Emotional Burden and Coping Strategies of Parents of Children with Rare Diseases

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Introduction

According to the European Council, a disease is defined as rare when it affects fewer than 1 in 2000 citizens («Regulation (EC) No 141/2000 of the European Parliament and of the Council of 16 December 1999 on orphan medicinal products», 1999). Rare diseases are a heterogeneous group of chronically debilitating and, in some cases, even life-threatening diseases.

The conceptual analysis of stress and coping offered by Lazarus and Folkman (Lazarus & Folkman, 1984) is a cornerstone of the psychological and medical approach to trauma and life–threatening conditions; they define coping as the process of executing the response to a threat, and argue that the severity of perceived stress in a subject with a life–threatening condition depends on the successful outcome of primary appraisal (the process of perceiving a threat to one's self) and secondary appraisal (the process of bringing to mind a potential response to the threat).

In an attempt to better understand the outcome of cognitive appraisal of a health threat, Leventhal (Leventhal, Diefenbach, & Leventhal, 1992) suggested an illness representation model based on five attributes of the illness: (1) identity (disease label and its symptom indicator); (2) time-line (the threat is acute, cyclic, or chronic); (3) physical, social, and economic consequences; (4) antecedent causes (injury, infection, genetic weaknesses); and (5) potential for cure and/or control.

Both rare and chronic diseases may have genetic hereditary cause, a chronic course of illness, and degenerative and debilitating physical consequences with social and economic impairment. However, because of the lack of medical knowledge, the identity of a rare disease is less definite and its clinical management is more difficult than in the case of chronic diseases, due to symptomatic treatment and the use of orphan drugs. These peculiar features of rare diseases, by hindering the illness representation process, may affect primary appraisal, and subsequently also secondary appraisal and the coping process.

Coping strategies and the psychological burden of parents of CCD have been analyzed in detail and reviewed (Clarke, McCarthy, Downie, Ashley, & Anderson, 2009; Grootenhuis & Last, 1997; Pai et al., 2007). Discordant results have been reported concerning levels of anxiety, depression, and quality of life in parents of CCD, the differences being at least partly due to different types of assessment and different study designs (i.e. validated vs. non-validated tool; cross-sectional vs. longitudinal approach). Similarly, inconclusive evidence showed that psychological distress was higher in mothers than fathers, even during early stages of the illness (Clarke et al., 2009; Grootenhuis & Last, 1997; Raina et al., 2005; Sloper, 2000). Most longitudinal studies not only confirmed

that anxiety, depression, and insomnia increased shortly after diagnosis (Grootenhuis & Last, 1997; Sawyer, Crettenden, & Toogood, 1986), but also reported that sleep disturbances and depression remained higher than in healthy controls several months after diagnosis (Magni, Carli, De Leo, Tshilolo, & Zanesco, 1986; Sawyer et al., 1986). Conversely, a few studies suggested that the emotional consequences decline over time (Fife, Norton, & Groom, 1987; Kupst et al., 1995)

As regards gender differences in parental coping, it has been reported that mothers of CCD more commonly cope using emotion-focused strategies, such as seeking of emotional social support (Goldbeck, 2001; Koch, Härter, Jakob, & Siegrist, 1996), whereas fathers are more likely to use problem-focused strategies such as seeking of instrumental social support (Goldbeck, 2001; Hoekstra-Weebers, Jaspers, Kamps, & Klip, 1998). Some studies have taken a different approach to the parental coping process, considering the couple's coping style as a whole; they differentiate between a synchronous type and a dissimilar type (Adams-Greenly, 1986; Christ & Flomenhaft, 1982). More recently, Goldbeck (Goldbeck, 2001) studied the coping dissimilarities between mothers and fathers in childhood cancer, and found that synchronicity or dissimilarity in different types of coping strategies influence the child's quality of life.

Focusing on the adaptation of parents of CRD, some studies have merely investigated the level of family stress and discomfort, suggesting a positive correlation with the severity of the illness, without assessing the adaptive process used to deal with the family's emotional burden (Brewer et al., 2008; Bruns & Foerster, 2011; Latessa & Frasier, 2007; Tabolli et al., 2010). Other studies have examined the factors that affect the parental emotional burden and perceived stress, reporting that a good relationship with the caregiver, especially with the physician, has a beneficial impact (Lammens et al., 2011; Zierhut & Bartels, 2012). Finally, some studies have stressed the crucial role of the exchange of information between physician and family, which parents and voluntary associations had already indicated as a distinctive feature of rare diseases (Gundersen, 2011; Mura, Bhat, Pisano, Licci, & Carta, 2012). However, a definitive and consistent assessment of the outcome of the primary and secondary appraisal processes in parents of CRD, in terms of residual emotional burden and coping strategies, has not yet been performed.

The present study aimed to describe differences, between parents of CRD and parents of CCD, in emotional burden (which includes quality of life, anxiety, depression, mood states profile) and coping strategies, and to evaluate gender differences in parents of CRD and in those of CCD, at a time distant from the impact of diagnosis.

Method

Partecipants

All 30 voluntary associations belonging to the *Rete Interregionale per le Malattie Rare del Piemonte e della Valle d'Aosta* (Italy) were asked to provide data of parents who both 1) had given their permission to be contacted and invited to participate in future research studies in the field of rare diseases and 2) had a child who met the inclusion criteria. All parents were initially contacted by telephone to obtain preliminary verbal consent to participation in the study; they subsequently signed a written informed consent form at the time of assessment.

All parents of CCD accessing to the Department of Pediatric Endocrinology and Diabetology, Regina Margherita Children's Hospital (Turin, Italy) who met the inclusion criteria were asked to participate in the study. The inclusion criteria for CRD and CCD were: 1) diagnosis of the illness during childhood; 2) age below 18 years; 3) communication of the diagnosis one year or more prior to enrolment.

A unique identification code was assigned to each couple of parents in order to maintain the collected data in an anonymous form. Assessment was in two stages: first, socio-demographic information was collected by means of face-to-face semi-structured interview (i.e. gender, age, education, employment) and, secondly, assessment tools were administrated separately to both parents.

Measures

The Satisfaction With Life Scale (SWLS) (Diener, Emmons, Larsen, & Griffin, 1985) is a 5-item self-report scale that measure quality of life in terms of subjective well-being and satisfaction with life. According to the Italian version of SWLS (Di Fabio & Ghizzani 2006), the parents enrolled were requested to rate each of the five statements of SWLS on a 7-point Likert scale (1=strongly disagree, 7=strongly agree). The SWLS assesses six levels of satisfaction with life: extremely dissatisfied (5-9), dissatisfied (10-15), slightly dissatisfied (16-19), slightly satisfied (21-25), satisfied (26-30), and extremely satisfied (31-35). A neutral point on the scale is located at the score of 20 and indicates that the participant is neither satisfied nor dissatisfied with life. The SWLS has shown similar validity to other quality-of-life assessment tools and good temporal stability (Pavot & Diener, 2008).

The Profile of Mood States (POMS) (McNair, Lorr, & Droppleman, 1981) is a self-report tool that assesses transient, distinct mood states by evaluating six affective-emotional components of well-being: tension-anxiety, depression-dejection, anger-hostility, vigor-activity, fatigue-inertia, and confusion-bewilderment. The Italian version

of POMS (Farnè, Sebellico, Gnugnoli, & Corallo 1991) contains 58 items that subjects rate on a 5-point Likert scale. The Total Mood Disturbance (TMD) index is provided by the total POMS score.

The Coping Orientation to Problem Experienced (COPE) is a self-report multidimensional scale developed by Carver et al. (Carver, Scheier, & Weintraub, 1989) to assess coping strategies, following Lazarus and Folkman's conceptualization of the appraisal process (Lazarus & Folkman, 1984). In the present study, we used the Italian validated version (Sica, Novara, Dorz, & Sanavio, 1997a) that consists of 60 items, corresponding to 15 discrete and most common coping strategies. Each item provides a 4-point answer (from 1="I usually do not" to 4="I mostly do"). The Italian version of COPE has shown good psychometric properties, which are comparable to those of the original version (Sica, Novara, Dorz, & Sanavio, 1997b).

The Hamilton Rating Scale for Depression (HAM-D) (Hamilton, 1960) is a 21-item clinician-administered interview. The first 17 items are scored, while the remainders provide additional clinical information. Total scores range from 0 to 30, with five levels of severity: normal (0-7), mild (8-13), moderate (14-18), severe (19-22), very severe (23 and above). The HAM-D has been widely used in research to evaluate the severity of depression (Hamilton, 1980).

The Hamilton Rating Scale for Anxiety (HRSA) (Hamilton, 1959) is a 14-item clinician-administered interview that assesses both psychic and somatic symptoms of anxiety. Total anxiety scores range from 0 to 56, with four levels of severity: normal (0-13), mild (14-17), moderate (18-24), severe (25 and over). In the present study we used the Italian version of HAM-D and of HRSA (Conti, 2000).

Statistical analysis

Categorical variables were compared using Pearson's $\chi 2$ test or Fisher's exact test, depending on expected frequencies in the two groups of parents. Mean differences in continuous variables were evaluated either by the independent-samples t test or by Mann-Whitney's U test, depending on whether the distribution of variables was normal or non-normal (determined by the Shapiro-Wilk test).

A correlation matrix was produced for the two groups of parents, using all burden features (SWLS, POMS TMD, POMS sub-scales, HRSA, HAM-D) and all coping strategies. Bivariate correlations were evaluated in terms of Pearson's r coefficient or Kendall's tau (τ) coefficient, depending whether distribution of the variables was normal or non-normal.

A p-value of 0.05 was used to designate statistical significance; p-values resulting from multiple comparisons were corrected by Bonferroni's method, in order to control the family error rate.

All computations were performed using the IBM SPSS Statistics for Windows software package (Version 19.0. Armonk, NY: IBM Corporation).

Results

25 out of 30 contacted rare disease voluntary associations responded but only 13 provided data of family who met the inclusion criteria. All the involved families (both of CRD and of CCD) gave their informed consent. Therefore the enrolled sample consist of 55 parents of CRD, equal to 30 mothers (of which 5 single) and 25 fathers, and 56 parents of CCD, equal to 30 mothers (of which 4 single) and 26 fathers.

Table 1 lists the diagnoses of the children of the recruited parents.

<Insert Table 1 about here>

Parents of CRD vs. parents of CCD

Comparison of general and socio-demographic characteristics showed no statistically-significant difference between parents of CRD and parents of CCD (Table 2).

<Insert Table 2 about here>

As regards parental burden (Table 3), parents of CRD neither showed significantly lower satisfaction with life on the SWLS scale, nor presented a higher level of depression on HAM-D, than those of CCD. Although the two groups showed no statistically-significant difference in TMD score, parents of CRD presented significantly higher confusion-bewilderment and significantly lower vigor-activity scores than those of CCD.

HRSA scores of parent of CRD were considerably higher than those of parents of CCD. In particular, parents of CRD had significantly higher scores on several HRSA items: fears ($U=954.50$, $p<.001$, $r=.38$), insomnia ($U=888.50$, $p<.001$, $r=.37$), depressed mood ($U=983.00$, $p<.001$, $r=.33$), somatic sensory ($U=1012.00$, $p<.001$, $r=.41$), respiratory symptoms ($U=1103.50$, $p=.001$, $r=.31$), and autonomic symptoms ($U=745.00$, $p<.001$, $r=.47$).

<Insert Table 3 about here>

With regard to the results of COPE (Table 4), parents of CCD showed higher scores for active planning, seeking of instrumental social support, positive reinterpretation and growth than those of CRD; conversely, turning to religion was higher among parents of CRD than those of CCD.

<Insert Table 4 about here>

As shown in Table 5 and in Table 6, the burden features showed closer correlations with the coping strategies among parents of CCD than those of CRD. In particular, among parents of CCD (Table 5), several problem-focused (i.e., active coping, planning, and seeking of instrumental social support) and emotion-focused coping strategies (i.e., seeking of emotional social support, focus on and venting emotion, and positive reinterpretation and growth) were correlated with vigor-activity.

Conversely, among parents of CRD (Table 6), focus on and venting emotion was correlated with HRSA, behavioral disengagement was correlated with depression-dejection, and mental disengagement was correlated with both tension-anxiety and TMD.

<Insert Table 5 and Table 6 about here>

Mothers vs. fathers of CRD

The SWLS, TMD and POMS sub-scales, HAM-D, and HRSA scores showed no significant differences between mothers and fathers of CRD, whereas, among CCD group, mothers showed higher HRSA scores than fathers (Table 7). As regards coping strategies, in CRD group, mothers had significantly higher scores than fathers in seeking of emotional social support and focus on and venting emotion coping strategies. Conversely, in CCD group, mothers showed higher scores in active coping, seeking of instrumental social support, positive reinterpretation and growth, and turning to religion (Table 8).

<Insert Table 7 and Table 8 about here>

Discussion

The study's primary aim was to compare family burden and coping strategies in parents of CRD with those in parents of CCD. As regards emotional burden, although parents of CRD were more anxious, confused and bewildered, and less active and vigorous, than those of CCD one year after diagnosis, both groups presented good levels of satisfaction with life, ranging from slightly satisfied to satisfied; low levels of depression, ranging from normal to mild; and no severe anxiety symptoms. As regards way of coping, parents of CCD more often appeared to deal with their child's chronic illness by taking active steps to circumvent it (active coping), construing it in positive terms (positive reinterpretation), and seeking advice, assistance or information about it (seeking of instrumental social support) than parents of CRD who, conversely, more often turned to religion in order to cope with their child's rare disease. According to Carver et al (1989) the religious coping is an alternative strategy to active coping,

seeking of emotional support, and positive reinterpretation and growth. The falling back on alternative coping strategies (turning to religion) by parents of CRD could be explained by the really problematic cognitive representation of child's rare disease.

According to Leventhal's five-attributes model (Leventhal et al., 1992), some common features of rare diseases, such as the lack of available medical knowledge, the indefinite identity of the disease, the uncertain course of the illness, and the difficulty of clinical management connected with symptomatic treatment and orphan drugs, could really hinder illness representation during the primary cognitive appraisal of child's rare diseases, and then they could lead parents to adopt alternative coping strategies (turning to religion) instead of more useful problem-focused (active coping, seeking of instrumental social support) and emotion-focused strategies (positive reinterpretation and growth) during secondary cognitive appraisal.

Although some previous studies reported both the positive correlation between religious coping and the mental-health-related outcome of several life–threatening conditions (Carver et al., 1989; Pargament, Smith, Koenig, & Perez, 1998) and the predictive value of religious coping for health and well-being regardless of nonreligious coping strategies (Nooney & Woodrum, 2002; Pargament, 2001), in the present study, the adoption of turning to religion as coping strategy by parents of CRD doesn't seem to affect any burden features, whereas the preferential use of active coping, seeking of instrumental social support, and positive reinterpretation and growth by parents of CCD appear to enhance their vigor-activity.

Moreover, considering also the strategies that did not differ between groups, the use of focus on and venting emotion strategy led to high level of vigor-activity in parents of CCD, whereas in parents of CRD it was correlated with high level of anxiety, suggesting that this emotion-focused strategy may be useful to deal with child's chronic illness than with child's rare illness. Nevertheless, only among parents of CRD the use of disengagement strategies was associated with emotional distress in term of depression-dejection, tension-anxiety and TMD. These finding are consistent with those of previous studies reporting the beneficial impact of a good relationship with the caregiver, especially with the physician, upon the parental emotional burden and perceived stress (Lammens et al., 2011; Zierhut & Bartels, 2012). Indeed, the lack of information about rare diseases should not result in a reduced availability to pay attention to emotional burden of parents of CRD.

Taken together the findings, concerning the first aim of the present study, seem to suggest that both parents of CRD and those of CCD achieved good overall adaptation one year after child's illness diagnosis, but they did so

in different ways, showing slightly differences in emotional burden, probably due to the lack of available medical knowledge about diagnosis, prognosis and treatment of rare diseases.

As regards the second aim of this study, mothers of CRD required greater use of emotion-focused coping strategies (i.e., seeking of emotional social support and focus and venting emotion) than fathers to achieve the same burden features. Conversely, in CCD group, although mothers tended to make greater use of active coping, seeking of instrumental social support, positive reinterpretation and growth, and turning to religion than fathers, they showed higher level of anxiety than fathers. The comparison with previous studies (Goldbeck, 2001; Hoekstra-Weebers, Jaspers, Kamps, & Klip, 1998; Koch, Härter, Jakob, & Siegrist, 1996), which reported more frequent use of emotional-focused strategies by mothers than fathers of child with cancer, is really difficult owing to differences in the assessment tools, design (longitudinal vs. cross-sectional), sample and time-lag from diagnosis. However, it is possible to argue that mothers of child with cancer, of CRD, and of CCD seems to make greater use of emotion-focused strategies than fathers, though this finding is more noticeable in mothers of child with cancer and in those of CRD than in those of CCD who also use problem-focused strategies more frequent than fathers. According to Lazarus and Folkman (Lazarus & Folkman, 1984) the emotion-focused strategies are more likely to be used when the health problem is appraised as uncontrollable and thus, applying the Leventhal's five-attributes model of illness representation, it is possible to suggest that child's rare disease as well as child's cancer is perceived by mothers as more threatening than child's chronic disease. Anyway, considering the couple's style as a whole according to Adams-Greenly and Christ & Flomenhaft (Adams-Greenly, 1986; Christ & Flomenhaft, 1982), both parents of CRD and CCD showed a dissimilar type of coping like parents of child with cancer (Goldbeck, 2001).

The main findings of the present study confirm the importance of training and information-providing in the field of rare diseases. The availability of more information might improve the perception of the illness by caregiver and parents of a CRD, as has been suggested by other researchers (Gundersen, 2011; Mura, Bhat, Pisano, Licci, & Carta, 2012), and may foster the use of problem-focused coping strategies. In particular, physicians' training may improve their leadership skills and thus facilitate the use of 'seeking of support' strategies by parents of CRD.

The most notable limitations of the present study are sample size and study design. The small sample size is common critical issue within the field of rare disease (Brewer et al., 2008; Bruns & Foerster, 2011; Lammens et al., 2011; Tabolli et al., 2010). Clearly, a larger group of parents of CRD would ensure greater and more adequate

statistical power for the gender comparisons. Moreover, as the sample was enrolled in NW Italy, it may not be properly representative of parents of Italian CRD; the results could therefore not be generalized.

Furthermore, a longitudinal study design would enable any changes in coping strategies and emotional burden over time to be evaluated, and the predictive value of coping strategies used immediately after diagnosis, for the emotional burden experienced during subsequent months, could also be examined.

Further studies might expressly investigate, in a larger sample, gender differences in the adaptive process of parents of CRD, in order to confirm the difference in coping strategies between mothers and fathers observed in this study, and to evaluate whether this dissimilarity may lead to a positive outcome in terms of emotional state and quality of life of the parents as a couple.

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