Research tools for quality of life assessment of patients with cystic fibrosis

Narzędzia badawcze do oceny jakości życia chorych na mukowiscydozę

Izabella Uchmanowicz, Beata Jankowska-Polańska, Marta Wleklik

Zakład Pielęgniarstwa Internistycznego, Katedra Pielęgniarstwa Klinicznego, Wydział Nauk o Zdrowiu, Uniwersytet Medyczny we Wrocławiu

AUTOR DO KORESPONDENCJI:
Izabella Uchmanowicz
Zakład Pielęgniarstwa Internistycznego
Katedra Pielęgniarstwa Klinicznego
Wydział Nauk o Zdrowiu
Uniwersytet Medyczny we Wrocławiu
ul Bartla 5
51-618 Wrocław
tel. 71 784-18-05, 71 341-95-33, fax: 71 343-20-86
e-mail:izabella.uchmanowicz@am.wroc.pl.

STRESZCZENIE

Narzędzia badawcze do oceny jakości życia chorych na mukowiscydozę

Mukowiscydoza jest genetycznie uwarunkowaną chorobą ogólnoustrojową, która jeszcze kilka lat temu była uważana za śmiertelną chorobę wieku dziecięcego. Kompleksowe, wielospecjalistyczne leczenie sprawiło, że obserwuje się wzrost liczby dorosłych chorych na mukowiscydozę, co rodzi potrzebę zminimalizowania interwencji medycznych na jakość wydłużającego się życia chorych. Dużą wartość nadaje się badaniom dotyczącym jakości życia uwarunkowanej stanem zdrowia, jej ciągłe i kompleksowe monitorowanie powinno stanowić stałe uzupełnienie oceny parametrów klinicznych. Analiza samooceny jakości życia umożliwiłaby zarówno zaplanowanie i zorganizowanie opieki nad pacjentem, jak i modyfikację podejścia terapeutycznego w oparciu o aktualne fizyczne, społeczne, psychologiczne potrzeby chorego. Pojęcie jakości życia uwarunkowanej stanem zdrowia, HRQoL zostało wprowadzone w 1900 roku przez Schippera i wsp. Jest to koncepcja wielowymiarowa, która ściśle wiąże się w holistycznym podejściu do człowieka z definicją zdrowia sformułowaną przez WHO mówiącą o zdrowiu jako pełnym dobrostanie fizycznym, psychicznym i społecznym. Od lat 90 obserwuje się nieustanny rozwój badań nad jakością życia osób dotkniętych chorobami. Wśród metod stosowanych do badania i analizy jakości życia znajdują się metody jakościowe i ilościowe. Do technik ilościowych zalicza się badanie kwestionariuszowe umożliwiające samoocenę jakości życia. Wśród chorych na mukowiscydozę istnieje możliwość zastosowania kwestionariuszy ogólnych i specyficznych, w tym specyficznych dla chorób układu oddechowego oraz specyficznych dla mukowiscydozy.

Słowa kluczowe: **mukowiscydoza, kwestionariusze, jakość życia**

ABSTRACT

Research tools for quality of life assessment of patients with cystic fibrosis

Cystic fibrosis is a genetic disorder which a few years back was considered a fatal childhood disorder. Comprehensive, multispecialty treatment resulted in the increase of the number of adults with cystic fibrosis which in turn brings about a need to minimize clinical interventions. That is why health related quality of life surveys are ever more important. Constant and comprehensive monitoring of HRQoL should complement the assessment of clinical parameters. The analysis of QoL could make planning and arranging patient's care possible and allow modification of therapeutic approach based on present physical, social and psychological needs of a patient. The term HRQoL was coined by Schippera et al. in 1900. It is a multidimensional concept which in its holistic approach to man is directly related to the definition of health developed by the WHO in which health is defined as a state of complete physical, mental, and social well-being. Since the 90s there has been an ongoing development of studies in the field of QoL of people suffering from various conditions. The research and analysis methods include qualitative and quantitative tools. Quantitative tools encompass QoL self-assessment surveys. General and specific questionnaires may be applied in patients with CF including respiratory diseases specific and cystic fibrosis specific questionnaires.

Key words: **cystic fibrosis, questionnaires, quality of life (QoL)**

INTRODUCTION

Cystic Fibrosis is a genetic systemic condition connected with exocrine glands disorders. Although CF is still incurable, it absolutely requires multispecialty symptomatic treatment aiming at delaying the disease's progress as well as preventing complications which significantly influence the length and quality of patients life [1,2]. Although a few years back CF was considered a fatal childhood disorder, contemporary statistical data shows that 50% of people with cystic fibrosis have a chance to live to 30 years of age, and a life span of children born in the nineties is to amount to at least 40 years. The increase in the number of adult patients will require not only the improvement of clinical management efficiency but also reduction of the impact of interventions on the quality of ever extending life [3,4,5,6]. In the case of chronic conditions, including cystic fibrosis, abandoning biomedical approach to patient's treatment and care in favour of more dimensional one and enabling the assessment of the impact of somatic condition on areas important to every individual are crucial [7]. Studies involving evaluation of health related quality of life are ever more important and HRQoL constant and comprehensive monitoring should complement the assessment of clinical parameters. The analysis of QoL could make planning and arranging patient care possible and enable modification of therapeutic approach based on current physical, social and psychological needs of a patient [6,8]. Studies on QoL of chronic patients show that even small changes of little significance for clinical staff, influence, to a great extent, patients and their families whereas those which clinical staff identify as significant taking into account health condition and patients' life situation, is of no importance for them [9].

Health Related Quality of Life was introduced by Schipper et al. in 1900 and described as 'the functional effect of an illness and its consequent therapy upon a patient, as perceived by a patient'. As early as in 1940s, these issues were examined as a part of social studies, and studies on quality of life were conducted by Campell, a forerunner in this field, in the 1970s [10,11]. On the other hand, Karnofski or Katz are believed to have been the authors of measuring scales developed for medical purposes [12].

HRQoL is a multidimensional concept taking into account such aspects of human life as their physical and mental condition, social situation, economic conditions, or spiritual domain thanks to which, in its holistic approach, it is strictly connected to the definition of health developed by the WHO according to which health is a state of complete physical, mental, and social well-being. Growing interest in quality of life in medicine, leads to the establishment of special WHO section dealing with studies on quality of life called WHO Quality of Life (WHOQOL) in the 1990s [7,12,13].

QUALITY OF LIFE ASSESSMENT TECHNIQUES

There are quantitative and qualitative methods among methods applied to the assessment and analysis of quality of life. Qualitative techniques are a multifactor way to assess a patient's well-being enabling, at the same time, a detailed and thorough analysis. Nonetheless, they require many research instruments this making them labour-consuming, and since only some of the results are expressed in points they cannot be used in comparative studies. Results obtained thanks to quantitative techniques including questionnaires are expressed in points this making the comparison of the conducted studies with the studies on other groups of patients or a control group of healthy people possible. Research tools for assessment of QoL may be divided into generic and specific [11,14,15].

Generic scales are used for global assessment of QoL in various diseases, among healthy people and for comparative analysis. However, unlike disease or dimension specific questionnaires, they do not take disease-specific-information into account. The disease- specific questionnaires enable the assessment of QoL in a given disease or group of diseases. They are also used for assessing changes in QoL in time since they are more clinically sensitive to changes in health condition than generic questionnaires. Nonetheless, they do not enable global assessment of QoL, they are one dimensional and less accurate and valid as compared with generic questionnaires. Disease-specific questionnaires may be divided into two groups. The first one i.e. domain-specific questionnaires focuses on evaluating certain domains of patient's functioning affected by a disease and subsequent treatment, the second category, on the other hand, i.e. disease-specific questionnaires focuses on factors stemming from a disease itself [9,14].

QoL assessment may be administered objectively by staff of long term care facilities or by family or be self-administered by a patient based on their experience or opinions. Subjective assessment obtained thanks to a questionnaire is believed to be more important in the QoL analysis. QoL self-assessment may change in time and under the influence of objective factors [11,16,17].

Selecting an appropriate study instrument should be based on the study's goal and patient's clinical situation. QoL assessment questionnaires have to be reliable, take into account basic psychometric parameters and meet the following criteria i.e. reliability, validity and sensitivity [14].

In the case of studies on patients with CF it is possible to apply both generic and specific questionnaires including respiratory system conditions specific and cystic fibrosis specific questionnaires.

REVIEW OF QUALITY OF LIFE ASSESSMENT QUESTIONNAIRES FOR PATIENTS WITH CYSTIC FIBROSIS

Generic questionnaires

1. **Quality of Well-Being Scale (QWB)** was developed by Robert Kaplan et al. from the University of California. This is a tool administered to assess quality of life in four domains: mobility (MOB *Mobility Scale*), physical activity (PAC – *Physical Activity Scale*), social activity (SAC – *Social Activity Scale*) and symptoms (CPX – *Symptom / Problem complexes*). In the ratings "0" following transformation means death, whether "1.0" is an asymptomatic full func-

tion. QWB is mainly used for measuring QoL of a population or groups of people. It is available in English [18].

2. **Nottingham Health Profile (NHP)** was developed by Hunt, McEwan and McKenn(1986). The NHP is a generic quality of life survey used to measure subjective physical, emotional, and social aspects of health. It enables subjective measuring QoL and its impact on daily functioning, however it focuses on poor functioning of patients without taking into account their well-being. It consists of two parts. Part I of the survey covers presently perceived problems and includes 38 simple statements concerning six dimensions of health: physical mobility, pain, social isolation, emotional reactions, energy, and sleep. Part II (complementary, does not have to be administrated) consists of seven statements about areas of life that are most affected by current health status such as: relationships at home, care of home, sex life, social life, job, hobby, taking holidays. These are 'yes/no' statements. The obtained results are added up (0-100 points), each subarea separately, the higher the score, the worse health status. The maximum score possible to obtain, and at the same time meaning the lowest QoL, amounts to 45. The NHP is a reliable and accurate instrument, administered in many countries, available also in Polish [19].

3. **Short Form 36 (SF-36) –** developed by Americans is one of the most commonly applied tools for self-assessment of quality of life in various groups of patients and general populations globally, useful for predicting the course of a disease as well. Moreover, SF – 36 meets minimal psychometric standards for conducting a comparative analysis between a study group and a control group. The questionnaire consists of 36 questions concerning 8 domains of life: Physical Functioning, Role Physical, Bodily Pain, Social functioning, Mental Health, Role emotional, Vitality, General Health perceptions. Adding all physical components PF, RP, BP, VT provides Physical Component Summary - PCS whereas adding all mental components SF, RE, MH, GH, provides Mental Component Summary - MCS. Questions relate to 4 weeks prior to the questionnaire. Scaling of items is diversified i.e. consists of 3-point, 5-point and 6-point Likert Scale. Each answer scores a certain number of points thus a result provided in a said category may amount to 0 to 100 points. The lower the score, the lower QoL. SF – 36 is available in Polish [20].

4. **World Health Organization Quality of Life Questionnaire (WHOOQoL-100)** was developed by the section dealing with studies on quality of life at the World Health Organization. It is applied in various groups of patients and general populations, in countries of different cultures, economic levels and healthcare systems. It consists of 100 questions relating to six domains: physical health, psychological health, level of independence, social relationships, environment, spirituality/religion/personal beliefs. Jaracz and Wołowicka prepared Polish version of WHOQoL-100. One subscale was added to the original version of the questionnaire encompassing four questions concerning overall quality of life and general health [21].

5. **Sickness Impact Profile (SIP)** was developed in the United States in 1972. The questionnaire enables self-assessment of health condition and the impact of a disease on physical, psychological and social functioning and activities of daily life. It consists of 136 questions divided into 12 categories: sleep and rest, emotional behaviour, body care and movement, home management, mobility, social interaction, ambulation, alertness behaviour, communication, work, recreation and pastimes, and eating. Respondents are asked about their well-being while filling out the questionnaire and provide their answers by marking 'yes' or 'no'. Results are measured from 1 to 100 [22].

Respiratory disease specific questionnaire

1. **Chronic Respiratory Disease Questionnaire (CRDQ)** – A questionnaire the aim of which is to measure the impact of chronic respiratory disease symptoms on emotional and functional condition. It was developed by Guyatt and consists of 20 questions divided into 4 domains: dyspnea, fatigue, emotional function, and mastery. The answers are measured in 7-point modified Likert Scale. The questionnaire is available in Polish [23].

2. **St. George's Respiratory Questionnaire (SGRQ)** – A questionnaire developed at St. George's Hospital in Great Britain by professor P.W. Jones. It is one of the most important instruments for QoL assessment in respiratory diseases particularly in patients with Chronic Obstructive Pulmonary Disease and patients with asthma. The questionnaire consists of 76 questions in three domains: the first one concerns respiratory symptoms i.e. bringing up phlegm, coughing, attacks of wheezing episodes of shortness of breath. The second one focuses on limitations in physical activity resulting from airways disease and the third one analyses the impact of disease on daily social and emotional life. The answers in the first category are expressed in 5-point Likert Scale whereas in the second and third category the answers are dichotomous 'yes' or 'no'. Scores range from 0 to 100, with higher scores indicating more limitations. The questionnaire is available in Polish version the author of which is Kuźniar et al. [24].

3. **Pulmonary Functional Status & Dyspnea Questionnaire (PFSDQ)** – The questionnaire consists of 164 questions divided into 2 components: functional ability and dyspnea as related to six categories: social functioning, recreation, mobility, home management, eating and self-care. Modified Likert 0 to 10 Scale with verbal descriptors is used; higher scores indicate worse functional status and/or dyspnea symptoms. It is available in English [25].

4. **Shortness of Breath Questionnaire (SoBQ) –** The questionnaire is a valuable assessment instrument in both clinical practice as in research in patients with moderate and severe pulmonary disease. It consists of 24 items that assess self-reported shortness of breath connected with activities of daily living (21 questions). Three questions concern limitations due to: shortness of breath, fear of harm from overexertion and fear of shortness of breath. Answers are provided on 6-point scale. Maximal score to be obtained amounts to 120 points. It is available in English [26].

Cystic fibrosis specific questionnaires

1. **Cystic Fibrosis Quality of Life Questionnaire (CFQoLQ 14 years) –** The questionnaire was developed in the UK by Gee et al. It is a multidimensional instrument focusing mainly on a patient's well-being and to a lesser degree on dysfunctions. It consists of 52 questions divided

into 9 subscales: Physical Functioning, Social Functioning, Emotional Functioning, Treatment Issues, Chest Symptoms Interpersonal Relationships, Body Image, Future Concerns, Career Concerns. Scaling of items: 6-point Likert Scale where 1 indicates the worst assessment and 6 the highest (in question 6 it is the other way round). Following transformation, the results are ranged from 0 to 100 points where 100 points indicates the most optimal quality of life. The questionnaire meets all requirements in the scope of reliability and validity and was adapted to Polish conditions by Dębska et al. [27,28].

2. **Cystic Fibrosis Questionnaire (CFQ 14 years, CFQ 6 years)** – Cystic Fibrosis Questionnaire version for children: 14 years and a parent and 6 years and a parent. It was developed especially for people with diagnosed CF. It consists of 44 questions concerning 9 domains: Physical Functioning, Vitality, Emotional state, Social limitations, Role Limitations/School Performance, Embarrassment, Body Image, Eating Disturbances, Treatment Constraints and 3 symptoms scales: Respiratory, Digestive, Weight. Scaling of items: 4-point Likert Scale (e.g. always/often/ sometimes/never). Scores range from 0 to 100, with higher scores indicating better health. The original questionnaire was developed in France, was adopted to Polish conditions by Sands et al. and is available in Polish [29].

3. **Questions of Life Satisfaction – Cystic Fibrosis FLZN-CF** – The questionnaire was presented by German researchers Goldbeck L, Schmitz TG, Henrich G. et al. It is used for subjective assessment of life satisfaction in the prior 4 weeks. It consists of 16 questions concerning the assessment of satisfaction and importance of disease symptoms such as breathing difficulties, digestive trouble, routine therapy, sleep, adherence to daily therapy, significance for others, understanding, and free from disadvantage. Answers are provided on a 5-point scale. The questionnaire is designated for patients over 15 years of age [30].

CONCLUSION

The objective of this paper was to present study tools for assessment and analysis of health related quality of life in patients with cystic fibrosis. CF is a chronic condition of progressive course therefore, despite of wide selection of questionnaires, the administration of questionnaires more sensitive to changes in health status i.e. disease specific is advised. Selecting an appropriate research instrument based on a study goal is of great importance since it enables correct analysis of QoL and thus showing the actual problems of patients.

PIŚMIENNICTWO

1. Mazurek H. Mukowiscydoza 610 – 615, [w:]. Szczeklik A, red. Choroby wewnętrzne, stan wiedzy na 2010 rok. Medycyna Praktyczna. Kraków 2010.
2. Zalecenia Polskiego Towarzystwa Mukowiscydozy 2009 Poznań–Warszawa–Rzeszów. Zasady rozpoznawania i leczenia mukowiscydozy. Standardy Medyczne/Pediatria. 2009; 6: 352-378.
3. Nowakowska A, Sands D, Piotrowski R i wsp. Okresy przeżycia chorych na mukowiscydozę. Pediatr.Pol. 1999; 74: 459-463.
4. Majka L, Pogorzelski A, Młynarczyk W i wsp. Sytuacja społeczna dorosłych chorych na mukowiscydozę w Polsce. Acta Pneumonologica et Allergologica Pediatrica. 1999;2: 1, 8-11.
5. Stężowska-Kubiak S. Charakterystyka epidemiologiczno-kliniczna polskiej populacji chorych na mukowiscydozę. Rozprawa doktorska. Poznań: Uniwersytet Medyczny. 2011.
6. Dębska G. Jakość życia chorych na mukowiscydozę. Acta Pneumonologica et Allergologica Pediatrica. 2003; (6) 3: 39-42.
7. Ostrzyżek A. Jakość życia w chorobach przewlekłych. Probl Hig Epidemiol. 2008; 89: 467-470.
8. Dziurowicz-Kozłowska A. Wokół pojęcia jakości życia. Psychologia jakości życia 2002; 1(2): 77 -99.
9. Kłak A, Mińko M, Siwczyńska D. Metody kwestionariuszowe badania jakości życia. Probl Hig Epidemiol. 2012; 93(4): 632-638.
10. Schipper H. Quality of life: Principless of the clinical paradigm. Journal of Psychosocial Oncology. 8; 2/3: 171-185.
11. Farnik – Brodzińska M, Pierzchała W. Znaczenie badań jakości życia w przewlekłych chorobach układu oddechowego w aspekcie holistycznego pojęcia medycyny. Wiad. Lek. 1998; 52: 7-8.
12. Wołowicka L. (red.). Jakość życia w naukach medycznych. Poznań; 2001.
13. De Walden – Gałuszko K, Majkowicz M. Jakość życia w chorobie nowotworowej. Gdańsk: Wydawnictwo Uniwersytetu Gdańskiego; 1994
14. Chrobak M. Ocena jakości życia zależnej od stanu zdrowia. Problemy Pielęgniarstwa. 2009; 17(2): 123-127.
15. Sokolnicka H, Mikuła W. Metody oceny jakości życia mające zastosowanie w medycynie. Med. Rodz. 2003;3(4): 129-131.
16. Trojanowska A. Znaczenie badań nad jakością życia w medycynie. Zdr. Publ. 2011; (121) 1: 99-103.
17. Dębska G. Badanie jakości życia pacjentów z przewlekłymi chorobami układu oddechowego. Acta Pneumonologica et Allergologica Pediatrica. 2003; (6) 2: 39-42.
18. Kaplan RM et al. The quality of well-being scale. Med Care. 1989; 27:1076-1083.
19. McEwen J, Hunt S, McKenna S. A measure of perceived health: the Nottingham Health Profile. [w:] Abellin T, Brzeziński ZJ, Carstairs VDI, red. Measurement in health promotion and protection. Copenhagen: World Health Organization. 1987: 590 -603
20. Ware JE, Kosinski M et al. SF -36 Health Survey: Manual and Interpretation Guide. Boston: The Health Institute, New England Medical Center; 1993.
21. The WHOQoL Group, The World Health Organization Quality of Life Assessment (WHOQoL): Development and general psychometric properties. Soc Sci Med. 1998; 46: 1569 -1585.
22. De Jong W. et al. Quality of life In patients with cystic fibrosis. Pediatri. Pulmonol. 1997; 23: 95-100.
23. Tobiasz-Adamczyk B, Owsiński J, Wojtowicz E. Jakość życia chorych z przewlekłymi chorobami układu oddechowego. Zdrowie Publiczne. 1999; 4: 133-137.
24. Jones PW et al. The St George's Respiratory Questionnaire. Respir. Med. 1991; 85 suppl. B.: 25-31.
25. Lareau SC. et al. Development and testing of the modified version of the pulmonary functional status and dyspnea questionnaire (PFSDQ). Heart Lung. 1998; 27: 159-68.
26. Kaplan RM Et al. Validation of a new dyspnea measure. The UCSD shortness of breath questionnaire. Chest. 1998; 113: 619-24.
27. Gee L et al. Development of a disease specific health related quality of life measure for adults and adolescents with cystic fibrosis. Thorax. 2000; 55: 946-954.
28. Dębska G, Mazurek H. Kwestionariusz CFQoL jako narzędzie oceny jakości życia chorych na mukowiscydozę. Pol. Merkuriusz Lek. 2007;23: 340-343.
29. Henry B, Aussage P, Grosskopf C et al. Development of the Cystic Fibrosis Questionnaire (CFQ) for assessing quality of life in pediatric and adult patients. Qual Life Res. 2003; 12(1): 63-76.
30. Goldbeck L et al. Questions on Life Satisfaction for Adolescents and Adults with Cystic Fibrosis. Chest. 2003; 123: 42-48.

Praca przyjęta do druku: 24.02.2013
Praca zaakceptowana do druku:03 .06.2013