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Assessment of Quality of Life of Parents of Children with Osteogenesis Imperfecta

Ocena jakości życia rodziców dzieci chorych na wrodzoną łamliwość kości

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Abstract

Objectives. The aim of the work was an objective assessment of the quality of life of parents of children with osteogenesis imperfecta (OI) and of its determinant factors.

Material and Methods. The survey answers of 25 parents were analyzed and contained demographic parameters, socioeconomic status information, quality of life of responses and type of support they have been receiving. In order to assess the effects of this children's disease on the quality of life of the parents, families were divided into two groups depending on the OI severity: group M – mild (type I and IV OI), group S – severe (type III OI). The objective of the work was carried out based on the WHOQOL-BREF quality of life questionnaire and measures of family status: education degree based on the International Standard Classification of Education (ISCED), a subjective assessment of the family's wealth (Perceived Family Wealth, PFW), and the family's financial resources (Family Affluence Scale, FAS).

Results. 56% of respondents assessed their global quality of life (Quality of Life, QL) as good, whereas 8% answered poor. Perception of general health status was similar. Life domains assessed in the WHOQOL-BREF questionnaire received the following mean values on a scale from 4 to 20 points: physical – 12.2 ± 1.2, psychological – 15.04 ± 2.2, environmental – 13.32 ± 2, social relationships – 14.28 ± 1.5. In the severe OI group, the environmental domain was assessed as worse than in the mild OI group and this assessment was statistically significant, despite the fact that the group of families with severe cases of OI received more support from the appropriate institutions. Indicators of socioeconomic status did not affect the respondents' assessment of their global quality of life.

Conclusions. In the tested group of families, the child's disease did not affect either the global quality of life assessment or health of the respondents or their quality of life in terms of physical and mental status and social relationships. The parents of children with severe OI assessed the life domain associated with the environment they live in as worse than the parents of children with mild OI. The global quality of life assessment of the respondents did not depend on the family's socioeconomic status and on the help they have been receiving with regard to care for the child (**Adv Clin Exp Med 2012, 21, 1, 99–104**).

Key words: osteogenesis imperfecta, quality of life, WHOQOL-BREF.

Streszczenie

Cel pracy. Obiektywna ocena jakości życia rodziców dzieci chorych na wrodzoną łamliwość kości (*osteogenesis imperfecta* – OI) oraz jej uwarunkowań.

Materiał i metody. Zanalizowano odpowiedzi 25 rodziców zawierające dane demograficzne, informacje o statusie socjoekonomicznym, jakości życia respondentów oraz rodzaju pomocy, z jakiej korzystają. Aby ocenić wpływ choroby dziecka na jakość życia rodziców, rodziny podzielono na dwie grupy w zależności od ciężkości przebiegu OI: grupa M – łagodny (typ I, IV OI), grupa S – ciężki (typ III OI). Cel realizowano za pomocą kwestionariusza oceny jakości życia WHOQOL-BREF oraz mierników statusu rodzin: stopień wykształcenia wg International Standard Classification of Education (ISCED), subiektywnej oceny zamożności rodziny (*Perceived Family Wealth* – PFW), zasobów materialnych rodziny (*Family Affluence Scale* – FAS).

Wyniki. Globalną ocenę jakości życia (*Quality of Life* – QL) 56% ankietowanych określiło jako dobrą, 8% – złą. Podobnie kształtowała się percepcja ogólnego stanu zdrowia. Oceniane w kwestionariuszu WHOQOL-BREF dziedziny życia uzyskały następujące wartości średnie w skali od 4–20 pkt.: fizyczna – 12,2 ± 1,2, psychologiczna – 15,04 ± 2,2, środowiskowa – 13,32 ± 2, dziedzina relacji społecznych – 14,28 ± 1,5. W grupie S dziedzina środowiskowa była istotnie statystycznie gorzej oceniana niż w grupie M, mimo iż ta grupa rodzin otrzymywała większe wsparcie ze strony instytucji pomocowych. Wskaźniki statusu socjoekonomicznego nie wpływały na globalną ocenę jakości życia ankietowanych.

Wnioski. W badanej grupie rodzin przebieg choroby dziecka nie wpływał na globalną ocenę jakości życia czy zdrowia ankietowanych, a także na jakość ich życia w aspekcie stanu fizycznego, psychologicznego i relacji społecznych. Rodzice dzieci z ciężkim przebiegiem OI gorzej niż rodzice dzieci z łagodnym typem choroby ocenili dziedzinę życia związaną ze środowiskiem, w jakim żyją. Globalna ocena jakości życia respondentów nie zależała od statusu socjoekonomicznego rodzin, a także od pomocy, z jakiej korzystają w opiece nad chorym dzieckiem (**Adv Clin Exp Med 2012, 21, 1, 99–104**).

Słowa kluczowe: *osteogenesis imperfecta*, jakość życia, WHOQOL-BREF.

Objective assessment of the quality of life and its determinant factors among parents of children with osteogenesis imperfecta (OI) may help study the effects of a child's disease on the functioning of a family.

In medicine, quality of life is most often analyzed in groups of patients, often with respect to its relation to health (Heath Related Quality of Life, HRQL) [1]. Such studies have also been conducted among children with OI, and, among other things, they have managed to determine the effects of bisphosphonate treatment on their quality of life [2].

The expression "quality of life" has many meanings and it can be defined in different ways. The World Health Organization and the World Health Organization Quality of Life Group (WHOQOL Group) has suggested a comprehensive definition [3]. According to this definition, "quality of life is the individuals' perception of their position in life in the context of the culture and value systems in which they live and in relation to their goals, expectations, standards and concerns" [3]. The WHOQOL Group has proposed some research tools to assess quality of life: the WHOQOL-100 questionnaire and its abridged version: WHOQOL-BREF. Using the latter, it is possible to assess quality of life in four domains (physical health, psychological, environment, and social relationships) plus a global quality of life assessment and a perception of global health status by the respondents [4]. Demographic and socioeconomic parameters are an important supplement when interpreting the questionnaire results.

Osteogenesis imperfecta is a connective tissue disease. The majority of types of this disease is caused by a defect in type I collagen synthesis and may be manifested in different clinical forms, from a mild (type I, IV) course with few bone fractures to a severe (type III) or even lethal (type II) [5]. OI classification is based on clinical symptoms, radiological signs and genetic determinants [6]. The main clinical features of this disease are as follows: brittle bones, small stature and progressive deformations of motor organs. Other symptoms described in children with OI include: blue sclera (type I, III), loose joints, loss of hearing during puberty (type I, IV) [7]. In many cases, patients with congenital bone fragility require interdisciplinary specialist medical care and support due to their significant levels of disability [8, 9]. This severe and chronic disease affects the functioning of patients' families that in many cases are involved in the treatment and rehabilitation processes.

The aim of the work was an objective assessment of the quality of life of parents of children with osteogenesis imperfecta and of its determinant factors. So far such a situational assessment of parents of children with osteogenesis imperfecta has not been found in the literature.

Material and Methods

Tested Subjects and Course of Studies

The study was performed from July to December 2009 among parents of 25 children with OI.

In the Department of Pediatric Propaedeutics and Metabolic Bone Diseases of UM in Łódź, the survey was performed among 21 parents, whereas 4 parents responded via the National Support Group for Children with Osteogenesis Imperfecta (in which case the survey was answered electronically). Before the start of the study, the respondents were informed of its aim, regulations, anonymity and voluntary participation.

Applied Methods

The objective of the work was carried out based on the WHOQOL-BREF quality of life questionnaire and measures of family status: education degree based on the International Standard Classification of Education (ISCED), a subjective assessment of the family's wealth (Perceived Family Wealth, PFW) and the family's financial resources (Family Affluence Scale, FAS).

The survey prepared for the purposes of the study was made up of 3 parts: 1 – a Polish version of the WHOQOL-BREF questionnaire, 2 – a demographic survey containing additional questions about the child's disease, 3 – a survey to assess the family's socioeconomic status

Part 1

The quality of life of the respondents was analyzed based on the WHOQOL-BREF questionnaire assessing 4 life domains (24 questions): physical health, psychological, environment and social relationships, plus a subjective global quality of life assessment and perception of global health status of the respondents (one question each) [4]. Questions in the WHOQOL-BREF questionnaire are closed questions, with a 5-point answer scale. Scores achieved after survey analyses were transformed into a 4–20 point target scale using the methods of the questionnaire authors. The higher the score the better the assessment of each life domain which was evaluated [4].

Part 2

The demographic survey included questions on the place of residence, parents' age and education, family structure (two or single-parent), and type of support received (medical care and/or support from an institution, relatives and friends).

Part 3

Depending on the education level, the respondents were classified into three groups: low, middle or high education according to the International Standard Classification of Education (ISCED) [10]. The assessment of professional activity included questions not only about having a job, but also regarding situations the which the child's disease resulted in a discontinuation of professional activity.

Subjective assessment of the family's wealth (Perceived Family Wealth, PFW) comprised answering the following question using a 5-point scale: "In your opinion, is your family wealthy, namely is it well off?"(1 – "very well off" to 5 – "very poor") [11].

The family's financial resources were assessed based on the FAS (Family Affluence Scale) scale with questions regarding the possession of the following items: own car, computer, child's own room and holiday travel. Families were classified into 3 groups with low, moderate or high affluence level based on scores achieved on a 0–7 scale [10].

In order to assess the effects of the child's disease on the quality of life of parents, the studied families were divided into two groups depending on OI severity: a mild OI group (type I OI – 7 families, type IV OI – 4 families) and a severe OI group (type III OI – 14 families). Questionnaire results were analyzed in both groups.

The statistical analysis included elements of descriptive statistics. Differences between the assessments of different life domains in mild OI and severe OI groups of families were analyzed with the non-parametric U Mann-Whitney test. Differences between the global quality of life assessment, perception of general health status, and socioeconomic indicators in both groups were analyzed with the chi^2 test with contingency tables. The chi^2 test was also used to analyze the relationship between the global quality of life assessment and indicators of family socioeconomic status. Statistical calculations were performed using SPSS software. The value $p \leq 0.05$ was accepted as significance level.

Results and Discussion

Parents of children with OI assigned the highest score to the psychological domain and the lowest to the physical health domain in the WHOQOL-BREF quality of life questionnaire. (Tab. 1). Mean scores in individual life domains, apart from the latter, were not significantly different from the values obtained by the WHOQOL Group among respondents including 11,830 inhabitants of 23 countries (general population).

In the WHOQOL-BREF questionnaire the results were as follows: physical health – 16.2 ± 2.9; psychological – 15.0 ± 2.8; environmental – 13.5 ± 2.6; social relationships – 14.3 ± 3.2 [4].

The environmental domain (including such aspects as a feeling of safety, accommodation, access to information, opportunity for fulfilling one's interests) received a lower score from parents in group S (Tab. 1), despite the fact that they received support from support institutions more often (Tab. 3). This observation can be explained by significantly more responsibilities and needs of these families with regard to care for a severely sick child. Other life domains were assessed similarly by both groups of parents (Tab. 1). The majority of parents (56%) assessed global quality of life as good and the score distribution was similar in M and S groups (Tab. 4). 60% of the respondents evaluated their general health status as good, and a similar observation was reported in group M and S (Tab. 4).

The analysis of demographic data from the tested families has indicated that two-parent families are predominant (1 single-parent family), with moderate education, living in a city (Tab. 2). A high rate of non-working mothers is of notice – in the majority of cases it is a result of the need to provide care for a sick child. Nearly one third of non-working fathers does not work for the same reason (Tab. 2).

Children from the majority of families were under specialist medical care but the support from

Table 1. Mean scores in different life domains in the WHOQOL-BREF questionnaire

Tabela 1. Średnie wartości punktowe w poszczególnych dziedzinach życia w kwestionariuszu WHOQOL-BREF

Domain (Dziedzina)	Whole group (Cała grupa)	Mild OI (Umiarkowane OI)	Severe OI (Ciężkie OI)	p
Physical (Fizyczna)	12.24 ± 1.23	12.45 ± 1.03	12.1 ± 1.38	0.56
Psychological (Psychologiczna)	15.04 ± 2.13	15.54 ± 1.86	14.64 ± 2.31	0.19
Environmental (Środowiskowa)	13.32 ± 2.03	14.5 ± 1.73	12.57 ± 2.06	0.025*
Social relationships (Relacje społeczne)	14.28 ± 1.46	14.36 ± 1.43	14.21 ± 1.53	0.81

* p value statistically significant.
* wartość p istotna stytystycznie.

Table 2. Demographic parameters of the studied families

Tabela 2. Dane demograficzne ankietowanych rodzin

Place of residence (Miejsce zamieszkania) n (%)	Urban (Miasto) 20 (80)			Rural (Wieś) 5 (20)		
Parents (Rodzice)	mothers (25)			fathers (24)		
Age – years – median ± SD (Wiek – lata – mediana ± SD)	34.3 ± 7.1			38.1 ± 8.5		
	low	moderate	high	low	moderate	high
Education level (Wykształcenie) n (%)	8 (32)	10 (40)	7 (28)	10 (42)	10 (42)	4 (16)
Professional activity (Aktywność zawodowa) n (%)	working	not working	not working due to child's illness	working	not working	not working due to child's illness
	13 (52)	12 (48)	11 (44)	18 (75)	6 (25)	2 (8)

appropriate institutions (families with children with more severe disease obtained this support more frequently) or from relatives or friends was unsatisfied.

According to the analysis of socioeconomic indicators, 99% of the respondents assessed their family's wealth (PFW) as average. However, the objective assessment of the families' financial resources (FAS) indicates that low affluence can be observed in almost half of the families (48%). A similar distribution of PEW and FAS indicators was observed in both groups of families (Tab. 4).

An analysis was performed to assess the relationship between the global quality of life assessment of the respondents and their socioeconomic status. Differences between groups of persons with different education level, financial resources or dependant on support received by the families were not observed (Tab. 5).

The relatively low number of respondents was a study limitation which results from that fact that osteogenesis imperfecta is a very rare disease. The Department of Pediatric Propaedeutics and Metabolic Bone Diseases in Łódź monitors a relatively numerous group of patients with OI, and more than ten are being treated with bisphosphonates. According to the anonymous character of the study, the authors didn't assess the influence of the disease's duration on the quality of life of parents of children with OI.

So far in Poland there has been no national registry of patients with osteogenesis imperfecta, and the understanding of this disease among physicians and society is still insufficient. This work may help provide more information on children with OI as well as information on families with OI children.

Table 3. Number of families that have received support with regard to care for the child

Tabela 3. Liczba rodzin, które otrzymywały pomoc w opiece nad chorym dzieckiem

Type of support (Rodzaj pomocy)	Whole group (Cała grupa) (n)	Mild OI (Umiarkowane OI) (n)	Severe OI (Ciężkie OI) (n)	p
Support from close persons (Wsparcie ze strony najbliższych osób)	3	1	2	$chi^2(1) = 0.16$ $p = 0.69$
Specialist medical care (Specjalistyczna opieka medyczna)	19	9	10	$chi^2(1) = 0.36$ $p = 0.55$
Institutional support (Pomoc instytucjonalna)	11	2	9	$chi^2(1) = 5.31$ $p = 0.021$*

* p value statistically significant.
* wartość p istotna stytystycznie.

Table 4. Analysis of distribution of selected quality of life indicators and socioeconomic indicators in mild OI group and severe OI group

Tabela 4. Analiza rozkładu wybranych wskaźników jakości życia oraz wskaźników socjoekonomicznych w grupie M (umiarkowane OI) i S (ciężkie OI)

Indicator (Wskaźnik)	Chi^2 test result (Wyniki testu chi^2)
Global quality of life assessment (Globalna ocena jakości życia)	$chi^2(3) = 3.3$; $p = 0.35$
Perception of general health status (Percepcja ogólnego stanu zdrowia)	$chi^2(3) = 4.4$; $p = 0.22$
PFW	$chi^2(1) = 1.33$; $p = 0.25$
FAS	$chi^2(2) = 3.4$; $p = 0.18$

PFW – Perceived Family Wealth.
PFW – subiektywna ocena zamożności rodziny.

FAS – Family Affluence Scale.
FAS – zasoby materialne rodziny.

Table 5. Analysis of global quality of life assessment of the respondents depending on socioeconomic indicators

Tabela 5. Analiza rozkładu oceny globalnej jakości życia ankietowanych w zależności od wskaźników socjoekonomicznych

Indicator (Wskaźnik)	Chi^2 test result (Wyniki testu chi^2)
Mother's education (Wykształcenie matki)	$chi^2(6) = 6.17$; $p = 0.45$
Father's education (Wykształcenie ojca)	$chi^2(2) = 3.39$; $p = 0.18$
FAS	$chi^2(6) = 3.24$; $p = 0.78$
Support from close persons (Wsparcie ze strony najbliższych osób)	$chi^2(3) = 4.03$; $p = 0.26$
Specialist medical care (Specjalistyczna opieka medyczna)	$chi^2(3) = 7.77$; $p = 0.051$
Institutional support (Pomoc instytucjonalna)	$chi^2(3) = 4.42$; $p = 0.22$

FAS – Family Affluence Scale.
FAS – zasoby materialne rodziny.

The authors concluded that in the tested group of families, the child's disease did not significantly affect either the global quality of life assessment or health of the respondents or their quality of life in terms of physical and psychological status and social relationships. The parents of children with severe OI assessed the life domain associated with the environment they live in as worse than the parents of children with mild OI. The global quality of life assessment of the respondents did not depend on the family's socioeconomic status or on the help they have been receiving with regard to care for the child.

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