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Original Article

# Comparison of health-related quality of life among children with cystic fibrosis and their parents in two Eastern European countries

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**Abstract**

*Background:* In the optimal care of cystic fibrosis (CF) patients, not only medical parameters are respected but also health-related quality of life (HRQOL). The aim of our study was to compare HRQOL of CF patients from two Eastern European countries.
*Methods:* 141 patients with CF (6–18 years) and 102 parents completed the Cystic Fibrosis Questionnaire-Revised (CFQ-R). Data about disease severity, type of children's education and questions about parents' employment status were collected.
*Results:* In the patient group, a significant difference was found only in *Treatment burden*, whereas in the parent group, there were significant differences in *Treatment burden, Emotional functioning, Eating* and *Digestive symptoms* between the two countries. School attendance was revealed as an important factor influencing HRQOL.
*Conclusions:* Observed differences in evaluation of HRQOL may be caused by different therapeutic and diagnostic challenges between countries. To identify possible presence of psychosocial problems, monitoring of HRQOL is recommended.
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*Keywords:* Health-related quality of life; *Cystic fibrosis*; Individual education; Eastern Europe

## 1. Introduction

Worldwide, increasing attention is focused on individualised and customised therapeutic approaches to patients with cystic fibrosis that aim to improve not only the longevity but the quality of life of these patients [1]. Health-related quality of life (HRQOL) is a very well-documented measure of patients' reported outcomes (PROs). PRO is any report of a patient's health condition that comes directly from the patient [2]. The Guidance of the US Food and Drug Administration recommends the use of PRO data measuring the treatment benefit of clinical trials [3]; in addition, the importance of PROs is increasing in the everyday care of chronically ill patients [4].

It is challenging to compare Western and Eastern European clinical practice patterns and, consequently, the outcomes of treatment strategies, because of the limited diagnostic tools in Eastern Europe. It causes declension in patient care which causes our patients' average age of survival and clinical status to be slightly worse than their Western counterparts. It is also well known that diagnostic delay can cause significant deterioration in CF patients' clinical status, resulting in malnutrition, severe chronic lung disease and bacterial colonisation with *Pseudomonas aeruginosa* (PA), or accelerating the decline in lung function in early childhood [5]. Suboptimal patient care and

*Abbreviations:* CF, Cystic fibrosis; HRQOL, Health-related quality of life; CFQ-R, Cystic fibrosis questionnaire-revised; $FEV_1$, Forced expiratory volume in one second; BMI Z-score, Body mass index standardised for age; PA, *Pseudomonas aeruginosa.*

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subsequent worse clinical status may lead to the patients' limited survival to early adulthood and deteriorated HRQOL.

There are some differences in CF care between Hungary and Poland which can lead to important differences in health perception of the patients. The health insurance and reimbursement systems have significant differences which can lead to more difficult availability of drugs free of charge or with allowance in Poland as in Hungary. Contrary to Poland, the availability of diagnostic tools are limited in Hungary, as newborn screening for CF and objective measures of pancreas sufficiency are not available. In Hungary, conductivity is still measured for the diagnosis of CF instead of sweat chloride method which is recommended according to the Guideline of the European Cystic Fibrosis Society [6]. Furthermore, there are numerous basic problems in relation to CF care in the Eastern European region, such as the limited possibilities to perform lung function testing on children younger than 6 years using full-body plethysmography or multiple-breath washout techniques. There is limited access for physiotherapists, nutritionists, and psychologists to be part of the CF team in specialised CF units in Poland and Hungary.

When our study was planned, we assumed that the quality of life of patients in the Eastern European region might differ from those living in countries with higher income. Published data about the framework of care in seven low-income countries (Czech Republic, Hungary, Latvia, Poland, Serbia, Slovakia and Ukraine) demonstrated that in many CF centres, there is a lack of a full-time CF nurse specialist, dietician, microbiologist, psychologist, social worker or secretarial support. The reasons for such inequalities may be the absence of appropriate funding, insufficient staff recruitment and training, and also inadequate political prioritisation [6,7].

In our study, we assessed and compared the HRQOL of Polish and Hungarian patients with CF and evaluated potential factors that influence HRQOL. In 2012 in Hungary, there was a total of 579 patients with CF (median age: 16.4 years) out of 10 million inhabitants [8]; in Poland in 2012, there were 1620 patients diagnosed with CF [9] out of 38 million inhabitants (median age: 14.0 years).

We evaluated social factors that can influence HRQOL. In relation to the children we should consider that a child with CF attends school activities along with their peers. This is of great importance in the development of both emotional and social functionings. Usually, a poor health condition is the reason to choose individual forms of learning at home. But sometimes children do not attend school due to problems in social functioning and also because of parental overprotection.

To our knowledge, this is the first comparative study of CF patients' HRQOL from Eastern Europe, as no published study was found by conducting a literature search in Medline using the following keywords: quality of life, cystic fibrosis and Eastern Europe until the date of 01/04/2014.

Our study objectives included:

1. To evaluate and compare the HRQOL in children with CF from two Eastern European countries: Poland and Hungary.
2. To evaluate the associations between the HRQOL and social factors.

## 2. Materials and methods

### 2.1. Participants and procedure

A single cross-sectional study was conducted over a one-year period in one paediatric outpatient CF centre in Poland and five in Hungary. Children who were between 6–18 years of age and had a confirmed diagnosis of CF according to national standards were eligible to be included [10,11]. Patients with clinical exacerbation, acute respiratory infection, other chronic illnesses, mental retardation or reading difficulties were excluded from the study.

All 161 patients with CF who met the inclusion criteria were asked to participate; 43 Hungarian and 98 Polish children were prospectively included in the study. In both countries, the recruitment process looked the same. The study was approved by the Semmelweis University Regional and Institutional Committee of Science and Research Ethics in Hungary and the Bioethical Committee of the Institute of Mother and Child in Poland. Written informed consent was obtained and the legal caregivers of children younger than 18 years were asked to give permission.

### 2.2. Instruments used in the study

Age, gender, type of genetic mutation, forced expiratory volume in one second ($FEV_1$% predicted), body mass index (BMI) Z-score, *P. aeruginosa* status, parents' employment status and children's school attendance were recorded.

HRQOL was measured using the Cystic Fibrosis Questionnaire-Revised (CFQ-R). The CFQ-R is a widely used, validated, disease-specific instrument for HRQOL measurement in patients with cystic fibrosis [12–16]. The Hungarian and Polish [17] versions of the CFQ-R were used to assess HRQOL from patients' (patient-report) and their parents' perspectives (parent-proxy report). The CFQ-R has both generic and disease-specific domains. Four versions of the CFQ-R were used: the CFQ-R Child 6–11 and the CFQ-R Child 12–13 contain 35 items divided into 8 domains; the CFQ-R Teen/Adult is a patient-report version with 50 items divided into 12 domains for patients $\geq$14 years; and the CFQ-R Parent is a proxy-report with 44 items divided into 11 domains for parents of children who are 6–13 years.

The questionnaire provides a summary score in each domain; scores range from 0 to 100 and higher scores indicate a better HRQOL. Response choices include ratings of frequency and difficulty on a 4-point Likert scale (1 = always to 4 = never, 1 = a lot of difficulty to 4 = no difficulty) or true or false responses (1 = very true to 4 = very false). Scoring was computed only if at least half the questions have been completed within each domain. Medical data of the participants were obtained from each patient's file and included current height, weight, infection with mucoid strains of PA during the last year, and type of mutation.