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ORIGINAL ARTICLE

Disease specific knowledge about cystic fibrosis, patient education and counselling in Poland

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Abstract

Introduction and objective. The presented study assesses levels of specific knowledge of the disease among cystic fibrosis (CF) patients and their families, and evaluates the effectiveness of a targeted, disease specific education programme.

Materials and methods. A cross-sectional survey among 462 families with a CF child evaluated their knowledge of the disease. A one year follow up survey among 200 families assessed the effectiveness of an educational programme developed to correct gaps, errors and misconceptions identified in the previously administered survey. Self-administered, comprehensive, 5-domains, 45-item multiple-choice CF Disease Knowledge Questionnaire (CFDKQ) was anonymously completed by 462 subjects.

Results. 228 respondents were male (49%), 234 female (51%). The level of disease-specific knowledge in the age groups 0–6 and 7–10 years, was significantly higher than in 11–14 and 15–18 years of age groups ($p<0.005$). General medical and Genetics/Reproduction knowledge was low in all patients. Significant predictors of patient and parental knowledge were age and domicile. Patients and parents rely heavily on doctors for information about CF (77%). The follow-up survey (CFDKQ) emphasized that special education programmes significantly improved levels of disease specific knowledge ($p<0.0001$).

Conclusions. If left uncorrected, the misconceptions, gaps and errors in CF knowledge identified in the presented study could result in inadvertent non-adherence to treatment, and impact on the progression and outcome of the disease. Secondly, the results demonstrate the effectiveness of targeted, disease specific information in improving disease knowledge of CF patients and their families, and highlights the value and need for the development of educational programmes for chronically ill patients and their families.

Key words

cystic fibrosis, disease knowledge, education, information

INTRODUCTION

Cystic Fibrosis (CF) is the most common autosomal recessive disorder of childhood in Caucasian populations, affecting 1 in every 2,500–3,000 live births with a carrier rate of one in 25–30 inhabitants [1, 2]. The disease is characterized by abnormalities in electrolyte and macromolecule secretions of exocrine glands, leading to chronic pulmonary obstructions, infections and digestive disorders, such as exocrine pancreatic insufficiency. CF is still fatal with a median survival age of about 30 years. The multi-faceted, chronic treatment imposes a great burden on the patients and their families. As life expectancy increases, patients and their families are undertaking increasing responsibility for medical management, and are assuming more control over their treatment. Information about CF specific knowledge levels among patients and their parents is inconsistent and/or lacking [3, 4, 5, 6, 7]. The presented study was undertaken to assess levels of disease specific knowledge among CF patients and their families, and to evaluate the effectiveness of a targeted, disease specific education programme. If left uncorrected, the misconceptions, gaps and errors in the knowledge of CF that were previously identified could potentially result in inadvertent non-adherence to treatment of the patient and thereby influence the progression of the disease.

This paper highlights the value and need for the development of an educational programme for CF patients and their families.

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MATERIALS AND METHOD

Participants. One-thousand (1,000) questionnaires were distributed to CF patients and their families, of which 462 were completed and returned (46% response rate). After one year, a follow-up was conducted which had a response rate of 43% (200 CF patients and their families).

Study design. The design of the presented study was approved by the Institutional Review Board at the University of Medical Sciences in Poznań, Poland. The study was explained and presented to CF patients and their families who signed informal consent for participation in the study.

The study was conducted in three phases:

Phase-1(cross-sectional): conducted among 462 CF patients and their families to asses their medical knowledge of the illness, using the Cystic Fibrosis Disease Knowledge Questionnaire (CFDKQ).

Phase-2(intervention): educational materials were prepared specifically targetting gaps and misconceptions previously identified in the cross-sectional phase. These were

subsequently distributed to CF patients via regular mail and using the cystic fibrosis web portal: cystic fibrosis virtual community: www.virtualcf.pl.

Phase-3(longitudinal): of the 462 patients who completed phase-1,200 patients completed a one-year follow-up assessment using CFDKQ, with the aim of measuring the effectiveness of the educational programme.

Measures. The CFDKQ was developed in two versions: one for parents of patients under 14 years of age and one version for CF adolescents. It consists of 45 multiple choice questions divided into 5 sections dealing with all aspects of CF: general medical knowledge (5 questions), general knowledge on CF (10 questions) [6], specific knowledge about symptoms and treatment of respiratory disorders (10 questions) [8], specific knowledge about genetics/ reproduction (10 questions) [3, 9] and specific knowledge about symptoms and treatment of gastrointestinal disorders (10 questions) [10].

Answers were evaluated as correct (+1 point), wrong (-1 point) and neutral "don't know" as (0 points) [7, 10]. The option of "don't know" was included in order to reduce guessing. Results were calculated as an average and a median of all questions in each domain and as well as a total score.

Table 1 displays CFDKQ.

Statistical methods. Statistical analysis was performed by the Biostatistics Department of Poznań University of Medical Sciences using the Statsoft Inc. package Statistica PL version 7.1. [11] Spearman rank order correlations were used to examine associations between the knowledge about CF and the sociodemographic and medical variables.

Questionnaire results and their medians were compared using non-parametric Mann Whitney test for two non-dependent groups defined by gender, concomitant diseases, presence of CF in the family, financial situation of family, domicile and type of CF. The results were also tested using non-parametric variance analysis ANOVA of Kruskal-Wallis. The results were considered to be statistically significant at significance level $p<0.05$.

RESULTS

Cross-sectional part. Completed questionnaires were received from 462 (n=462) adolescents and parents of CF children. The 462 subjects with CF consisted of 228 boys (49%) and 234 girls (51%) in the following age ranges: 0–6 years: 94 patients (20%), 7–10 years: 91 patients (20%), 11–14 years:78 patients (17%), 15–18 years: 108 patients (23%), 91 patients (20%) aged over 18.

122 patients (26%) lived in big cities > 100,000 inhabitants, 153 (34%) lived in towns <100,000 inhabitants and 187 patients (40%) lived in villages.

The majority of children were diagnosed with CF when they were under one year of age: 345 patients (75%) were diagnosed between the age of the 2nd and 3rd month of life, between 1–3yrs: 49 (10%), between 4–6yrs: 35 (7%), between 7–15 yrs:26 (5%), and 7 patients (3%) above the age of 15.

17% (n=80) of patients had predominantly respiratory symptoms, 15% (n=64) had predominantly abdominal symptoms, and 68% (n=318) had a mixture of both. 42% (n=165) of CF patients had (an)other chronic illness(es) requiring daily treatment.

Table 1. Examples of items included in the Cystic Fibrosis Disease Knowledge questionnaire (CFDKQ)

General Medical Knowledge	1	Common cold is caused by:	
		A	Bacteria
		B	Viruses
		C	I don't know
	2	Antibiotics are effective against infections caused by:	
		A	Bacteria
		B	Viruses
		C	I don't know
Cystic Fibrosis specific Knowledge	3	Sputum examination is important for:	
		A	Establishing the right dose of antibiotic
		B	Detection of bacteria and assessment of their susceptibility on antibiotic
		C	I don't know
	4	Organs affected by CF:	
		A	Lungs, pancreas, liver, reproduction system
		B	Brain, heart, stomach
		C	I don't know
Genetic & Reproduction specific Knowledge	5	Does the carrier of *CF gene* have clinical symptoms?	
		A	Yes
		B	No
		C	I don't know
	6	Risk of siblings as carriers:	
		A	25%
		B	50%
		C	I don't know
Respiratory Symptoms specific Knowledge	7	Outdoor exercise and sport for children with CF should be:	
		A	Forbidden, because they worsen the cough
		B	Encouraged, because they facilitate airway clearance
		C	I don't know
	8	Children with CF may suffer from frequent pneumonia because:	
		A	They lack Vitamin 'C':
		B	Germs become trapped in the mucus of their lungs
		C	I don't know
Gastrointestinal Symptoms specific Knowledge	9	Children with CF need to put on weight, which of the following has highest calorificc charge:	
		A	Toasted bread with fruit juice
		B	Bacon and eggs
		C	I don't know
	10	Pancreatic enzymes should be taken:	
		A	3 times daily, before meals
		B	At the beginning and during each meal
		C	I don't know

There were 15% of single parent families (n=69); 1% of families with grandparents (n=7), while the majority of families had both parents (84%, n=386).

The main source of knowledge about CF for patients and their parents was a physician (77%), followed by medical literature (75%). A substantial part of knowledge was obtained through peer-to-peer contact among parents of CF patients (47%). New sources of information, e.g. the Internet, were indicated by 35% of CF patients and their parents (Tab. 2). Although sources of information on CF are relatively widely

Table 2. Sources of CF knowledge (% response)

Physician	77 %
Medical literature	75 %
Other parents	47 %
Internet	35 %

available and accessible there were still unmet needs among the subjects concerning information on detection of flares and exacerbations (68%), information on diagnostics (68%), rehabilitation methods (61%), nutrition (58%) and drug treatment (55%).

The subjects expressed a relatively low interest in information on proper parent-patient relationship (39%), genetics and reproduction (26%) (Tab. 3).

Table 3. Knowledge needs among CF patients (% response)

Detection of flares and exacerbations	68 %
CF diagnostics	68 %
Rehabilitation methods	61 %
Nutrition	58 %
Drug treatment	55 %
Parents-patient relationship	39 %
Genetics & reproduction	26 %

Assessment of knowledge levels measured by the CFDKQ allows both evaluation of a total test score and domain/sub-domain specific scores. The total score of knowledge levels related to the age groups is presented in Table 4.

Table 4. Summary data on CF Knowledge Total Scores

Age	N	Mean	Q25	Median	Q75
0–6*	94	0.76	0.71	0.78	0.84
7–10*	91	0.75	0.69	0.78	0.87
11–14	78	0.67	0.60	0.69	0.80
15–18	108	0.66	0.56	0.67	0.78
>18	91	0.72	0.62	0.76	0.87
Total	**462**	**0.71**	**0.62**	**0.73**	**0.82**

* p<0.005

Mean values were at the level 0.71, while median values were at the level 0.73; the small difference indicating an absence of extreme results. A closer analysis shows that age group 0–6 and 7–10 have significantly higher values of total knowledge score compared to age group 11–14 and 15–18 yrs (p<0,005) (Tab. 5).

Table 5. Summary data on CF Knowledge Questionnaire (% of correct answers)

Knowledge Domains	< 14 yrs	> 14 yrs
General Medical Facts	40%	41%*
General Cystic Fibrosis	92%	89%
Genetics & Reproduction	56%*	46%
Respiratory Symptoms	80%*	76%
Gastrointestinal Symptoms.	84%*	78%

*p<0.005

Assessment of knowledge on the domain levels reveals that the population of CF patients under 14 years of age have significantly higher knowledge levels in the domain Genetics and Reproduction (p<0.0001), Respiratory Symptoms (p<0.03) and in the domain Gastrointestinal Symptoms than the population (adolescents) above the age 14. (p<0.02). In contrast, the General Medical knowledge of patients >14 yrs was significantly higher in comparison with the population < 14 years of age (parents), (p<0.03).

In the total assessment knowledge levels in both domains: General Medical knowledge, and Genetics and Reproduction was significantly lower than in other domains (p<0.05). Table 6 describes the Cystic Fibrosis Disease Knowledge Questionnaire.

Table 6. Cystic Fibrosis Disease Knowledge Questionnaire

	Domain Genetics & Reproduction	% correct answers
1	Is CF a hereditary disease?	95
2	Does a CF carrier transmit the CF gene to all his children?	91
3	Must both parents of CF child be carriers?	85
4	Is a CF carrier affected (ill)?	95
5	What is the risk of a future sibling of a CF child being a carrier?	21
6	What is the risk of having a CF child if both parents are carriers?	35
7	What is the risk of having a CF child if only one parent is a carrier?	60
8	What is the risk of having a CF child if one parent has CF?	26
9	Are CF male patients all fertile?	88
10	Can CF women become pregnant?	97
	Domain Respiratory Symptomatology	% correct answers
1	Outdoor exercise and sport for children with CF should be… …..?	94
2	CF children can suffer from pneumonia more frequently because…	100
3	Increased volume of green colored sputum is a symptom of…	91
4	A frequent cause of pulmonary infection is bacteria …	78
5	Effective treatment for pulmonary infections are…	97
6	Mucolytics are drugs which…	87
7	Inhaled antibiotics should be administered before physiotherapy…	41
8	CF children who are coughing should stay at home…	92
9	Physiotherapy is not necessary when a patient is feeling well…	84
10	Physiotherapy shall be practiced with a minimum …	93
	Domain Gastrointestinal Symptomatology	% correct answers
1	CF patients should use a no-fat diet…	84
2	CF children should gain weight. Which of listed menus is right?	85
3	Smelly stools can be a symptom of fat malabsorption…	97
4	Abdominal pain can be caused by lack of pancreatic enzymes…	94
5	Which has a higher caloric value, sandwich with jam or cheese?	91
6	A diet rich in fat has a higher caloric value than carbohydrates…	64
7	Should CF children eat a low-salt diet ?	86
8	Should CF children take pancreatic enzymes with every meal?	99
9	Pancreatic enzymes should be taken before, during or after the meal?	100
10	Abdominal pain and bloating can be caused by…	100

An additional significant predictor was domicile. Patients living in cities have significantly higher scores in CFDKQ test than those living in villages (p<0.001). Especially in the domains: General Medical Knowledge (p<0.02), Genetics and Reproduction (p<0.03) and Respiratory symptoms (p<0.001).

There were no statistically relevant correlations between level of knowledge and gender, financial situation of the family, type of CF (respiratory predominant, gastrointestinal predominant or mixed form), another chronic illness, or existing CF in the family.

Follow-up after 12 months. In the follow up phase 12 months after the initial test with the CFDKQ, a follow-up CFDKQ was administered. 200 (n=200) subjects responded. 88 (n=88) questionnaires were from adolescents above 14 years of age, and 112 (n=112) concerned patients below the age of 14 (parents). The implemented educational programme resulted in a significant improvement (p<0.0001) in the total CFDKQ score, as well as in the single sub- domains (Tab. 7, 8).

Table 7. Total scores of follow up CFDKQ (median). Population < 14 yrs (N=112)

Knowledge Domains	Median phase 1	Median phase 3
General Medical Facts	2.00	3.00*
General Cystic Fibrosis	10.00	10.00
Genetics & Reproduction	6.00	8.00*
Respiratory Symptoms	8.00	9.00*
Gastrointestinal Symptoms	8.00	10.00*
Total	**7. 80**	**8.70***

*p<0,0001

Table 8. Total scores of follow up CFDKQ (median). Population > 14 yrs (N=88)

Knowledge Domains	Median phase 1	Median phase 3
General Medical Facts	2.00	3.00*
General Cystic Fibrosis	10.00	10.00
Genetics & Reproduction	4.00	8.00*
Respiratory Symptoms	8.00	10.00*
Gastrointestinal Symptoms	8.00	10.00*
Total	**7.10**	**8.20***

*p<0,0001

In both groups (<14 years and > 14 years), a significant improvement was observed in the domains: General Medical Facts, Genetics and Reproduction, Respiratory Symptoms and Gastrointestinal Symptoms. The lack of improvement in the domain General CF Knowledge can be explained by the fact that maximum scores were already obtained in the initial test.

DISCUSSION

Information source and value. In general, subject (patient and parent) disease specific knowledge on chronic childhood illnesses is considered important to ensure subsequent effective treatment. Examples are: better disease control in asthma patients [12], better maintenance of remission in Crohn's disease following nutritional education [13], and positive correlation between children's understanding of their illness and its treatment in a paediatric oncology unit [14]

Until now, there have been published only a few, non-controlled trials demonstrating that CF specific knowledge is an important factor determining therapy success [6], contributing to development of patient independence [15] and treatment adherence [16]. Since CF is primarily diagnosed in early childhood, it is important that parents and patients receive all necessary knowledge for effective treatment, nutrition and rehabilitation at an early time point.

The results of the presented study reveal that CF patients and their families who live in cities have significantly higher knowledge levels than their peers living in villages, especially concerning the areas of General Medical Facts, Genetics and Reproduction and Respiratory Symptoms. This can be attributed to the lower educational level of their parents, lower accessibility to medical specialists, and lower opportunity of contact between parents and other CF patients in more rural areas. The availability of computers and access to the Internet is also lower in this population.

The presented study confirms the role of the physician as the dominant source of information for patients and their families. It is important to realize that especially on the physician-parent level much misunderstanding and errors can occur. It has been previously observed [17] that up to 32% of parents are not in a position to fully understand the physicians' recommendations for their children. If patients/parents are not fully versed in the specifics of the daily medical regimen, they cannot be expected to adhere to it. Thus, the notion that lack of understanding can lead to poor treatment adherence makes intuitive sense. Not without blame is the disproportional use of professional, medical terminology, which is rarely understood by parents and patients. [18]. In our opinion, the specialist's terminology should not be avoided, but thoroughly explained to patients and parents alike.

It has been demonstrated here that the knowledge needs of patients and their parents has been improved by specially developed educational materials containing general information. In these materials, specific medical terminology was mentioned, accompanied, however, by a detailed explanation of all the terms used.

The presented study reveals that new sources of information, e.g. the Internet, are still not commonly used. Only 35% of patients and their families mention the Internet as a source of information.

Since many patients complaining about difficult access to specialists an alternative way of communication, like internet, can be helpful in obtaining relevant information. Current trends show that virtual environments, such as the virtual community portal www.virtualcf.pl can be effective platforms for communication [19].

Disease specific knowledge. Although this study has demonstrated that CFDKQ scores (median=0,73) of CF patients and their families are relatively satisfactory; the identified errors, gaps and misconceptions in disease related knowledge should be a cause for concern.

For patients under 14 years of age, the test results actually reflect the knowledge of their parents, contrary to the CF population above 14 years of age. Hence, parents were more knowledgeable about Genetics and Reproduction,

Respiratory Symptoms and Treatment, and in the domain Gastrointestinal Symptoms and Treatment. These findings confirm other publications on this topic. [6]

The substantially lower scores for both tested groups in the domain General Medical Facts is mainly caused by difficulties in discerning between the symptomatology of bacterial and viral infections and treatment This confirms results published by L. D. Henley [4]. Viral infections like the common cold frequently occurring in the general population can be wrongly interpreted as a sign of disease progression, or may even be attributed to treatment non-adherence.

Knowledge about Genetics and Reproduction was shown to be very low (parental score: 56%, children > 14 years (46%). Nonetheless, there was little interest expressed in more information on Genetics and Reproduction – only 26% expressed interest.

As information on Genetics and Reproduction tends to be difficult and quite complicated, patients and their parents lack the ability to make use of and value this information. This is confirmed in our questionnaire where the most difficult questions to be answered correctly were the ones related to reproduction, sexual life, or the possibility of having a healthy child. Most respondents were not aware that the majority of males with CF are sterile.

Clarification of complex disease-specific knowledge domains can potentially influence the quality of life of cystic fibrosis patients.

In the Respiratory Symptoms domain only 40% patients and parents knew that nebulized antibiotics should be administered after physiotherapy. 22% indicated difficulty in discerning between bacterial and viral infections and accompanying symptomatology and treatment.

Given that a high calorific intake is desirable for all cystic fibrosis patients, the identified gaps in the Gastrointestinal Symptoms domain were related, surprisingly, mainly to the nutritional part. Another question which was difficult to answer correctly was related to the need for replenishment of salt during hot weather or following intense physical exercise.

The results of the presented study reiterate the importance of knowledge about the practical aspects of the daily life of CF patients and their parents, i.e. on physiotherapy, nebulization, nutrition and proper pancreatic enzyme supplementation, sports and exercise. Sound knowledge is a prerequisite for optimal treatment adherence.

This study highlights the value and effectiveness of targeted, disease specific information in improving disease specific knowledge of CF patients and their families, and such material should be further developed, thereby limiting the risk of treatment non-adherence and possibly influencing the course of disease.

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