



# Human genetics



Last update: 18 Jun 2002

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Document reference: WHO/ICF(M)A/IACFA/99.6, Distr.: General, English Only

## Services for adults with cystic fibrosis

### **Report of a joint WHO/ICF(M)A/IACFA meeting**

*The Hague, The Netherlands, 7-8 June 1999*

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## **EXECUTIVE SUMMARY**

**Cystic Fibrosis (CF) is the most common life-threatening single gene inherited disorder in people of European descent worldwide. Increasingly, cases are identified in other Caucasians in the Middle East and on the Indian subcontinent.**

**Until recently, CF has been regarded as a children's disease because few patients survived into adult life. However, better prevention and management of its complications, particularly chronic lung disease, has led to longer survival. Thus, in some industrialised countries, there are as many affected adults as CF children; and the proportion of CF children surviving to become adults is rapidly increasing throughout the world.**

**Provision of services for CF adults has not kept pace with this improved prognosis. Many adult pulmonologists have neither the necessary training to manage this complex disease, nor, individually, do they have enough patients to encourage them to develop a special interest in CF and ensure provision of the range of services patients need. Some CF-related conditions are specific to affected adults (e.g. fertility problems) while others (e.g. diabetes) may affect children but become more frequent and severe as the disease progresses.**

**WHO, the International CF (Mucoviscidosis) Association ICF(M)A and the (patients') International Adult CF Association (IACFA) therefore convened a joint meeting (The Hague, 7-8 June 1999) to produce recommendations for the provision of adult medical and associated services for CF adults. The group did not consider or make detailed clinical recommendations; because these recommendations would soon become dated and can be found in specialist textbooks and journals. Instead, its scope was limited to service provision. This report provides a reference for health care providers by outlining the range and types of specialist services required by CF adults, in the hope that the momentum of the improving prognosis can be maintained or accelerated. It includes a checklist of adult medical, psychological and social problems associated with CF, the composition of the specialist team, physical accommodation and facilities, recommended arrangements for transitional, community and terminal care, training of CF specialists and funding arrangements. It reflects the experience of the professionals and patients who comprised the working party.**

## **1. Background**

Previous joint meetings between World Health Organization (WHO) Human Genetics Programme and the International Cystic Fibrosis (Mucoviscidosis) Association (ICF(M)A) have addressed the need for education concerning diagnosis and management of cystic fibrosis (CF) [1-3].

Cystic fibrosis is a genetic disease with significant variations in incidence, morbidity and mortality around the world. Although most often reported in white Caucasians, since discovery of the mutated gene in 1989 [4] it is also being diagnosed with increasing frequency in other populations, often with variant clinical expression. Better understanding and treatment of CF has led to an improved survival in countries where CF services are well developed [5-8]. Published data from these countries report a similar mean life expectancy of greater than thirty years, compared with survival less than a decade 30 years ago [9]. Today many CF patients reach adult life with the consequent need for specialist care different from pediatrics. For example in the United Kingdom it was calculated that at the turn of the century there would be as many adults as children with CF [4].

The major cause of morbidity and mortality is chronic respiratory disease, but CF is a

complicated disease affecting multiple organs which therefore requires the involvement of a variety of medical and other specialists. Optimal care for CF consequently needs a team approach which can only be coordinated in specialist centres. Such centres have developed in many countries and the benefits of centre care have been clearly demonstrated [8], although differences in survival between individual centres have also been observed [10]. Because CF has historically been regarded as a children's disease, CF centres have grown up in the context of pediatric hospitals which in many instances have continued to provide care into adult life. However many countries have strict regulations or customs for transfer of patients to adult medicine at a defined age, which has highlighted the need for adult CF services. Health care providers and the medical profession have not always kept pace with the demographic changes in the disease, and may benefit from guidance on the development of adult CF services.

WHO Human Genetics Programme, ICF(M)A and IACFA have convened this joint meeting to make recommendations for services for adults with CF around the world.

## **2. Size of problem**

With continually improving survival reported from industrialized countries, there are now many adults with this disease. In Scandinavia for example, over 45 % of the patients are over 18 years of age [7]. This demographic change requires services appropriate for an adult CF population.

The requirement for separate services for CF adults will depend upon patient numbers, and will vary between and within countries according to the expertise and resources available. There is a training requirement for adult physicians interested in this disease and willing to devote their time and energy to providing these services.

Because CF is a progressive disease, it follows that a disproportionate number of adults will be at the severe end of the clinical spectrum. Thus, adult CF services can be anticipated to be relatively more expensive than their pediatric counterparts.

In some countries and cities it may be appropriate to provide pediatric and adult care at the same centre; but it even so it is necessary to provide specific services for adults for which there is no pediatric equivalent. It is recommended that an adult CF clinic should be established when numbers of adults in a centre are 50 or above.

## **3. Transition**

Adolescents and young adults with chronic conditions share the same developmental, social, and emotional needs as their healthy peers. There are multiple transitions that the young healthy adult has to go through: family home to independent living, romantic partnership and education for a career. All these transitions help the person to become increasingly self-

sufficient.

In addition, persons with a chronic disease need to make a transition within the health care system to gain greater personal responsibility for their illness. Improving the sense of control in the CF child, by giving children time alone with the specialist, should be introduced from an early age.

Problems arise in transition because there may be limited access to adult caregivers due to lack of training and funding. There is no model which is proven superior to others, however the physiological and psychological development from a child into an adult must be taken seriously.

If a combined clinic is more appropriate to the national culture, population size or resources, access and possible transfers to other doctors in the same clinic should be available once adulthood has been reached.

It is essential to treat each adolescent as an individual and to retain flexibility in the age at which transfer is arranged according to the physical and emotional development of the patient.

It is also helpful if the adult specialist is present in certain pediatric clinic sessions so that transfer does not imply a total break from the familiar services.

Occasionally the diagnosis of CF is delayed until teenage or adult life. These patients may find it more difficult to adjust to the diagnosis and its implications than those diagnosed in childhood.

## **4. Components of adult cf service**

### **4.1 Multidisciplinary care**

The care for adult CF patients must be multidisciplinary since the disease affects the whole body. It also has major psychological and social consequences for the individual. Older patients with CF are striving for independence, control of their situation and major input into health management decisions. Expectations are high because they have become accustomed to a high standard of pediatric care with individual attention.

### **4.2 Adult medical problems**

New medical issues might arise for the CF patient in adult life. Most of these can also affect children, but occur with greater frequency as they become older. These include:

- increased frequency of respiratory exacerbations;

- emergence of new pathogens;
- haemoptysis;
- pneumothorax;
- allergic bronchopulmonary aspergillosis;
- cardiac failure;
- respiratory failure;
- malnutrition;
- distal intestinal obstruction syndrome (DIOS/meconium ileus equivalent);
- gallstones;
- pancreatitis;
- malignancy of the gastrointestinal tract;
- progressive liver disease;
- portal hypertension and oesophageal varices;
- diabetes mellitus;
- nasal polyposis and sinusitis;
- osteoporosis;
- arthropathy;
- vasculitis;
- male infertility;
- pregnancy;
- pre/post lung transplantation.

### 4.3 Psychosocial problems

The transfer of responsibility from the parents to the CF individual involves both psychological and social adjustments (see Section 7).

### 4.4 Core CF team

The following personnel are required as core members of the CF team:

- Respiratory physician and medical team;
- Clinical Nurse Specialist and home care nurse if appropriate (see 5.5);
- Specialist physiotherapist/respiratory therapist;
- Specialist dietitian;
- Social worker.

Although not part of the core CF team, other health care providers such as general practitioners and university, college or occupational health services may contribute to the care of individuals with CF and appropriate liaison arrangements should be made.

### 4.5 Consultants

The skills of the following specialists will also be needed:

- Expert laboratory support services: microbiology, biochemistry, immunology, pathology, genetics;
- Pharmacist;
- Gastroenterologist;
- General surgeon;
- Thoracic surgeon;
- Radiologist;
- Endocrinologist;

- Otorhinolaryngologist;
- Anaesthetist/Intensive care specialist;
- Infectious disease specialist;
- Psychologist with psychiatric support;
- Obstetrician/gynecologist;
- Rheumatologist;
- Transplant services team;
- Geneticist;
- Pain relief team;
- Terminal care team including chaplain/religious advisor where appropriate.

#### 4.6 Facilities

Provision of a comprehensive CF service requires extensive organisational arrangements. These include:

- Inpatient accommodation;
- Outpatient accommodation;
- Lung function testing with facilities to prevent cross-infection between patients;
- Facilities for isolating patients with transmissible organisms such as *Burkholderia cepacia* and methicillin-resistant *Staphylococci (MRSA)*;
- Home care services;
- Shared care arrangements at outlying hospitals where appropriate;
- Liaison with pediatric clinic including transition arrangements.

### 5. Patterns of health care delivery

## 5.1 Established adult CF clinics

Independent adult Cystic Fibrosis Clinics have been established for the past 20-30 years in some countries. At these centres, all inpatient and outpatient care is provided by a multidisciplinary team similar to that described above.

## 5.2 Parents and partners

In an Adult CF Clinic, patients are encouraged to take more responsibility for their own medical care. Parents should normally not be completely excluded (if only because with advancing disease the patient may revert to a greater degree of dependence), but patients in an adult clinic need the opportunity to be seen independently. This facilitates frank and open discussion about relationships, contraception, adherence to therapy, possible alcohol or substance abuse and other concerns. The family will continue to have a major role as supporters and advocates, and in many cases adult patients involve life partners in their health management.

## 5.3 Independent adult CF clinics

Wherever there are sufficient adult patients and adequate resources a specialized adult clinic would seem to be the preferred model. **An adult physician and team that is both interested and capable of providing good quality care is fundamental to such a model. Motivation, commitment and dedication of time by the team are more important than any other consideration. If these conditions cannot be met no attempt should be made to establish an independent clinic.** Appropriate professional training of all members of the adult multidisciplinary team is required.

## 5.4 Other models

There are a variety of models of adult care ranging from an adult physician attending the pediatric clinic through to the provision of a full and comprehensive service being provided by an independent adult team. The appropriate model for each clinic will depend on numbers of adult patients, availability of skilled adult team members and the provision of adequate resources (financial, infrastructure, allocated time). If there are no adequately trained and motivated personnel to provide an adult CF team, the pediatric CF clinic will need to develop its services and facilities to cope with the needs of adults. This has certain advantages, in respect of continuity of care and the expert knowledge and experience of the pediatric CF team, but may be perceived by adult patients as continuing a parental relationship and diminishing their autonomy.

## 5.5 Home care and self management

Home care consists of any type of therapy provided in the home that attempts to prevent or

reduce secondary complications of CF.

Components of home treatment involve: intravenous antibiotics, intensified physiotherapy and nutritional support. In addition, psychosocial support should be offered in the home setting.

The evolution of home care options as alternatives for traditional hospital based care highlights increased expertise and expectations of progress in CF treatment. Benefits and risks must be weighed carefully before home treatment can be advised and instituted. There are considerable differences in uptake of home care programmes between CF-care centers which may also be influenced by national health care provision systems including reimbursement for home care costs. Aspects of legal responsibility for care and care complications may arise in those under age or in those with severe advanced disease.

The **advantage** of home care is that it leads to less interruption of regular life. Home treatment might also reduce the risk of cross infection between CF patients, and it decreases the financial burden for the patient and the community since a hospital bed is not needed. Many consider maintenance of individual independence the most important factor offered in home care, and patients may be less inclined to minimize symptoms if they know that intervention does not necessarily involve admission to hospital. However, few data exist on true cost effectiveness or cost utility.

The **disadvantage** of home care is that medical treatment is less controlled. Physiotherapy and nutritional support may be less consistent and intensive. Furthermore, the family continues to carry the burden of CF when the patient is sick, which may lead to increased psychological stress.

- Conditions for home care;
- adequate facilities in the home;
- adequate teaching of patient and family;
- adequate physical and psychological strength of care givers;
- medical suitability of patient;
- regular home surveillance/guidance from the CF center;
- availability of community doctor/nurse specialist in case of emergency.

**It must be clearly stressed that home care is not an ideal choice for all patients and careful advice and assessment is needed.**

**Terminal care** of the dying patient at home will depend on local settings. If attempted, psychological strength of caregivers must be ensured and medical and nursing support systems must be available.

## **6. The adult with CF in society**

The adult CF subject desires as normal a life as possible.

Occasional patients are not diagnosed before adulthood. These patients may experience a period of psychological adjustment with similar responses (e.g. emotional shock, denial, relief, anger guilt depression etc.) to those which are more usually encountered in the parents of a newly diagnosed child, and these reactions need to be recognized and accommodated by caregivers.

### **Psychological characteristics of CF adults may include the following:**

Many adults with CF are dissatisfied with their physical appearance and feel unattractive. Emotional disturbances are mainly presented in the form of anxiety and depression.

Normal peer relations can be difficult and CF subjects have often met together. Recent trends have reduced this possibility, especially in CF centres that have instituted segregated care for CF patients. CF camps for children and adolescents have been withdrawn due to the risk of crossinfection. Patients may thus become socially isolated.

There may be a delay in achieving independence from parents with regard to both separate living and financial support.

Children with CF may have missed significant periods of school, sometimes through overprotection, and higher education may not have been contemplated. Impaired health during adolescence and adult life makes sedentary occupations more appropriate than manual work, but patients often have difficulty in obtaining suitable training. Attendance at universities and colleges away from home may involve transfer to a new and unfamiliar clinic, and requires a degree of emotional independence which some may not have achieved.

Adults with CF may have difficulty finding a partner and are often reluctant to reveal their illness to others. There is therefore a significantly smaller proportion of CF adults married or cohabiting than in the normal healthy population.

Adults with CF may become biological parents with the help of modern reproductive technology. This raises new ethical, emotional and medical issues for both patients and

caregivers. Before considering pregnancy, it is essential that the subject is discussed in depth with the couple in the best interest of themselves and the potential child.

CF adults are less likely to be in paid employment, the reason appearing to be predominantly health related. Daily activities, such as looking after the home or pursuing a hobby may also be impaired due to ill health.

Psychosocial support is needed to help CF adults to face the challenges of all these problems, and is an indispensable component of the service provision. However, it must be stressed that the majority of CF adults are psychologically stable and well-adjusted, even in the presence of advanced disease.

All team members, but most importantly the physicians, must be trained to understand these worries and struggles, so that they may help and encourage the patients and families to attend to them and not to be embarrassed by the team's awareness of their existence.

Dealing with end of life issues still remains an extremely important psychosocial issue for patients, family members, and team members alike. It must be recognized that giving care to chronic sick and terminally ill patients is psychologically demanding, and an important function of the CF psychologist is to counsel and support other members of the team.

## **7. Training of adult cf physicians**

Training will depend on the specialty from which the trainee is coming, and will depend on the individual's previous level of involvement with CF adults. Although trainees may be recruited from respiratory medicine, general internal medicine, infectious disease, gastroenterology and immunology, the majority will be trainee pulmonologists.

The basic training of pulmonologists differs between countries. Consequently this leads to a difference in knowledge of internal medicine. Therefore programmes to train pulmonologists in CF specialised care need to be tailored with respect to the educational background of pulmonologists in each country. With increasing age, other CF-related non-pulmonary complications like exocrine pancreatic insufficiency, diabetes, hepatobiliary disease, distal intestinal obstruction and musculo-skeletal disorders will contribute significantly to morbidity and require specialised care.

Although there are no definite models for physicians wishing to train to be centre directors in cystic fibrosis units, relatively informal arrangements are available in Europe, North America and Australia usually involving a 2-year period spent as a trainee in an Adult CF Unit.

The following areas should be considered for training:

- general internal medicine, respiratory medicine and gastroenterology;
- management of diabetes and nutrition;
- attachment to a pediatric CF unit for a period of time;
- trainees should have appropriate understanding of laboratory support services;
- involved in the CF care, particularly in medical genetics and microbiology;
- understanding the role of clinical psychology;
- a period of at least 2 months should be spent in a transplantation centre;
- performing or coordinating a research project.

For other core CF team members a period of attachment to an established CF center elsewhere is highly desirable before taking up their appointment.

## 8. Economics

**Adult services will be relatively more expensive but funding cannot be withdrawn from pediatric clinics if present good survival into adult life is to be maintained.** Hospitals offering centre care for CF are confronted with the financial burden of treating these patients. This financial burden is mostly caused by in patient expenditure. In-hospital cost is predominantly caused by intravenous antibiotic treatment for exacerbations of *Pseudomonas aeruginosa* lung infections. Additionally with increasing age, resistance becomes more prominent requiring newer and more expensive antibiotics.

Including the CF services in the general hospital medical budget leads to complaints and antagonism among staff when it is realised that the CF services consume a disproportionate amount of money. It is therefore strongly advised that the funding for CF services should be separately identified and provided.

Because days in hospital account for most of the CF expenditure the most effective way to reduce costs is to keep patients out of the hospital.

Home I.V. (intravenous) therapy with antibiotics for treatment of infectious exacerbations is the best example. Multiple studies have shown that home I.V. antibiotic treatment is a viable option for selected patients who are evaluated, trained and monitored by a CF team experienced in home care. It may lead to a reduction in costs and is less disruptive to the patients and their families.

New therapies may decrease the number of hospitalisations but they tend to be very expensive when first introduced. It is therefore important that they are properly evaluated before gaining general acceptance and such studies can only be undertaken by collaboration between CF units. Although they may eventually reduce the long-term cost of treatment they inevitably increase costs in the short to medium term. Cost effectiveness should be part of the evaluation of new therapies. Useful patient outcomes when undertaking economic evaluation of CF care should include pulmonary function, nutrition, quality of life and long term survival.

It is recognised that not all patients have equally severe disease and consideration can be given to costing patient care according to grades of patient disease severity [11].

## **9. Recommendations**

1. The medical profession and health care providers should be educated about the requirement for adult cystic fibrosis services in their country.
2. The distinctive medical, emotional, social and economic needs of adolescent and adult CF patients should be recognised.
3. Adult CF services are best provided in special, adequately staffed and funded adult-oriented clinics.
4. Where independent adult CF clinics cannot be justified because of insufficient numbers of patients, appropriate services and facilities for CF adults should be provided within the framework of an all-age service led by the paediatric CF clinic director.
5. Adult CF clinics should be located in large medical centres where the expertise of a multidisciplinary team of consultants is available.
6. Physicians, nurses, physiotherapists, dietitians and social workers providing CF care for adults should be given adequate training at established CF centres. Training programs for CF centre directors are urgently needed.
7. Adult CF patients must take the major responsibility for compliance with their health care programs but parents and partners should not be excluded because they have vital supportive roles.
8. Wherever possible, for both medical and economic reasons, care should be provided at home by CF team personnel, for suitable patients.
9. Adult patients should have clearly defined access and input into management policies of their clinic.
10. Adult CF clinics should have a budget clearly separated from the general budget of the

hospital.

11. In the expectation that future drugs designed to promote a cure for CF will be expensive, the cost and benefit of new treatments should be rigorously evaluated.
12. WHO, ICF(M)A and IACFA should keep the provision of adult CF services under review and make further recommendations if indicated.

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