

Psychological status of mothers of children with cystic fibrosis and primary ciliary dyskinesia

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Abstract

Objectives: We aimed to investigate depression, burnout, attitude, and burden of caregivers of children with cystic fibrosis (CF), and especially caregivers of children with primary ciliary dyskinesia (PCD) due to limited number of studies on this topic, and to compare them according to their children's clinical status. **Methods:** Clinical features and demographic data of children and their families were asked to caregivers in four pediatric pulmonology centers. Beck Depression Inventory (BDI), Maslach Burnout Inventory (MBI), Zarit Caregiver Burden Scale (ZCBS), and Parental Attitude Research Instrument (PARI) were administered to caregivers in both groups. Results were compared between the two groups. **Results:** In total, 131 children with CF and 39 with PCD and their caregivers were involved in study. All primary caregivers were mothers in both groups. Depression, burnout, and burden scores of mothers of children with CF were significantly higher than mothers of children with PCD ($p=0.017$, $p=0.024$, $p=0.038$, respectively). Burnout was higher in both CF and PCD groups with low family income ($p=0.022$, $p=0.034$). Number of hospital visits in the previous six months was correlated with burnout in both CF and PCD groups ($r=0.207$, $p=0.034$; $r=0.352$, $p=0.044$). **Conclusions:** Although mothers of children with CF have higher depression, burnout, burden, and negative attitudes towards children than mothers of children with PCD, it is disregarded in mothers of children with PCD. Psychological problems of mothers of children with CF and PCD may increase with frequent hospital visits, hospitalizations, low family income, number of children, and chronic disease in another child.

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Objectives: We aimed to investigate depression, burnout, attitude, and burden of caregivers of children with cystic fibrosis (CF), and especially caregivers of children with primary ciliary dyskinesia (PCD) due to limited number of studies on this topic, and to compare them according to their children's clinical status.

Methods : Clinical features and demographic data of children and their families were asked to caregivers in four pediatric pulmonology centers. Beck Depression Inventory (BDI), Maslach Burnout Inventory (MBI), Zarit Caregiver Burden Scale (ZCBS), and Parental Attitude Research Instrument (PARI) were administered to caregivers in both groups. Results were compared between the two groups.

Results : In total, 131 children with CF and 39 with PCD and their caregivers were involved in study. All primary caregivers were mothers in both groups. Depression, burnout, and burden scores of mothers of children with CF were significantly higher than mothers of children with PCD ($p=0.017$, $p=0.024$, $p=0.038$, respectively). Burnout was higher in both CF and PCD groups with low family income ($p=0.022$, $p=0.034$). Number of hospital visits in the previous six months was correlated with burnout in both CF and PCD groups ($r=0.207$, $p=0.034$; $r=0.352$, $p=0.044$).

Conclusions: Although mothers of children with CF have higher depression, burnout, burden, and negative attitudes towards children than mothers of children with PCD, it is disregarded in mothers of children with PCD. Psychological problems of mothers of children with CF and PCD may increase with frequent hospital visits, hospitalizations, low family income, number of children, and chronic disease in another child.

Introduction

Chronic diseases in childhood affect the lives of the children as well as their caregivers. Although caregiving is a normal part of being a parent, the level of care needed by children with chronic diseases increases the burden and stress on the caregiver and affects the routines of family life and socialization.¹Caregiver's psychological status, such as depressive symptoms, can also negatively affect the child's condition and outcomes.²

Cystic fibrosis and PCD have a significant impact on health and quality of life of patients.³ The pathogenesis in both primary ciliary dyskinesia (PCD) and cystic fibrosis (CF) includes impaired mucociliary clearance leading to intermittent or chronic pulmonary infections. Although PCD has similar characteristics to CF,

lung disease is usually milder than in CF.⁴ Patients with PCD have been shown to have similar respiratory function and magnetic resonance imaging findings as patients with mild CF.⁵ Patients with PCD generally have a longer life span and better prognosis.⁶ On the other hand, while CF can be diagnosed before clinical findings begin with newborn screening, patients with PCD can be diagnosed late with clinical findings.⁷ The frequency of chronic respiratory symptoms can potentially result in lifestyle restrictions in PCD.⁸ The caregivers of children with CF experience depression, unwillingness, dissatisfaction, and sometimes even negative feelings for the child due to long-term care. These emotions cause stress, which leads to physical, emotional, and social exhaustion.⁹ As most of the morbidity and mortality in CF is associated with respiratory infection and disease, clinical severity of lung disease plays a significant role in determining the caregiver's mood. The psychological status of caregivers of children with CF affects their daily tasks and their children's compliance with treatment regimens.¹⁰ In addition to the stress of managing daily care, parents also have to deal with health system issues.¹¹ Family relationships and the psychological functioning of the child with CF and family members may be affected. Social relationships can also be difficult to maintain, given the commitment to daily care and the uncertainty of the disease. Employment issues can contribute to financial distress, which has far-reaching effects on the functioning of the family system.¹² Although many studies are evaluating the psychological status of caregivers of children with CF, studies on caregivers of children with PCD are very limited.

The aim of this study was to evaluate the depression, burnout, caregiver burden, and attitude of caregivers of children with CF, and especially the caregivers of children with PCD due to the limited number of studies on this topic, and the relationship between the clinical characteristics of children, sociodemographic characteristics, and the psychological status of their caregivers.

Materials and Methods

All children with CF and PCD and their primary caregivers who were referred to four pediatric pulmonology centers and agreed to participate in the study were recruited between July 2017 and September 2019. Informed consent forms were obtained from the primary caregivers before data collection. The study was performed according to the principles of the Declaration of Helsinki and approved by the Ethics Committee of the Faculty of Medicine (No. 2017/984; June 16, 2017). CF was diagnosed with genetic analysis and/or sweat chloride test, and PCD was diagnosed with genetic analysis and/or high-speed video microscopy and/or transmission electron microscopic examination, and clinical findings.

Questionnaires regarding the children's clinical features and demographic data about the children and their families were administered to the caregivers. Information collected about the children included age, sex, ethnicity, body mass index (BMI), pulmonary function tests (PFT) results, history of sibling death, follow-up duration, number of hospital visits (both emergency and outpatient clinics) and hospitalizations in the previous 6 months, duration of hospitalizations in the previous 6 months, and the number of exacerbations in the last year. Information on the presence of bacterial colonization in the sputum culture of children both during hospitalization or outpatient clinic visits were obtained from their medical records. Colonization was defined as bacterial growth in a sputum sample with no clinical signs or symptoms.¹³ In the children's PFT results who were adaptable to PFT technique, FEV₁ (forced expiratory volume in the first second), FVC (forced vital capacity), FEF₂₅₋₇₅ (25-75% of forced expiratory flow) percent predicted by age, and FEV₁/FVC values were obtained from their medical records. The PFTs were performed according to the American Thoracic Society-European Respiratory Society ATS-ERS guidelines.¹⁴

Data of the caregivers were obtained from the demographic questionnaire. Information on factors that might affect the psychological status of the caregiver was collected such as their ages, ethnicity, higher education levels, occupational status, family income, physical or psychiatric disorders and treatments, consanguinity, the number of children, chronic disease in another child (if any), family type (nuclear or extended), the family members who were the primary caregivers, any persons helping with the children's care (defined as assistants), and any work or family problems (such as dismissal, inability to care for other children, parental problems) related to the children's hospitalization. Monthly family income levels were classified as below the hunger threshold, between the hunger and poverty thresholds, and above the poverty threshold according

to 2016 data of the Turkish Statistical Institute.¹⁵ The occupational status of the mothers was classified as homemakers, and blue and white-collar workers. Salaried professionals, which refers to general office workers and management, were defined as white-collar workers. Workers who performed manual labor, earned hourly wages or were paid piece wages according to the amount of work done were defined as blue-collar workers.

The Beck Depression Inventory (BDI), Maslach Burnout Inventory (MBI), Zarit Caregiver Burden Scale (ZCBS), and the Parental Attitude Research Instrument (PARI) were used for psychological measurements and attitudes towards the children of the primary caregivers. The Turkish versions of these scales have been validated and used in many previous studies.¹⁶⁻¹⁹ The modified Shwachman-Kulczycki Score (mSKS) was used for evaluating the clinical status of children with CF.

The BDI consists of 21 items and measures symptoms related to emotional, cognitive, physical, and motor functions in depression. The overall depression score was obtained by summing all the items' scores. Cronbach's alpha value of the inventory was found as 0.80.¹⁶ Scores between 0-9 were classified as no depression, between 10-18 as mild depression, between 19-29 as moderate depression, 30 and above as severe depression. The scores that can be obtained from the scale vary between 0-63.²⁰

The MBI assesses burnout and consists of 22 items divided into three subscales: emotional exhaustion, depersonalization, and personal accomplishment. Emotional exhaustion indicates the stress dimension of burnout and refers to a decrease in the individual's emotional and physical resources. Depersonalization represents the interpersonal dimension of burnout and refers to negative, rigid attitudes toward people, and unresponsiveness to work. Personal accomplishment refers to the sense of competence and achievement in one's work. A low score indicates that the person tends to evaluate themselves negatively. Cronbach's alpha value of the inventory was found as 0.93.^{17,21} Each subscale was scored according to the reference ranges of the MBI: for emotional exhaustion, between 0-16 as low, between 17-26 as moderate, 27 or above as high; for depersonalization, between 0-6 as low, between 7-12 as moderate, 13 or above as high; for personal accomplishment, 31 or less as low, between 32-38 as moderate, and 39 or above as high. The total scores obtained range from 0-54 for emotional exhaustion, 0-30 for desensitization, and 0-48 for personal accomplishment. Burnout was defined using the updated Maslach-recommended criteria of "high emotional exhaustion and high depersonalization" or "high emotional exhaustion and low personal accomplishment".²²

The ZCBS consists of 22 items and assesses caregivers' perceived burden. Each item is scored on a 5-point Likert scale (0=*never*, 1=*rarely*, 2=*sometimes*, 3=*quite often*, 4=*almost always*). A total score between 0-20 points indicates no burden, between 21-40 illustrates mild burden, between 41-60 shows moderate burden, and between 61-88 points indicates severe burden. Cronbach's alpha value of the ZCBS was found as 0.83.^{18,23}

PARI was used to determine the attitudes of caregivers towards their children. PARI, which is a Likert-type scale, consists of 60 items and five subscales. These subscales are over-protectiveness, democratic attitude, rejection of the homemaker role, marital conflict and strictness. For each statement, one of the options "I find it very appropriate" (4), "I find it quite suitable" (3), "I find it somewhat appropriate" (2), "I do not find it suitable at all" (1) is selected. The option number marked for each item is evaluated as the item score. Responses to items 2, 29, and 44 are scored in reverse. A separate score is obtained for each subscale. There is no total score. A high score from the subscale indicates that the attitude reflected by that dimension is approved. The highest scores that can be obtained from the subscales are 64 for over-protectiveness, 36 for democratic attitude, 52 for rejection of the homemaker role, 24 for marital conflict, and 64 for strictness. Except for democratic attitude, high scores from the subscales indicate the presence of a negative parental attitude. The Cronbach's alpha coefficient of the scale was found as 0.89.^{19, 24}

The mSKS is calculated in four domains, including general activity, physical examination findings, nutritional status, and radiological findings. Each parameter was scored from 5 (i.e. severely impaired) to 25 (i.e. normal), which resulted in a total score categorised as excellent (86-100), good (71-85), mild (56-70), moderate (41-55) and severe (<40).^{25,26}

In the descriptive statistics section, categorical variables are presented as numbers, percentages, and con-

tinuous variables as mean \pm standard deviation and median (range). Pearson's Chi-square test and Fisher's exact test were used to evaluate categorical variables. The Mann-Whitney U test was used for comparative analysis between two independent variables for data that did not conform to normal distribution, and the independent sample t-test was used for data with normal distribution. In comparisons of three and more variables, one-way analysis of variance (ANOVA) was performed where parametric test conditions were ensured, and the Kruskal-Wallis H test was performed where parametric test conditions were not ensured. The relationship between data that did not conform to normal distribution was evaluated using Spearman's correlation test, and data with normal distribution were evaluated using Pearson's correlation test. P-values less than 0.05 were considered statistically significant.

Results

One hundred thirty-one children with CF and 39 children with PCD and their primary caregivers were enrolled in the study. Ethnicity of all patients were Caucasian. Sixty-six (50.4%) of the children with CF and 20 (51.3%) of the children with PCD were female ($p=0.087$). The mean age of the children was 6.5 ± 5.3 years in the CF group, and 10.4 ± 4.6 years in the PCD group ($p=0.001$). The numbers of hospital visits and hospitalizations were higher and the length of hospitalizations was longer in the previous 6 months in the CF group than in the PCD group ($p=0.006$, $p=0.011$, and $p=0.001$ respectively). The sociodemographic features and clinical characteristics of the children in the CF and PCD groups are displayed in Table 1.

All children's primary caregivers were their mothers. Fifty-seven (43.5%) of the mothers in the CF group and 19 (48.0%) in the PCD group had an assistant – in all cases, a relative. Consanguinity were present in 67 of the mothers (51.1%) in the CF group and 17 (43.5%) in the PCD group. The sociodemographic characteristics of the children's mothers are shown in Table 2.

The median mSKS of the CF group was 90 (min: 10, max: 100). The mSKS was excellent in 66.7% of children, good in 17.5%, mild in 10.6%, moderate in 3.0%, and severe in 2.2%.

Only one mother in the PCD group with moderate depression and two mothers in the CF group with mild and severe depression, respectively, were receiving antidepressant treatment. A comparison of the mothers' mean BDS, MBI, ZCBS, and PARI scores is given in Table 3.

There were no significant differences in BDI, MBI, ZCBS, and PARI scores according to the mothers' educational levels and occupational status, family type, the number of children, presence of an assistant, and work and family problems related to their children's hospitalization in each group ($p>0.05$). The BDI scores were higher in mothers who had another child with chronic disease ($p=0.046$), low family income ($p=0.014$), and children with colonization ($p=0.008$) in the CF group. Burnout was higher in both the CF and PCD groups with low family income ($p=0.022$ and $p=0.034$, respectively). The ZCBS scores were higher in mothers with a history of sibling death in the CF group ($p=0.002$). The mSKS was lower in children with CF with bacterial colonization ($p=0.028$). Democratic attitude was lower in mothers with consanguinity in the CF group ($p=0.016$). Over-protectiveness and strictness scores were higher in mothers with high family income in the CF group ($p=0.041$ and $p=0.024$, respectively). Rejection of the homemaker role scores was higher in mothers who had another child with chronic disease in the PCD group ($p=0.001$).

There were no correlation between the ages of children with CF and PCD and BDI, MBI, ZCBS, and PARI scores of their mothers. The number of hospital visits, hospitalizations, and the duration of hospitalizations in the previous 6 months were negatively correlated with the children's FEV₁ ($r= -0.443$, $p=0.001$; $r= -0.469$, $p=0.001$; $r= -0.478$, $p=0.001$, respectively) and FVC ($r= -0.496$, $p=0.001$; $r= -0.362$, $p=0.007$; $r= -0.361$, $p=0.007$, respectively) in the PFT results of children with CF. Also, the number of hospitalizations in the previous 6 months were negatively correlated with BMI in the PCD group ($r= -0.238$, $p=0.043$). Correlations between children's hospital visits and length of hospitalization with mothers' scores are given in Table 4.

In the CF group, the BDI scores of mothers were correlated with mSKS, burnout, ZCBS and over-protectiveness scores ($r= -0.311$, $p=0.004$; $r=0.483$, $p=0.001$; $r=0.259$, $p=0.010$; $r= -0.267$, $p=0.005$, respectively). The mSKS of children was correlated with burnout, democratic attitude, and marital conflict

scores of mothers ($r = -0.232$, $p = 0.033$; $r = 0.290$, $p = 0.007$; $r = 0.348$, $p = 0.001$ respectively). The burnout scores of mothers were correlated with ZCBS, democratic attitude, and strictness scores ($r = 0.468$, $p = 0.001$; $r = -0.242$, $p = 0.015$; $r = -0.222$, $p = 0.026$, respectively). The ZCBS scores of mothers were correlated with over-protectiveness and strictness scores ($r = -0.243$, $p = 0.014$; $r = -0.339$, $p = 0.001$, respectively). The over-protectiveness scores of mothers were correlated with rejection of the homemaker role, marital conflict, and strictness scores ($r = 0.431$, $p = 0.0001$; $r = 0.474$, $p = 0.001$; $r = 0.713$, $p = 0.001$ respectively).

In the PCD group, the BDI scores of mothers were correlated with burnout and ZCBS scores ($r = 0.582$, $p = 0.001$; $r = 0.622$, $p = 0.001$, respectively). The ZCBS scores of mothers were correlated with burnout and marital conflict scores ($r = 0.488$, $p = 0.009$; $r = 0.388$, $p = 0.031$, respectively). The burnout scores of mothers were correlated with marital conflict scores ($r = 0.407$, $p = 0.026$). The democratic attitude scores of mothers were negatively correlated with over-protectiveness and strictness scores ($r = -0.394$, $p = 0.025$; $r = -0.617$, $p = 0.001$, respectively). The rejection of the homemaker role of mothers was correlated with marital conflict and strictness scores ($r = 0.457$, $p = 0.010$; $r = 0.657$, $p = 0.001$, respectively). The marital conflict of mothers was correlated with strictness scores ($r = 0.483$, $p = 0.006$).

Discussion

Although there are many studies investigating the psychological status of caregivers of children with CF, data on caregivers of children with PCD are limited. This study showed that depression, burnout, burden, and negative attitudes of mothers of children with CF were higher than mothers of children with PCD, but these problems were also quite common in mothers of children with PCD. Mothers of children with CF and PCD with lower family income had more burnout. Rejection of the homemaker role was higher in mothers of children with PCD who had another child with chronic disease. Frequent hospitalizations were negatively associated with BMI in children with PCD. Frequent hospital visits were correlated with burnout of mothers in children with CF and PCD.

In a study evaluating the depression and anxiety levels of patients with CF and their caregivers, it was found that 20-34% of mothers had depression in the evaluations made with different scales. Mothers' depression has been found to be associated with children's recent intravenous antibiotic therapy and physiotherapy.²⁷ In our study, the depression level of the mothers of children with CF was higher than the mothers of children with PCD. Depression in mothers of children with CF was associated with the presence of chronic disease in another child, low family income, and colonization in their children. In a study of 326 caregivers of children with CF, a higher burden was found in the caregivers of older patients and patients with *P. aeruginosa* infection.²⁸ In our study, the caregiver burden of mothers of children with CF was higher than mothers of children with PCD. It was observed that as the number of hospital visits and duration of hospitalization of patients with CF increased, their respiratory functions were also negatively affected. The caregiver burden was increased in the mothers of children with CF who had a history of sibling death and frequent hospitalizations. Frequent hospital visits, long and frequent hospitalizations, and lower clinical scores may affect the psychological status of the mothers of patients with CF and may affect the care of their children.

Carotenuto et al.²⁹ found that mothers of children with PCD have higher levels of stress compared with mothers of healthy children. PCD is highly stressful and depressing for affected children and their families. In our study, all mothers of children with PCD had caregiver burden, approximately one in three mothers had depression, and one in ten mothers had burnout. As the depression level of mothers of children with PCD increased, burnout and caregiver burdens are also increased. Like children with CF, children with PCD need frequent clinical evaluation and treatment, including chest physiotherapy and inhaled therapies, and antibiotics for recurrent respiratory infections. All these daily practices can lead to caregiver burden, burnout, and depression. In mothers caring for an ill child, burnout increases due to an increased sense of responsibility, concerns about the health of the child, limited social activities, and the inability to find time for their own needs. Due to burnout and caregiver burden, mothers may feel insignificant and withdrawn from their environment and have an introverted and depressed mood. These negatively affected moods of mothers may affect the care of their children, adherence to treatment, and the course of the disease. The higher depression, burnout, burden and negative attitudes of mothers of children with CF than mothers of

children with PSD may be related to the younger age of children with CF and their greater need for maternal care for all daily practices.

Burnout caused by prolonged stress is quite common among parents of children with chronic illnesses.³⁰ Children with PCD usually have a normal life span. Lung functions usually deteriorate more slowly than in CF.³¹ In our study, the level of burnout was associated with low family income and the frequency of hospital visits of their children in mothers of both children with CF and PCD. Anxiety about the unmet daily financial needs of their children due to low family income may increase the burnout level of mothers. Hospital visits and hospitalizations of children with PCD are, as expected, less than for children with CF, and their respiratory function is also significantly better. The BMIs of children with PCD who had frequent hospitalizations were decreased. Growth monitoring is important, especially in children with PCD who have frequent hospitalizations.

Caring for a child with a chronic illness can pose significant challenges for caregivers, due to uncertainty about the aggravation of the disease, financial costs associated with care, compliance with treatment programs, impact on family life due to frequent outpatient visits, and hospitalization.²⁸ In a study conducted with caregivers of 44 children with CF and 19 with PCD, the burden of caregivers of children with CF was higher than for PCD in relation to pulmonary function and quality of life.³² In our study, mothers of all children with CF and PCD had caregiver burden. Families have to cope with daily life difficulties, implementation of complex treatment regimens, regular clinic visits, care costs, disruptions in plans, as well as uncertainties about the progression of chronic disease. In addition, problems in the marital relationship, the decrease in the life expectancy of the child, and the decrease in the time allocated to the family are significant sources of stress.³³ In a study evaluating 36 children with CF, mothers of children with CF with frequent hospital visits or hospitalizations were found to be more overprotective, rejecting the role of homemaker, and experiencing marital conflicts.³⁴ In our study, marital conflict was higher in mothers of children with PCD with burnout and caregiver burden. Rejection of the role of homemaker was more common in mothers who had other children with chronic disease and in mothers with marital conflict. The time the mother spends with their children can reduce the time spent with other family members, which may lead to rejection of the role of homemaker, affecting family relationships and thus increasing conflict. Mothers who have strict rules can cause unrest in the home and marital problems due to the increased stress associated with their child's illness and compliance with their treatment. This situation can be exacerbated, especially in mothers who have other children with chronic disease.

The nature of overprotection has been defined as exhibited protective behavior that is excessive in light of the child's developmental stage and has been associated with parental stress. Parental stress has been found to be associated with a child's disease outcomes.³⁵ In our study, mothers of children with PCD who were overly controlling, intrusive, and strictly disciplined had a negative democratic attitude. Fears and anxieties of mothers about the future of the child may lead to overprotective, stricter, and authoritarian behaviors. Democratic attitude was lower in the mothers of children with PCD when the mothers had more children. The fact that mothers spend more time with their ill children and protect them more may cause them not to show the same attention to their healthy children.

The mean of the consanguineous marriages is 23.2 % (8.9-42.6) in our country according to 2016 data of the Turkish Statistical Institute.¹⁵ In our study, consanguinity were present in 51.1% of CF and in 43.5% in PCD families. The higher rates of consanguineous marriages in our study compared to the general population may be associated with the increased incidence of both diseases in consanguineous marriages.

Previous research on well-being and psychological issues of PCD and CF has focused on the emotional burden of the condition, including patients' concern about current and future health. The issue has been recently further enriched with the contribution of several studies conducted during the current SARS-CoV-2 pandemic, an event that has had an impressive impact on the quality of life of patients with chronic conditions and their families.^{36,37}

The limitations of our study were that all primary caregivers were mothers and the questionnaires were

administered cross-sectionally.

In conclusion, mothers of children with CF and PCD had various difficulties related to the children's disease in our study. Although the psychological status of mothers of children with PCD was better than mothers of children with CF, their problems may increase with frequent hospitalizations, low family income, number of children, and chronic disease in another child. It is important not to neglect the psychological problems that mothers of children with PCD may experience due to difficulties associated with care. Screening the psychological status of mothers of children with CF and PCD and providing them with psychological and social support may help reduce their anxiety about their children's health and benefit the care of their children.

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