

QUALITY OF LIFE, CAREGIVER BURDEN AND ASSOCIATED FACTORS WITH REGARD TO PARENTS OF CHILDREN WITH INHERITED METABOLIC DISORDERS

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ABSTRACT

Parents of children with inherited metabolic diseases have many difficulties compared to parents of healthy children. The aim of this study is to determine quality of life, caregiver burden and associated factors with regard to parents of children with inherited metabolic disorders. The parents of 121 children diagnosed with inherited metabolic disorders who were admitted to the pediatric metabolic clinics of the university hospital constituted the sample of the study. The sample of the study was determined by Power Analysis. Data were collected using Quality-of-Life Scale and Caregiver Burden Inventory. Data were statistically analyzed using IBM SPSS V23. One-way MANOVA was used to compare the scores obtained from the scale in terms of demographic characteristics and significance levels for the differences between any pair of means were evaluated using the Duncan's Multiple Range Test. The mean score of parents in the Caregiver Burden Inventory was found to be 37,6±13,1. Sub-dimension scores of the Quality-of-Life Scale, in this study, were found as follows: mean value of overall health status was 43,6±18, physical health status was 42,7±12,7, psychological health status was 51,3±15, social relations was 47,5±19,9 and environment was 46,9±16,4. It was concluded that quality of life of caring parents is affected in many dimensions, particularly physical health status, and the caregiver burden is mild/moderate. It was found that the child's gender, need for a specific diet, the diagnosis of the child, parent's educational background and having another sick/disabled child did not affect Quality of Life Scale scores of the parents.

Key words: Inherited metabolic disorders, Parent, Quality of life, Caregiver burden

KALITSAL METABOLİK HASTALIĞI OLAN ÇOCUĞA SAHİP EBEVEYNLERİN YAŞAM KALİTESİ, BAKIM YÜKÜ VE ETKİLEYEN FAKTÖRLERİN BELİRLENMESİ

ÖZ

Kalitsal metabolik hastalık tanısı olan çocukların ebeveynleri sağlıklı çocukların ebeveynlerine kıyasla birçok yönden güçlükler yaşamaktadır. Bu çalışmanın amacı kalitsal metabolik hastalık tanısı almış çocukların ebeveynlerinin yaşam kalitesini, bakım yükünü ve etkileyen faktörleri belirlemektir. Araştırmanın örneklemini bir hastanenin çocuk metabolizma kliniğine başvuran, kalitsal metabolik hastalık tanısı olan 121 çocuğun ebeveyni oluşturmuştur. Örneklem sayısı güç analizi ile belirlenmiştir. Veriler Tanımlayıcı Özellikler Soru Formu, Yaşam Kalitesi Ölçeği ve Bakım Verme Yükü Ölçeği kullanılarak elde edilmiş, IBM SPSS V23 ile analiz edilmiştir. Demografik özelliklere göre ölçek puanlarının karşılaştırılmasında Tek yönlü MANOVA kullanılmış ve çoklu karşılaştırmalar Duncan testiyle gerçekleştirilmiştir. Ebeveynlerin bakım yükü puan ortalaması 37,6±13,1 bulunmuştur. Yaşam kalitesi ölçeği alt boyut puanları; genel sağlık durumu ortalama değeri 43,6 ± 18, fiziksel sağlık ortalama değeri 42,7±12,7, psikolojik ortalama değeri 51,3±15, sosyal ilişkiler ortalama değeri 47,5±19,9, çevre ortalama değeri 46,9±16,4 olarak bulunmuştur. Bakım veren ebeveynlerin yaşam kalitesinin, fiziksel sağlık başta olmak üzere birçok alanda etkilendiği ve bakım yükünün ise hafif/orta derecede olduğu tespit edilmiştir. Çocuğun cinsiyeti, özel bir diyetle ihtiyaç duyması, çocuğun tanısı, ebeveynin eğitim durumu ve başka bir hasta/engelli çocuğa sahip olmanın ebeveynlerin Yaşam Kalitesi Ölçeği puanlarını etkilemediği bulunmuştur.

Anahtar Kelimeler: Kalitsal metabolik bozukluk, Ebeveyn, Yaşam kalitesi, Bakım yükü

INTRODUCTION

In accordance with the classification in 2021, inherited metabolic disorders (IMD) include 1450 disorders in 24 categories (1). IMDs refer to a group of rarely encountered diseases characterized by impaired biochemical functions in metabolism. Although data on its incidence and prevalence are limited, the records of 2631 patients reviewed for the purpose of a study in Austria conducted between 1921 and 2021 revealed that the rate was as 9/100,000 by 2021 (2). It was reported that the disease is more common in male patients (2), the hospitalization rate is the highest in patients under the age of ten (3) and the most common types are amino acid and peptide metabolism disorders (2,3).

Inherited metabolic disorders (IMD) can manifest itself over the course of a wide timespan, starting with the child's life in the intrauterine period and ending with adulthood. Diagnosis of IMD is quite difficult, therefore it is often diagnosed at a later stage. The reasons for this are that these diseases are rarely encountered and they typically manifest with signs and symptoms that are not unique to metabolic disease, such as trouble with latching on or sucking and inability to gain weight. The age of the child and the degree of metabolic disorder at which clinical signs and symptoms may be diagnosed vary and almost all of these diseases may affect all organs (4). The symptom burden of children diagnosed with this disease is quite high and symptoms typically affect neurological, respiratory and gastrointestinal systems. Most of these symptoms are quite difficult to treat. The care that will be provided to these children requires special attention (5).

Rapid improvements with regard to the opportunities concerning the diagnosis and treatment affect the incidence of IMDs, improve survival rates and allow IMDs to be listed among chronic diseases (6). Despite

this, surviving children may suffer functional disabilities and developmental retardation. Extra care required by these children is mainly provided by their parents, who are obligated to adopt this challenge in their lives. In addition, IMD can affect the daily routine of patients, as it requires strict dietary control and frequent hospitalization. Undergoing a restrictive diet treatment due to IMD may often increase the dependence of children on their care giving parents and may significantly affect the quality of life of children diagnosed with an IMD and their families (7). The burden of parenting a child diagnosed with IMD is expressed with a significantly lower health-related quality of life compared to the parents of healthy children (8). Taking into account these specific circumstances, caregiver parents face major obstacles in coordinating the care of their children. As parents need to constantly monitor their children diagnosed with IMDs, they have to allocate quite a long time for caring their children (9). For these reasons, familial and social lives of caregiver parents are seriously affected in many respects (8). IMDs and the treatment procedures necessary for these diseases often negatively affect familial functions. IMD restricts the social life of parents, may cause them to have difficulty in meeting their own needs as well as the needs of other children and may expose them to a financial burden (10-12). These challenges elevate the caregiver burden of parents and their quality of life may also be significantly affected (8,13). Studies revealed that the health status of parents of children with metabolic diseases is not good, and further studies are needed to improve and support the quality of life of these parents (7). This study is notable in terms of determining the quality of life, caregiver burden and associated factors of parents with a child diagnosed with IMDs.

Research Questions:

- What is the level of parents' score in Quality of Life Scale?
- What is the level of parents' score in Caregiver Burden Inventory?
- Is there a significant difference between parents' Caregiver Burden and Quality of Life scores based on some characteristics of the child and parent?

MATERIALS AND METHODS***Population and Sample***

This research is executed between October 2020 - April 2021 in a southeastern province of Turkey, with the parents of children who were admitted to the pediatric metabolic polyclinic and clinics of the University Hospital which serves as the only center providing care for children diagnosed with IMDs. The sample of the study was determined by Power Analysis in the G Power program taking into account similar studies in the literature (17,20). Considering the correlation value in the power analysis; the number of cases that should be included in the study was determined as 46 with 95% confidence (1- α), 80% test power (1- β) and $r= 0.50$ effect size. As children with metabolic diseases constitute a specific group and due to high probability of experiencing data loss, parents who can be reached in person are included in the study.

Data Collection Tools

Data were collected via Descriptive Characteristics Form, a Quality-of-Life Scale (WHOQOL- BREF) and a Caregiver Burden Inventory.

Descriptive Characteristics Form

Descriptive Characteristics Form consists of a total of 23 questions aimed at determining the sociodemographic characteristics of children diagnosed with IMD and their parents. The form was developed from the

literature (17,20). Independent variables of the study are the age and the gender of the child, the diet followed, type of disease (diagnosis), duration of treatment, age and education of the parent, presence of a sick/disabled child, income level, and the number of children. Mean Caregiver Burden Inventory and Quality of Life Scale scores are the dependent variables of the study.

Quality of Life Scale (WHOQOL- BREF)

WHOQOL- BREF, consisting of 26 items, is a short version of the WHOQOL-100 scale. WHOQOL- BREF is a Likert-type scale and each question is scored between 1-5. Higher scores indicate a better quality of life. The scale provides information about four sub-dimensions (domains): Physical, Psychological, Social Relations and Environmental. "Cronbach alpha" values calculated for the internal consistency of the scale in the Physical Domain, Psychological Domain, Social Relations Domain and Environmental Domain were found to be 0.83, 0.66, 0.53 and 0.73 respectively (14). Turkish validity and reliability study was conducted by Eser et al (14).

Caregiver Burden Inventory

The scale was developed by Zarit, Reever and BachPeterson (1980) in order to determine the impact of care giving process on care giving individuals' own lives. This is a 22-item instrument self-report scale with a Likert-type assessment, ranging from 0 to 4 that is verbalized with the expressions never, rarely, sometimes, often or almost always. Minimum and the maximum scores that can be obtained from the scale is '0' and '88' respectively. Higher score indicates that the distress/burden experienced by the caregiver throughout the care giving process is high. Scores obtained were graded as (0-20) little/no burden, (21-40) mild/moderate burden, (41-60) moderate/severe burden and (61-88) excessive burden. Internal

consistency coefficient of the original scale was found to be between 0.87 and 0.94 (15). Turkish validity and reliability study was conducted by İnci in 2006.

Data collection

As the data collection process coincided with the pandemic period, the data were collected by taking the necessary precautions. Due to the COVID-19 pandemic, participants' HES (*Hayat Eve Sığar*) code was questioned before each interview and hygiene rules such as wearing a mask and physical distancing were observed during the interviews. The parents were duly notified about the purpose and significance of the study and interviews were held upon obtaining the verbal consent of each parent. Each interview took an average of 15-20 minutes. After the data

collection forms were applied, the individual questions posed by the patients were answered. Data were collected via face-to-face interview.

Data Analysis

Data were statistically analyzed using IBM SPSS V23. Kolmogorov-Smirnov and Shapiro-Wilk tests were used to evaluate the suitability of the data for normal distribution. One-way MANOVA was used to compare the scores obtained from the scale in terms of demographic characteristics and significance levels for the differences between any pair of means were evaluated using the Duncan's Multiple Range Test. The significance level of mean \pm sd for the quantitative data obtained in the analysis was considered as $p < 0.050$.

RESULTS

Table 1. Descriptive Characteristics of the Child and the Parent (N=121)

Characteristics	Number (n)	Percent (%)
Age of the child (year)		
0-1	19	15.7
1-3	45	37.2
3-6	24	19.8
6-12	26	21.5
12-18	7	5.8
Gender		
Female	59	48.8
Male	62	51.2
Child's diagnosed disorder		
Type of intoxication	70	57.5
Energy Metabolism diseases	24	19.8
Complex molecular diseases	27	22.3
Stay in hospital for treatment		
Don't stay	28	23.1
Stay once	27	22.3
Stay twice and more	66	54.5
A specific diet		
Yes	79	65.3
No	42	34.7
Caregiver parent		
Mother	104	86.0
Father	17	14.0
Parent's age		
19-30	60	49.6
31-40	46	38.0
41 and over	15	12.4

(Continuation of Table 1)

Educational status		
Illiterate	14	11.6
Literate	22	18.2
Primary school	57	47.1
Middle school	15	12.4
High school	9	7.4
University	4	3.3
Employment status		
Yes	20	16.5
No	101	83.5
Income Status		
Expenses more than income	11	9.1
Income is equal to expenses	73	60.3
Income more than expenses	37	30.6
Marital status		
Single	4	3.3
Married	117	96.7
Family structure		
Nuclear family	88	72.7
Extended family	33	27.3
Consanguineous marriage		
Yes	103	85.1
No	18	14.9
Number of children		
1	13	10.7
2	22	18.2
3	32	26.4
4	34	28.1
5 and more	20	16.5
Having another sick/disabled child		
Yes	40	33.1
No	81	66.9

Table 2. Scores obtained from the Quality of Life Scale and Caregiver Burden Inventory (ZCBI) (N=121)

Scale Scores	$\bar{X} \pm SD$	Median (Min-Max)
ZBCI		
Caregiver Burden Inventory	37.6 ± 13.1	39 (7 - 64)
Quality of Life Scale		
Overall Health	43.6 ± 18	50 (0 - 75)
Physical Health	42.7 ± 12.7	42.9 (10.7 - 78.6)
Psychological Health	51.3 ± 15	50 (0 - 87.5)
Social Relations	47.5 ± 19.9	50 (0 - 100)
Environment	46.9 ± 16.4	46.9 (3.1 - 96.9)

Scores obtained from the Quality-of-Life Scale and a Caregiver Burden Inventory are exhibited. Table 2 reveals that mean total score obtained from Caregiver Burden Inventory was 37.6 ± 13.1 , whereas minimum and maximum scores were 7 and 64 respectively. Sub-dimension scores of the Quality-of-Life Scale, in this study,

were found as follows: mean overall health status score was 43.6 ± 18 , mean physical health status score was 42.7 ± 12.7 , mean psychological health status score was 51.3 ± 15 , mean social relations score was 47.5 ± 19.9 and mean environment score was 46.9 ± 16.4 .

Table 3. Some Demographic Characteristics of Children Sub-dimension scores of the Quality of Life Scale and Caregiver Burden Inventory (N=121)

Characteristics of the child	Scale Sub-dimension	Sum of square.	Sd	Mean Square	K	p	Partial Eta square
Age of the child	Caregiver Burden Inventory ¹	609.3	4	152.3	1.039	0.392	0.043
	Overall Health Status ²	4150.9	4	1037.7	5.331	0.001	0.187
	Physical Health ³	223.8	4	55.9	0.380	0.822	0.016
	Psychological Health ⁴	687.0	4	171.7	0.982	0.421	0.041
	Social Relations	1217.4	4	304.3	1.218	0.309	0.050
	Environment ⁶	472.3	4	118.0	0.692	0.600	0.029
Gender of the child	Caregiver Burden Inventory	55.0	1	55.0	0.375	0.542	0.004
	Overall Health Status	76.7	1	76.7	0.394	0.532	0.004
	Physical Health	21.6	1	21.6	0.147	0.702	0.002
	Psychological Health	28.9	1	28.9	0.165	0.685	0.002
	Social Relations	21.5	1	21.5	0.086	0.770	0.001
	Environment	37.7	1	37.7	0.221	0.639	0.002
Diet	Caregiver Burden Inventory	23.6	1	23.6	0.161	0.689	0.002
	Overall Health Status	14.0	1	14.0	0.072	0.789	0.001
	Physical Health	81.3	1	81.3	0.553	0.459	0.006
	Psychological Health	2.6	1	2.6	0.015	0.903	0.000
	Social Relations	213.9	1	213.9	0.856	0.357	0.009
	Environment	106.9	1	106.9	0.627	0.431	0.007
Diagnosis of the child	Caregiver Burden Inventory	968.1	2	484.0	3.301	0.041	0.066
	Overall Health Status	36.0	2	18.0	0.093	0.912	0.002
	Physical Health	32.1	2	16.0	0.109	0.897	0.002
	Psychological Health	491.5	2	245.7	1.405	0.250	0.029
	Social Relations	280.0	2	140.0	0.560	0.573	0.012
	Environment	138.0	2	69.0	0.404	0.669	0.009
Duration of the treatment	Caregiver Burden Inventory	486.2	5	97.2	0.663	0.652	0.034
	Overall Health Status	4221.6	5	844.3	4.337	0.001	0.189
	Physical Health	491.6	5	98.3	0.668	0.648	0.035
	Psychological Health	267.1	5	53.4	0.306	0.908	0.016
	Social Relations	1885.6	5	377.1	1.509	0.195	0.075
	Environment	1255.1	5	251.0	1.470	0.207	0.073

¹R²=0,370, ¹corrected R²=0,187; ²R²=0,533, ²corrected R²=0,398; ³R²=0,291, ³corrected R²=0,085; ⁴R²=0,399, ⁴corrected R²=0,224; ⁵R²=0,513, ⁴corrected R²=0,372; ⁵R²=0,505, ⁵corrected R²=0,362.

Table 3 shows the parents' quality of life and caregiver burden in terms of some characteristics of the child and parent

whereas Table 4 presents descriptive data regarding the variables examined.

Table 4. Some Demographic Characteristics of parent's Sub-dimension scores of the Quality of Life Scale and Caregiver Burden Inventory(N=121)

Characteristics of the parents	Scale sub-dimension	Sum of square	Sd	Mean square	F	P	Partial eta square	
Parent's age	Caregiver Burden Inventory	130.4	2	65.2	0.445	0.642	0.009	
	Overall Health Status	176.8	2	88.4	0.454	0.636	0.010	
	Physical Health	416.5	2	208.2	1.416	0.248	0.030	
	Psychological Health	1082.8	2	541.4	3.096	0.050	0.062	
	Social Relations	2142.6	2	1071.3	4.286	0.017	0.084	
	Environment	1842.6	2	921.3	5.395	0.006	0.104	
	Educational status	Caregiver Burden Inventory	748.3	5	149.6	1.021	0.410	0.052
		Overall Health Status	694.9	5	138.9	0.714	0.614	0.037
Physical Health		1196.4	5	239.2	1.626	0.161	0.080	
Psychological Health		186.1	5	37.2	0.213	0.956	0.011	
Social Relations		893.5	5	178.7	0.715	0.614	0.037	
Environment		936.7	5	187.3	1.097	0.367	0.056	
Having another sick/disabled child		Caregiver Burden Inventory	6.4	1	6.4	0.044	0.834	0.000
		Overall Health Status	287.1	1	287.1	1.475	0.228	0.016
	Physical Health	0.1	1	0.1	0.001	0.971	0.000	
	Psychological Health	73.6	1	73.6	0.421	0.518	0.005	
	Social Relations	270.5	1	270.5	1.082	0.301	0.012	
	Environment	0.0	1	0.0	0.000	0.993	0.000	
	Income status	Caregiver Burden Inventory	816.8	2	408.4	2.785	0.067	0.057
		Overall Health Status	4529.0	2	2264.5	11.63	0.000	0.200
Physical Health		943.3	2	471.6	3.206	0.045	0.064	
Psychological Health		2370.4	2	1185.2	6.778	0.002	0.127	
Social Relations		3876.9	2	1938.4	7.755	0.001	0.143	
Environment		4175.6	2	2087.8	12.22	0.000	0.208	
Number of children		Caregiver Burden Inventory	1673.1	4	418.2	2.852	0.028	0.109
		Overall Health Status	3618.1	4	904.5	4.646	0.002	0.167
	Physical Health	2003.7	4	500.9	3.405	0.012	0.128	
	Psychological Health	989.0	4	247.2	1.414	0.235	0.057	
	Social Relations	5396.0	4	1349.0	5.397	0.001	0.188	
	Environment	1417.0	4	354.2	2.075	0.090	0.082	

¹R²=0,370, ¹corrected R²=0,187; ²R²=0,533, ²corrected R²=0,398; ³R²=0,291, ³corrected R²=0,085; ⁴R²=0,399, ⁴corrected R²=0,224; ⁵R²=0,513, ⁴corrected R²=0,372; ⁵R²=0,505, ⁵corrected R²=0,362.

The effect of the child's diagnosed disorder on the Caregiver Burden Inventory scores was found to be statistically significant

(p=0.041). Mean score differs depending on child's diagnosed disorder. Highest caregiver burden was measured in the Intoxication type

of IMD (Table 4). The effect of the total number of children of a parent on the Caregiver Burden Inventory scores was found to be statistically significant ($p=0.028$, Table 3). Highest mean score was obtained in parents with five and more children, while the lowest mean score was obtained in parents with a single child. (Table 4). It was concluded that other demographic characteristics did not have a statistically significant effect on the scale scores ($p>0.05$). (Table 3, Table 4)

Parents' quality of life sub-scale scores were further examined in terms of independent variables. A statistically significant difference was found between the parents' **overall health status score** when measured in terms of the age of the child ($p=0.001$), duration of the treatment ($p=0.001$), parent's income ($p=0$) and the total number of children of the parent ($p=0.002$) (Table 3). Mean overall health status score differs depending on the age of the child. Lowest mean score was obtained in parents with five and more children, while highest mean score was obtained in parents with only one child. The highest mean score was obtained in parents with higher income status, while the lowest mean score was obtained in parents with poorer income status (Table 4).

A statistically significant difference was found between the parents' **physical health status score** when measured in terms of their income level ($p=0.045$) and number of children ($p=0.012$). (Table 3). The effect of the total number of children of a parent on physical health status score was found to be statistically significant; highest mean score

was obtained in parents with four children, while the lowest mean score was obtained in parents with five and more children (Table 4). The effect of parent's income status on **psychological health status** score was found to be statistically significant ($p=0.002$, Table 3). Highest mean score was obtained in parents with higher income status, while the lowest mean score was obtained in parents with poorer income status (Table 4).

A statistically significant difference was found between the parents' **Social Relations scores** in the measurements performed on the basis of the parents' age ($p=0.017$), income level ($p=0.001$) and the number of children ($p=0.001$) (Table 3). Highest mean score was obtained in parents with 19-30 age. Highest mean score was obtained in parents with higher income status, while the lowest mean score was obtained in parents with poorer income status. Highest mean score was obtained in parents with one child, while the lowest mean score was obtained in parents with five and more children (Table 4).

The effect of the parent's age on **environment score** was found to be statistically significant ($p=0.006$, Table 3). The effect of parent's income on their environment score was found to be statistically significant ($p=0$). Highest mean score was obtained in parents with higher income, while the lowest mean score was obtained in parents with poorer income (Table 4). It was concluded that other demographic characteristics did not have a statistically significant effect on the scale scores ($p>0.05$). (Table 3, Table 4).

Table 5. Some Demographic Characteristics of Children and Parents Sub-dimension scores of the Quality of Life Scale and Caregiver Burden Inventory (N=121)

	Caregiver Burden Inventory	Overall Health	Physical Health	Psychological Health	Social Relations	Environment
Age of the child						
0-1 age	35.2 ± 11.8	48 ± 17.3 ^b	47.7 ± 12.1	51.3 ± 12.7	57 ± 22.1	54.9 ± 13.6
1-3 age (13-36 month)	37.4 ± 13.1	47.2 ± 17 ^b	42.1 ± 12.7	52.4 ± 14.6	47 ± 19.8	47.9 ± 16.5
4-6 age (37-72 month)	34.6 ± 14.4	45.8 ± 15 ^b	41.4 ± 12.9	49.7 ± 18.7	44.4 ± 20.1	46.6 ± 17.5
7-12 age	39.7 ± 14.8	35.1 ± 20.3 ^a	41.9 ± 12.8	51.8 ± 15.4	46.2 ± 17.4	40.7 ± 16.5
12-18 age	35 ± 11.8	32.1 ± 14.2 ^a	40.3 ± 13	47.6 ± 10.7	40.5 ± 20.7	42 ± 10.7
Gender						
Female	37.1 ± 12.7	41.7 ± 16.8	42.9 ± 12.3	49.6 ± 14	45.2 ± 18.5	44.6 ± 15
Male	36.6 ± 14.2	45.4 ± 19	42.5 ± 13.1	52.9 ± 15.9	49.7 ± 21.1	49 ± 17.4
A Specific Diet						
Yes	38.4 ± 13	44.5 ± 18.2	42.3 ± 13.5	52.2 ± 16.3	49.8 ± 19.4	48.4 ± 15.5
No	34 ± 13.8	42 ± 17.6	43.5 ± 11.2	49.6 ± 12.2	43.3 ± 20.5	44 ± 17.7
Diagnosis of the child						
Type of intoxication	38.7 ± 13 ^b	44.8 ± 18.9	43.4 ± 13.6	53.8 ± 15.7	50.1 ± 19.6	48.4 ± 16.6
Energy metabolism diseases	31.5 ± 12 ^a	46.4 ± 14.5	42.9 ± 8.7	48.4 ± 8.7	50.3 ± 12.9	48.4 ± 10.8
Complex molecular diseases	36.7 ± 14.8 ^{ab}	38 ± 17.8	40.9 ± 13.3	47.2 ± 16.7	38.3 ± 23.6	41.4 ± 19
Duration of treatment						
0-1 year	35 ± 11.5	51 ± 16.5 ^c	47.6 ± 13.3	51.5 ± 11.1	57 ± 20.8	53.9 ± 12.9
1.1 - 2 year	35.8 ± 12.3	47.8 ± 18.3 ^{bc}	42.5 ± 10.8	54.9 ± 15.1	47.1 ± 21.5	50.6 ± 16.9
2.1 -3 year	40.1 ± 12.6	40.1 ± 12.2 ^{ab}	39.5 ± 12.2	45.6 ± 12.2	37.7 ± 17.2	37.5 ± 16.2
3.1 - 4 year	34.7 ± 20.4	43.8 ± 23 ^{abc}	43.6 ± 14.7	54.6 ± 26.7	48.3 ± 22.8	53.1 ± 21.8
4.1 - 5 year	37.5 ± 15.2	35 ± 18.4 ^a	41.1 ± 11.1	46.3 ± 10.8	42.5 ± 13.3	40.6 ± 6.8
>5.1 year	37.9 ± 13.8	37.9 ± 17.8 ^{ab}	41.1 ± 13.9	51.8 ± 15.1	47.6 ± 17.7	43 ± 15.1
Parent's age						
19-30	34.8 ± 13.6	48.3 ± 17.1	44.2 ± 13	55.6 ± 14.2	54.6 ± 18.8 ^b	53.3 ± 15 ^b
31-40	39.7 ± 13.3	39.9 ± 17.4	40.7 ± 13.6	44.7 ± 14.9	38.4 ± 19.4 ^a	39 ± 15.5 ^a
41 and more	36.3 ± 12.3	35.8 ± 18.8	43.1 ± 7.2	54.2 ± 11.9	47.2 ± 15 ^b	45 ± 13.9 ^a
Educational statu						
Illiterate	44.9 ± 11.8	48.2 ± 16.2	41.8 ± 12.6	47 ± 9,6	45.2 ± 15.2	45.1 ± 15.8
Literate	36.9 ± 14.1	40.9 ± 15	42.4 ± 12.5	47.7 ± 11.5	46.6 ± 19.4	46.6 ± 15.8
Primary school	36.7 ± 13.2	39.7 ± 19.6	41.4 ± 12	50.1 ± 15.9	44.4 ± 21.7	43.6 ± 16.3
Middle school	28.1 ± 13.6	52.5 ± 15.8	41.7 ± 11.4	59.2 ± 17.2	55.6 ± 18.8	57.1 ± 14.5
High school	41.1 ± 8.3	48.6 ± 15.9	48.4 ± 15.5	55.6 ± 16.5	52.8 ± 15.6	46.9 ± 15.3
University	33.8 ± 13	53.1 ± 12	57.1 ± 17	63.5 ± 8.6	62.5 ± 16	62.5 ± 16.7
Having another sick/disabled child						
Yes	41.1 ± 13	36.9 ± 17.9	41.1 ± 12.2	45 ± 15	39.2 ± 18.9	40.9 ± 15.5
No	34.7 ± 13.2	46.9 ± 17.2	43.5 ± 12.9	54.4 ± 14.1	51.6 ± 19.2	49.8 ± 16
Income Status						
Expenses more than income	32.7 ± 13.3	63.6 ± 11.8 ^a	44.8 ± 10.3 ^b	65.5 ± 16.6 ^a	66.7 ± 13.9 ^a	66.5 ± 12.2 ^a
Income is equal to expenses	34.5 ± 12.6	46.2 ± 14.8 ^b	45.4 ± 12.7 ^b	53.1 ± 12.2 ^b	50.2 ± 18.5 ^b	49.3 ± 14.5 ^b
Income more than expenses	42.8 ± 13.4	32.4 ± 18.3 ^c	36.9 ± 11.6 ^a	43.5 ± 15.6 ^c	36.5 ± 18.3 ^c	36.2 ± 13.5 ^c
Number of children						
1	26.5 ± 9.5 ^a	62.5 ± 13.5 ^c	47 ± 11.1 ^b	66 ± 16.2	71.8 ± 16.5 ^c	66.1 ± 12.3
2	36.4 ± 11.8 ^{bc}	45.5 ± 13.1 ^b	41.6 ± 10.3 ^{ab}	53.4 ± 11.7	48.9 ± 15.1 ^b	45.7 ± 11.5
3	33.9 ± 14.5 ^b	44.1 ± 13.8 ^b	43.2 ± 14.7 ^{ab}	48.2 ± 14.8	42.2 ± 20 ^{ab}	44.4 ± 16.3
4	40.1 ± 12.7 ^{bc}	41.5 ± 17.6 ^b	45.3 ± 12.7 ^b	49.5 ± 13.4	49 ± 18 ^b	45.5 ± 16.2

^{a-c} There is no difference between tenses with the same letter within each group.

DISCUSSION

Lowest Quality of life score in this study was determined in the physical health status sub-dimension. Lower scores imply that care practices provided to children diagnosed with IMD adversely affect the physical health status of the parent, as these require more time and effort compared to a healthy child and since all care responsibility is assumed by the parent. Highest QoL score was determined in the psychological health status sub-dimension. Thomas et al. (2017) examined the QoL of parents (n = 72) with children diagnosed with a metabolic disease. This study revealed that the lowest quality of life score of the parents was obtained in the overall health status (16). As reported in the systematic review of the studies conducted with parents of the children with IMD were examined to determine the difficulties experienced by parents and their quality of life was reported to be low (7).

There are different results in the literature with regard to the factors affecting quality of life of parents of children with IMD. The child's age and the duration of the treatment were determined in this study to be the factors significantly affecting only the general health status score (Table 3). No significant difference was measured between the quality of life of the parents based on the child's gender, diagnosis, diet, parent's education and the presence of another disabled/sick child. In another study it was reported that the child's gender, age, diagnosis and the duration of the treatment did not affect the parent's quality of life (17).

It was further reported that the quality of life of children and their parents with dietary restrictions in their treatment was negatively affected (7,18,19). The studies conducted by Eminoğlu (2013) and Hatzman (2009) reported that the diagnosis of IMD affects the quality of life of parents (8, 18). This

difference is thought to affect the quality of life of the parents due to the fact that the patients' diagnosis of IMD have a risk of progression and adverse effects may deteriorate. The duration of treatment of the child was found to significantly affect the overall health status sub-dimension score of the parents. Highest mean score thereof was obtained with parents of children whose treatment lasts between 0-1 year, while the lowest mean score was obtained with parents of children whose treatment lasts between 4.1-5 years. On the other hand, the study conducted by Turan (2021) reported that the diagnosis of the child does not affect the parents' quality of life score (17). The findings of this study revealed that the age of the parent significantly affected the social relations and the environment sub-dimension scores of the QoL scale. These findings support the results of the studies conducted by Hatzman (2009) and Thomas (2016) (8,16). Educational background of the parent did not affect the quality of life. The research conducted by Hatzman (2009) and Thomas (2016) reported that educational background of the parents affected the quality of life of the parent. It has been stated that quality of life scores of parents decreases as their level of education decrease. The difference between the results of the studies reviewed in the literature and the results in this study is attributed to the fact that level of education of majority of the parents participating in our study were low. It was further concluded, in this study, that the income status of the parent significantly affected the scores with regard to the overall health status, physical health status, psychological health status, social relations and environment sub-dimensions of the QoL. It is argued that the quality of life of the parents were negatively affected as parents have difficulty meeting the high caring costs of their children with IMDs with their income. Similarly, Thomas (2016) and Hatzman (2009) revealed in their

studies that income status is an effective parameter on the quality of life of parents (8,16).

It was further concluded, in this study, that the number of children by the parent affected the scores with regard to the overall health status, physical health status and social relations sub-dimensions of the QoL. Highest mean score thereof was obtained with parents with 5 and more children whereas the lowest mean score was obtained with parents of a single child. It is argued that as the number of children in the family increase, not only the time that parents devote both to their children diagnosed with IMDs and other children will increase but also the qualified time that parents will allocate for themselves will decrease. As a result, parents fail to pay attention to their own health and their social relations are considered to be negatively affected.

In this study, mean caregiver burden score of parents with a child diagnosed with IMDs was found to be 37.6 ± 13.1 . This value indicates that the parents experience mild/moderate burden of care. Mean caregiver burden score of parents was found to be consistent with the findings derived by Arpacı et al (2020) (20). However mean caregiver burden score of parents contradict with the findings derived by Bozyer (2019), Turan et al. (2021) and Kim et al. (2010) (13,17,21). The scores obtained in the studies in the literature are thought to be due to the sample structure. Metabolic diseases include a wide range of diseases, however different conditions manifest with each disease. Caregiver Burden Inventory scores are thought to vary due to this difference.

Considering the variables affecting the caregiver burden score, it was found that the diagnosis of the child and the number of children of the parent significantly affected the results, however other variables did not (Table 3). It was determined herein that the

diagnosis of the child affected the parents' caregiver burden. Highest caregiver burden score was determined in the parents of children with intoxication-type inborn errors of metabolism (IT-IEM). The study conducted by Bozyer (2019) also reported that the type of metabolic disease affects the caregiver burden of the parents (21). On the contrary, the study conducted by Turan and Yayan (2020) revealed that the diagnosis of metabolic disease did not affect the caregiver burden of the parents (17). It was further concluded, in this study, that the total number of children of the parents affected Caregiver Burden Inventory scores. It was found that Caregiver Burden Inventory scores increased in line with the number of children in the family. Highest caregiver burden score was measured in the parents with five and more children. In this respect, the findings of this research are consistent with Bozyer (2019) (21).

It was concluded, in this study, that the age of the child and gender did not affect the parents' Caregiver Burden Inventory scores. Similarly, the findings of the studies conducted by other study (17,20). It was concluded, in this study, that duration of the treatment did not affect the parents' Caregiver Burden Inventory scores. However Arpacı et al. (2020) put forward that duration of the treatment affected the parents' Caregiver Burden Inventory scores (20). It is suggested that this difference is attributable to the lower ages of the patients included in the study and shorter duration of treatment.

It was concluded, in this study, that the age of the parent and educational background did not affect the parents' Caregiver Burden Inventory scores. This finding is in consistent with other study in the literature (13,20,21).

It was concluded, in this study, that having another sick/disabled child did not affect the parents' Caregiver Burden Inventory scores. Although the results were not statistically

significant, mean Caregiver Burden Inventory scores of parents with another sick/disabled child were higher than those who did not. The findings with regard to the parents' Caregiver Burden Inventory scores were similar with the studies conducted by other study in the literature (17,20,21).

It was further concluded, in this study, that income status of the parent did not affect the parents' Caregiver Burden Inventory scores. However Bozyer (2019) and Arpacı et al. (2020) reported that income status affected the parents' Caregiver Burden Inventory scores (20,21). This difference is explained by the fact that majority of the parents included in our study expressed their income status as 'moderate'.

CONCLUSION AND SUGGESTIONS

It was found that the child's gender, need for a specific diet, the diagnosis of the child, parent's educational background and having another sick/disabled child did not affect Quality of Life Scale scores of the parents with children diagnosed with IMDs.

Caregiver burden of parents is mild/moderate and diagnosis of IMDs along with total number of children by the parent significantly affected.

We further suggest that certain interventions addressing the parents, aiming to facilitate the child's care, should be developed by the nurses; parents with more than one child should be supported and informed about the care of other children; social support mechanisms should be strengthened in this respect; parents should be referred by the nurses to institutions where they can receive social and economic assistance for the care and treatment of their children, policies addressing children with IMDs should be developed.

Limitations of the research

This study was conducted only in the health center where patients in the province were admitted. Therefore, patients who reside in the province however who cannot be reached as their health status are followed up in other provinces constitute the limitation of the study.

Conflict of interest

The authors declared that they have no conflict of interest.

Ethical considerations

For the purpose of the research, written permission was obtained from the Ethics Committee of Harran University, the health institution where the research was conducted and from the relevant authors for using their scales in the study. The research was carried out upon obtaining the verbal consent of the parents who will participate. The parents were duly notified about the purpose of the study, the voluntary basis was observed and the participants were informed that the study data will be used for scientific purposes.

Author Contributions

Concept and design: HK, FA

Data collection: FA

Data analysis: HK, FA

Writing: FA, HK

Critical Review: FA, HK

All authors reviewed the results and approved the final version of the manuscript.

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