Challenges in care of adult CF patients – the specialist cystic fibrosis team

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Abstract

Cystic fibrosis (CF) is the most common life-limiting, autosomal, recessive genetic disorder. The gene which is responsible for the symptoms of this disease is located on the long arm of chromosome 7 and encodes the protein called Cystic Fibrosis Transmembrane Conductance Regulator (CFTR), an apical chloride channel in epithelial cells. CF is a "multi-system" disease. It affects many parts of the body and it has a varied clinical expression. All patients with CF should have access to specialist services and the treatment must be comprehensive and multidisciplinary. The multidisciplinary team approach is important when trying to optimize care given to the patient and their family. The cystic fibrosis team may include personnel from the following specialist areas: medical, nursing, physiotherapy, dietetics, psychological, social/supportive. Close coordination is vital. Ideally, 'all members of the team' should have had CF care-related training. The specialist team approach ensures that such specialized multidisciplinary expertise is applied in all aspects of care, better knowledge of individual families, continuity, knowledge of treatment advances and the ability to apply these in daily management. Doubtlessly CF team ensures families a specific point of contact and they know who to talk to.

In 1997 The Department of Pulmonology Diseases in Poznań started running a programme for CF adults at our University – the second CF adult centre in Poland. Members of our CF team have experience in the management of adults who have cystic fibrosis. Its members include physicians, nurses, a physiotherpist, a dietitian, a social worker and a psychologist. We must to build the team approach in CF care and use effectively talents of multiprofessional team members as fully as possible to deliver better services to patients.

Key words: cystic fibrosis, CF team, multidisciplinary approach, adult CF patients.

Cystic fibrosis (CF) is the most common life-limiting, autosomal, recessive genetic disorder in Caucasian populations, particularly those of northern European origin.[1]. The gene which is responsible for the symptoms of this disease is located on the long arm of chromosome 7 and encodes the protein called Cystic Fibrosis Transmembrane Conductance Regulator (CFTR), an apical chloride channel in epithelial cells. Nowadays there are identified over 1000 mutations of the CFTR gene. The mutation of the gene mentioned above causes the pathology of chloride, sodium, potassium and water conductance in and out of cells which changes the composition of secretion in many organs. Normally, these secretions are thin and slippery, but in CF, the defective gene causes secretions to become thick and sticky. Instead of acting as a lubricant, secretions plug up tubes, ducts and passageways, especially in the pancreas and lungs.

CF is a "multi-system" disease which affects many parts of the body and it has a varied clinical expression. In a classic (full-symptoms) presentation it affects the respiratory system (e.g. recurrent and chronic sinusitis, nasal polyposis, chronic bronchopulmonary disease, bronchiolitis, bronchitis, bronchiectasis), the gastrointestinal system (pancreatic insufficiency, recurrent pancreatitis, meconium ileus, the distal intestinal obstruction syndrome (DIOS), rectal prolapse, intussusception, multilobular, biliary liver cirrhosis, cholelithiasis, portal hypertension, oesophageal varices, splenomegaly) the endocrine

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system (diabetes mellitus, delayed pubertal development), the reproductive system (azoospermia caused by congenital bilateral absence of the vas deferens, too thick cervical mucus), the urinary system (incontinence of urine), sweat glands (elevated sweat chloride concentration >60 mEq/l), and many others. Although most of the patients with CF suffer from multi-organ disorders, a respiratory failure is the most common cause of morbidity and mortality.

The progress in therapy and care of people with CF, in the last 50 years, have transformed the prognosis for this group of patients by increasing the median age of survival from less than 1 year to slightly more than 40 years of age [2]. All patients with CF should have access to specialist services, and the treatment must be comprehensive and multidisciplinary. The multidisciplinary team approach is important when trying to optimize care given to the patient and their family as cystic fibrosis is a complex multi-system disorder requiring diverse understanding and knowledge. The advances of the latter are rapid and so treatment protocols change regularly. Moreover, patients and families have a high level of knowledge and growing expectations of their management [3]. The exact staffing levels vary from unit to unit but all of the team members should be experienced in working with cystic fibrosis patients. Based on the strong association between the establishment of comprehensive CF Care Centers and improved patient outcomes, The Cystic Fibrosis Foundation Committee strongly recommends the multidisciplinary approach modeled on the highly successful pediatric CF care system.

The primary objectives of the adult health care team are to: ensure optimum care; facilitate access to pertinent medical resources; coordinate care among specialists and primary care practitioners; support quality of life and independence for each patient. A frequent patient contact with the Center is necessary to accomplish these objectives. In general, quarterly visits are sufficient, although some patients with special needs or an advanced disease may require more frequent attention. The Adult CF Care Team may function in a primary care capacity or in concert with an independent primary care practitioner. Coordination and communication with other medical professionals involved in the patient's care are essential [4].

The cystic fibrosis team may include personnel from the following specialist areas:

1. Medical – primary care physicians, pediatricians, adult physicians (respiratory and gastroenterology), surgeons (ENT, gastroenterology and transplant), gynecologists and obstetricians.

2. Nursing – hospital staff, community nurses, clinical nurse specialists. The important nursing issues include supporting the family and patient from the moment of diagnosis to the terminal stage of the disease; counseling and educational needs of the family and patient, examining the implications of patient's nursing needs during admissions to hospital, outpatient visits and in the community. Caring for the patient with CF can be challenging and gratifying for the nurse. As part of the health care team nurses can do a considerably much to help patients and their parents to overcome their numerous problems [5].

3. Physiotherapy – a specialist respiratory physiotherapist. Physiotherapy is recognized as integral part of the management of patients with CF and is one of the aspects of treatment that contributes to improvement in quality of life. Assessement and reassessement of patients are essential for effective management. Hence each patient should be reviewed by a physiotherapist with an interest in cystic fibrosis at 2-3 monthly intervals, and more often during the first year of diagnosis. Physiotherapy programs must be discussed with and accepted by patients. They also should be realistic to optimaze adherence and obtain a balance between the sufficient treatment and quality of life [6].

4. Dietetics – a specialist dietician. The importance of the nutritional status in the long-term survival of patients with CF is well-documented. Potential nutritional problems are multifactorial, and include maldigestion with subsequent malabsorption, increased requirements and poor dietary intake result in malnutrition. There is a strong indication that improvement in nutritional status leads to an improvement in the prognosis for these patients. The prevention of malnutrition should be a primary goal of the health care team. A regular contact with a dietitian, who can offer simple practical dietary advice will help to prevent a decline in the nutritional status, and may have the added advantage of improving the quality of life and a prognosis for some patients. In addition, supervised nutritional support, either oral, enteral or parenteral, might be actively offered to patients with a severe lung disease, who are struggling to achieve their individual dietary requirements [7].

5. Psychological – child guidance/adult support teams, psychiatrists. Inevitably, the burden of living with any chronic life-limiting disease creates immense stress for the individual and their family. Physical health and psychological health are naturally interdependent and psychological concerns should never be neglected. Some difficulties may require the specialist attention of a psychologist or psychitrist. The importance of establishing a good relationship with the family from the outset cannot be underestimated and can prove valuable when attempting to help at a particular time of the need [8].

6. Social/supportive – social workers, chaplains/religious support, family and friends, administrative staff. The social work brief will inevitably be dictated by prevailing social attitudes, social and economic policies and the legislstive framework that governs health and social care. Supporting people and their carers to live independently in the community makes economic sense and usually provides better opportunities for people to lead more fulfilling lives. The social worker seeks to consider the individual and his carers in order to support their choices, and to act as an advocate for them. Within a specialized clinical framework the social worker is in a unique position to develop an understanding of the needs of people with cystic fibrosis and to convey these to social services' departments and other resource providers in the community [9].

Ideally, 'all members of the team' should have specific training in CF care. Back-up personnel should be available in the event that a team member is unable to perform his or her duties. The pulmonary and GI/nutritional manifestations of the disease predominate in adults as well as in children, but several other issues also emerge. The optimal management of CF requires input from all members of the health care team. Evaluation and intervention by team members should be individualized to suit each patient's conditions. However, the minimum of one comprehensive evaluation per year by each team member is recommended. These evaluations should encompass an assessment of adherence to therapies and the identification of relevant psychosocial issues as well as specific medical issues [20], such as: reviewing the past year's events, examining all aspects of the patient's needs, screening for complications, developing treatment plans for the coming year, ensuring continued patient education. Ideally, the center should organize a case management conference or other in place of it in order to provide for a periodic review of the status of each patient and the formulation of a treatment plan. These assessments should be documented in the medical record and passed on to other health care professionals involved in the care of the patient because close coordination is vital. Many teams are very devolved, but others work better with clear leadership. Individual roles frequently overlap and good communication avoids duplication. The specialist team approach ensures that: specialized multidisciplinary expertise is adopted in all aspects of care; better knowledge of individual families; continuity – especially when team members change; families have a specific point of contact and know who to talk to; knowledge of treatment advances and the ability to apply these in daily management [3].

Regular team meetings are very helpful. It is easier to plan ongoing care for individual patients, assess the impact and effectiveness of current treatment and update on psycho-social issues affecting the patient and their family. When members of CF team meet they may discuss research, new treatment protocols and share their experiences.

Whilst team meetings would normally only include the specialist team, consideration must be given to involving a wider range of team members such as those in related specializations, the community, and other units. It often takes a considerable effort to keep lines of communication open, but such measures can prove invaluable.

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People who receive their care at the accredited CF Center such as The Department of Pulmonology Diseases in Poznań, benefit from access to cutting edge research and development, clinical trials, and a health care team that is dedicated to caring for CF adult patients. It is also essential to distinguish CF from conditions with similar presentations, such as asthma, chronic bronchiectasis, dyskinetic (immotile) cilia, pancreatitis, alfa 1-antitrypsin deficiency, male infertility, or malabsorption. CF diagnostic evaluation may clarify the clinical picture.

Members of our CF team have experience in the management of adults with cystic fibrosis. Adults, generally, have a more severe pulmonary disease, a higher prevalence of DM, and face more complex financial and psychosocial problems. Therefore, a relatively higher intensity of respiratory therapy, and endocrine, nutritional, and psychosocial services may be needed. In addition, adult patients have unique needs, including vocational counseling, contraceptive and reproductive services, and obstetric care. There is a consensus that a multidisciplinary team with training and experience in adult CF care should oversee the care of adults with CF. Our multidisciplinary team consists of physicians with internal medicine training and additional expertise in CF, respiratory physicians, nurses, a dietitian, a physiotherpist, a psychologist and a social worker. Care is provided in inpatient and outpatient facilities.

According to recommendations each patient is seen at our CF Center at least four times a year. Complete nutrition and social work assessment, as well as a full pulmonary function study and chest x-ray is performed annually. Annual laboratory testing of blood and sputum is also part of comprehensive CF care.

Many people are involved in the management of the patient with CF. The patient, parents and relatives must adhere to the actual treatment which has been prescribed by the CF team at hospital. Their understanding of treatment protocols is absolutly essential if the patient and those at home are to be motivated to comply with the treatment advised. The better the patient and relatives understand why a particular treatment is needed, the more efficiently it will be carried out. We aim at teaching the patient and his family as much as possible about the medical aspects of CF and welcome any questions on the "whys and wherefores" of our advice, or on any aspects of the disease.

Within the years of existance of our CF centre we have faced many problems, often caused by the unstable health care system and administrative and financial restrictions. Although there are many areas of treatment where we are reasonably certain that our advice is correct, there are many other areas where there is no proof of efficacy. Last years brought some promising changes, which influenced the functioning of CF centres in Poland. We worked out close cooperation with other departments at University (gynecology, diabetology, anasthesiology, genetics, otorhinolaryngology and palliative care) expanding significantly the number of people interested in CF. We should improve our cooperation with such importent specialists as gastroenterologists, endocrinologists and surgeons. Another very important problem for our centre is difficult access to a lung transplant centre and some of our patients have indications for heart-lung or double-lung transplantation and express their interest in this procedure. The further growth of the centre dependes greatly on the financial security of the health system. Lack of money can be, however, greatly compensated by the devotion and growing expertise of people involved in CF care.

We must develop the team approach in CF care using the talents of a multiprofessional team as fully as possible in order to provide patients with better services which is a primary challenge for us.

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