relative cessation of active grieving and a reorientation to current realities and future possibilities. This is characterized by acknowledgment of feelings, a change in the quality and intensity of emotional response, and an assertion of coping and moving on in life. Unresolved parents display an absence of these characteristics and a coping style that reflects the ongoing effects of trauma. Common features include cognitive distortion, emotional disorientation, and a continuing search for an explanation or existential reason for the child’s condition. The concept of resolution has similarities to notions of “rebuilding world assumptions” (Janoff-Bulman, 1992) and “redefining situations” (Tunali & Power, 1993), which involve the realignment of beliefs, values, and living patterns following a traumatic event to accord with new realities.

Research on resolving the diagnosis has been conducted with mothers of children diagnosed with cerebral palsy or epilepsy, at 2–4 years postdiagnosis (Marvin & Pianta, 1996; Sheeran, Marvin, & Pianta, 1997). Resolved and unresolved parents were reliably identified through a brief standardized assessment known as the Reaction to Diagnosis Interview (RDI) (Pianta & Marvin, 1993). Approximately equal numbers of resolved and unresolved mothers were found. Given the challenges presented by PKU, it is likely that a substantial proportion of parents of children with PKU will also experience difficulty in resolving the diagnosis. (Pianta et al., 1996) found no association between the parent’s resolution classification and the time since the diagnosis. This was interpreted as indicating that resolution occurs soon after the diagnosis and that parents move into a relatively stable coping style. However, there has been relatively little research into implications of resolution versus nonresolution for longer term adjustment of children and parents.

Resolution of the Diagnosis and Child Adjustment

The concept of resolution has parallels with the attachment theory notion of secure caregiving internal working models, which provide the foundation for secure parent–child attachment relationships (Bowlby, 1988). Secure caregiving internal working models are defined by an acknowledgment and integration of positive and negative thoughts and feelings about the parent as caregiver and about the child in the attachment relationship. In contrast, the concept of lack of resolution captures features of insecure caregiving internal working models where thoughts and feelings are not integrated and where, in the most insecure form, thoughts and feelings are sufficiently distorted to make it difficult for parents to adequately care for their child (George & Solomon, 1999). Marvin and Pianta (1996) suggest that resolving the diagnosis involves the integration of internal working models in a way that allows acceptance of the condition and enables the parent to organize their caregiving behavior. This implies that resolved parents are more likely to respond sensitively to their child’s emotional needs and to display confidence, clarity, and consistency in setting behavioral limits.

There is limited empirical research supporting an association between parent resolution and positive child adjustment. In the Marvin and Pianta (1996) study of mothers of preschool children with cerebral palsy,
Resolution of the Diagnosis and Other Coping Processes

Resolving the diagnosis can be viewed as a resilience factor for dealing with the stressful impact of the child’s condition within a stress and coping framework based on risk and resilience factors, such as that proposed by Wallander and Varni (1998). As a resilience factor, the construct of resolution has similarities to recognized coping processes described in the wider literature. First, the “coping styles” described within the Marvin and Pianta (1996) model have similarities to “coping strategies” that mediate the impact of stress on health (Carver, Scheier, & Weintraub, 1989; Lazarus & Folkman, 1984). Resolved coping styles may be emotion-, cognition-, or action-based. These coping styles have similarities to adaptive coping strategies, such as support seeking, reappraisal, and problem-solving, which have been identified in wider research on childhood chronic conditions (Beresford, 1994). Some of the unresolved coping styles, such as those classified as “emotionally overwhelmed” or “neutralizing”, have similarities to palliative coping strategies that involve self-blame and avoidance (Beresford, 1994). Second, resolution is characterized by a balanced outlook with positive expectations of being able to manage the challenges that lay ahead (Marvin & Pianta, 1996). In this regard, resolution implies the formation of a hopeful outlook, which has been identified as an important coping resource in recovery from traumatic events (Nunn, 1996).

Although resolution implies the use of adaptive coping strategies and a hopeful outlook, the construct is essentially based on emotional processing of an event that is deemed to be traumatic. Marvin and Pianta (1996) suggest that this occurs through cyclical phases of the stress response (Horowitz, 1997) that enable the parent to incorporate painful new information and move forward. Resolved parents are seen as able to recall events surrounding the diagnosis and acknowledge distress associated with it, but are more oriented to the present and future needs of their child. In contrast, unresolved parents are described as emotionally “stuck in the past” and more preoccupied with negative aspects of their child’s condition (Marvin & Pianta, 1996). Lack of resolution indicates that the diagnosis continues as a major source of stress and is likely to be a strong predictor of stress symptomology. Further study is needed to clarify the relationship between resolving the diagnosis, coping strategies, and hopefulness, and to determine whether resolving the diagnosis contributes independently to the explanation of parent and child outcomes.

Aims of the Study

The present study had three aims. The first was to examine the relationship between resolution of the diagnosis and adjustment outcomes for parents (stress symptoms) and children (behavior problems) in families of children with PKU. The second was to examine relations between parent resolution of the diagnosis and other coping variables, specifically coping strategies, and personal hopefulness. The third aim was to compare the variance in parent and child outcomes explained by resolution of the diagnosis with that explained by personal hopefulness and coping strategies. It was hypothesized that resolved parents will have fewer stress symptoms and that children with resolved parents will have fewer behavior problems. It was also hypothesized that resolved parents will have higher levels of personal hopefulness and tend to use active rather than palliative coping strategies. It was further hypothesized that parent resolution classification will predict variance in parent and child outcomes over and above measures of personal hopefulness and coping strategies.

Method
Participants

Participants were recruited through the PKU Clinic at an Australian pediatric referral hospital, which is linked to a newborn screening service and provides management for all children with PKU in the home state. The sample consisted of one or both parents from 55 two-parent families with a child with PKU aged 2–12 years. If families had more than one child with PKU within the age range, data were collected in relation to the older child with PKU. Participating families represented 83% of the clinic population. The nonparticipating families (n = 11) did not differ significantly from the study sample when compared on available demographic data (child age...
and gender), and their family names indicated they were from a range of cultural backgrounds. Reasons for refusals were mostly either the time commitment required for the study or the personal nature of the information sought.

The Parent Sample
From 55 families, data were available for 52 mothers and 47 fathers. The mean age of mothers was 35.3 years ($SD = 5.2$, range 24–51 years), and the mean age of fathers was 38.0 years ($SD = 5.7$, range 25–51 years). Parents were from various cultural backgrounds; 31% ($n = 16$) of mothers and 26% of fathers ($n = 12$) had migrated to Australia, mainly from South East Asia, the Middle East, and Northern and Southern Europe. A minority of families (13%, $n = 7$) spoke a language other than English at home, and 76% ($n = 42$) were residing in metropolitan areas. Most fathers (94%, $n = 44$) were in the paid workforce, 60% ($n = 28$) had completed tertiary education, and 51% ($n = 24$) were in occupations requiring technical or professional qualifications. More than half of the mothers (56%, $n = 29$) were in the paid workforce, 50% ($n = 26$) had completed tertiary education, and 23% ($n = 12$) were in technical or professional occupations.

The Child Sample
Of the 55 children, 28 were male and 27 were female. The mean age of the children was 6.6 years ($SD = 3.2$, range 2.0–12.7 years). All children had a form of PKU. Most (93%, $n = 51$) had a diagnosis of classic PKU requiring dietary treatment and regular blood tests. Three children had a mild form of PKU requiring limited dietary adjustment and monitoring, and one had a diagnosis of BH4 Synthesis Defect, requiring medication and monitoring. In most families there were three or fewer children ($SD = 2.0$, range 2–12.7 years). All children had a form of PKU. Most (93%, $n = 51$) had a diagnosis of classic PKU requiring dietary treatment and regular blood tests. Three children had a mild form of PKU requiring limited dietary adjustment and monitoring, and one had a diagnosis of BH4 Synthesis Defect, requiring medication and monitoring. In most families there were three or fewer children (95%, $n = 52$), and the child with PKU was either the first- or second-born (89%, $n = 49$).

Measures
Resolution of the Diagnosis
The RDI (Pianta & Marvin, 1993) is a brief, structured interview that aims to elicit the affective and cognitive responses of parents to receiving their child’s diagnosis, changes in responses over time, and the meaning the parents attribute to the experience. The RDI is comprised of five questions and takes ~15 min to administer. Interviews are video-recorded and later coded (resolved vs. unresolved) using the RDI Classification System (Pianta & Marvin, 1993). Resolution is determined by identification of key characteristics (acknowledgment of feelings, change in emotional response, and assertion of moving on).

In the present study, all except three interviews were video recorded. In these cases, either an audio recording or hand-written notes were made. Coders were the first and second authors, both of whom obtained formal certification (from R. Marvin, University of Virginia) in the use of the classification system. Reliability was established by independently coding 20 interviews. Initial agreement was 90%, and differences were conferenced to agreement. All RDI classifications were assigned while the coders were blind to the questionnaire data.

Parent Adjustment
The Malaise Inventory (MI) (Rutter, Tizard, & Whitmore, 1970) assessed emotional and physical stress symptoms. The MI is self-administered and consists of 24 items, with a ‘yes/no’ response format. The score is derived by totalling the ‘yes’ responses. A score of six or more is considered to represent a clinical level of distress. Internal reliability (Cronbach’s α) was .82 for mothers and .87 for fathers. The MI correlates well with other stress scales, and test–retest reliability over short periods has been reported at $r = .91$ (Hirst & Bradshaw, 1983). The measure has been extensively used with parents of children with chronic conditions (Sloper & Turner, 1993).

Child Adjustment
The Child Behavior Checklist (CBCL) Problem Scale (Achenbach, 1991) assessed the emotional and behavioral adjustment of the children. The relevant age-related scale versions were used, and $T$-scores were derived to allow comparison across age and sex groups. Achenbach (1991) reports a high test–retest reliability ($r = .93$) for the Problem Scale. $T$-scores above 63 represent a clinical level of emotional disturbance, while $T$-scores of 60–63 are in the borderline range. Some reports use $T \geq 70$ as the clinical range, and the results are reported for both cut-off points.

The CBCL has been extensively used in developmental and clinical research (Achenbach, 1991). Items capturing somatic symptoms have been shown not to confound behavioral assessment in children with chronic medical conditions (Holmes, Respess, Greer, & Frentz, 1998). The Problem Scale was, therefore, used.

Coping Variables
The Hunter Opinions and Personal Expectations (HOPE) Scale (Nunn, Lewin, Walton, & Carr, 1996) assessed the parents’ personal hopefulness. The measure consists of 20 items, each scored on a 5-point Likert scale, and
comprises two subscales, Hope and Despair. Internal reliability (Cronbach’s α) for the Hope subscale was .82 for mothers and .76 for fathers, and for the Despair subscale was .84 for mothers and .77 for fathers. A Global Personal Hopefulness (GPH) score is computed from combining the subscales, and a higher GPH score means a more hopeful and less despairing outlook. The GPH score was used in the present study. GPH has been shown to have good test–retest reliability over a period of 64 weeks (r = .71) and to correlate well with measures of trait anxiety (r = -.64) and depression (r = -.54) (Nunn et al., 1996).

The Ways of Coping Questionnaire (WCQ) (Folkman & Lazarus, 1988) assessed the parents’ coping strategies. Parents completed the measure in relation to a challenging situation associated with their child’s PKU that occurred in the previous month. The WCQ has 66-items, each with a 4-point Likert scale indicating the frequency with which the strategy was used. The measure comprises eight scales representing theoretically different coping strategies: Confrontative Coping, Distancing, Self-Controlling, Seeking Social Support, Accepting Responsibility, Escape-Avoidance, Problem Solving, and Positive Reappraisal. In the present study, the relative score method was used. Internal reliability (Cronbach’s α) ranged from .65 to .84 in the mothers’ data and .71 to .86 in the fathers’ data. The WCQ has been widely used in studies of childhood conditions (Beresford, 1994).

Demographic Variables
Data were collected in relation to the child’s age, gender, birth order, number of siblings, and number of children in the family with PKU. Parent demographic data were collected in relation to their age, education level, type of occupation, country of birth, and language background (English speaking vs. non-English speaking).

Severity of PKU
Information about the child’s type of PKU was collected from the treating doctor for use as a control variable. Severity of PKU was treated as a binary measure reflecting whether or not the child required a special diet.

Procedures
The research ethics committees of the hospital and the associated university approved the study. Participating parents provided written consent and attended a single assessment session lasting 60–90 min in which they were interviewed and completed a set of self-report questionnaires. Assessments were conducted by the first author or a qualified social worker at either the hospital or the parent’s home. Parents were seen separately, and language interpreters were used on three occasions with parents who were not fluent in English.

Data Analysis
The mothers’ and fathers’ data were analyzed separately, and the distributions of scores for all measures were examined. To test the first and second hypotheses, the bivariate relationship of resolution classification with MI, CBCL Problem Scale, HOPE Scale, and WCQ coping scales was examined using t-tests. To test the third hypothesis, a hierarchical regression strategy was used to examine coping variables (HOPE Scale and WCQ scales) and resolution classification as predictors of parent and child outcomes, with the Malaise score as the dependent variable in the parent models and the CBCL Problem Scale as the dependent variable in the child models. Preliminary analyses were conducted to examine the bivariate relationship of the predictor and dependent variables with each other and with the severity of PKU and demographic variables (i.e., child’s age, gender, birth order, number of siblings, parent’s age, parent’s language background, parent’s education, and parent’s occupation), using t-tests, ANOVA, chi-square, or Spearman correlations as appropriate. None of the predictor variables was confounded with the severity of PKU or demographic variables (r ≤ .61). Demographic and coping variables were retained for the multiple regression analyses where they were related to the dependent variable at an α-level of .05. The severity of PKU was entered first as a control variable, and the retained demographic variables were entered second. The retained coping variables were then tested in the model using the stepwise procedure with α at .05. In the fourth step, resolution classification was entered as a dummy variable. Finally, an exploratory mediation analysis was conducted to examine whether the influence of resolution on Malaise and CBCL scores was mediated by the other coping variables.

Results
Resolution of the Diagnosis and Adjustment Outcomes
Resolution Classification and MI
Coding of RDIs indicated that 69% of mothers (n = 36) and 77% of fathers (n = 36) were resolved to the diagnosis. Although a larger percentage of fathers than mothers was resolved, the difference between the groups in the ratio of resolved to unresolved cases was not significant, [χ² (1, 45) = .79, p > .05]. No association was found between resolution classification and the child’s age in either the mothers’ or fathers’ data.
The mothers’ scores on the Malaise Inventory ranged from 0 to 20, with a mean of 4.73 (SD = 4.34). Although most mothers reported a relatively low number of stress symptoms, 33% (n = 17) scored 6 or more, indicating a high level of distress. Resolved mothers had significantly lower Malaise scores, t (50) = −3.74, p < .001, although there was greater variability in scores among the unresolved group (Table I).

Fathers’ Malaise scores ranged from 0 to 18 with a mean of 3.49 (SD = 3.94), and 17% (n = 8) scored 6 or above. The difference in scores for mothers and fathers was not significant when compared using a paired t-test. Resolved fathers had significantly lower scores than unresolved fathers, t (45) = −2.15, p < .05, and like mothers there was greater variability in scores among the unresolved group (Table I).

Resolution Classification and Child Behavior
The T-scores for the CBCL recorded by mothers were normally distributed (M = 51.33, SD = 11.78), with 13% (n = 7) in the borderline clinical range (T ≥ 60), 11% (n = 6) in the clinical range (T ≥ 64), and three T-scores > 70. The CBCL T-scores recorded by fathers were also normally distributed (M = 50.62, SD = 10.68), with 16% (n = 8) in the borderline clinical range, 10% (n = 5) in the clinical range, and with two T-scores above 70. The correlation between the mother’s and father’s T-scores was .64 (p < 0.001).

Resolved parents tended to report fewer behavior problems with their child. However, the difference in CBCL T-scores recorded by resolved and unresolved parents approached significance only in the mothers’ data, t(50) = −1.99, p = .05 (Table I).

Resolution of the Diagnosis and Parent Coping
Resolution classification was not significantly related to the HOPE Scale score in either the mothers’ or fathers’ data. Resolved mothers had significantly lower scores for Escape-Avoidance coping, t(50) = 3.37, p < .01. However, resolution classification was not significantly related to the other coping strategy scales in either the mothers’ or fathers’ data.

Resolution of the Diagnosis, Parent Coping, and Stress Symptoms

Mothers
Mothers’ Malaise scores were unrelated to the severity of PKU or demographic variables other than their language background. Mothers born in a non-English speaking country had significantly higher Malaise scores, t(50) = −2.93, p < .01. The correlation of Malaise scores with the coping variables and resolution classification is presented in Table II.

In the hierarchical regression analysis, the severity of PKU was entered first, and the retained demographic variable (mother’s language background) was entered second. In the third step, the retained coping variables (HOPE Scale, Problem Solving scale, and Escape-Avoidance scale) were tested using the stepwise procedure and the HOPE Scale entered the model (p < .01). The mother’s resolution classification was then entered and found to be significant (p < .01). In the final equation, non-English speaking background (β = .28), lower levels of personal hopefulness (HOPE Scale, β = −.38) and lack of resolution of the diagnosis (β = .40) were associated with higher Malaise scores. The model explained 44% of the variance in mothers’ Malaise scores, with language background accounting for 15%, the HOPE Scale adding 14%, and resolution classification adding a further 15% (Table III).

Fathers
Fathers’ Malaise scores were unrelated to the severity of PKU or demographic variables other than their occupation level. Fathers in lower occupation groups had
significantly higher Malaise scores, $t(45) = 2.13, p < .05$. The correlation of fathers’ Malaise scores with the coping variables and resolution classification is presented in Table II.

In the hierarchical regression analysis, higher Malaise scores for fathers were predicted by lower levels of personal hopefulness (HOPE Scale, $\beta = -.43$) and lack of resolution of the diagnosis ($\beta = -.25$). With these variables in the model, the fathers’ occupation level lost significance. The model explained 37% of the variance in fathers’ Malaise scores, with occupation level accounting for 10%, the HOPE Scale adding 21%, and resolution classification adding a further 6% (Table III).

### Resolution of the Diagnosis, Parent Coping, and Child Behavior Problems

#### Mothers

Mothers’ CBCL Problem Scale scores were significantly related to the severity of PKU, $t(30) = -2.34, p < .05$, but unrelated to the demographic variables. The correlation of mothers’ CBCL scores with the coping variables is presented in Table II.

In the hierarchical regression analysis, CBCL Problem Scale scores were predicted by the severity of the child’s PKU ($\beta = .38$) and the mother’s level of personal hopefulness (HOPE Scale, $\beta = -.41$). These two variables together explained 28% of the variance in CBCL Problem Scale scores recorded by mothers. Mothers’ resolution classification, however, was not a significant predictor of their child’s behavior problems (Table IV).

#### Fathers

Fathers’ CBCL Problem Scale scores were unrelated to the severity of PKU or the demographic variables. The correlation of fathers’ CBCL scores with the coping variables is presented in Table II.

In the hierarchical regression analysis, CBCL Problem Scale scores were predicted by the severity of the child’s PKU ($\beta = .36$), and the father’s Escape-Avoidance coping ($\beta = .25$) and level of personal hopefulness (HOPE Scale, $\beta = -.43$). These three variables together explained 33% of the variance in CBCL Problem Scale scores. Fathers’ resolution classification was not a significant predictor of children’s behavior problems (Table IV).
The higher proportion of resolved cases in this study did not appear to be a function of time since birth, despite the wide age range of the children (2–12 years). This finding supports the assumption that the affective and cognitive responses of parents to their child’s diagnosis tend to remain constant over time (Pianta et al., 1996). The results raise the question of whether some childhood disorders are intrinsically more difficult for parents to resolve than others. It has been argued that the impact varies according to features such as the duration and course of the condition, intensity of treatment, and level of functional impairment (Silver, Westbrook, & Stein, 1998). However, controllable conditions also present particular adjustment challenges. For PKU, these include not only the underlying threat to the child’s brain development and the rigorous dietary management required, but also the difficulties of living with an “invisible” condition that might not be socially recognized or validated. On the other hand, the predictable management and relatively positive outlook for PKU might aid parents in resolving the emotional impact of the diagnosis.

The results for the MI were consistent with those for the RDI and with previous studies of distress among parents of children with chronic conditions (Sloper & Turner, 1993; Wallander & Varni, 1998). A minority of parents had Malaise scores in the clinical range, and a significant association was found between Malaise scores and resolution classification for both mothers and fathers. The results suggest that lack of resolution contributes to stress symptomatology over time, although the cross-sectional design of this study prevents any conclusion regarding the direction of influence. It is also possible that parents with more stress symptoms were more impacted by sources of stress other than PKU, either prior to or following the diagnosis, which may have impeded efforts to resolve the diagnosis.

Contrary to expectations, resolution classification was not significantly associated with personal hopefulness. Personal hopefulness is an integrative construct that combines several elements of personal functioning, including self-efficacy, capacity to control events, and purpose in life (Nunn, 1996). The HOPE Scale captures a broad range of life expectations, and GPH is seen to represent a relatively enduring trait that might not be socially recognized or validated. On the other hand, the predictable management and relatively positive outlook for PKU might aid parents in resolving the emotional impact of the diagnosis.

### Discussion

The study examined the proportion of resolved versus unresolved cases and the association of resolution classification with parent adjustment (stress symptoms) and child adjustment (behavior problems). The study also compared the strength of association of resolution classification with coping-related variables (hopefulness and coping strategies) to parent and child adjustment. The coping variables were selected because of their conceptual similarity to the notion of resolving the diagnosis.

The proportions of resolved cases found in the present study (69% for mothers and 77% for fathers) were higher than the proportions of resolved cases (46–48% for mothers) found in previous studies (Marvin & Pianta, 1996; Sheerin et al., 1997). The higher proportion of resolved cases in this study did not appear to be a function of time since birth, despite the wide age range of the children (2–12 years). This finding supports the assumption that the affective and cognitive responses of parents to their child’s diagnosis tend to remain constant over time (Pianta et al., 1996). The results raise the question of whether some childhood disorders are intrinsically more difficult for parents to resolve than others. It has been argued that the impact varies according to features such as the duration and course of the condition, intensity of treatment, and level of functional impairment (Silver, Westbrook, & Stein, 1998). However, controllable conditions also present particular adjustment challenges. For PKU, these include not only the underlying threat to the child’s brain development and the rigorous dietary management required, but also the difficulties of living with an “invisible” condition that might not be socially recognized or validated. On the other hand, the predictable management and relatively positive outlook for PKU might aid parents in resolving the emotional impact of the diagnosis.

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### Coping Variables as Mediators of Resolution of the Diagnosis

Only one coping variable in the mothers’ and fathers’ data met the criteria for examination as a mediator of the influence of resolution on the outcomes of interest. In the mothers’ data, the Escape-Avoidance scale had a similar correlation to both resolution classification ($r_s = .43$, $p < .01$) and Malaise score ($r_s = .43$, $p < .01$). When tested in a regression analysis, while controlling for the severity of PKU and mother’s language background, Escape-Avoidance reduced the variance in Malaise scores explained by resolution classification from 20% to 13%, thus, indicating partial mediation.

### Table IV. Final Step of Hierarchical Regression Analyses for Variables Predicting Child Behavior Problem Scores reported by Mothers and Fathers

<table>
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<th>Variables</th>
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<th>SE B</th>
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<td>50.70</td>
<td>21.54</td>
<td>.32*</td>
<td>24</td>
</tr>
<tr>
<td>HOPE Scale</td>
<td>-3.35</td>
<td>.16</td>
<td>-0.29*</td>
<td>33</td>
</tr>
<tr>
<td>Father’s resolution classification</td>
<td>-3.28</td>
<td>3.19</td>
<td>-0.13</td>
<td>35</td>
</tr>
</tbody>
</table>

*p < .05.
**p < .01.
experience, which is linked to its roots in attachment theory.

Resolution classification was also unrelated to parent coping strategies other than mothers’ use of Escape-Avoidance. This scale includes items that focus on wishful thinking, anesthetizing negative feelings, and avoidance of people (Folkman & Lazarus, 1988). The association of Escape-Avoidance with lack of resolution for mothers is consistent with previous research on palliative coping strategies (Sloper & Turner, 1993; Thompson, Gil, Burbach, Keith, & Kinney, 1993) and the notion that unresolved parents tend to use more avoidant strategies to defend against painful feelings associated with the diagnosis. Fathers’ resolution of the diagnosis was not associated with their coping strategies, which most likely reflects the less prominent role played by fathers in managing childhood chronic conditions (Beresford, 1994). Overall, the results in relation to coping strategies most likely reflect the broad range in the type of stressful events selected by parents when completing the WCQ. Coping strategies generally vary according to both the perceived degree of threat and situational appraisal of control (Folkman & Lazarus, 1988), and the results are consistent with previous studies indicating that parents of children with chronic conditions tend to use a wide range of coping strategies in response to varying problems (Beresford, 1994). Future research might investigate whether resolved and unresolved parents respond differently to specific kinds of challenges in managing their child’s condition.

The models predicting parent adjustment indicated that resolution classification was a relatively strong predictor of level of stress symptoms compared with the coping variables. Resolution classification independently explained 15% of the variance in Malaise scores for mothers and 6% for fathers. These findings highlight the relevance of the issues explored in the RDI to parent wellbeing and the importance of resolving the diagnosis in the adjustment process. They also support the validity of the RDI in capturing diagnosis-related distress. Personal hopefulness (GPH) was also a strong predictor of mothers’ and fathers’ level of stress symptoms. Hopefulness represents expectations of being able to deal positively with future events, based on a sense of value and purpose in one’s activities (Nunn, 1996). This result is consistent with a number of previous studies that have found hope to be an important resilience factor for parents in enduring the demands of childhood chronic conditions (Horton & Wallander, 2001; Mednick et al., 2007).

The association of parent resolution with stress symptomology appears to be relatively independent of other coping processes, as indicated by the exploratory mediation analysis. Escape-Avoidance coping by mothers was the only coping variable found to partially mediate the influence of resolution on stress symptomology. This suggests that coping strategies may not be the mechanism by which resolution is associated with parent and child adjustment.

Language and cultural factors appeared to play a role in mothers’ adjustment, as mothers of non-English speaking background tended to report more stress symptoms. This might relate to perceptions of PKU, the cultural significance of the diet, or difficulties accessing support services. However, this result needs to be treated with caution given the small sample sizes and without further evidence of the reliability of the MI with parents of non-English speaking background. Fathers’ level of stress symptoms was more strongly associated with their occupation level than cultural or language factors. This might represent a variety of influences, including self-appraisal, income, and occupational stress.

The models predicting child adjustment examined parent resolution and other coping variables as predictors of child behavior problems. It should first be noted that ~90% of children were functioning in the normal range, and the percentage of children with clinical levels of behavior disturbance was similar to that in the general community (Achenbach, Hensley, Phares, & Grayson, 1990). This suggests that metabolic control was satisfactory for most of the children, as the neuro-toxic effects of high phenylalanine (PHE) levels can give rise to problems in intellectual and behavioral functioning (Sullivan & Chang, 1999). Resolved mothers tended to report fewer child behavior problems. However, resolution classification accounted for only 7% of variance in mothers’ CBCL Problem Scale scores, and the trend was weaker in the fathers’ data. The parent’s level of personal hopefulness (GPH) was a stronger predictor of child behavior problems reported by both mothers and fathers. As well as supporting personal resilience, hopefulness may contribute to more effective parenting and more positive perceptions of the child’s behavior. Fathers’ engagement in the management of their child’s condition was also highlighted, as fathers who relied on Escape-Avoidance coping tended to report more child behavior problems.

It is important to recognize that the wide developmental range of the children and the reliance on parent-report measures represent limitations to the study. For parents, the challenges of managing PKU change as the
child develops, and lack of parental resolution may be reflected in different behavioral issues at different developmental stages. In early childhood, for example, the challenges include helping the child accept the special formula, restricted diet, and blood testing. Through middle childhood, managing the PKU diet in the context of school and peer relationships becomes important. As resolution appears strongly linked to attachment style (Marvin & Pianta, 1996), the implications of parent resolution for the child’s behavioral adjustment warrant further exploration. Future studies should also be based on assessment of more specific behavioral issues associated with the condition, such as eating or demanding prohibited foods and refusal to take the prescribed dietary formula. It is also important to note that the cross-sectional design and the small sample size limit the interpretation and generalization of the findings. The outcomes examined may have also been influenced by other variables, such as the parent’s appraisal of PKU and the support available through the partner and social networks. Nevertheless, the study adds to the limited available data on adjustment to PKU.

The results have implications for intervention with families. This study highlights the importance of acknowledging the emotional impact of the diagnosis on parents and supporting hopefulness by recognizing positive aspects of the child’s condition. Pediatricians and dieticians in PKU clinics need to be aware of indicators of lack of resolution, such as persistent distress, avoidance of discussion of the diagnosis and related problems, and a continuing preoccupation with the cause or reason for the child’s condition. An understanding of these issues will assist clinicians in referring parents appropriately for psychosocial support. Ideally, psychological assessment of parent reactions to the diagnosis should be an integrated component of clinical care. Although the RDI is not suitable for routine use in clinical settings, due to the recording and coding required, the interview can be readily adapted for incorporation into psychosocial assessments with parents. Parents assessed as unresolved should be offered counseling aimed at relieving diagnosis related grief and promoting balanced expectations of the child’s future (Ormerod & Huebner, 1988). An important avenue for future research is the evaluation of counseling and other supportive interventions in assisting unresolved parents in coming to terms with the diagnosis.

Conflicts of interest: None declared.

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References


