



Research Paper: Health-Related Quality of Life of Mothers of Children With Phenylketonuria



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ABSTRACT

Background: Quality of life is one of the pivotal notions of the World Health Organization's perspective. Regarding the complications of Phenylketonuria (PKU) disease in children, it seems that this disease affects the quality of life of these mothers.

Objectives: Investigating the quality of life of mothers with PKU children and to compare it with that of mothers of normal children.

Materials & Methods: This case-control study was conducted on 65 mothers with PKU children and 122 mothers with normal children in Hamadan City, Iran, in 2018 (selected randomly from the general population). For data collection, mothers filled a demographic profile questionnaire and the Iranian version of the World Health Organization Quality of Life-Brief. The obtained data were analyzed using the independent t test and univariate analysis of variance ($\alpha=0.05$) in SPSS V. 16.

Results: The Mean±SD total scores of quality of life of mothers with PKU children and mothers with normal children were 11.74±1.91 and 14.37±1.58, respectively and the difference between the two groups was statistically significant ($P<0.001$). There was also a significant relationship between the quality of life of mothers with PKU children and the age of the child at the onset of treatment ($P<0.05$).

Conclusion: According to the current study results that indicated the low quality of life of the mothers of PKU children, it is recommended that rehabilitation and psychological interventions be included in pediatric treatment programs to improve the quality of life of mothers.

Keywords: Quality of life; Phenylketonuria; Mothers

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Highlights

- The quality of life of the mothers of PKU children is low.
- The quality of life of mothers with PKU children that the onset of treatment is before 6 months is better than others.

Introduction:

Phenylketonuria (PKU) is a rare autosomal recessive disorder of Phenylalanine (PHE) metabolism arising from a shortage of the phenylalanine hydroxylase enzyme (PAH; EC 1.14.16.1). This defect leads to increased blood condensation of PHE and its toxic store in the brain. Its incidence in Europe is around one in 10000 to 15000 births and in Iran around 1.5 to 1.7 in 10000 births [1, 2]. In this regard, the family should spend plenty of time, energy, and cost (concerning the nature of the disease) for diet planning, periodic blood tests for PHE monitoring, and regular visits to the pediatric metabolic specialist and rehabilitation [3]. Failure to treatment causes severe intellectual disability and neurological abnormalities, but newborn screening tests, provide the early detection and treatment [4].

Many studies have been conducted on neurocognitive, psychosocial, psychological, and behavioral outcomes in children with Phenylketonuria (PKU); the results of the research showed that caregivers/parents of children with PKU reported significant mental health and general health problems [5, 6]. Some studies have recently highlighted certain critical areas of caregiver management [7-9]. With such a high difficulty arising from PKU, questions are being raised about its impact on the Quality of Life (QOL), not only of the sufferers but also of their family caregivers and most importantly the mothers. Wilson et al. found that even oxygen consumption at home in premature infants affected the quality of life of the parents [10].

Dadkhah et al. (2009) suggested that the mean scores on QOL were significantly lower in mothers of disabled children than those of the other group [11]. In line with the goals of the World Health Organization by 2020 and promoting a healthy lifestyle in the community, effective strategies for improving individual and social life should be placed on the agenda of the national health programs [12]. Also, in the 20th century, the main health concern was “survival” and the challenge of the present century is “better quality of life” [13].

Today, with the development of societies and the improvement of standards of living, the QOL has become remarkably notable, so that this issue has been raised as a new approach to social and medical research. It has also been mentioned that the improvement of QOL as one of the goals of community development has influenced the policies of many countries [14, 15]. The health of the general population has been considered necessary following the definition of the WHO regarding the health in 1948: “health is a state of complete physical, mental and social well-being and not merely the absence of disease or infirmity” [16]. According to WHO, the QOL is defined as “the individual’s perception of their position in life in the context of the culture and value systems in which individual lives and concerning one’s goals, expectations, standards, and concerns, which is completely individual and not understandable to others and is associated with individual’s perception of different aspects of their life [17].

The QOL at the individual level is feelings of physical and mental health and related issues, including the risk of health conditions, functional status, social support, and socioeconomic status [13, 17]. It is therefore obvious that the disease of children is one of the factors affecting the parental quality of life. The birth of a child with intellectual and physical problems causes social, economic, and emotional problems for parents, and especially mothers due to their traditional role in caring for and responsibility of children mostly with restrictive, destructive, and inclusive nature [18, 19]. The effective use of time and related strategies varies in these mothers [20-22]. Considering PKU as a chronic disease of children and QOL as one of the components of mental health in women, the present study was conducted to evaluate and compare the quality of life of the mothers of PKU children with mothers of normal children in Hamadan City, Iran.

Materials and Methods

This analytical case-control study was conducted to investigate the quality of life of mothers with and without PKU in Hamadan in 2018.

Study population

A total of 65 mothers of PKU children who referred to the association of PKU patients in Besat Hospital of Hamadan were enrolled in the study. Besides, 122 mothers of normal children were selected through convenience sampling method from the community (the cinema, parks, schools, shopping centers, mosques in different parts of the city) and were included in the study as a control group. The mothers were assured of the confidentiality of their information. The study questionnaire of mother included age, occupation, marital status, educational level, health status, cousin marriage, family size, the number of PKU children in the family and their gender, socioeconomic status (family income), history of the time to diagnose a child's disease and the time to treat a child.

The inclusion criteria in two groups were as follows: Iranian mothers living in Hamadan, having at least one child with PKU diagnosis, age under 18 years, and living with the mother. Unwillingness to participate in the study at any stage and the presence of any PKU-associated acute medical illness were the exclusion criteria. A written consent form was obtained from all individuals before entering the study.

The World Health Organization Quality Of Life (WHOQOL)-BREF questionnaire

This valid and reliable questionnaire (The Cronbach alpha of 84%) evaluates the quality of life in general. In this study, the reliability of the QOL questionnaire calculated by the Cronbach alpha method was found as 0.918. The quality of life questionnaire consisted of 24 questions on four areas of physical health (3 questions), mental health (6 questions), social relationships (7 questions), and environmental health (8 questions). The questionnaire has two other questions that do not belong to any areas and assess the health status and QOL in general. It should be noted that the physical health field deals with issues such as the power of movement, daily activities of life, work capacity, energy, pain, and sleep. In the area of mental health, self-confidence, thinking, learning, memory and concentration, religion, and psychological state are questioned. The area of social relationships evaluates personal communication, social support, and sexual life.

The environmental health area asks about the financial resources, the physical security of the health and social care, the physical environment of the residence, the available opportunities for new skills and information, recreational status, physical environment (pollution, noise, weather, etc.) and transportation. Each area can get a score

of 4-20, in which 4 shows the worst and 20 the best QOL in the desired area. This questionnaire has been translated and validated in over 40 countries. The scores were categorized as poor QOL (0-29), inadequate QOL (30- 59), the average QOL (60-89), and the appropriate QOL (90-120).

In the current study, face validity and construct validity of the questionnaire were confirmed by a group of health and rehabilitation therapists Content Validity Ratio (CVR) was greater than 0.8 and the content Validity Index (CVI) for all the questions was >0.85 ; importance score was greater than 0.85). Also, the reliability was over 0.918 with the Cronbach alpha coefficients of all constructs. SPSS version 16 was used in this study for data analyses. Descriptive statistics was done to provide information as frequency tables, percentage, mean and standard deviation, and inferential analysis for finding the relationship between variables. Statistical tests, including the t test, Chi-square, and ANOVA were conducted. The study Type I error test was considered 0.05, so the lower values were considered significant.

Results

Most of the mothers with PKU children (50%) were under the age of 35 years, and most of them were housekeeper (93.40%), married (92.10%), living with the spouse (76.30%), illiterate or semi-literate (67.10%), with monthly income less than IRR5000000 (44.70%) and living in families consisting of two or three members (35.50%). In this group, about 80.30% of mothers had physical and mental health and had no certain illness, and generally, most of them had cousin marriage (64.50%). Other demographic information of the participants in this study is presented in [Table 1](#).

The Chi-square test was used to compare the demographic characteristics between the two groups of mothers with PKU and normal children. The results of this test showed no statistically significant difference between the two groups in terms of age ($P=0.05$), marital status ($P=0.20$), and marital life status ($P=0.08$). Therefore, the two groups were homogeneous in terms of these demographic characteristics of mothers and adjusted scores.

There was a significant difference between the two groups in terms of other demographic characteristics such as occupation, educational level, monthly income, number of family members, physical and mental health of the mother, and parental cousin marriage ($P<0.05$).

The Mean \pm SD of the children with PKU was 93.63 ± 60.75 months; the Mean \pm SD age of diagnosis of phe-

Table 1. The demographic characteristics of mothers of normal children and mothers of PKU children

Factor	Group	No. (%)	
		Mothers With PKU Children	Mothers With Healthy Children
Age, y	18-35	38 (50)	67 (54.9)
	36 -55	37 (48.7)	45 (36.9)
	≥56	1 (1.3)	10 (8.2)
Work status	Self-employed and private centers	5 (6.6)	20 (16.4)
	Housekeeper	71 (93.4)	82 (67.2)
	In government centers	0 (0)	19 (15.6)
	Student	0 (0)	1 (0.8)
Marital status	Married	70 (92.1)	117 (95.9)
	Single	6 (7.9)	5 (4.1)
Marital situation	By agreement	58 (76.3)	107 (87.7)
	Confrontation	14 (18.4)	8 (6.6)
	Divorce	2 (2.6)	4 (3.3)
	Wife's death	2 (2.6)	3 (2.5)
Education	Illiteracy and reading writing	51 (67.1)	31 (25.4)
	Diploma and above diploma	24 (31.6)	61 (50.0)
	College education	1 (1.3)	30 (24.6)
Monthly income, IRR	5000000 ≤	34 (44.7)	13 (10.7)
	5000000-15000000	31 (40.8)	45 (36.9)
	≥ 15000000	11 (14.5)	64 (52.5)
Number of family members	1 -3	27 (35.5)	73 (59.9)
	4 -6	25 (32.9)	40 (32.8)
	≥ 6	24 (31.6)	9 (7.4)
Maternal physical and mental health	Healthy mother	61 (80.3)	117 (95.9)
	Sick mother	14 (18.4)	4 (3.3)
Parental kinship ratio	yes	49 (64.5)	31 (25.4)
	No	27 (35.5)	91 (74.6)

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Table 2. The quality of life of mothers of children with phenylketonuria and mothers of healthy children

Variable	No. (%)	
	Mothers With Phenylketonuria Children	Mothers With Healthy Children
Unsuitable	14 (18.4)	1 (0.8)
Moderate	56 (73.7)	75 (61.5)
Appropriate	6 (7.9)	46 (37.7)

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Table 3. The Quality Of Life of mothers of normal children and PKU children

Variable	Mean±SD		P	t test
	Mothers With Phenylketonuria Children	Mothers With Normal Children		
Quality of Life	11.74±1.91	14.37±1.58	0.001	-10.47
Physical	12.97±1.97	15.48±1.84	0.001	-9.12
Psychological	11.31±2.22	13.79±1.75	0.001	-8.68
Social	11.86±2.89	14.01±2.69	0.001	-5.31
Environments	10.94±2.28	13.95±2.13	0.001	-9.42



nylketonuria was 25.72±29.67 months and the Mean±SD age of initiation of treatment was 32.93±49.28 months.

The level of QOL in two groups of mothers showed that the appropriate QOL was 7.90% in mothers with PKU children and 37.70% in the control group. The QOL level was medium in 73.70% of mothers with PKU children and 61.50% in the control group. The poor and inappropriate QOL was observed among 18.4% of mothers with PKU children and 0.80% of the control group (Table 2).

The independent t test was applied to compare the mean QOL of mothers with PKU children and mothers with normal children. The mean QOL score in mothers of PKU children (58.82) was lower than in mothers with normal children (71.60) and this difference was statistically significant (P=0.001). There was a difference in QOL in mothers of PKU children treated before 6 months of age and treated after 6 months of age (P<0.05), QOL of the first group was better than the others.

No statistically significant difference was found in the study of the effect of variables, including the number of PKU children in the family, the maternal disease, occupation, and educational level, and marital life status on the QOL of the mothers with PKU children.

The mean QOL score in mothers of PKU children showed a statistically significant difference in terms of income levels (P=0.04).

Considering the significance of the result of the analysis of variance, we used LSD (least significant difference) post hoc test to compare the mean quality of life. Following pairwise comparisons of the mean QOL at different income levels of the families with PKU children using LSD, the results showed that the mean QOL of the mothers of PKU children with a monthly income level below IRR5000000 was statistically significant compared to a monthly income level of over IRR1500000 (P=0.017).

The independent t test was used to compare the Mean±SD score of the QOL of mothers with phenylketonuria (11.74±1.91) and the QOL of mothers with normal children (14.37±1.58). The mean score of QOL among mothers of children with PKU was lower than the mean score of QOL of mothers with normal children and this difference in the overall score of QOL of the two groups was statistically significant (P=0.001).

According to the results of the independent t test, the mean difference in other dimensions (physical, psychological, social, and environmental) is also statistically significant (P=0.001) (Table 3).

Discussion

The results suggest that the QOL of mothers of children with phenylketonuria was lower than that of mothers of normal children. The results indicated that QOL was medium in 73.7%, appropriate in 7.9%, and inappropriate in 18.4% of the mothers of PKU children. Authorities should pay more attention to coping strategies and rehabilitation programs for these mothers to promote the QOL as a major aspect related to the life of an individual. This result is confirmed by other studies such as Etemad and associates. They reported the low level of QOL in parents of children with PKU and problems that happen in the families of children with PKU [23].

These results are interpretable and the cause of the decline in the quality of life of mothers due to several reasons, including the time spent by the family, especially by the mother for child care [24, 25], the severity of the disability and illness of the child [24], the particular diet of these patients [26], the stigma and negative labels [27], the lack of coordination and weakness of care services in providing appropriate services to children and families [19, 28]. As Taylor stated, the mothers of these disabled children often sacrifice themselves to their children and prefer them to their own [29].

Also, in a family with higher income, the quality of life of mothers is better. From when a baby with PKU is very small and through all of its growing stages, breastfeeding or the infant formula to protein intake is calculated daily and the number of grams or units of phenylalanine is determined by the concentration of phenylalanine in the blood [26]. This process is difficult, time-consuming, and costly for mothers. Another reason may be the severity of the child's illness in low-income people due to non-compliance with the diet. Non-compliance with diet in low- and middle-income families is due to the lack of support centers, large families, and financial restrictions [23, 30]. Gunduz et al. reported that low-income families suffer from stress and depression due to the pressure from the high cost of the child's diet and the limitations of low income that lowers the QOL of the mother [30].

In this study, there was a significant difference in the QOL of mothers of PKU children treated before 6 and after 6 months of age. To explain the findings of current and similar studies we can state that early diagnosis of the disease followed by rapid treatment with dietary intake reduces the level of phenylalanine and its metabolites in body fluids, prevents serious brain damage [31], causes natural growth of the baby [32], and naturally reduces the complications of the PKU disease, including high costs of medication and special treatments [23] and devoting less time special care of the child [33]. Given that early treatment reduces the side effects of the disease, the QOL of mothers can be better with the early treatment of children.

Also, the QOL in healthy mothers with PKU children was higher than that in the sick mothers with PKU children. Although the type of maternal disease has not been specifically assessed. Consistent with our results, Xiang et al. (2009) study showed the lower QOL for the parents of children with attention deficit hyperactivity disorder who have specific medical conditions themselves compared to the control group [34].

In this study, the educational level of the mother, the number of PKU children, the mother's job (employee and homemaker), the status of marital life (interpersonal understanding, conflict, widow and divorced) did not affect the quality of life of the mothers of PKU children. However, Elkodoos et al. stated that higher education provides the opportunity for parents to adapt to the conditions [35]. Alaei et al. showed that the fewer number of children in a family has a direct relationship with better care of the mother of the PKU child [36].

The reason for the heterogeneity between the result of the present and the mentioned studies can be attributed

to the small sample size in the present study, in which 92.1% of the patients had only one PKU child and 9.7% had more than one ill child [37]. About the status of marital life, 79.3% of the mothers lived by agreement and 18.4% had a conflict. In similar studies, about three-quarters of the PKU's caregivers were married and few were widows and divorced women [35].

Limitations of this research included a small sample size for generalization of the results. The wide age ranges in the study limited more specific analyses for the impact of the illness and each developmental stage upon the families. We suggested that future research focus on collecting larger longitudinal samples to survey specifically the impact of PKU on QOL of the mothers for each developmental stage of their children and assess the specific stressor and challenge factors. This study had a cross-sectional design and did not measure causality. Certain demographic variables such as job and income were missing in some cases, due to a lack of desire to express them.

Conclusions

According to the results, the QOL of mothers with PKU children is lower compared to the mothers of normal children. Also, mothers of PKU children often have a medium quality of life. Respective planning, education, and treatment in this area are recommended given the importance of QOL in promoting health.

Ethical Considerations

Compliance with ethical guidelines

The approval for the study was obtained from the Ethics Review Committee of the Research Deputy of Hamadan University of Medical Sciences, Hamadan, Iran (UMSHA. REC.1394, 29). Before the study, all participants signed a written informed consent, approved by the Ethics Committee of the Hamadan University of Medical Sciences.

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Authors contributions

Conceptualization, investigation, writing the original draft: Zahra Mortazavi, Saideh Sadat Mortazavi; Writing, review, and editing: Minoo Dabiri Golchin, Saideh Sadat Mortazavi; Resources: Saideh Sadat Mortazavi, Minoo Dabiri Golchin; Methodology: Leili Tapak, Said-

eh Sadat Mortazavi; Funding acquisition: Leili Tapak; Supervision: Saideh Sadat Mortazavi.

Conflict of interest

The authors declared no conflicts of interest.

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