Coping Strategies, Stress, and Support Needs in Caregivers of Children with Mucopolysaccharidosis

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Abstract The mucopolysaccharidoses are a set of rare, inherited conditions that can have a catastrophic impact on those affected and their families. Because of the rarity of these disorders, little is known regarding the challenges faced by families of those affected and what coping mechanisms are commonly used. Coping is a way to manage demands that occur in one's environment or within oneself. Medical social workers historically have facilitated this process while providing support to patients who are responding to pressures of their diagnosis and the system.

A questionnaire of demographics and qualitative questions, along with the Pediatric Inventory for Parents (PIP) and Brief COPE, was sent by electronic survey to caregivers of children with MPS. The results of Brief COPE showed that problem-focused coping was more frequently used than emotion-focused ($p < 0.001$) or dysfunctional coping ($p < 0.0001$). Acceptance was the most frequently used coping strategy ($p < 0.05$). The results of PIP showed that emotionally distressing events were the most difficult ($p < 0.001$), while events related to medical care occurred at the highest frequency ($p < 0.001$). Psychosocial support provided by medical social workers significantly increased acceptance of caregivers ($p = 0.04$). Guidance on what to expect provided by any member of the care team increased denial ($p = 0.02$) and the difficulty of emotional distress

($p = 0.04$). This study identified commonly used coping strategies and measured stress among caregivers of children with MPS, as well as access to and use of psychosocial support services. Results highlight the urgency to improve the coverage and quality of psychosocial support and other support services.

Introduction

The mucopolysaccharidoses (MPSs) are a group of rare, inherited metabolic disorders, which belong to a family of more than 40 identified lysosomal diseases (Schultz et al. 2011). Major symptoms of MPS diseases include cardio-pulmonary disease, growth delay, skeletal dysplasias, organomegaly, and shortened life span. Patients with severe MPSs I, II, III, or VII exhibit progressive neurodegeneration and mental retardation. Current therapies for treatment include hematopoietic stem cell transplantation (HSCT) and enzyme replacement therapy (ERT). HSCT has a high rate of mortality (10–15%) and severe morbidity. ERT has negligible neurological improvements because the blood-brain barrier prevents intravenously infused enzyme from entering the brain.

The impact of parenting children with MPS can be “devastating” with major challenges being emotional distress, financial strains, and severe behavior difficulties (Nidiffer and Kelly 1983). A study conducted in the UK reported that families with children with MPS withdrew from social outings and restricted visitors to their homes, reducing the social support they could access (Bax and Colville 1995). Characteristics, such as child behavior difficulties or levels of social support, may contribute to parental distress. Grant et al. (2013) showed that caregivers

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of children with MPS III were less future-oriented and goal-directed than caregivers of children with intellectual disabilities. Due to extreme heterogeneity in symptom presentation and lack of established phenotype-genotype correlation (Ou et al. 2017), it is difficult for professionals to predict disease progression (Valstar et al. 2008). Previous findings suggest that the unpredictability of MPS may contribute to increased stress levels.

Medical social workers have a long history of providing psychosocial support for physical and mental health concerns (Globerman and Bogo 2002). Regarding patient experience, Gibbons and Plath (2009) determined that medical social workers' relationship- and rapport-building skills and ability to provide empathy, practical assistance, and advocacy were essential to their role on the multidisciplinary team. Medical social workers utilize strong psychosocial assessment skills that incorporate systems and community knowledge to solve problems and provide emotional support. They practice holistically as they work with psychosocial issues that may be a barrier to achieving optimal health and help address the relationship between diagnosis and environment (Craig et al. 2015; Lazarus 1993). By gathering data to inform best practices in psychosocial intervention for caregivers, it would be expected that children have better outcomes in coping with their health-care needs. Medical social workers historically have facilitated development of coping skills while providing consistent support to patients who are responding to pressures of their diagnosis and the system (Craig et al. 2015; Lazarus 1993).

Objectives

Identify coping strategies used and measure stress among caregivers of children with MPS, and determine how these caregivers access and use psychosocial support services. Each type of MPS has differences in presentation; variation in stress and coping among syndromes was not measured in this study.

Methods and Materials

Methods

A mixed methods design was used. Two validated measures, the Brief COPE and the Pediatric Inventory for Parents (PIP), were adapted to measure coping strategies and stress among caregivers of children with MPS, respectively. Participants were asked a mix of open- and closed-ended questions about their experiences with the medical system and access to and use of psychosocial support services.

Participants

A survey link was sent through the National MPS Society to caregivers of children with MPS. Eligible participants were over the age of 18 years, and a primary caregiver of a child with an MPS disorder aged 18 years or younger.

Brief COPE

The Brief COPE (Carver 1997) is a self-report questionnaire with 14 subscales assessing coping techniques including self-distraction, active coping, denial, substance use, use of emotional support, use of instrumental support, behavioral disengagement, venting, positive reframing, planning, humor, acceptance, religion, and self-blame. Items are rated on a Likert scale (1 = I haven't been doing this at all to 4 = I've been doing this a lot). Reliability and validity of this measure have been documented within medical populations (Yusoff et al. 2010). Two subscales had poor internal consistency in the current study ($\alpha = 0.477$ for behavior disengagement and 0.438 for self-distraction). Internal consistency for the remaining subscales ranged from 0.519 to 0.904.

The subscales were grouped into three composite scores: problem-focused coping (active coping, planning, use of instrumental support), emotion-focused coping (acceptance, positive reframing, humor, religion, use of emotional support), and dysfunctional coping strategies (behavior disengagement, denial, self-distraction, self-blame, substance use, venting). The internal consistency of each category was analyzed ($\alpha = 0.575$ for problem-focused, 0.557 for emotional-focused, and 0.548 for dysfunctional coping strategies).

Pediatric Inventory for Parents

The PIP (Streisand et al. 2001) is a self-report measure of parenting stress related to caring for a child with an illness. There are four subscales with frequency and difficulty scores: communication, medical care, emotional distress, and role function. This measure was selected given the life-limiting nature and high level of physical health difficulties of MPS diseases. The reliability and validity of PIP were reported to be high (α range, 0.8–0.96) in a previous study (Grant et al. 2013). Since some participants skipped questions in this inventory, the total score of each subscale could not accurately reflect the data. Therefore, the average frequency and difficulty scores of each subscale were used for analysis. All subscales had good internal consistency (Cronbach's α 0.623–0.896).

Questionnaire

Participants were asked a mix of open- and closed-ended questions. A checklist was provided about services offered at time of diagnosis, including guidance on what to expect related to disease process by any provider on the care team (GE), education on behavioral changes that may occur related to the diagnosis and appropriate interventions (EDB), assistance with practical concerns such as insurance and finances (AIF), and opportunity to process emotions (EMS). Participants were asked about access to a licensed social worker and in what type of setting. Open-ended questions were included for deeper understanding of overall experience, stress, and support needs.

Data Analysis

Less than 5% of data was missing on any individual scale. For each scale, Little's MCAR test was not significant (all p values > 0.05) indicating that the data were missing completely at random. SPSS 20.0 was used to analyze data, p value < 0.05 (two-tailed tests) to denote statistical significance. The assumption of normality was not met for a number of outcome variables; therefore nonparametric tests were used.

Results

Demographic Data

Eleven of 93 participants did not provide enough information for analysis and were excluded in further analyses. Twenty-one participants (25%) were aged 25–34, 42 (50%) were aged 35–44, and 16 (22.6%) were aged 45–64. Sixty-two participants (81.7%) were Caucasian, eight (9.8%) were Hispanic, four (4.8%) were African, and three (3.7%) were Asian. In terms of education background: 24 (29.3%) undergraduate degree, 12 (14.6%) graduate degree, 11 (13.4%) high school degree, and 8 (9.8%) associate degree. Sixty-seven (81.7%) were married, six divorced, six single, and three separated. Sixty-six participants (80.5%) had one child with MPS, while 16 had two children with MPS. Out of 100 children with MPS, 34 were female (age, 8.7 ± 4.8 years old) and 56 were male (age, 8.2 ± 4.5 years old). The participants were caregivers to children with various MPS disorders: 21 MPS I, 19 MPS II, 14 MPS IIIA, 9 MPS IIIB, 7 MPS IVA, and 9 MPS VI.

Support Services

Participants were asked about experiences at time of diagnosis and were provided with a checklist inquiring about services provided to them. Of 63 respondents, 38

(58%) were offered general guidance on what to expect (GE) related to disease process. Nineteen (29%) were offered education on behavioral changes that may occur related to the diagnosis and appropriate interventions (EDB), 18 (28%) were offered assistance with practical concerns such as insurance and finances (AIF), and 13 (20%) were offered opportunity to process emotions (EMS). Of those not offered these services, 53 (81.5%) participants would have been interested in EMS, 46 (70.8%) interested in EDB, 40 (61.5%) in AIF, and 35 (53.9%) in GE.

Of 75 respondents, 57 (76%) preferred that help be offered, while 18 (24%) preferred to ask for help. Fifty-nine of 72 respondents (77.6%) participated in online support networks (e.g., Facebook), 30 (39.4%) attended in-person patient/family meetings specific to MPS or lysosomal storage disease (LSD), 17 (22.4%) participated in counseling services, and 3 (3.9%) participated in in-person support groups.

Of 73 respondents, 9 (12.3%) had access to a medical social worker that works specifically with children with MPS. Out of these nine, five had regular contacts with this individual, indicating the rarity of social work professionals working specifically with children with MPS. Alternatively, 30 (41.7%) out of 72 participants had access to a social worker in other settings: 8 in primary care clinic, 14 from the child's school, and 4 who were private therapists or counselors.

Coping Strategies

The individual scales of Brief COPE were evaluated and illustrated in Fig. 1. Acceptance had the highest score (3.4 ± 0.84) and was significantly higher ($p < 0.05$) than other scales except active coping (3.0 ± 0.96) and planning (3.1 ± 0.92). Denial, substance use, and behavior disengagement have the lowest score: 1.4 ± 0.80 , 1.2 ± 0.51 , and 1.3 ± 0.65 , respectively. The 14 scales were classified into three categories as previously described (Cooper et al. 2008). There was significant difference between these categories: problem-focused coping the highest (2.8 ± 0.56) and dysfunctional coping the lowest (1.7 ± 0.42) (Fig. 1). The correlation of individual scales was evaluated by Pearson's correlation (Table 1). Further, the correlation between the three categories was analyzed: emotional-focused coping was significantly correlated with problem-focused coping ($r = 0.508$, $p < 0.0001$).

To evaluate the relationship between psychosocial support and coping strategies, the individual scales of the Brief COPE and psychosocial support were analyzed by a general liner model. Five participants had regular contact with a social worker that specifically works with children with MPS; therefore we compared all participants that had

Fig. 1 Comparison of scales of Brief COPE and stress in caregivers of MPS children. (a) Average score of each individual scale. For simplicity consideration, the statistical significance was not marked. (b) Average score of each category. (c, d) Indicate stress difficulty and

frequency, respectively. *CM* communication, *MC* medical care, *ED* emotional distress, *RF* role function. Data were represented as mean $\pm$ standard errors. ** $p < 0.01$, *** $p < 0.001$, **** $p < 0.0001$

access to social workers across settings with those who had no access to social workers. Multiple factors including age, marriage status, education level, number of children with MPS, and types of initial support received were included. Psychosocial support from a social worker significantly increased acceptance ($p = 0.043$) but had no significant impacts on other individual scales or categories. These results demonstrated that psychosocial support increased acceptance of caregivers but required refinement in improving other positive coping strategies.

Self-blame decreased as education levels of caregivers increased ($p = 0.014$), while positive reframing increased as number of children with MPS increased ($p = 0.008$). Denial decreased as age of caregivers increased ($p = 0.039$), and EDB significantly decreased denial ($p = 0.018$).

Unexpectedly, GE increased denial ($p = 0.021$), indicating the necessity to improve this in the future.

Stress

To investigate the stressors of caregivers of children with MPS, the PIP was employed. There are four subscales with frequency and difficulty scores: communication (CM), medical care (MC), emotional distress (ED), and role function (RF). A total of 45 participants provided enough information for analysis. Emotional distress was related as significantly more difficult than any other category ($p < 0.0001$) (Fig. 1c). In terms of frequency, emotional distress and medical care were significantly higher than the other two categories ($p < 0.0001$) (Fig. 1d). These results

Table 1 Correlation between individual scales of Brief COPE

Correlations	Self-distraction	Behavior disengagement	Use of instrumental support	Planning	Denial	Self-blame	Substance use	Venting	Religion	Active coping	Use of emotional support	Positive reframing	Humor	Acceptance
Self-distraction														
Behavior disengagement														
Use of instrumental support					0.256*	0.521**	0.256*	0.297*					0.368**	
Planning														
Denial									0.292*	0.527**	0.408**	0.278*		
Self-blame						0.341**								-0.363**
Substance use														
Venting								0.424**					0.429**	
Religion														
Active coping												0.530**		
Use of emotional support										0.265*		0.458**		
Positive reframing														0.354**
Humor														0.257*
Acceptance														

For simplicity consideration, the correlation between two factors was only shown once. * $p < 0.05$, ** $p < 0.01$

showed that emotional distress was the major contributor to stress of caregivers. Pearson's correlation was performed among frequency and difficulty of all categories. As shown in Table 2, there were significant correlations in difficulty and frequency between each category.

To determine the relationship between psychosocial support and stress, the individual scales in the PIP and psychosocial support were analyzed by a general liner model. Multiple factors including age, marriage status, education level, number of children with MPS, and types of initial support received were included. Psychosocial support had no significant impact on stress levels in the individual scales or the aforementioned four categories. These results indicate a necessity to improve psychosocial support to decrease stress of caregivers. When offered education on behavioral changes and interventions, communication difficulty significantly decreased ($p = 0.041$). The difficulty of role function decreased as ages of caregivers increased ($p = 0.028$). The frequency of emotional distress decreased as education levels and ages of caregivers increased ($p = 0.009$ and 0.031 , respectively). The difficulty of emotional distress decreased as education levels of caregivers increased ($p = 0.019$). However, when provided guidance on what to expect related to disease progression, emotional distress increased ($p = 0.044$), strongly suggesting the need to improve that discussion.

Correlation between coping strategies and stress was also evaluated. As shown in Table 3, emotional-focused coping was negatively correlated with the difficulty of medical care stress ($r = -0.31$, $p = 0.04$), while total coping scores were positively correlated with the frequency of role function stress ($r = 0.31$, $p = 0.04$). Individual scales of coping strategies showed that most dysfunctional coping strategies were correlated with stress.

Qualitative Questions

Qualitative questions were included for a deeper understanding of the unique needs and concerns of caregivers. Twenty-one percent of respondents rated online support groups and networking with other families as the most helpful service. Notably, 10.7% respondents received no support services. In terms of what services caregivers would like more of, 19 respondents (36.5%) mentioned emotional support services, and 28 respondents (53.8%) mentioned information on community resources. Finally, many respondents reported feelings of sadness, loneliness, and concerns about lack of support, information, and public awareness.

Discussion

Previous studies have identified unique challenges involved in parenting a child with MPS (Nidiffer and Kelly 1983; Bax and Colville 1995; Grant et al. 2013; Cooper et al. 2008; Ucar et al. 2010). This is the first study to measure stress, coping, and access to and use of social work support and other psychosocial services among caregivers of children with MPS.

Coping and Stress

Caregivers who used more problem-focused coping used more emotion-focused coping (and vice versa). In individual scales, positive correlations mainly existed between scales within the same category (e.g., self-distraction with substance use and venting, behavior disengagement with denial and self-blame, planning, and active coping). There was a negative correlation between denial and acceptance,

Table 2 Correlation between individual scales of PIP

Correlations	CM_F	MC_F	ED_F	RF_F	CM_D	MC_D	ED_D	RF_D
CM_F		0.554*	0.635*	0.611*	0.764*	0.550*	0.534*	0.453*
MC_F			0.642*	0.669*	0.450*	0.667*	0.575*	0.557*
ED_F				0.787*	0.731*	0.698*	0.867*	0.776*
RF_F					0.633*	0.628*	0.710*	0.849*
CM_D						0.786*	0.706*	0.668*
MC_D							0.699*	0.714*
ED_D								0.752*
RF_D								

For simplicity consideration, the correlation between two factors was only shown once

CM_F communication stress frequency, CM_D communication stress difficulty, MC_F medical care stress frequency, MC_D medical care stress difficulty, ED_F emotional distress frequency, ED_D emotional distress difficulty, RF_F role function stress frequency, RF_D role function difficulty

* $p < 0.01$

Table 3 Correlation between coping strategies and stress

	<i>r</i> value	<i>p</i> value	
Dysfunctional coping	0.6	<0.001	TSF
	0.65	<0.001	TSD
	0.47	0.001	CM_F
	0.63	<0.001	CM_D
	0.5	<0.001	MC_F
	0.62	<0.001	MC_D
	0.64	<0.001	ED_F
	0.57	<0.001	ED_D
	0.43	<0.001	RF_F
Emotional-focused	0.48	<0.001	RF_D
	−0.31	0.04	MC_D
Total coping	0.31	0.04	RF_F
Behavior disengagement	0.3	0.048	TSD
	0.33	0.026	CM_D
	0.31	0.041	ED_F
Self-distraction	0.3	0.041	MC_F
	0.3	0.047	MC_D
	0.3	0.048	RF_F
Venting	0.37	0.012	CM_D
	0.31	0.037	MC_D
Self-blame	0.62	<0.001	TSF
	0.56	<0.001	TSD
	0.5	<0.001	CM_F
	0.5	0.001	CM_D
	0.5	<0.001	MC_F
	0.44	0.003	MC_D
	0.65	<0.001	ED_F
	0.56	<0.001	ED_D
	0.47	0.001	RF_F
Denial	0.48	0.001	RF_D
	0.41	0.05	TSF
	0.48	0.001	TSF
	0.4	0.007	CM_F
	0.49	0.001	CM_D
	0.33	0.028	MC_F
	0.5	<0.001	MC_D
	0.43	0.003	ED_F
	0.38	0.01	ED_D
	0.03	0.032	RF_D

CM_F communication stress frequency, *CM_D* communication stress difficulty, *MC_F* medical care stress frequency, *MC_D* medical care stress difficulty, *ED_F* emotional distress frequency, *ED_D* emotional distress difficulty, *RF_F* role function stress frequency, *RF_D* role function difficulty, *TSF* total stress frequency, *TSD* total stress difficulty

which is unsurprising considering the incompatibility of these two strategies. Additionally, dysfunctional coping was

positively correlated with total stress and each category of stress (Table 3). Several dysfunctional coping strategies including venting, self-distraction, self-blame, behavior disengagement, and denial were correlated with stress indicating that dysfunctional coping may significantly increase stress. Further, emotion-focused coping was negatively correlated with the frequency of medical care stress, suggesting a positive impact of emotion-focused coping. Interestingly, there was a significant correlation between total coping scores and the frequency of role function stress ($p = 0.04$), which may be contributed by dysfunctional coping.

Psychosocial Support

Psychosocial support provided by medical social workers had positive impacts on acceptance. This could be because increased emotional support may help individuals develop ways to cope with MPS. EDB (education on behavioral changes and appropriate interventions) decreased denial and the frequency of communication stress; this kind of conversation provides practical tools to help manage behaviors. This supports the need for early intervention that provides psychosocial services and concrete support to caregivers. Services around emotional support services and education on community resources were specifically identified areas of need by caregivers. Furthermore, specific guidance on disease progression to help minimize denial and emotional distress should be provided.

Guidance on what to expect in disease progression requires remarkable improvements because it increased denial and emotional distress. These discussions need to be tailored with individual caregiver coping styles or stress levels in mind. Considering the impact of EDB on decreasing communication stress, more resource and attention should be allocated to this type of service. EDB offers practical advice for problem-solving and intervention which may increase caregiver confidence.

Other Factors

This study evaluated the impacts of several other factors (e.g., education level, age) on coping and stress. Higher education decreased self-blame and emotional distress; while older caregivers had less denial, the difficulty of role function stress and the frequency of emotional distress decreased. These are important factors to consider in terms of service outreach and provision. Younger and less educated caregivers may need more concrete and ongoing support from a social worker, while older and those with higher education may need less intense guidance. Although previous studies showed the importance of marital support (Grant et al. 2013), in this study marital status had no

impact on coping and stress. Collectively, these factors should be considered in data analysis of future studies.

Implications for Clinical Practice

Considering severe stress and lack of support, it is suggested that clinicians screen for anxiety and depression in caregivers and refer to appropriate psychological services as deemed appropriate. Screening tools provide a starting point for clinical interventions. Because those who had access to social workers reported an increase in the use of acceptance as a coping skill, inclusion of comprehensive social work services to assess and treat psychosocial needs is an important consideration. Respondents preferred proactive assessment and intervention. Early interventions to increase emotional- or problem-focused coping and decrease dysfunctional coping could be offered.

Online support groups and connection with other parents were rated as most helpful by caregivers. Online support groups are a cost-effective consideration to help reduce feelings of isolation and connect caregivers from geographically diverse locations. Social workers can help establish or be actively involved in such groups.

Social workers need to be better trained in MPS, in providing individualized caregivers services and psychosocial supports. Additionally, program development to better support caregivers in a variety of areas including one-to-one and group counseling and increased availability of social workers to caregivers is needed.

Future Research

A weakness of this study is selection bias. Those who participated may have an unknown reason for selecting and completing the questionnaire. Also, a larger sample size would have permitted separate analysis by gender, age, or type or phase of the disorder. Specifically, because stress and coping may look different among MPS syndromes.

Researchers should be aware of the impact of such studies on psychological functioning of caregivers. Future studies should consider applying a combination of psychometrically strong tools and qualitative interviews in a sensitive manner that respects the role of parent as expert-by-experience.

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Synopsis

Unique patterns of stress and coping strategies of caregivers of children with MPS were identified, as well as gaps in psychosocial support services to help inform best practice for the future.

Details of Author Contributions

Amy Schadewald was involved in conception and design of this study, critically revising the article, and interpretation of the qualitative data. Ericka Kimball was involved in conception and design of this study and critically revising the article. Li Ou was involved in analysis and interpretation of the data, drafting, and critically revising the article. All authors are in agreement with submission of this draft to JIMD.

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Competing Interest Statement

Amy Schadewald, Ericka Kimball, and Li Ou have no competing interests to declare.

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Details of Ethics Approval

This study was approved by the University of Minnesota Institutional Review Board (IRB, study #1702S0831) and supported by the National MPS Society.

Patient Consent Statement

Patient data was not used for this study.

Institutional Committee for Care and Use of Laboratory Animals

This article does not contain any studies with animal subjects performed by any of the authors.

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