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Problems of patients with cystic fibrosis during transition to adulthood

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Abstract

The proper care of cystic fibrosis patients extends over their lifetime. More than half of the children with the disease die before adulthood. An important element in the patient's care is a time of transition from a paediatric to the care of an internist and the patient's acceptance of this necessity. Transition from paediatric care to an internist should be adequately prepared. It is not only a question of transfer of medical records, but also careful preparation of patients for such transition. The patients expect not only continuity of care but also the introduction to the management with the disease. The creation of a base for specialist hospital treatment for exacerbation of the disease at the adulthood is an important element in the care of these patients. The problem has been solved in the children group, but is still waiting for solution in adults with cystic fibrosis. It has been proven that care in the centres carried out by a specialized team ensures longer life and better quality of life of these patients. The paper is an overview of these two important elements of care of adults with cystic fibrosis.

Key words: cystic fibrosis, childhood to adulthood

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Introduction

In Poland, cystic fibrosis (CF) affects approximately one in every 4,400 births [1]. In statistical terms, there are around 80 babies with cystic fibrosis born each year. The Polish population of patients with CF is estimated at around 1,500 (according to the CF Patients Register of the Polish Cystic Fibrosis Society; some of the listed paediatric patients are found to have unknown mutations and are subject to clinical observation).

However, the statistics only include diagnosed patients. Until recently, CF was diagnosed within the first two years of life in a clear majority of cases. Since mid-2009, newborn screening for selected congenital diseases, including cystic fibrosis, has been under way all over Poland. The currently used highly sensitive screening model makes it highly unlikely to detect any cases of CF in the paediatric population who have tested negative. However, there are still cases where CF is diagnosed in individuals who have not been screened (born before July 1st 2009), including adults (which represent around 2.5% of all diagnosed cases).

Overall, the annual incidence rate of CF is in Poland is estimated at around 70–80 new cases. In 2011, adult patients represented over one third of the total CF population of over 500 patients in Poland, which makes it clear that physicians specifically trained in the management of adult patients should have at least 60 new cases per year.

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A mean age of CF population is 14.4 years; around 35% of cystic fibrosis patients are adults [2].

A median age of patients with CF in 2003 was 16.3 years ± 4.9 and differed between countries. In UE countries it amounted to 17 years, in non-EU countries it amounted to 12.1y. Data were collected from 35 countries on the basis of information about 29,025 patients. In the UE, patients aged 18–40 years accounted for 42% of all patients, in countries outside the EU — 26%. Accordingly, patients over 40 years of age accounted for 5% of all patients in the UE and 2% of all patients in the countries outside the EU, Poland has not been included in the analysis [3].

Other data confirmed the increase in the average lifespan time of patients with CF in recent decades.

Based on data collected from 10 countries in the years 1980 to 1994, a median age of patients who died in 1974 was eight years and 21 years in 1994. These results vary between countries and have been shown to largely depend on the CF patient care system [4].

It is estimated that in 2025, in comparison with 2010, the number of patients over 18 years in Poland will increase by approx. 15.4%. Rating for our country is difficult due to incomplete epidemiological data [5].

In Denmark, where 75% of CF patients are managed at a single CF specialist centre, around 80% of patients are predicted to survive until 50 years of age due to aggressive treatment they receive [6].

Apart from new treatment options, there are two other factors that play a pivotal role in increasing the life expectancy of CF patients.

Transitional care is one of the basic issues, with a focus on patient preparation for the transition from paediatric to adult care (and its acceptance by the patient).

There can be also certain organisational problems involved in adult CF patient management.

The challenges of chronic care for patients during adolescence

For many years now, comprehensive paediatric care for CF patients has been well-established and accessible at several CF centres within paediatric hospitals all over Poland.

Just like in any other chronic condition, health-related quality of life in cystic fibrosis is of particular significance in early adulthood when the simultaneous developmental and psychosocial transitions from childhood to adulthood are taking place. Patients continue university education or enter the labour market, establish new relationships, and to address their new needs, they are transferred to a new healthcare facility dedicated specifically to adults patients. Unless it is managed in a skilful and professional manner, the transition from paediatric to adult care can add extra stress to patients, or increase the risk of patient mismanagement, and therefore may have some adverse consequences for patient health [7–9]. The transitional care is defined as a pre-planned, comprehensive, and active healthcare process offering young patients with chronic illness the chance to develop their healthcare-related skills and knowledge during the difficult period of adolescence. As such, patient care in the transitional period should be well adapted to psychosocial, educational and professional needs of adolescents [8–12].

Although a number of healthcare centres where CF patients are managed have the necessary experience in transitional care, or even dedicated transitional care programmes, what is missing is a uniform approach to patient support in adolescence and to teaching patients the skills of self-care [13]. The few studies that have been published have shown a number of shortcomings in the transition from paediatric to adult care that can negatively affect the experience of adolescent patients in this critical period of life [7–10, 12].

The basic assumptions of transitional care programmes for CF adolescents are basically consistent with other programmes designed for chronically ill patients. These are in particular:

- adapting the patient care system in an age -appropriate way
- supporting the development of communication and decision-making skills to boost self-assurance and assertiveness of patients
- developing the sense of control and independence, both in the healthcare system and in personal life
- improving the life opportunities and the quality of life [11].

Health-related effects on the quality of life during adolescence

According to paediatric care, adolescence is the period of transition from childhood to adulthood taking place between 12 and 19 years of age. Young people between 16 and 19 years of age develop their own unique identity. During that time, a chronic condition can have a significant impact on the physical, emotional and mental development of a young individual. Therapeu-

tic management can be negatively affected by behaviours that can be typical of adolescents (sluggishness, forgetfulness, absent-mindedness, non-adherence, nonconformist approach). The priorities of adolescents and their parents or carers start to diverge. Adolescents typically strive for independence, and do not perceive the warnings of complications, permanent impairment, or even death from non-adherence as any imminent threats. Therapeutic problems may arise, which makes it even more difficult to make young patients aware of the importance of regular treatment [14]. The discipline and sacrifices of life of patients suffering from a chronic condition collide with the challenges of adolescence. In the first years of adolescence, when young people are preoccupied with the physical development of their body, some CF patients may be seriously concerned over their delayed growth and pubertal development. Their personal independence can be limited by frequent pulmonary exacerbations, and the resulting intensive treatment and hospitalization, which may accentuate the feeling of being different from their peers. Young patients with CF find it difficult to plan for the future due to frequent cases of infertility, genetic implications of CF, and shorter life expectancy [11].

Certain practical questions arise in the face of the challenges of the transitional period. There are issues such as when and how to prepare patient for the transition from paediatric to adult care, what is the best time of transition, and what is the role of a paediatrician, internists, and other healthcare professionals involved in the transition process.

Preparation of patients for the transition from paediatric to adult care

The patient transition should start as early as possible, preferably at the age of 11/12 years. The transition process should be supervised by a multidisciplinary team of healthcare professionals: physicians, nurses, physiotherapists, social assistants, and occupational therapists. The process should also involve the families of patients, but only upon prior patient's consent. The transition process should preferably take place in the period of remission. Adolescent patients should be comprehensively informed of the nature of the disease, its treatment and prognosis, as well as therapy-related benefits and risks. Plans for the future should be discussed with the patient, taking into account the patient's interests. Patients should be also made aware that they should become self-dependent. Another important step is to provide patients with psychological support and convince them to visit their doctor regularly [8, 9, 11, 14]. An example of progressive development of patient's independence in self-care is provided in Table 1.

The preferable time of transition from paediatric to adult care is 16 to 18 years of age. In any case, account must be taken of the patient's personality and health status, especially in terms of physical, psychosexual, social and professional maturity [14, 15]. In the conditions of healthcare and education systems in Poland, it must be borne in mind that the preferred age of transition coincides with intensive school education, and the high level of anxiety related to the Matura examinations examinations, as well professional or educational choices. 19-year old patients typically change school and move home, and become more self-dependent. Thus, 19 years of age appears to be the best time for transition, preferably in the period of remission (Table 1).

Recommendations for transitional care

Recommendations for patient care in adolescence can facilitate transition to adult care [11, 13, 15]. Mutual trust and respect in the patient-physician relationship is one of the cornerstones of proper patient care. Many teenagers perceive the qualities and personality of a physician as a key determinant of transitional care. It is difficult to win the trust of adolescent patients if they suspect some information can be shared with their parents instead of being discussed with them alone. Notwithstanding the different behaviours which can be expected of adolescent patients, any type of prejudice towards patients should be avoided. Patient care should be continuous and coherent. Young patients need around 4 or 5 visits to make sure that a doctor can be trusted. Visits should be arranged in absence of parents, and the scheduled appointment time should be longer than the time typically devoted to adults and paediatric patients. Also, the relevant health problems, such as addictions, sexuality, emotional and psychological instability should be taken seriously, which is another precondition for establishing mutual trust in the patient-physician relationship. Adolescent patients should be preferably made personally responsible for monitoring their health condition, and should be made prepared to take medications and manage rehabilitation in an unsupervised manner. In essence, young patients should be made aware of the importance of regularity and discipline in managing their condition [14]. Also, they should

Table 1. Self-esteem scale of CF patients [adapted from Town 2011]

Disease management	Level 1 always dependent 10–12 yrs	Level 2 often dependent 12–14 yrs	Level 3 sometimes dependent 14–16 yrs	Level 4 always dependent > 16 years
Physiotherapy and practice: — Physiotherapy — PEP mask — Flutter	Involvement in assisted / supervised treatment Parents in charge of organising routine exercise and chest physiotherapy School ± sport	Initiating and developing an effective technique through incentives Introducing routine practice accompanied by incentives/support from parents School ± sport; personal interests	Effective physiotherapy techniques with minimum supervision or assistance Supporting independence from parents, e.g. sport of physical exercise	Independence and responsibility for physiotherapy or seeking assistance for techniques that need third-person involvement Regular physical activity/sport
Nutritional management/enzyme supplementation	Understanding: — why they need enzyme supplementation — how enzymes work — why they need extra salt — basic rules of nutrition, the difference between fats, hydrocarbons and proteins	Enzyme supplementation with minimum supervision, also at school Being familiar with: — supervised enzyme dose adjustment to dietary intake — demand for extra calorific intake — demand for high fat foods	Independence in: — using enzyme supplementation, enzyme dose adjustment to dietary intake — replenishing salt losses Understanding nutritional needs Initiating the selection of high fat foods to increase calorific intake	Independence in enzyme and salt supplementation Actively discussing nutritional needs and ways to satisfy them Being familiar with the issues of nutrition in cystic fibrosis
Medication	— Being familiar with the drugs used — Maximum parental supervision	— Being familiar with the drugs used and how to use them — Being familiar with the side effects — Moderate parental supervision	— Being responsible for taking the medicines, unsupervised or with minimum supervision	— Unsupervised drug taking — Being responsible for getting the drugs — Being familiar with the drug therapy principles
Disease management	— General knowledge of the disease and disease management	— Asking questions — Being familiar with genetic aspects	— Being aware of circumstances where medical assistance needs to be sought — Being familiar with the puberty issues	— Being aware of how to seek medical assistance — Discussing the relevant problems

understand that effective disease management crucially depends on their active involvement in therapy.

HEADSS for Adolescents [16] is an interview instrument which can be used in daily clinical practice and can be adapted accordingly to examine and discuss adolescents' family relationships, family support and household chores (H — home), education, work and expectations for the future (E — education), rehabilitation (E — exercise), personal interests (A — activities), friendship and aspirations (A — affect, ambitions), dental care, drugs, diet, body weight control (D — dental care, diet, drugs), sexuality (S — sex), and sleep patterns (S — sleep).

Transfer of medical records

Patient transition from paediatric to adult care is essentially based on the transfer of medical records. Patient records, the so-called Cystic Fibrosis Passport, should include full patient history: type of disease, date of diagnosis, examination results, disease progress, complications, therapies, medical procedures, rehabilitation recommendations, and the current health status, together with the latest medical check-ups (diagnostic imaging) and a treatment plan. The transition procedure should be harmonized and unified [8, 9]. Paediatricians can be expected to prepare adolescent patients and their closest relatives for the transition and self-care, and should inform internists in advance (6–12 months) of the planned patient transition, which should be scheduled to take place, to the extent possible, during the period of CF remission. It should be accompanied by the transfer of medical records (CT Passport), establishment of a treatment plan, and provision of contact details (phone number, e-mail address of the paediatrician). At best, physicians / patient management team should liaise closely to review the case [15]. Also, patients should not be excessively and repeatedly investigated during the transitional period. On patient transition, the internist should focus on continuity of care, and should progressively prepare the patient for independence and self-reliance in the disease management.

The challenges of adult patient care in cystic fibrosis

At present, adult patient care is in the majority of cases continued at paediatric units (the paediatric care centre in Rabka, Poland, has around 166 adult patients, in Gdańsk — 48, in Karpacz — 24).

Adult CF patients can be provided with continuous treatment at only a few pulmonology units for adults: 85 patients in Poznań — at a teaching hospital, 100 patients at the Institute of Tuberculosis and Lung Diseases in Warsaw, and a small group of patients in Białystok and Zabrze, along with single cases managed at pulmonology wards scattered all over Poland.

Apart from the continuity of outpatient care, a major difficulty arises with respect to hospitalization of patients with exacerbation of the chronic obstructive bronchopulmonary disease. The two biggest pulmonology units for adults (Poznań and Warsaw) are unable to meet the demands and needs of such patients for reasons of: 1) limited number of beds, 2) contractual limitations, 3) and lack of inpatient isolation rooms for adults with CF.

It is worth noting, however, that no distinction is made between specific diagnoses under the National Health Fund (NFZ, *Narodowy Fundusz Zdrowia*) contracts with hospitals, and the treatment of CF exacerbations (lasting more than 2 days) is awarded 188 points (E84 is the underlying diagnosis!). In terms of patient isolation, this precaution is intended to protect CF patients against infectious agents transmitted by other patients (patients with infectious tuberculosis) as well as vulnerable (immunosuppressed) patients against pathogens transmitted by CF patients. The current epidemiological standards can be assumed to provide proper conditions for safe hospitalization of CF patients in any inpatient pulmonary unit. Another benefit of inpatient isolation rooms is that patients with CF are offered better comfort of care (a single room with private en suite toilet and bathroom).

To provide for optimum adult CF patient care in the years to come, it would be necessary to establish hospitalization options and comprehensive patient care at several pulmonary centres for adult patients all over Poland. Healthcare centres in Lublin, Zabrze, Białystok, Gdańsk, and Radom have been interested in developing this type of care. In the coming months, the major challenge would be to encourage physicians and other healthcare professionals, along with all decision-makers, to express their willingness and invite the existing healthcare centres to set up satellite CF units. The new CF units should be established as an integral part of the existing pulmonary centres and should operate, for example, as semi-independent entities within hospitals for patients with pulmonary insufficiency suffering from other underlying pulmonary diseases,

although other organizational challenges will definitely arise in the long run. As evidenced from the organizational experience of other countries with well-established CF patient care system, an optimum solution is where specialist patient care is managed by a multidisciplinary team of experienced healthcare professionals at CF centres [17]. Specialist care in dedicated CF centres is associated with improved survival and quality of life [18].

This strategy has been based on the *Standards of care for patients with cystic fibrosis: an European consensus* [19, 20]. A CF centre should normally care for a minimum of 50 patients. There are ready-made organizational solutions and requirements for both the staff (multidisciplinary teams) and healthcare facilities already in place.

Apart from inpatient isolation rooms for adults with CF, CF centres dedicated to adult patients should offer:

— an inpatient ward and a day-care ward,
— outpatient care facilities,
— an intensive care unit (basic condition),
— a radiology department (X-ray, CT),
— laboratory and microbiological facilities,
— expertise in bronchoscopy and other endoscopic techniques,
— expertise in nutrition support techniques,
— expertise in endovascular techniques (obliteration of bronchial arteries) in at least 2 selected centres (currently available in Lublin and Poznań)
— full diagnostic capability, including functional tests of the respiratory system,
— expertise in implantation and maintenance of vascular access ports,
— physiotherapy facilities,
— expertise in home oxygen therapy and mechanical respiratory assistance facilities,
— patient preparation for lung transplant and post—transplant care.

As a rule, inpatient pulmonary wards in Poland meet the conditions listed. Apart from standard pulmonary care (over 90% of respiratory insufficiencies can be life-limiting conditions), pulmonary clinics should also offer consultations in ENT, gastroenterology, nutrition, diabetology, gynaecology and obstetrics, and andrology (adapted to the individual patient's needs).

Effective patient care can be only provided by healthcare facilities having appropriate space and equipment, as well as trained and dedicated physiotherapists, nurses, dieticians, psychologists, other professionals offering spiritual and social support, as well as office staff. Pharmacists must be also involved in the establishment of new CF centres.

To operate successfully, CF centres must obviously employ physicians who are fully committed to CF patient care, but must be also supported by the hospital's executive staff, who must be fully aware of the challenges and capable of providing support. On the other hand, decision-makers who work to set up a CF centre should be conscious of the actual financial context of the project, and should be co-responsible for its financial condition.

CF centres should provide inpatient care, outpatient care and the combined inpatient and home care, as well as home care options. Antibiotic therapy options available in home care (following slight modifications in the currently proposed model) can offer patients the chance to stay at home in the exacerbation period, unless hospital care is essentially required, which could take the burden off hospital.

Pulmonologists who have never dealt with adult patients with CF may find it difficult to understand the level of difficulty associated with patient transition from paediatric to adult care. They simply fail to fully comprehend the organizational and psychological aspects of patient care in a chronic disease, requiring full engagement of patient and family, often involving close relationships with paediatricians and other healthcare professionals, built on shared experienced and the special (and well-deserved) privileged position of chronically ill patients.

Moreover, it cannot be denied that some pulmonologist may fear that the problems of CF patients, most notably the organizational challenges and the financial burden of CF therapy, may dominate their working environment where the needs of other groups of patients cannot be left unanswered.

It needs to be clearly stated that there is no chance of an effective and comprehensive care for adolescents with cystic fibrosis, while maintaining the relations which typically dominate at adult wards, where we deal with patients with mature and established personality. A CF centre cannot be successfully operated unless a sensitive approach to CF patients is adopted, and full dedication is always given to both patients and their families. This approach needs to be adopted not only by physicians who often feel overwhelmed by the challenges of managing such complicated and time-consuming cases (and can feel intimidated by the extent to which patients and their families are familiar with this multidimensional

condition), as well as all healthcare professionals engaged in comprehensive medical care.

It is hardly conceivable to abandon efforts to provide CF patients with optimum care or to offer them special preferences in the face of the challenges they have before them, although their life is often very short.

The challenge ahead — which is currently one of the basic tasks of pulmonologists managing adult patients in Poland, and one of the most challenging healthcare issues in patient care – will involve organizational zeal, a multitude of concerted initiatives, and institutional support (for example within the framework of the national CF patient care programme), taken independently or as part of the national pulmonology programme. The initiative to create and accept projects related to CT patient care can be supported by national and regional consultants, scientific societies, patients' associations, as well as non-governmental organisations.

Conflict of interest

The authors declare no conflict of interest.

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