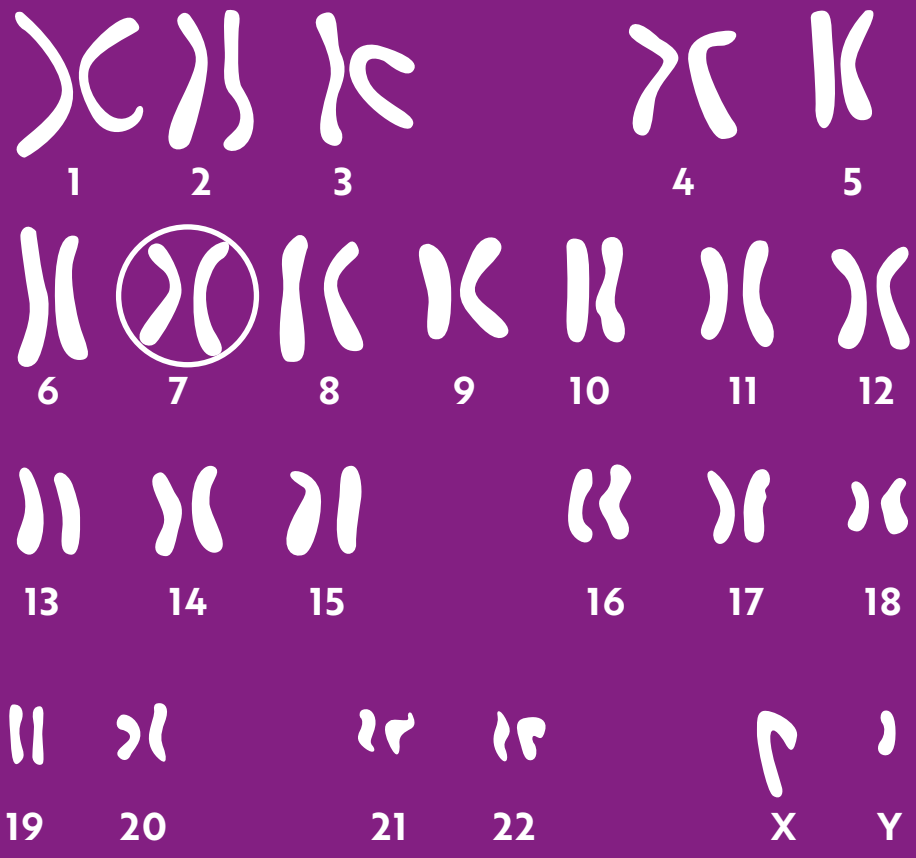

CYSTIC FIBROSIS

a multisystem disease

Editor
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PREFACE

Dear Readers,

We were very pleased with the positive response to the first edition of our monograph, and that it sold out very quickly. We were delighted to learn that the book is being used both by physicians dealing with cystic fibrosis on a daily basis and those preparing for their specialty examinations. It is also being used by dieticians, physiotherapists, and psychologists involved in the treatment of patients as members of multidisciplinary teams.

The rapid development of new CFTR variant-specific therapies in cystic fibrosis has encouraged us to work swiftly on the second edition, which is supplemented with the latest developments in biotechnology. Responding to the interest in the book among specialists outside the field of pulmonology, we have significantly expanded the sections on gastroenterology, radiology, and physiotherapy. With hospital management staff in mind, we have prepared a practical chapter on how to organize a cystic fibrosis center in accordance with current European standards.

This second edition of the monograph, available as an e-book, has allowed us to overcome the size constraints of a printed book and also to modernize the chapter presenting the results of diagnostic imaging.

We wish you enjoyable reading!

Prof. Dorota Sands
Scientific Editor

PART I

EPIDEMIOLOGY AND PATHOPHYSIOLOGY OF CYSTIC FIBROSIS

CHAPTER 1

EPIDEMIOLOGY AND ORGANIZATION OF MEDICAL CARE IN POLAND ACCORDING TO EUROPEAN STANDARDS

Dorota Sands, Andrzej Pogorzelski, Anna Skoczylas-Ligocka, Łukasz Woźniacki

1.1. Introduction

Cystic fibrosis (CF) is a multi-system disease clinically characterized by chronic bronchial obstruction and respiratory infections, as well as digestive disorders and their consequences. In the Caucasian population, it is one of the most common genetically determined diseases, inherited as a monogenic, autosomal recessive trait.

The usual reported prevalence for Caucasians, calculated in the 1980s, is 1 : 2500 live births, but due to the widespread use of screening programs around the world, which has allowed more accurate estimation of the prevalence, it is believed to be around 1 : 4000-5000. In Poland, the incidence of the disease calculated on the basis of data from the screening program (2006-2010) is 1 : 4394 in relation to live births.

CF registries are very helpful tools to help improve the quality of patient care (approximately 100,000 patients worldwide). They provide extremely valuable data on factors affecting the length and quality of life of patients. In the case of CF, these rates have improved significantly over the last 50 years. It used to be an exclusively pediatric disease, but now more than half of the patients in highly developed countries are adults. The Canadian Registry reported that the median survival in 2019 was 54.3 years.

The life expectancy of CF patients and the percentage of the adult population affected are **very sensitive indicators of the quality of the health care system**.

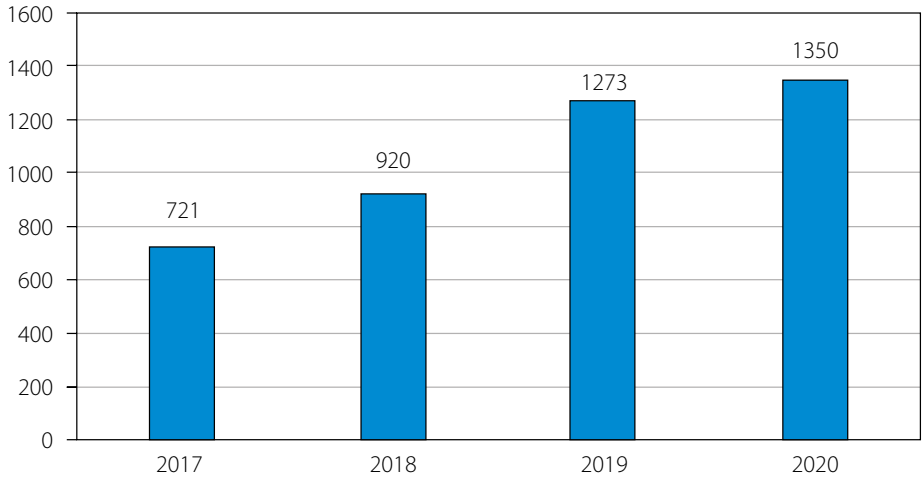
Unfortunately, Poland ranks unfavorably compared to other developed countries in these key statistics for CF (the ratio of adult patients in Poland is only about 33%, whereas in Canada it is 68%).

1.2. Population of patients with cystic fibrosis in Poland

So far, it has been estimated that the number of CF patients in Poland is about 2000, but after a thorough analysis, it transpired that this number is overestimated. In recent years, precise epidemiological data on the patient population have not been available because the register of these patients has not been actively maintained. The Polish Cystic Fibrosis Registry, in operation until the end of 2012, which did not include full data, contained the records of 1661 patients (including 1552 living patients), of whom 34.5% (535 people) were adults.

This situation changed when Poland joined the European Cystic Fibrosis Society Patient Registry (ECFSPR) in 2018. Since then, epidemiological information has been provided anonymously by centers dealing with the treatment of patients with CF once a year for the previous year (for the first time in 2018 for 2017). The most up-to-date data come from June 2023 and cover 2022. According to these data, in Poland there were 1663 CF patients, 66.7% of whom were children, and 38.2% were people over 18 years old. The gender distribution was even: 49.9% female, 50.1% male, and the median age of patients in the registry was 14.4 years. The oldest patient was 62 years old. In 2022, 10 patients died – the median age at death was 19.1 years, and the average was 16.8 years.

Currently, out of 13 CF centers, 7 care for adults, but not all patients (mainly adults) have been entered into the register. This is indicated by data from databases of beneficiaries of foundations and associations that support CF patients from all over Poland. According to the data of the Polish Society Against Cystic Fibrosis, at the end of November 2021 the database of beneficiaries included over 1700 people, 52% of whom were adults. In 2020, the society recorded 23 deaths of patients; the median age



Data presented by the national coordinator of the ECFSPR during the 20th Scientific and Training Conference of the Polish Society of Cystic Fibrosis entitled "Cystic fibrosis – new challenges", September 2-04-2021, Brzeg

FIGURE 1.1. The number of Polish patients in the register of the European Cystic Fibrosis Society in 2017-2020

at the time of death was 26 years. In a few years, it will be possible to assess the survival rate of Polish patients better; it should increase significantly with improved care and access to CFTR modulators and lung transplantation.

The authors predict that the number of Polish patients in the ECFSPR registry will increase in the coming years, but it should not exceed 1700 people (Figure 1.1).

1.3. Organization of care for patients with cystic fibrosis – model solutions

1.3.1. European standards of treatment of cystic fibrosis

CF is a chronic disease that requires constant supportive treatment throughout the life of the patient, as well as regular outpatient check-ups aimed at preventing the progression of lung disease, monitoring the nutritional status, and early diagnosis of complications. The standard applicable in highly developed countries is the care of CF patients organized in specialist centers and carried out by a specialized team, which seems to be an optimal solution. Providing such a way of caring for patients translates into a longer and higher quality of life.

The European Cystic Fibrosis Society standards published in 2014, describing the diagnosis, treatment, organization of the CF center, and quality control methods, is a consensus containing a description of model solutions for the care of patients with CF. The standards in their original version of 2004 enabled the implementation of a modern and effective model of care for CF patients in many European countries, allowing for a significant extension of their survival time, increasing the population of patients reaching adulthood, and improving the quality of life of patients.

In accordance with the European Standards for the Treatment of Cystic Fibrosis, the structure of a CF treatment center is based on a multidisciplinary team, cooperation with other medical and surgical specialists, available facilities, as well as equipment and software, which together will allow the multidisciplinary team to provide care at a level that meets the complex medical challenges of the disease using effective diagnostics and holistic treatment programs.

According to the standard, to maintain the appropriate level of knowledge and experience, a specialist center should have a minimum of 100 adult or pediatric CF patients. Occasionally, the geographical location of a CF specialist center or the low prevalence of the disease in some populations may mean that the number of patients at the center is also lower, but never less than 50. Centers with less than 100 patients should be connected to a larger center until there are an appropriate number of patients, and experience and resources sufficient to provide services independently.

CF center facilities must be suitable for each age group. There should be enough space for patients requiring an urgent follow-up visit in the outpatient clinic or overnight ward, or during their stay on the ward. The number of hospital beds should be sufficient to accommodate standard patients within 7 days and emergency patients within 24 hours.

Care for a CF patient requires regular follow-up as part of outpatient care or single-day hospitalizations. According to the European Cystic Fibrosis Treatment

Standards, it is the outpatient clinic, not the hospital ward, that is the basis for patient care, and patients should use it with a frequency adequate to their individual needs. People with a stable disease should come in for routine check-ups every 2-3 months, depending on the severity of the disease. Newly diagnosed newborns should have check-ups more frequently (initially weekly). All patients should be reviewed annually to ensure that a full medical, dietary, physiotherapy, and psychosocial review is performed at least once a year and that all follow-up examinations have been ordered.

Patient care should be the responsibility of a multidisciplinary team of qualified and experienced CF healthcare professionals. It should include a number of people commensurate with the size of the population served by the center and should include the following CF specialists and support staff:

- a pediatrician or internist specializing in respiratory diseases or a pulmonologist,
- clinical microbiologist,
- medical interns,
- a nurse with a clinical specialty,
- a physiotherapist,
- a dietitian,
- a clinical psychologist,
- a social worker,
- a pharmacist,
- a clinical geneticist,
- secretarial service,
- a database coordinator.

Prevention and control of respiratory tract infections play an extremely important role in the care of patients with CF. According to European standards, all CF centers

TABLE 1.1. Full-time work equivalent depending on the size of the clinic (pediatric patients treated by full-time employed HCPs)

Multidisciplinary team	50 patients	150 patients	≥ 250 patients
Consultant 1.	0.5	1	1
Consultant 2.	0.3	0.5	1
Consultant 3.	–	–	0.5
Medical interns	0.8	1.5	2
Specialist nurse	2	3	4
Physiotherapist	2	3	4
Dietician	0.5	1	1.5
Clinical psychologist	0.5	1	1.5
Social worker	0.5	1	1
Pharmacist	0.5	1	1
Secretary	0.5	1	2
Database coordinator	0.4	0.8	1

Source: European Cystic Fibrosis Society Standards of Care: Guidelines for Cystic Fibrosis Treatment Centres, 2015

must have a clear infection prevention and control policy, and facilities must allow for proper triage of patients to prevent cross-infection. While at the facility, patients should not share rooms, bathrooms, or toilets, and should not come into contact with each other in waiting areas, such as clinics, CF units, pharmacies, and radiology departments. Detailed recommendations for infection prevention and control were published by the US National Cystic Fibrosis Foundation in 2013.

1.3.2. Polish cystic fibrosis audit – where are we?

In May 2015, organizations operating in the field of CF conducted a survey among CF treatment centers aimed at verifying the housing conditions, employment of specialists, and access to services, as well as key problems that need to be solved. The results of the study were compiled in the study "Audit of Cystic Fibrosis Treatment Centers".

The analysis of information obtained from CF treatment centers revealed that the inability to provide comprehensive care, taking into account its multiple aspects, and the unsuitability of centers for adults are the main problems for most centers. Also, the lack of isolation facilities for patients at bacteriological risk was a key challenge and problem.

The infrastructure of most centers did not allow for providing CF patients with adequate isolation during treatment adequate to their needs. The survey showed that less than half of the Polish centers had developed and implemented rules for preventing cross-infections. Low availability of hospital beds in single rooms with separate sanitary facilities was also significant, which made it difficult (and in some centers even impossible) to isolate patients during hospitalization.

Another important problem was the fact that the outpatient clinic was not adapted to ensure the isolation of patients during outpatient care. In most centers, this was caused by housing problems – lack of separate entrances, narrow corridors, and an insufficient number of consultation and treatment rooms.

1.3.3. Number of hospital beds allocated to patients in cystic fibrosis centers

The results of the Polish CF audit also showed that almost all centers dealing with CF treatment did not have formally established multidisciplinary teams dedicated to working with CF patients. The availability of consultations with a dietitian, physiotherapist, or psychologist often resulted solely from the good will of hospital managers and the devotion of physicians, because there was no requirement to impose an obligation on centers to provide such care or financial solutions that would allow such teams to finance medical care as part of specialist outpatient care or hospital treatment.

There was a significant deficit of physiotherapists and dietitians providing consultations as part of outpatient care. Only single centers had access to a social worker.

The conclusion of the audit was that, unfortunately, at that time in Poland there were no medical facilities that could meet the definition of a CF treatment center in accordance with the European Standards for the Treatment of Cystic Fibrosis. The audit also showed a heterogeneous distribution of patients across

the centers and a significant shortage of adult-only facilities. The audit concluded that although the existing network of pediatric centers dealing with the treatment of CF allowed for sufficient access to specialist care for this group of patients, it was not possible to provide patients with treatment at the same high level.

1.3.4. Proposals of model solutions for the organization of care for patients with cystic fibrosis in Poland

The CF community has been striving for many years to adapt the treatment system for patients with CF in Poland to European standards. Over the past years, several centers have successfully completed renovations that have significantly improved the adherence to infection prevention and control recommendations for CF. However, investments in modern infrastructure are insufficient to improve the system of treating this disease in Poland. In Poland, the package of guaranteed health services lacks solutions that would allow for the financing of proper care for patients with CF, adjusted to the requirements of European standards, including inpatient, outpatient, and home care.

So far, in the Polish system, care for patients with CF has focused on the treatment of exacerbations of lung disease, which is reflected in the appropriate pricing of hospital treatment services. Unfortunately, the pricing of outpatient care, which is inadequate to the needs, has not yet allowed patients to be consulted by a multidisciplinary team. Moreover, it did not allow the costs of necessary tests, e.g. microbiological tests, to be covered. For CF patients, for formal and financial reasons, home care services in the form of physiotherapeutic care or home intravenous antibiotic therapy are practically unavailable.

To implement systemic changes in the treatment of patients with CF, since 2014, in discussions with the Ministry of Health, the CF community has postulated the implementation of a new solution in the form of a "product" separate from the package of guaranteed health services, under the name "**Comprehensive, coordinated care for patients with CF**".

As part of the new service, it was postulated that a system of coordinated care be created for patients suffering from CF provided by reference centers. The creation of two categories of centers dealing with patients with this condition was proposed – depending on the size of the population under treatment and the capacity of the center. For each level of reference, detailed requirements regarding infrastructure, personnel, and the scope of services provided, including outpatient and home care, were defined.

The most significant changes concerned outpatient care for patients with CF, which so far has not provided for a comprehensive, multidisciplinary approach.

In the proposal submitted as part of the new service in the field of outpatient treatment, it was agreed that basic examinations and consultations should be performed during a routine patient check-up to monitor the progress of lung disease and nutritional status, namely the following:

- physical examination,
- microbiological examination,
- spirometry test,
- medical consultation,

- consultation with a physiotherapist,
- dietitian consultation,
- psychologist consultation,
- therapeutic education.

Once a year, patients should undergo assessment, during which, in addition to routine examinations and consultations, tests are performed to diagnose CF complications and comorbidities. A CF treatment center should remain in formalized cooperation with centers of other specialties (gastroenterology, laryngology, diabetology, etc.), to which patients requiring consultation should be referred.

Establishing the health service "Comprehensive, coordinated care for patients with cystic fibrosis" would mean more formal requirements than before and increase of the organizational and financial burden for the centers; therefore, the key element determining the success of the implementation of such a solution would be, above all, its appropriate valuation. There is no doubt, however, that the introduction of such a model of care would make it possible to extend the survival time and improve the quality of life of patients with CF and the rational use of health care resources.

1.4. Conclusions

Although the process of extending life expectancy and improving the quality of patient care is observed, the treatment options for patients with CF in Poland still differ from the standard of many developed countries. However, the successes include reimbursement of CFTR modulators since 2022, and dynamically developing lung transplantation programs at the Silesian Center for Heart Diseases in Zabrze, at the Teaching Department of Thoracic Surgery and Transplantation of the Medical University of Szczecin in Szczecin-Zdunowo, and at the Teaching Department of Cardiac Surgery and Vascular Surgery of the Medical University of Gdansk, where over 100 lung transplantations have been performed in patients with CF.

Modern solutions have been introduced in Poland in recent years, such as universal neonatal screening, due to which, since 2009, early diagnosis has been possible, and thus starting treatment before the disease develops and irreversible changes occur. Patients in Poland have access to most drugs and foodstuffs for special dietary purposes used in the treatment and prophylaxis of CF; however, many of them are not reimbursed, which is associated with a high financial burden for the families of patients.

To a large extent, due to efforts and resources of the community of physicians and patients, some of the centers have recently carried out investments in renovation and reconstruction of infrastructure, which has significantly improved the conditions for patients and increased the possibilities of effective infection control. The launch of the Cystic Fibrosis Treatment Center in Dziekanow Lesny at the beginning of 2017 proved be a spectacular success. With its multidisciplinary treatment team, it is now a model CF treatment center in Poland.

It is necessary to create new centers for adult patients based on this model.

Currently, however, the most urgent need is for the introduction of systemic and financial solutions that will guarantee patients with CF access to comprehensive,

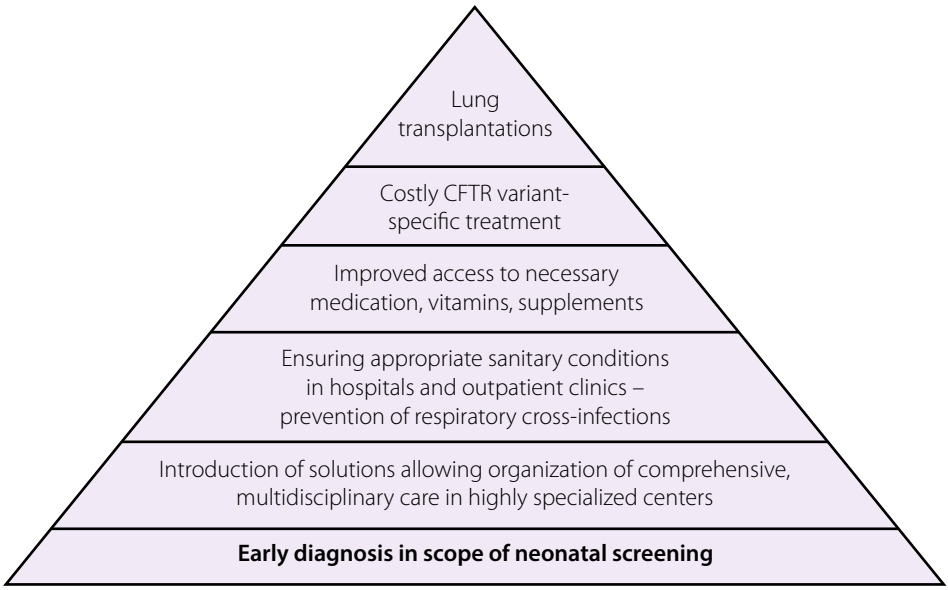


FIGURE 1.2. Pyramid of needs of cystic fibrosis patients in Poland

multidisciplinary medical care, compliant with European standards, provided in all centers specializing in the treatment of CF.

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Prof. Dorota Sands, MD, PhD, specializes in pediatrics, allergology, pulmonary disorders, and pediatric pulmonary diseases. She has authored over 100 scientific publications. During her extensive tenure at the Institute of Mother and Child in Warsaw, she has completed multiple internships at leading pulmonology centers, including the Royal Brompton Hospital in London and the Division of Pediatric Pulmonology and Allergology at the Medical University of Graz, where she held a scholarship from the American Austrian Foundation. Prof. Sands pursued her postdoctoral research during a fellowship at the University of Leuven in Belgium. She shares the knowledge gained at the pioneering cystic fibrosis treatment center at the Institute of Mother and Child in Warsaw with members of her multidisciplinary team. This team, recognized across Poland, is widely acknowledged internationally for its expertise. Since January 2017, Prof. Sands has been affiliated with the Children's Hospital in Dziekanów Leśny. In this capacity, she serves as the head of the Cystic Fibrosis Center (CLM). As a researcher at the Institute of Mother and Child in Warsaw, she also heads the Cystic Fibrosis Department. Being a renowned international expert with extensive experience in conducting clinical trials, Prof. Sands has contributed to the development of European cystic fibrosis standards of care and the organization of cystic fibrosis centers. These standards are successfully implemented at the Dziekanów Leśny Cystic Fibrosis Center, which serves as a model center for the treatment of this condition in Poland.

I am dedicating this book to
Anna Nowakowska, MD, PhD,
my invaluable mentor who, in addition to teaching me clinical skills, instilled in me a deep respect for fellow human beings.

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