

STANDARDS OF CARE



**Standards for the
Clinical Care of
Children and Adults
with Cystic Fibrosis
in the UK
2001**

*A revised, expanded and referenced
version of the Cystic Fibrosis Trust's
1996 Guidelines*

*The CFTrust's
Clinical Standards
and
Accreditation Group*

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The Cystic Fibrosis Trust's Clinical Standards and Accreditation Group

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STANDARDS FOR THE CLINICAL CARE OF CHILDREN AND ADULTS WITH CYSTIC FIBROSIS IN THE UNITED KINGDOM

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I. INTRODUCTION

I.1 Cystic fibrosis - incidence, prevalence and survival

Cystic fibrosis (CF) is the most common, life threatening, recessively inherited disease of Caucasian populations, with a carrier rate of 1 in 25 and an incidence of 1 in 2,500 live births. There are reports of CF in those of Asian origin both from the UK (Bowler et al, 1993 [III]) and N. America (Powers et al, 1996 [III]) but it is rare in people of Afro-Caribbean origin (Hamosh et al, 1998 [III]). In 1992 there were over 6500 people with CF in the UK with 65% under 16 years. Births of slightly over 300 per year outnumber deaths by 160, which suggest an estimated population of over 7500 patients in the UK at present (Dodge et al, 1997 [III]).

A health authority with a population of 250,000 has approximately 35 resident patients with CF of whom 15 are adults. The proportion and number who are adults is increasing and by 2002 will equal or exceed the number of children. Median survival has improved dramatically and has been predicted to be at least 40 years for children born in the 1990s (Elborn et al, 1991). However, many of the people with CF, known to the CF Trust, who die each year, are adolescents and still some children die. Population carrier screening is feasible and if introduced nationally may influence patient numbers; in the one area where antenatal screening has been routine it appears to have reduced patient numbers (Cunningham & Marshall, 1998 [III]). Neonatal screening allows early diagnosis and treatment and some 20% of newborns in the UK are currently screened for CF. The accumulated evidence tends to favour neonatal screening leading to early diagnosis and treatment as a means of preventing malnutrition and permitting early treatment of respiratory infection before irreversible pulmonary damage occurs (Murray et al, 1999 [III]); also early genetic counselling can be given to the parents and other family members of childbearing age (Super et al, 1994 [IV]). The Cystic Fibrosis Trust reiterates its strongly held beliefs on neonatal screening and welcomes the Government's recent support for a national neonatal CF screening programme.

I.2 Clinical manifestations

Cystic fibrosis is the result of mutations affecting a gene, which encodes for a chloride channel known as the cystic fibrosis transmembrane conductance regulator (CFTR), which is essential for the regulation of salt and water movements across cell membranes (Rommens et al, 1989; Riordan et al, 1989; Kerem et al, 1989). Improper or faulty regulation results in thickened secretions in organs such as the lung and pancreas. In the respiratory tract this impairs the clearance of microorganisms leading to recurrent infection, bronchial damage, bronchiectasis and eventually death from respiratory failure. In the pancreas, the exocrine ducts become blocked and there may be severe damage even before birth (Andersen, 1938 [III]; Imrie et al, 1979 [III]). Most men with CF are infertile due to failure of the *vas deferens*, seminal vesicle, ejaculatory duct and body and tail of the epididymis to develop (Oppenheimer & Esterly, 1969 [III]).

I.3 Presentation

There is a wide range of clinical presentation and severity. The majority present in early childhood with respiratory tract infections which are slow to clear, or persistent, intestinal malabsorption and failure to thrive. Some infants (c.15%) present at birth with meconium ileus. A minority of

patients is diagnosed in adult life - 10% in the US CF Foundation database were over 16 years when diagnosed.

I.4 Pathology and treatment

Most of the morbidity and more than 90% of the mortality of CF is related to chronic pulmonary sepsis and its complications (Hoiby & Frederiksen, 2000 [III]). The newborn baby with CF has normal lungs but there are minor obstructive changes in the submucosal glands of the trachea and major bronchi (Oppenheimer & Esterly, 1975 [III]). Infection occurs early and it has been suggested that inflammation may precede infection (Kahn et al, 1995 [III]). The initial infecting organisms are *Staphylococcus aureus* and *Haemophilus influenzae* for which patients are treated with oral antibiotics either long-term (Weaver et al, 1994 [I b]) or when respiratory cultures are positive (Szaff & Hoiby, 1981 [III]). Eventual infection with *Pseudomonas aeruginosa* is almost inevitable for reasons that are ill understood. However, although the establishment of chronic infection after the initial colonisation was previously thought to be inevitable, it is now established that early *P. aeruginosa* infection in most patients can be eradicated by the early use of nebulised antibiotics (Littlewood et al, 1985 [III]; Wiesemann et al, 1998 [I b]) or a combination of nebulised and oral anti-pseudomonal antibiotics (Valerius et al, 1991 [I b]; Frederiksen et al, 1997 [II a]). Further recurrences are commonly with a different strain of *P. aeruginosa* (Munck et al 1998 [III]) and can also be successfully eradicated in some patients (Frederiksen et al, 1997 [II b]). Chronic *P. aeruginosa* infection and respiratory deterioration are therefore not inevitable and can be prevented or delayed for years in some patients. A small but increasing proportion of patients transferred to adult CF clinics are free of chronic infection.

Treatment is directed at identifying and eradicating bacterial infection from the airways, for it is now clear that prevention of chronic *P. aeruginosa* infection will prevent or lessen deterioration of lung function (Kerem et al, 1990 [III]; Pamukcu et al, 1995 [III]) and improve survival (FitzSimmons, 1996 [III]; Frederiksen et al, 1996 [III]). If and when chronic *P. aeruginosa* infection becomes established, regular nebulised antibiotics will stabilise the patient's condition and slow the decline of respiratory function (Hodson et al, 1981 [II b]; Mukhopadhyay, 1996 [1a]; Ramsay et al, 1999 [1b]). In one study, using historical controls, patients receiving three monthly elective intravenous antibiotics had better survival and reduced mortality compared with controls treated symptomatically (Szaff et al, 1983 [III]). A three-monthly intravenous antibiotic regime has been adopted by a number of centres although its value has not been established by a satisfactory controlled trial. In a more recent study in the UK comparing three-monthly and elective intravenous antibiotics, no advantage was apparent for those having three-monthly treatment; in fact their mortality was slightly increased. However, the numbers were small and the study considered to be underpowered; also the symptomatic group did require 3 courses of intravenous antibiotics per year - only one less than the elective group (Elborn et al, 2000 [I b]).

Viral and Mycoplasma infection may also be responsible for exacerbations of the chest infection (Wang et al, 1984 [III]; Conway et al, 1992 [III]).

Thus, the provision of optimal respiratory care is essential from an early age if infections are to be identified promptly, treated early and effectively to avoid chronic infection, and consequent respiratory deterioration. The maintenance of a good nutritional state also plays a major part in maintaining respiratory health (Corey et al, 1988).

1.5 Complications

With increasing age patients may suffer a variety of complications, including nasal polyposis, pneumothorax, haemoptysis, allergic bronchopulmonary aspergillosis (ABPA), respiratory and cardiac failure, episodic and chronic distal intestinal obstruction syndrome (DIOS), gallstones, oesophageal reflux and oesophagitis, chronic liver disease, portal hypertension, colonic stricture, inflammatory arthritis, osteoporosis and male infertility. A high proportion of older patients develop diabetes mellitus. Behavioural and psychological problems are common in children and adults.

1.6 The need for specialist care

Cystic fibrosis is a multi-system disease requiring a holistic approach to care. Care should aim to prevent, for as long as possible, chronic infection and later stabilise the pulmonary infections to minimise deterioration in respiratory function and to maintain a good nutritional state. The aim is to maintain the patient's independence, improve their quality of life and extend their life expectancy. It is now generally agreed that a team of trained experienced health professionals in a Specialist CF Centre best provides this type of care (Neilsen et al, 1982 [III]; Phelan & Hey, 1984 [III]; Walters et al, 1994 [III]; Mahadeva et al, 1998 [II b]; Frederiksen et al, 1996 [III]). Alternatively, care for children may be provided at a general hospital if there is a CF Clinic there, which undertakes a recognised form of shared care with the staff of a Specialist CF Centre.

1.7 Support for specialist care

The need for Specialist CF Centres has been recognised by the Royal College of Paediatrics and Child Health (RCPCH - previously the British Paediatric Association), the British Thoracic Society (BTS) and the Royal College of Physicians of London (RCP) (see Appendix A). Most CF adults and parents of CF children favour having input to their CF care from the staff of a Specialist CF Centre (Clinical Standards Advisory Group, 1993 [III]; Clinical Standards Advisory Group 1996 [III]; Walters et al, 1994 [III]).

1.8 The CFTrust now recognises two levels of CF care

People with CF receive their care at either a **Specialist CF Centre** (previously Level I and II Centres), usually in a teaching hospital or a **CF Clinic** (previously Level III and IV) in a local general hospital, working in conjunction with a Specialist CF Centre. Regular contact with the Director and multi-disciplinary team of a Specialist CF Centre should ensure that all patients receive optimal care. The improved treatment resulting in better survival has been initiated and evaluated at Specialist CF Centres.

SPECIALIST CF CENTRE. The staff at Specialist CF Centres usually treats either adults or children. Patients are referred from other hospitals to a Specialist CF Centre for their expertise in CF care; CF is a special interest of the medical and paramedical staff. Usually there are over 100 children or adults attending for full care, permitting the staff to maintain and improve their

expertise. The team at a Specialist CF Centre has sufficient patients, interest, enthusiasm and time to carry out and publish research. The unit is a training, resource and research centre for the region. It provides full care for local patients and also for some distant patients who prefer to travel there rather than attend the CF Clinic at their local hospital. The Specialist CF Centre also provides shared care for other distant patients who attend the CF Clinic at their local hospital for most of their care.

CF CLINIC. A clinic, usually in a general hospital, where CF patients who live locally receive most of their hospital care. However, patients attending a local CF Clinic are also seen periodically by the staff of the Specialist CF Centre for some form of **shared care**. Although there may be only 20 or 30 patients attending the local CF Clinic, it is desirable that there are special clinic sessions set aside for CF patients and there must be a special interest in CF among clinicians and paramedical staff.

The essential requirements for routine outpatient and inpatient care and home support should be of the same standard as those at the Specialist CF Centre.

1.9 Number of patients

For geographical reasons, some Specialist CF Centres may have less than 100 patients yet may provide a comprehensive CF specialist service. However, it is generally agreed that a minimum of around 50 adults or 50 children on full care are required to develop and maintain a specialist team with the necessary experience and expertise to run an adult or paediatric Specialist CF Centre.

1.10 Staff and facilities

Specialist CF Centres usually provide either paediatric or adult services. Their specialist teams of health professionals include a paediatrician or adult physician, a clinical nurse specialist, physiotherapist, dietitian, social worker, psychologist, pharmacist, clerk and secretary. The team requires the support of staff from other specialties, together with full facilities for investigation, inpatient and outpatient treatment and clinical research. It is also important that there is adequate clinic and office accommodation for them to see not only their own patients and but also those referred from other hospitals for advice, assessment and shared care. Purpose built or adapted accommodation greatly facilitates the work of staff; inadequate accommodation may seriously compromise the service provided.

1.11 Full or shared specialist care for all patients

The Cystic Fibrosis Trust recommends, and it is widely accepted, that regular contact with the staff of a Specialist CF Centre is a necessary component of adequate care at a local paediatric CF Clinic. On the whole, good paediatric CF Clinics exist and shared care between them and the Specialist CF Centres can be an acceptable alternative to all care at a Specialist CF Centre (Littlewood & Kelleher, 1989 [IV]; Simmonds et al, 1998 [IV]; Simmonds et al, 1998 [IV]). Shared care has developed as a means of providing specialist care for larger numbers of patients whilst minimising travel for patients and improving local expertise and community support.

1.12 Adult care

With increasing age care becomes increasingly complex. There is great variation in the treatment required and developments and improvements are occurring all the time e.g. the management of CF related diabetes mellitus, osteoporosis, liver disease, fertility and pregnancy issues, and post-transplantation treatment. For these reasons, as a rule, adult patients are more effectively dealt with where the staff have experience of many patients; in practice, shared care for adult patients has proved difficult to implement.

Unfortunately, Specialist CF Centres for adults do not yet exist everywhere. They have to be developed as a priority and in sufficient numbers to cater for a growing population. Adult service development is not as advanced as paediatric care in some parts of the UK and expertise in adult requirements does not exist in many district general hospitals to the same extent as it does for paediatrics.

For these reasons it is recommended that the team at a Specialist CF Centre care for adults with cystic fibrosis, although there may be an opportunity to develop some innovative arrangement whereby a contribution to care can be made by the local hospital clinic. In some remote areas geography will necessitate the use of shared care. In exceptional circumstances, with good communication and an interested and appropriately trained local physician, this model can reduce the need for unnecessary patient travel while protecting the quality of the patient service.

1.13 Shared care for children

"Shared care" occurs when routine care comes predominantly from a CF Clinic in the patient's local general hospital with regular input from the staff of the paediatric Specialist CF Centre.

There are three means of providing shared care as follows -

1.14 Annual Review at the Specialist CF Centre

Patients receive most of their care at a recognised CF Clinic at their local hospital but they attend a Specialist CF Centre for an Annual Review including specialist investigations, dietary and physiotherapy assessment and for a full report including care plans for the coming year and advice for the local consultant and paramedical staff. This is a good system and the patients and families find the day at the Specialist CF Centre rewarding – particularly the time spent with the physiotherapist and dietitian (Littlewood, 1993 [III]; Carr & Dinwiddie, 1996 [III]).

1.15 Alternate visits to the local CF Clinic and Specialist CF Centre

Patients attend their local hospital CF Clinic and the Specialist CF Centre for alternate visits. A disadvantage of this arrangement is that patients often see several different doctors in a year and trends may be missed. Good communication, including patient/parent held records (or certainly copies of relevant correspondence from previous visits and Annual Reviews), is necessary to prevent confusing advice being given to parents/patient. There is also the potential problem of the parents/patient not being entirely clear which senior doctor is ultimately responsible for their care - a worry to many families (Data on file, Cystic Fibrosis Trust, 2000 [IV]).

I.16 Joint CF Clinics at the local hospital

Patients attend their local CF Clinic for all visits and the Consultant from the Specialist CF Centre, ideally with one or more members of the CF multi-disciplinary team, visits at regular intervals to run a clinic with the local health professionals. For patients from remote areas, this arrangement reduces the need for them to travel long distances. Patients are seen when they are relatively well so that deterioration can be prevented. It is an advantage if the joint clinic visit can be made soon after the patient's Annual Review, the investigations of which can be undertaken at the local hospital and the results made available for the visiting Specialist CF Centre staff's visit. Any specialised investigations not available at the local hospital can be sent to the Specialist CF Centre providing shared care and be available for the joint clinic. This system works well if it is well organised and if there is adequate staff at the Specialist CF Centre for them to be absent on a regular basis.

I.17 Arrangements for shared care

It is recommended that shared care arrangements between a CF Clinic and a Specialist CF Centre are covered by an agreement setting out the arrangements and responsibilities along the following lines: -

- The frequency with which the CF Consultant and members of the multi-disciplinary CF team see the patient e.g. at least once and ideally twice a year.
- An Annual Review is performed (Section 3.5). It should be specified where the Annual Review would be done i.e. at the Specialist CF Centre or the local CF Clinic.
- The CF Consultant from the Specialist CF Centre sees the results of the Annual Review and is responsible for reporting the results to the UK Database.
- It is desirable that the CF Consultant should write to the patient/parents summarising the results of the Annual Review and detailing any recommendations arising from the results.
- Arrangements for telephone contact between the local CF Clinic Consultant and the CF Consultant are agreed.
- Communication is improved if copies of the clinic data sheets and letters from the local CF Clinic are sent to the CF Consultant and reviewed by him/her before filing at the Specialist CF Centre.
- Events requiring notification to, discussion with or referral to the Specialist CF Centre are agreed (suggestions are listed in this document, Section 3.10).
- Prescribing arrangements for drugs are agreed as to whether they are obtained from the Specialist CF Centre, the local CF Clinic or the General Practitioner.
- The most appropriate method of contact for the patient/parents to obtain advice should be specified. This would usually be the Consultant at the local CF Clinic but may be the CF

Consultant when the local Consultant is on leave or otherwise absent, particularly if there is no local senior medical cover.

- Arrangements for regular clinical audit utilising data stored in the UK CF Database.
- The ultimate responsibility for the care of patients receiving shared care usually lies with the Consultant at their local CF Clinic but must be clearly defined in the agreement and made known to the patient/parents.

2. SPECIALIST CYSTIC FIBROSIS CENTRES

The essential component of a Specialist CF Centre, which differs from a local CF Clinic, is people – both sufficient patients with CF (ideally over 100 children for a paediatric centre or 100 adults for an adult centre and always over 50 on ‘full care’ - in contrast to ‘shared care’) and adequate numbers of suitably experienced staff to advise them on treatment. Also there are numerous other people in the hospital, some of whom may never come into direct contact with patients and families, who nevertheless are essential and contribute to the smooth running and expertise of the Specialist CF Centre’s service.

2.1 Functions of a Specialist CF Centre

Specialist CF Centres would usually be expected to provide most of the following services –

- i) *Care and Annual Review of their own patients* including the early assessment and management of infants detected by antenatal and neonatal screening. The availability and offer of open access service gives necessary support to patients and their families as well as to general practitioners.
- ii) *Comprehensive assessment and advice on diagnostic problems* and management for patients referred for shared care or on an *ad hoc* basis from smaller local CF Clinics.
- iii) *Provision and co-ordination of a wide range of treatments and services which are either not readily available at the smaller local CF Clinics or which require special expertise in patients with CF.* These would include treatment of newborn infants with meconium ileus, massive haemoptysis, pneumothorax, unusual and complex respiratory and gastrointestinal problems, liver and biliary complications, enterostomy feeding, thoracic surgery, ENT surgery, major elective surgery, cystic fibrosis related diabetes mellitus, liver disease, obstetric management, rheumatology and psychology. Counselling about fertility and pregnancy.
- iv) *Provision of expertise in specialised procedures often required by people with CF.* For example, inserting totally implantable venous access devices, upper gastrointestinal endoscopy, gastrostomy, non-invasive ventilation, bronchial artery embolisation and fibreoptic bronchoscopy on infants and children.
- v) *Provision of diagnostic and specialised laboratory facilities* including genetic investigations and counselling, antenatal diagnosis, neonatal screening investigations, sweat testing, CT and V/Q scanning, specialised cardiovascular investigations, and specialist investigation and evaluation of pancreatic function, gastrointestinal and liver disease. Detailed respiratory function tests.
- vi) *Psychosocial support for problems specific to CF.* Education, employment financial benefits, personal and family support particularly around the time of diagnosis, problems with burden of treatment, adapting to progressive disease, pre-transplant psychosocial assessment and counselling, terminal care and bereavement counselling. Treatment of depression, anxiety and a range of emotional difficulties, disordered eating and non-adherence problems.

- vii) *Ensuring the smooth transition of patients from paediatric to adult care.* Usually by running a regular adolescent/transition clinic with the appropriate paediatric and adult CF unit staff.
- viii) *Liaison with transplant centres and assessment of patients.* This would include those attending the Specialist CF Centre for all their care and also the patients from neighboring CF Clinics who are receiving shared care, prior to their referral to the Regional Transplant Centre.
- ix) *Ensuring professional colleagues in the region with responsibility for patients with CF are kept up to date.* Including the latest advances in management by providing study days, lectures, visits, educational material and discussion on mutual patients; also by arranging educational sessions for patients, parents and relatives.
- x) *Monitoring CF services in the catchment area receiving shared care and regular clinic audit.*
- xi) *Providing opportunities for patient involvement in the monitoring and development of the service and support for patient advocacy.*
- xii) *Responsibilities for entering data of all patients on full or shared care into the UK CF Database.*
- xiii) *Performing clinical research aimed at increasing the understanding of CF and introducing and evaluating new treatments. Collaboration with multi-centre trials.*
- xiv) *Attendance of the staff at national and international CF meetings. Presentation and publication of the results of research.*
- xv) *Additional specialised investigations are available at some Specialist CF centres e.g. complex lung function tests particularly for young children and infants. Nasal potential difference measurements.*

2.2 Facilities necessary for the care of people with cystic fibrosis

2.3 Outpatient facilities which should be available at all Specialist CF Centres and CF Clinics

Sufficient clinic sessions designated for patients with CF are required to allow adequate time for discussion and to avoid overcrowding in the waiting areas.

Ideally, there will be an area in each Specialist CF Centre or CF Clinic where patients and families attending for the day ('day cases') can be seen and treated on an *ad hoc* basis. In a Specialist CF Centre this area can also be used for patients coming from other hospitals for consultations, Annual Reviews, initiation of courses of home intravenous antibiotics and specialist investigations.

Relatives and patients require some accommodation, other than the waiting area, as they will often be spending many hours at the unit with young children and need a base for the day.

- 2.31** There should be separate rooms available for medical staff, physiotherapists, dietitians and psychologist/social worker and the nurse specialist – consulting and treatment rooms, where patients can be seen and minor procedures performed.
- 2.32** There must be a high standard of hygiene practiced by all concerned, particularly washing or disinfection of the hands (Vandenbroucke-Grauls, 2000; Pittet et al, 2000 [II a]) and regular expert microbiological surveillance ("*Pseudomonas aeruginosa* Infection in People with Cystic Fibrosis". Cystic Fibrosis Trust. 2001).
- 2.33** Respiratory function equipment should be available and appropriately calibrated and adequate to cater for patients with transmissible infections.

2.4 Inpatient facilities which should be available at all Specialist CF Centres and CF Clinics

- 2.41** Specialist CF Centres and CF Clinics should have a clear infection control policy. Patients with cystic fibrosis are at particular risk of harbouring multi-resistant organisms e.g. methicillin resistant *S. aureus* (MRSA), *P. aeruginosa*, and *Burkholderia cepacia* (Pedersen et al, 1986 [III]; Cheng et al, 1996 [III]; Jones et al, 2000 [III]; Armstrong et al, 2000 [III]). This poses a potentially fatal risk to themselves, to other patients with CF and also to susceptible patients with other illnesses (McCallum et al, 2000 [III]; McCallum et al, 2001 [II b]).
- 2.42** The beds should be in single rooms (the main reason being to reduce the likelihood of cross-infection), ideally with en-suite bathrooms or separate toilet and bathroom facilities according to the patient's microbiological status [C].

Children should be in single room accommodation, which has adequate observation and appropriate facilities for parental sleeping [C].

The equivalent of 3 to 5 beds is required for every 50 patients who are receiving full care at the Specialist CF Centre or CF Clinic (Various Working Parties. Appendix A. [IV]). The number of beds required would vary according to whether the patient population is mainly composed of relatively well children or adults at a Specialist CF Centre, a significant proportion of whom may be severely affected.

- 2.43** Facilities should be available for education and recreation taking account of the potential risks of cross-infection.
- 2.44** Equipment for measurement of height and weight should be available [C].
- 2.45** It is inappropriate for patients who have an exacerbation of their chest infection to join a waiting list. They should be admitted the same day if the doctor considers this is necessary. Arrangements for direct admission to the ward should be in place to avoid long delays in the Accident and Emergency Department [C].

2.5 Staffing

These staffing requirements have been developed by a number of working groups and committees (Appendix A) and have been accepted as representing an overview of expert opinion [C]. The requirements vary depending on the age of the patients, the severity of their condition and the amount of shared care and community support that a Specialist CF Centre provides. The numbers may have to be increased where there is a constant higher proportion of very unwell patients with more highly complex needs. It is suggested that an individual will require approximately half or more of their time working in CF patient care to maintain the necessary expertise. Necessary adjustments to these recommendations for certain categories of staff may be required in the case of exceptionally large clinics with over 200 patients.

The suggested number of whole time equivalent staff (WTE) required for every 50 patients on full care. Patients on shared care would require approximately 50% of the allocation for those receiving full care.

Staff Member	Local Clinic (< 50 patients)	Specialist Paediatric Centre	Specialist Adult Centre
Consultant 1	0.5	0.5	0.5
Consultant 2	Nil	0.2 – 0.3	0.2 – 0.3
Staff Grade	Nil	0.4	0.6
CF Sp. R	Nil	0.5	0.5
CF Nurse	1.0	1.0 – 1.5	1.0 – 1.5
Physiotherapist	1.0 – 2.0	2.0	2.0
Dietitian	0.4	0.4	0.4
Social Worker	0.4	0.4	0.4
Psychologist	0.4	0.4	0.4
Secretary	0.5	1.0	1.0
Data clerk	Nil	0.1	0.1
Pharmacist	0.2	0.3	0.3

Consultant 1 has a major commitment to CF care (Clinic Director or Lead Clinician in Cystic Fibrosis or Consultant with an interest at a CF Clinic); Consultant 2 is involved with routine CF care but to a lesser extent. At Specialist CF Centres, there is a Specialist Registrar (CF Sp. R) with full commitment to CF, in addition to the usual junior staffing of the paediatric/medical team and also a Staff Grade doctor.

It is recommended that the staff should be members of their appropriate CF interest group -

UK CF Nurse Specialist Group

Association of Chartered Physiotherapists in Cystic Fibrosis

UK CF Dietitians' Interest Group

British Psychosocial Professionals Group in Cystic Fibrosis

UK CF Pharmacists Group

3. STANDARDS OF CARE

3.1 Diagnosis

Diagnosis rests on the following -

- Clinical suspicion and the demonstration of excess sodium and chloride concentrations in sweat or on positive antenatal or neonatal screening. Laboratory staff experienced in the technique should perform two sweat tests using a recommended procedure. If two CF mutations are present, one sweat test is adequate. If only one mutation can be identified, it is preferable that the second sweat test is performed at the Specialist CF Centre (Wallis, 1997 [III]; Rosenstein & Cutting, 1998 [III]).
- Demonstration of intestinal malabsorption and of a pancreatic abnormality supports the diagnosis and confirms the need for pancreatic enzyme treatment.
- Diagnosis is not always straightforward and incorrect diagnoses are not uncommon (Wallis, 1997). When there is any doubt about the diagnosis, the patient should be referred to the Consultant at the Specialist CF Centre (rather than referring directly to the laboratory for a sweat test).

3.2 Genotyping

All patients should be genotyped (Lane et al, 1997 [III]). However, failure to detect a recognised mutation on either chromosome does not exclude the diagnosis. It is advisable to refer patients who have clinical and investigation findings suggesting a diagnosis of CF, but no demonstrable mutations, to the Specialist CF Centre for confirmation of the diagnosis.

3.3 Following diagnosis

The patient/parents should be seen by the Consultant, who will be responsible for their medical care, within 24 hours of the diagnosis being confirmed.

- The CF Consultant should give detailed and sensitive explanation of the diagnosis. If only one parent is present it is advisable that a friend or relative accompanies them to ensure they retain the information (Jedlicka-Kohler et al, 1996 [III]). A similar detailed explanation should be given to newly diagnosed adults who will be referred to a Specialist CF Centre for adult patients. It is helpful to stress the improving prognosis, the prospects of new treatments and the need for long-term follow-up.
- The commitment to long-term care by the clinical team should be emphasised and the local arrangements for access to 24-hour advice explained. Contact telephone numbers for appropriate team members, including in many Specialist CF Centres the Consultant, are provided.
- Access to appropriate information is made available.
- Families are given details of the Cystic Fibrosis Trust.

- An explanation of the symptoms and implications of CF is repeated and reinforced by the CF Nurse Specialist. The CF Nurse Specialist from the Specialist CF Centre (or the nurse performing the role of the CF Nurse Specialist at the local CF Clinic if she/he has the necessary knowledge and experience) will assess and document the physical and emotional needs of the patient and family. Ideally, a CF Nurse Specialist from the Specialist CF Centre, who has experience in neonatal CF screening, will see the families of infants detected by neonatal screening after liaison with the General Practitioner and Health Visitor (Appendix C).
- Explanations to the patient and family will be required during childhood, adolescence and adult life.
- All patients newly diagnosed in local CF Clinics are notified to the Consultant at the Specialist CF Centre that will share the care of the patient.
- The patient's General Practitioner(s) and Health Visitor are kept closely informed of these developments.
- It is advisable to admit to hospital infants with any respiratory symptoms or significant failure to thrive at the time of diagnosis as lower respiratory viral infections can be severe (Armstrong et al, 1998 [III]) and significant bacterial infection with an inflammatory response is common and occurs early (Armstrong et al, 1997 [II b]). Establishing a good nutritional state is of paramount importance and reversal of failure to thrive should be a priority during the first year of life (Farrell et al, 2001 [I b]).
- Infants presenting with meconium ileus and referred for neonatal surgery are referred to the CF paediatrician within 24 hours and gene tests arranged which are likely to confirm CF as the cause.
- The families of patients with CF are referred to the genetic service for advice and relevant family members offered genetic advice (Super et al, 1994; Lane et al, 1997).
- At approximately six months after diagnosis some assessment of the emotional well being of the parents and/or patient. This would identify any potential adaptive difficulties.
- Siblings of known CF patients should be sweat tested.
Newborn siblings should be considered as having CF until the gene results become available or a sweat test is performed at about a week of age. If the sweat test and genotyping is inconclusive, they should have another sweat test in the second month and, ideally, a faecal pancreatic elastase 1 (Cade et al, 2000 [II b]) or a faecal chymotrypsin (Brown et al, 1988 [II b]) for pancreatic abnormality and faecal microscopy (Walters et al, 1990 [II b]) or similar test to detect excessive faecal fat.

3.4 Routine outpatient reviews

Regular routine outpatient reviews should take place frequently, ideally every 6 to 8 weeks and at least every 3 months (Wood & Piazza, 1988 [II b]). Infants are initially seen every 2 weeks and when thriving every 3 to 4 weeks [C].

The following procedures are recommended at every clinic visit whether at a Specialist CF Centre or a local CF Clinic –

- *Measurement of weight and height (children only) by experienced staff.* The children's values will be plotted on a growth chart usually by the doctor, nurse or dietitian. In adults weight is recorded at each visit (Dupuis et al, 1998 [III]) and height is measured at every visit until growth has ceased. Some clinics measure the head circumference in children less than 2 years of age (Ghosal et al, 1995 [III]) [C].
- *Culture of sputum using appropriate media for the detection of B. cepacia also S. aureus and methicillin resistant S. aureus (MRSA).* It may be necessary to refer to a microbiologist with expertise in cystic fibrosis. In non-expectorating infants and children, cough swabs, nasopharyngeal aspirate or deep throat swabs may be substituted. Where a change in treatment based on the results of the culture and antibiotic sensitivity is indicated, this should be communicated to the patient or parent and to their family doctor as soon as possible [C].
- *An unhurried consultation and physical examination with a doctor experienced in management of CF.* This will include a review of the respiratory and gastrointestinal symptoms, a detailed review and confirmation of the drug treatments being taken (oral and nebulised antibiotics, bronchodilators, inhaled steroids, DNase, pancreatic enzymes and vitamins, treatment for CF related diabetes mellitus, oral contraceptives). Any additional treatment since the last appointment is noted. Any changes in therapy are made and discussed [C].
- *Spirometry.* All adults and co-operative children aged 5 years and over have respiratory function tests performed at every clinic attendance (Corey et al, 1997 [III]; Dupuis et al, 1998 [III]) [C].
- *Chest X-ray.* Patients have a chest X-ray if there is a significant fall in lung function or an unexplained increase in respiratory symptoms [C].
- *Review by a Physiotherapist.* At every attendance there should be an assessment of current respiratory status in relation to current physical therapy. This includes a review and/or re-education of airway clearance techniques, inhaler and nebuliser techniques and a review of their physical activity and exercise programmes as indicated. The physiotherapist will obtain a sputum/cough swab (Worthington & Kelman, 1996 [III]) [C].
- *Review by Dietitian.* A review of the nutritional and gastrointestinal state is made at each attendance including assessment of dietary intake including use of dietary supplements and enteral tube feeds (Collins et al, 1999 [III]). Enzyme use including dose, timing and method of administration and adjustment to fat content of the meal (Littlewood & Wolfe, 2000 [III]). Review of diabetic treatment and control. Review of vitamin supplementation [C].
- *Access to the CF Nurse Specialist.* The CF Nurse Specialist is pivotal in the clinic and will see all patients attending. In local CF Clinics in District General Hospitals, it is important that an 'outreach' or 'home' nurse from the local hospital has responsibility for the home care of patients with cystic fibrosis. It is particularly important that there is the expertise to

organise home intravenous antibiotic treatment - if necessary, by liaison with the CF Nurse Specialists from the Specialist CF Centre (Appendix C).

- *The Social Worker* should be available in the clinic for immediate access if required.
- *Access to a psychologist, experienced in CF*, if patients or families are having emotional difficulties.
- *Regular servicing of specialised equipment*, e.g. nebuliser/compressors systems, is organised either by the CF Clinic Nurses or Physiotherapists and carried out by qualified personnel – usually the hospital’s Medical Physics Department [C].
- *A clinic report should be sent within 10 working days to the patient’s General Practitioner and all relevant colleagues.*

3.5 Annual Reviews at the Specialist CF Centre or CF Clinic.

The Annual Review is a detailed assessment of every aspect of the patient’s treatment and condition to assess progress and identify areas where treatment could be improved (Crozier, 1974 [III]; Littlewood et al, 1993 [III]; Carr & Dinwiddie, 1996 [III]). Annual Reviews are now regarded as useful in achieving the best care of a changing progressive complex multisystem disorder such as CF to ensure that nothing is overlooked.

The Annual Review identifies suboptimal treatment and improvable abnormalities, permits formulation of an individual treatment plan, provides an update for patients, parents and colleagues, provides data for research and audit and presents an opportunity for review by the Consultant at the Specialist CF Centre responsible for long term care. If the Annual Review is performed at the Specialist CF Centre it allows the patient/family to have contact with the experienced clinic staff there.

It is good practice if the CF Consultant writes the final report (not a training grade doctor – even if training in cystic fibrosis) and sends to the GP or where appropriate to the referring hospital Consultant with copies to other colleagues involved in the Annual Review or the patient’s care. Many clinics now send a copy to the patient/parents. The Physiotherapist and Dietitian discuss their findings with their colleagues at the local CF Clinic and send them separate reports of their own findings.

If the investigations are performed at the local CF Clinic and reviewed by the Consultant from the Specialist CF Centre, it is advisable that he should write a report after reviewing the final results. Finally, it is wise to arrange a date for the next Annual Review so that it is not delayed or overlooked.

The Annual Review usually includes all or some of the following, in addition to the information obtained at the routine clinic visit. Items marked [*] are considered to be necessary for a thorough Annual Review.

- i) [*] Detailed assessment of progress and review of the patient’s and family’s knowledge of the condition by an experienced doctor. If appropriate, fertility issues could be discussed.

- ii) [*] Detailed assessment by an experienced physiotherapist (Worthington & Kelman, 1996 [III]).
- iii) If the patient is visiting the Specialist CF Centre, a review by the Psychologist or Social Worker (Schechter & Margolis, 1998 [III]).
- iv) [*] Review by the CF Nurse Specialist.
- v) [*] Full assessment by the Dietitian (Collins et al, 1999 [III]). The need for further nutritional intervention should be considered e.g. dietary supplements, enteral feeding by nasogastric tube or gastrostomy.
- vi) [*] Clinical measurements. In addition to those done as part of a routine outpatient review (i.e. weight and height in children, weight in adults also height until growth has ceased, sputum culture, oximetry, spirometry) the following are usually included as part of the Annual Review: -
 - [*] Sputum culture for non-tuberculous mycobacteria (Oliver, 1998 [III]; Torrens et al, 1998 [III]).
 - Shwachman Clinical Score (Shwachman & Kulczycki, 1958; Conway & Littlewood, 1996) will be required if the data is to be entered into the UK CF database. Details of the score are in the UK Database Annual Review forms. The puberty status is recorded.
 - [*] Chest X-ray score using a recognised scoring system e.g. Northern CF X-ray Score (Conway et al, 1994) or Chrispin Norman score (Chrispin & Norman, 1974).
 - Reversibility test to bronchodilator using either a nebuliser or inhaler and spacer device when appropriate (Eggleston et al, 1988 [III]; Konig et al, 1998 [III]). Changes are measured using the usual clinic spirometer.
 - When appropriate, more detailed lung function tests, depending on local availability, including lung volumes, CO transfer, exercise testing and challenge testing.
 - Blood gases in adults, if indicated, and only if the SaO₂ is less than 92% in air.
 - [*] Full blood count, urea and electrolytes, liver function tests including serum albumin, clotting studies, fat soluble vitamin levels A, D and E also K if available. Aspergillus species RAST and precipitins, IgA, IgG, IgM and IgE.
 - Pseudomonas antibody levels, if in routine use at the clinic, as an indication of chronic *Pseudomonas aeruginosa* infection (Brett et al, 1992 [III]; Pond et al, 1994 [III]).
 - [*] Assessment of the adequacy of intestinal absorption by a combination of clinical and laboratory methods. The latter include a semi-quantitative estimation of faecal fat e.g. by microscopy (Walters et al, 1990 [II a]) or acid steatocrit (Tran et al, 1994 [II a]). Estimation of 3-day faecal fat is useful, in addition to other relevant gastrointestinal investigations, if there are significant gastrointestinal problems.

- [*] Random or fasting blood glucose in all patients and some assessment of glucose metabolism in patients over 12 years or if clinically indicated (Koch & Lanng, 2000 [III]). This is most reliably achieved by an oral glucose tolerance test (Milla et al, 2000 [II b]) Patients with established diabetes should have the adequacy of their control measured by serum HbA1c.
 - Electrocardiogram if clinically indicated.
 - [*] An assessment of liver status using a combination of one or more clinical, biochemical or imaging methods (Sokol & Durie, 1999 [III]). Annual ultrasound of the liver and biliary system is recommended by some paediatric hepatologists and is now routine in many Specialist CF Centres (Colombo et al, 1998 [II b]).
 - [*] Urinalysis for glucose (Koch & Lanng, 2000 [III]), protein (Melzi et al, 1991 [III]) and blood (Patier et al, 1998 [III]).
- vii) [*] Review of results and planning of future therapy and immunisations including annual influenza immunisation. Discussion with other members of the CF multi-disciplinary team.
- viii) [*] Report and recommendations are sent to the Consultant at the referring CF Clinic, the General Practitioner, and an increasing number of clinicians send a copy to the patient or parent.
- ix) [*] Collection of data for audit and research. The results are entered into the Specialist CF Centre's database and also reported to the UK CF database (*www.cystic-fibrosis.org.uk*). The patient's and/or a parent's written permission must be obtained after explaining that the information, although anonymised, will to be used for routine audit and medical research.

In addition, if it is the first visit or contact with the Specialist CF Centre

- [*] The diagnosis is reviewed. It is advisable to repeat the sweat test if two CF mutations have not been identified.
- [*] Genotype if not already done.
- [*] Confirm pancreatic status by faecal pancreatic elastase 1; there is no need to stop the patient's pancreatic enzyme treatment (Cade et al, 2000 [II b]).
- Introduction to CF team members; it is an opportunity to see facilities at the Specialist CF Centre.
- Provision of the CF management information from the Specialist CF Centre.
- Ensure patient knows of the Cystic Fibrosis Trust.

3.6 Inpatient care

Patients require direct access to the CF service at all times. A bed in a ward suitable for CF inpatient care should always be available for an emergency admission. An inpatient admission is an opportunity to review all aspects of CF care.

3.7 Essential services

The following inpatient services are essential

- Full range of professional respiratory care including a knowledge of the potential risks of cross-infection: doctors trained in venous access techniques: doctors and nurses trained in the management of totally implantable venous access devices; physiotherapist with experience in CF; dietitian with experience in CF and pharmacist supervision.
- Oximetry and spirometry on admission and regularly throughout inpatient stay to determine progress and duration of treatment. Blood gases (arterial or capillary) where indicated.
- Medical, surgical and diagnostic services to deal with distal intestinal obstruction syndrome.
- Nutritional support as needed during medical and surgical admissions including additional snacks, high calorie meals, oral nutritional supplements, enteral tube feeding and total parenteral nutrition.
- Management of CF related diabetes mellitus.
- Instruction of patients in the management of home intravenous antibiotic therapy, enteral feeding and oxygen therapy.
- Review of all aspects of on-going medical and psychological care prior to discharge.
- Providing patients on discharge with a clear treatment plan, including all drugs and follow-up arrangements: a summary posted within 10 working days.

3.8 Home care

Home care is often cost effective and usually favoured by the patient and family. In addition to their clinical value, home visits by the CF Nurse Specialist and other members of the CF team provide additional psychological support. The complexity of modern CF care makes close supervision from the Specialist CF Centre or CF Clinic essential. When the Specialist CF Centre is distant, it is essential that an adequate local support service be in place supervised by the local CF Clinic, ideally with additional support from the staff of the Specialist CF Centre.

- Home intravenous antibiotic therapy, where it is properly resourced and when the patient and family desire it and are properly supported to attend to it, is an important alternative to inpatient therapy. Efficient supply arrangements must be in place. It is important that the CF Nurse Specialist ensures the home conditions are appropriate for giving intravenous treatment. This includes assessment of response to treatment e.g. by measuring oxygen

saturation and spirometry and aminoglycoside levels. Standards of home care must be as good as those in hospital. The CF Nurse Specialist is a central figure for an effective home intravenous antibiotic service.

- Appropriate physiotherapy support should be available.
- Communication with the General Practitioner and other community professionals. It is important to keep the GP informed at the time of diagnosis. He/she will have a role in the treatment of undercurrent illness, arranging immunisations and annual influenza vaccination (Section 3.9).
- Instruction and support of the family and patients, especially those receiving home oxygen or nasal ventilation or total parenteral nutrition, and for those with terminal disease.
- Visits to the school should be made by the CF Nurse Specialist to inform the relevant staff about cystic fibrosis.
- Flushing totally implantable venous access devices.
- Management of ileostomies.
- Management of overnight nasogastric and gastrostomy feeding including the use of a pump.
- Supervision of diabetic management appropriate to cystic fibrosis.

3.9 Role of the General Practitioner

The family's General Practitioner(s) has an important role in communication and ongoing support varying with geographical factors and prior knowledge of the patient and family. At the time of diagnosis the CF Nurse Specialist contacts the General Practitioner.

The General Practitioner is responsible for prescribing much of the routine treatment recommended by the Specialist CF Centre including oxygen therapy. The Specialist CF Centre or CF Clinic must ensure that the General Practitioner is adequately informed about the medication recommended, some of which may be unfamiliar or occasionally used out of product license. It is reasonable to expect the General Practitioner to prescribe adequate amounts of medication (at least one month's supply). Local chemists may be able to co-ordinate the ordering and delivery of repeat medication.

The General Practitioner ensures that patients are fully immunised, including arranging for appropriate influenza vaccine every winter.

The General Practitioner who is likely to be familiar with the viral-type illnesses that are prevalent in the community at any one time commonly deals with upper respiratory tract infections.

Other specific areas of involvement are fertility issues, appropriate referrals of parents and relatives for genetic counselling, and the provision of sick notes for adults.

There may be an important role in home care including terminal care and later bereavement counselling.

3.10 Management of specific problems

The following specific conditions are indications for referral to, or discussion with, the staff at the Specialist CF Centre.

- Prolonged exacerbations (>2-3 weeks) not responding to conventional treatment – particularly in patients infected with *B. cepacia*, MRSA, multiresistant *P. aeruginosa*, other multiresistant organisms, non-tuberculous mycobacteria, *Stenotrophomonas maltophilia*
- Deterioration in pulmonary function not responding to treatment
- Pneumothorax
- Significant haemoptysis (>20 ml)
- Allergic bronchopulmonary aspergillosis (ABPA)
- Significant or persistent atelectasis
- Respiratory or cardiac failure
- Meconium ileus
- Intussusception and acute surgical procedures
- Elective major surgical procedures
- Gastrointestinal bleeding (usually variceal)
- Complications of liver disease and gall bladder disease
- Jaundice
- Insertion of totally implantable venous access devices
- Formation of gastrostomies
- Significant or unexplained weight loss or failure to thrive
- Onset of glucose intolerance
- Vasculitis
- Arthropathy
- Pregnancy
- Serious psychosocial problems

- Lung/heart-lung transplantation assessment
- Fertility issues
- Osteoporosis
- Renal disease
- Hearing and ENT problems related to CF or its treatment

3.11 Management of advanced cystic fibrosis

All aspects of treatment should be reviewed.

- Nasogastric or gastrostomy feeding will usually be required to maintain and improve nutritional status.
- Management of cardiorespiratory failure: this must include assessment for oxygen therapy - daytime, nighttime and mobility. Recommendations should include an effective delivery system to match the patient's lifestyle and lung disease.
- Ventilatory support: intermittent positive pressure ventilation may be appropriate under some acute circumstances but not to prolong the process of dying. Non-invasive ventilation may be appropriate in some patients as a bridge to transplantation.
- A transplant (heart-lung or double lung) should be considered in any patient with very limited life expectancy (e.g. 2 years as judged by the rate of decline of respiratory function) and severely impaired quality of life in spite of optimal medical therapy. Not all patients wish for or are suitable for transplantation. Initial assessment should be undertaken at the Specialist CF Centre providing full or shared care and, at an early stage, would include a full psychosocial assessment. The final assessment is by the staff of the transplant centre. It is important that patients on the transplant waiting list are maintained in as good a nutritional and physical state as possible. Expert advice should be offered and readily available to assist this decision.
- Care of the dying may be given at home or in hospital. At an early stage those involved in this aspect of care should be clearly identified. This is particularly necessary for patients on the transplant waiting list. All involved should realise that this phase of care has been reached. Symptom relief is paramount. Appropriate support should be provided for patients and families, with liaison with primary health care teams and palliative health care teams. Bereaved families will usually require on-going support.
- Psychological support should be available throughout for the patient and family.

4. CONTRACTS AND DELIVERY OF SPECIALIST CARE

This document is intended to provide a framework for commissioning of CF services to ensure there is a uniformly high standard of care available throughout the country. To achieve the best possible health, quality of life and survival, CF care must be of the highest standard whether at a Specialist CF Centre or a local paediatric CF Clinic. Shared care can work for children but due to the variable interpretation of "shared care" it must be strictly defined and the details agreed between the Specialist CF Centre and the local CF Clinic.

In the context of increased life expectancy and the greater complexity of adult care and also the increasing numbers of adults, the service for them at local level is usually not sufficiently developed for shared care to be an acceptable option.

The Cystic Fibrosis Trust intends to use the present document as a basis for clinic accreditation and to highlight the standards required and also to draw them to the attention of Commissioning Authorities.

4.1 Essential services at all Specialist CF Centres and CF Clinics

- Adequate numbers of medical and paramedical staff with expertise [Section 2.5]. It is particularly important that there is adequate staff for the community [Section 3.8].
- Designated outpatient facilities and sessions that allow sufficient time in a suitable environment for patients and families to discuss problems with the doctor and other members of the care team [Section 2.3].
- Inpatient facilities should always be available when needed. They should include single cubicles for reasons of cross-infection, as there are risks to the CF patient, to others with CF and other ward patients. The burden of nursing care and physiotherapy should not fall on families requiring respite from daily care [Section 2.4].
- Clinical and laboratory investigations and genetic counselling relevant to the care of cystic fibrosis [Section 2.1].
- Other specialist services with experience in management of CF will be required for the provision of adequate care of CF patients [Section 2.1].
- Access to a transplantation programme, involving assessment, heart-lung or lung transplantation, liver transplantation and comprehensive postoperative care [section 2.1 viii and Section 3.11].
- Provision of adequate staff and specific equipment for treatment in the community [Section 3.8].
- Identification of resources for the provision of shared care [Section 1.13].
- Provision of adequate General Practitioner support [Section 3.9].

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APPENDIX A

Reports of Committees and Working Groups on Cystic Fibrosis

British Paediatric Association. Cystic Fibrosis in the United Kingdom 1977-85; an improving picture. Report of a working party. *BMJ* 1988; 297:1599-1602.

[First meeting held in 1982. Recommended the UK Cystic Fibrosis Survey]

Report to the British Thoracic Society on organisation of care for adult patients with cystic fibrosis. 1988.

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Clinical Guidelines for Cystic Fibrosis Care. Recommendations of a Working Group of the Cystic Fibrosis Trust, British Paediatric Association, British Thoracic Society.

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[A summary of the 1996 Clinical Guidelines for Cystic Fibrosis Care]

Clinical Practice Guidelines for Cystic Fibrosis. US Cystic Fibrosis Foundation. 1997.

Cystic Fibrosis Foundation Guidelines for Patient Services, Evaluation, and Monitoring in Cystic Fibrosis Centers. *Am J Dis Child* 1990; 144:1311-1312.

A Statement on *Burkholderia cepacia*. Cystic Fibrosis Trust's Infection Control Group. Cystic Fibrosis Trust, 1999.

[Up to date guidelines for patients, families and professionals]

Antibiotic Treatment for Cystic Fibrosis. Cystic Fibrosis Trust's Antibiotic Group. Cystic Fibrosis Trust, 2000.

[Detailed review of antibiotic treatment primarily written for professionals]

Pseudomonas aeruginosa infection in people with cystic fibrosis. Suggestions for prevention and infection control. Cystic Fibrosis Trust's Control of Infection Group. Cystic Fibrosis Trust, 2001.

[Review of cross-infection problems primarily for professionals]

APPENDIX B

Grading scheme for recommendations in the Standards for Clinical Care of Children and Adults with Cystic Fibrosis

The criteria for the grading of recommendations in this document are based upon a paper by Petrie et al published on behalf of the Scottish Intercollegiate Guidelines Network.

Levels of evidence

Level *Type of evidence (based on AHCP, 1992)*

- I a Evidence obtained from meta-analysis of randomised controlled trials
 - I b Evidence obtained from at least one randomised controlled trial
 - II a Evidence obtained from at least one well designed controlled study without randomisation
 - II b Evidence for at least one other type of quasi-experimental study
 - III Evidence obtained from well-designed non-experimental descriptive studies, such as comparative studies, correlation studies and case control studies
 - IV Evidence obtained from expert committee reports or opinions and/or clinical experience of respected authorities
-

Grading of recommendations

Grade *Type of recommendation (based on AHCP, 1992)*

- A (levels I a, I b) Requires at least one randomised controlled trial as part of the body of literature of overall good quality and consistency addressing the specific recommendation
 - B (levels I a, I b, III) Requires availability of well conducted clinical studies but no randomised clinical trials on the topic of the recommendation
 - C (Level IV) Requires evidence from expert committee reports or opinions and/or clinical experience of respected authorities. Indicates absence of directly applicable studies of good quality
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Petrie GJ, Barnwell E, Grimshaw J, on behalf of the Scottish Intercollegiate Guidelines Network. *Clinical guidelines: criteria for appraisal for national use*. Edinburgh: Royal College of Physicians, 1995.

Agency for Health Care Policy and Research. *Acute pain management, operative or medical procedures and trauma 92-0032. Clinical practice guidelines*. Rockville, Maryland, USA: Agency for Healthcare Policy and Research Publications, 1992.

APPENDIX C

Summary of National Consensus Standards for the Nursing Management of Cystic Fibrosis

UK Cystic Fibrosis Nurse Specialist Group

These consensus standards were compiled by cystic fibrosis clinical nurse specialists (paediatrics and adults) working in the community and the hospital and based in regions around the UK. It is intended that by referring to these consensus standards all nurses involved in the care of cystic fibrosis will provide an optimum level of care for patients and their families.

Thanks go to all members of the working party who helped compile this document. We are also grateful for the comments and suggestions from parents, patients and other professionals who reviewed this document.

This is a summary document, the full document is available from the Cystic Fibrosis Trust or the UK Cystic Fibrosis Nurse Specialist Group.

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Continued overleaf

Scheme used for grading strength of evidence and recommendations¹

Levels of Evidence

Level I	Randomised controlled trials, meta-analysis or systematic reviews
Level II	Cohort or case controlled studies
Level III	Uncontrolled studies or consensus

Recommendations

*	Based on Level I evidence
**	Based on Level II or extrapolated recommendations from Level I
***	Based on Level II or extrapolated recommendation from level I or II

I. Introduction

Caring for people with cystic fibrosis (CF) can be complex and demanding and nurses finding themselves in this situation must ensure that they liaise closely with the nearest Specialist CF Centre. This document is offered as a guideline to nurses, both specialist and general, who care for people with CF, although throughout the text reference is made to the nurse specialist. This document is not designed to be a textbook on CF or to be used as a detailed care plan or protocol, rather as a guide only. Nursing is a partnership between the patients, carers and nurses, if the suggestions given are used within the parameters of advocacy, clinical management, education, support, advice and research, a holistic plan of care can be made. Although this document does not follow the strict criteria required for guidelines¹, it does offer examples of evidence with some recommendations made on the basis of these levels.

I.1 Specialist care and the multi-disciplinary team

Advances in medical management over the last few years have improved both quality of life and longevity for people with cystic fibrosis², due to the complexity of the disease it is widely recognised that people with CF should be cared for using a multi-disciplinary team approach. Cystic fibrosis teams in the UK include doctors, nurse specialists, dietitians, physiotherapists, psychologists and/or social workers. In addition it is recommended that these teams be supported by staff such as respiratory function technicians and ward nurses skilled in caring for people with cystic fibrosis³. The multi-disciplinary team works very closely together to ensure a holistic approach to the care of their patients. Good communication is paramount and the team must allow time to regularly meet. Nurses working in areas where there is no opportunity to work within a team must communicate with the nearest Specialist CF Centre, both for their own support and to assure optimum care for their patients⁴.

I.2 The cystic fibrosis clinical nurse specialist

The cystic fibrosis nurse specialist has a five-fold responsibility to patients and their families and the staff that are involved in the care of those patients. These areas include advocacy, clinical management, advice and support, education, research and management^{5,6,7}.

- *Advocacy:* There are many demands made on both the patient and the CF team therefore patient and family advocacy is the most important role for the nurse specialist. Patient wellbeing and satisfaction with care are paramount and successful advocacy can ensure this⁷.
- *Clinical management:* The nurse specialist must take part in decision making and monitoring of care. In addition to the practical, day to day care that the nurse specialist offers, their responsibility is to be aware of all treatment modalities used in the management of CF and to ensure that each patient receives optimum care for their individual needs. The role of the nurse specialist is also to act as a link between patient and family, community services and the hospital multi-disciplinary team.
- *Support and advice:* Both patients and their families and professional colleagues will benefit

from the support, liaison and advice available from the nurse specialist⁸. This can be as a consistent carer, counsellor or as a confidante. Problems are often resolved more easily if pathways of communication are well established for all parties⁹.

- *Education for patient, families and carers:* There are many treatment regimens that have to be learnt throughout the patient's life. Successful teaching of the patient and all concerned (parent, carer, school, work colleagues) and their understanding of the disease process, will ensure that treatment is carried out safely and effectively and that issues surrounding adherence to treatment can be more successfully dealt with.
- *Education, research and management:* Nurse specialists are responsible for developing their own professional practice through reflection, participation on post registration courses and attendance at conferences and meetings. This also ensures that they keep up to date with new advances in treatment and new developments in the world of CF research enhancing their own research-based practice^{9,10}.

2. Recommendations for best practice

2.1 Diagnosis

Diagnosis of a life limiting disease can be likened to bereavement and needs to be handled with honesty and sensitivity by skilled personnel. A diagnosis of CF may be made at any age and although most patients are diagnosed within the first year of life it is not unusual for adults, especially males, to be diagnosed later on in life. When clinical evidence leads to suspicion of CF diagnosis can be made using a number of methods¹¹:

- Diagnosis should be carried out in a Specialist CF Centre or CF Clinic experienced in the techniques being used and if CF is confirmed immediate referral must be made to a CF team¹².
- The nurse specialist will ensure that pre-diagnosis support and counselling is available¹³.
- The nurse specialist should be present when the diagnosis is being given either to a child and family or to an adult¹⁴.
- The nurse specialist will help to determine the appropriate timing (to avoid inappropriate referral) for the introduction of information regarding¹⁴:
 - Contact numbers and availability of the CF team.
 - Involvement and liaison with the primary health care team.
 - Support agencies available, both local and national.
 - Government support.
- The specialist nurse will offer continued psychosocial advice, support and counselling to the patient and their family with particular reference to:
 - Continued education about the disease.
 - The genetic implications to the immediate and extended family.
 - Expectations of care and day-to-day treatment regimens.
- The nurse specialist will be available to offer advice, education and support to the patient, their families/carers and all staff involved⁹.

2.2 Inpatient care

- Ward nurses will have access to a CF nurse specialist.
- Each patient will be cared for or supported by specialist nurses who have a knowledge and experience of CF¹⁵, the disease process and the clinical and psychological outcomes, with specialist knowledge of:
 - psychological issues - living with a life limiting disorder

- issues surrounding diagnosis
 - nutritional requirements
 - enteral feeding
 - CF related diabetes
 - intravenous therapy
 - respiratory complications and support
 - nebuliser therapy
 - care of indwelling venous devices
 - infection control
 - osteoporosis
 - liver disease
 - terminal care and symptom management
- Nurses caring for patients with CF and supported by a nurse specialist must assess, plan, implement and monitor care according to the needs of each patient at every admission, taking into account the variability of the disease.
 - The nurse specialist will ensure that each patient has access to all members of the multi-disciplinary team as often as their needs dictate.
 - The nurse specialist will ensure a partnership in care with the patient and their family/carers¹⁶.
 - The nurse specialist will be available to offer advice, education and support to the patient, their families/carers and all staff involved¹⁷.
 - The nurse specialist, in liaison with ward staff, will ensure that discharge planning needs are met with special reference to the GP, the primary health care team, and the shared care hospital and school (where appropriate).

2.3 Outpatient care

- The nurse specialist will ensure that at every routine appointment each patient has access to a¹²:
 - doctor
 - nurse specialist
 - social worker and/or psychologist
 - physiotherapist
 - dietitian
- The nurse specialist will ensure that at every routine appointment each patient has measured (as indicated)¹²:
 - lung function
 - oxygen saturation

- weight and height
- sputum or cough swab for microbiology
- The nurse specialist, in conjunction with appropriate members of the CF team, will be responsible for specific aspects of clinical management such as¹²:
 - nutritional needs
 - nebuliser therapy
 - care of indwelling venous devices
 - self administration of intravenous therapy
 - enteral feeding
 - oxygen therapy
 - psychosocial and emotional support
- The nurse specialist will liaise with patient and family on issues surrounding cross-infection and infection control¹⁸.
- The nurse specialist will ensure that all patients receive a comprehensive Annual Review in a recognised Specialist CF Centre or CF Clinic¹⁹.
- The nurse specialist will liaise with the CF team, the primary health care team, shared care hospital, work and school (where appropriate)²⁰.
- The nurse specialist will be available to offer advice, education and support to the patient, their families/carers and all staff involved⁹.

2.4 Community

The nurse specialist will continue to support the patient in the community to ensure an equality of care to that offered in hospital.

- Each patient will have access to a CF nurse specialist¹².
- Nurses caring for patients with CF in the community will be supported by a nurse specialist who has knowledge and experience of CF. This will include the disease process, the clinical and psychological outcomes of the treatment modalities to ensure safe and effective care meeting the minimum nursing standards of care²¹.
- The nurse specialist will ensure that there is close liaison between the CF team, the GP, the primary health care team, the shared care hospital and work or school²¹.

- The nurse specialist will ensure that advice and support is available for the management of complications associated with CF such as¹²:
 - CF related diabetes
 - pneumothorax
 - haemoptysis
 - pain
 - arthropathy
 - breathlessness
 - liver disease
 - osteoporosis
- The nurse specialist, in liaison with colleagues, will support and co-ordinate:
 - home supplementary nutrition
 - home intravenous therapy
 - respiratory support
- The nurse specialist will assess and monitor the response to treatments carried out in the community²¹.
- The nurse specialist, in liaison with colleagues, will offer psychosocial and emotional support surrounding issues of living with a genetic, life limiting disease to families and patients of all ages.
- The nurse specialist will be available to offer advice, education and support to the patient, their families/carers and all staff involved⁹.

2.5 Issues surrounding adolescence and adulthood

Adolescence and young adulthood is a particularly difficult time for those having to deal with both the psychological and practical burden of cystic fibrosis. Many treatment related issues can become a problem and the nurse specialist must be alert to problems, especially those surrounding adherence and conflict between teenager and parents²².

- The nurse specialist will be instrumental in promoting self-care and responsibility in the young adult and offering support and advice to the parents²³.
- The nurse specialist will liaise with schools and colleges to support continuing education⁸.
- The nurse specialist, in liaison with colleagues, will ensure that adolescents receive appropriate knowledge regarding issues such as²⁴:
 - fertility
 - pregnancy
 - contraception/safe sex
 - cross-infection
 - further education/employment
 - smoking/substance abuse

- The nurse specialist should use experience and knowledge to advise on the appropriate time (however long) for transition and transfer to adult care for each patient. This will include²⁵:
 - Liaison and communication between the paediatric and adult Specialist CF Centres about all aspects of care, e.g. level of knowledge of CF at time of transfer.
 - The co-ordination of joint transition clinics, parallel care and visits to the adult Specialist CF Centre where appropriate.
 - Provision of adequate information and ongoing support for patients and parents during the transition period.
- The nurse specialist will allow opportunity for discussion between the patient, parents and CF team members when problems arise²⁶.
- The nurse specialist will be available to offer advice, education and support to the patient, their families/carers and all staff involved⁹.

2.6 Advanced cystic fibrosis

Although children still die, CF is becoming a disease of adulthood. However, pre-terminal grief at diagnosis onwards must be acknowledged throughout the patient's life. Treatment choices - the dilemma of aggressive management versus palliative care, is difficult for both the families and staff. The pre-terminal and terminal stages must be handled with sensitivity and compassion together with sound clinical judgement and involve the patient, their family and staff.

- The specialist nurse will recognise the patient's complexity of care and changing needs and will be able to offer support to the patient and their family to help them come to terms with and adapt to the changes²⁷.
- The nurse specialist must be aware of the concerns involved with heart/lung transplantation and liver transplantation as a treatment option, these include²⁸:
 - The stress of waiting.
 - The complications of transplantation.
 - The loss of a recognisable end point.
 - Death with dignity.
- The nurse specialist will advocate on the patient's and family's behalf. This will involve accepting new ways of coping, recognising denial, respecting their wishes and decisions about treatment, and allowing discussion around issues of dying²⁹.
- The nurse specialist will ensure that the patient and family receive sufficient knowledge to make informed decisions about treatment and where possible allow flexibility of choice. These treatment decisions will include^{27,30}:
 - Terminal care - hospital or home (within available resources).
 - Continuation of enteral feeding and intravenous therapy.
 - The options for respiratory support.

- Pain management and symptom control.
 - Dealing with complications such as haemoptysis, liver disease, pneumothorax, transplantation and management of cardiorespiratory failure.
 - Bereavement management.
- The nurse specialist will be able to recognise and support the healthcare team - both hospital and community based - in accepting the outcome²⁷.
 - The nurse specialist will involve other support and specialist agencies where appropriate.
 - The nurse specialist will maintain ongoing liaison and communication with the primary healthcare team.
 - The nurse specialist, in liaison with colleagues will ensure that appropriate bereavement support is offered to the family both in the terminal stages and after death.

Caring for the dying patient is complex and stressful, however, the nurse specialist should recognise that with support from the CF team and the available support network it can become a rewarding and challenging experience.

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APPENDIX D

Summary of the Clinical Guidelines for the Physiotherapy Management of Cystic Fibrosis. Recommendations of a Working Group. October 2000.

Association of Chartered Physiotherapists in Cystic Fibrosis (ACPCF)

These clinical guidelines were produced on behalf of the Association of Chartered Physiotherapists in Cystic Fibrosis. They are intended as a guideline for all physiotherapists working in paediatric and adult Specialist CF Centres, CF Clinics, and in the community, who are involved in the care and treatment of patients throughout the UK.

The Association would like to thank the Working Party and all its members for their work in the preparation of this document. It would also like to acknowledge the contributions made by other members of the CF multi-disciplinary teams working in cystic fibrosis.

This is a summary document of the *Clinical Guidelines for the Physiotherapy Management of Cystic Fibrosis*, the recommendations of a working group, produced by the Association of Chartered Physiotherapists in Cystic Fibrosis (ACPCF). The full document is available from the Cystic Fibrosis Trust, 11 London Road, Bromley, Kent. BR 1 1BY.

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Grading scheme for recommendations in the guidelines

Recommendations were formulated, and where possible, were graded A, B or C according to the level of evidence ^{1,2}.

The criteria for the grading of recommendations in the guidelines are based on the following by Petrie et al and published on behalf of the Scottish Intercollegiate Guidelines Network ^{1,2}.

Grading of recommendations

<i>Grade</i>	<i>Type of recommendations</i>
A (levels Ia, Ib)	Requires at least one randomised controlled trial as part of the body of literature of overall good quality and consistency addressing the specific recommendation
B (levels IIa, IIb, III)	Requires availability of well conducted clinical studies but no randomised clinical trials on the topic of recommendation
C (level IV)	Requires evidence from expert committee reports or opinion and/or clinical experience of respected authorities. Indicates absence of directly applicable studies of good quality.

Levels of Evidence

<i>Level</i>	<i>Type of evidence</i>
Ia	Evidence obtained from meta-analysis of randomised controlled trials
Ib	Evidence obtained from at least one randomised controlled trial
IIa	Evidence obtained from at least one well designed controlled study without randomisation
IIb	Evidence obtained from at least one other type of well designed quasi-experimental study
III	Evidence obtained from well designed non-experimental descriptive studies, such as comparative studies, correlation studies and case control studies
IV	Evidence obtained from expert committee reports or opinions and/or clinical experience of respected authorities

Introduction

Pulmonary dysfunction is the main contributory factor to the morbidity and mortality associated with CF. Bronchial damage results from persistent infection and inflammation within the airways. Physiotherapy is an integral part of the management of this disorder. Airway clearance techniques aim to reduce airway obstruction, improve ventilation and delay the progression of the pulmonary disease process. Physiotherapy treatments can be tiring and time consuming, therefore when formulating treatment programmes attention should be paid to minimising the treatment related burden on the patient and family. With improved longevity and an increasing population of adults with CF attention must be given to independence and quality of life.

Provision of Physiotherapy Services

All centres should aim to provide:

- Treatment by qualified physiotherapy staff throughout a 24 hour period as required
- Protected physiotherapy time for outpatient clinics
- Administrative support
- An opportunity for continuing professional development education and training
- Provision of, or access to domiciliary or community physiotherapy when required.

Specialist CF centres should aim to provide

- Physiotherapy staffing levels in a ratio of approximately two whole time equivalent physiotherapist per 50 patients. These physiotherapists should have specific expertise in the care of CF patients³ (C, IV)
- Physiotherapists with knowledge of and working to the published *Standards for respiratory care*⁴ of the Association of Chartered Physiotherapists in Respiratory Care (ACPRC)
- An open access service (C, IV)
- Specialist advice and support to local/shared care centres and community teams
- A service to satellite/peripheral clinics
- Exercise testing and exercise facilities
- Safe and appropriate equipment for patient and therapist use
- An age appropriate service ensuring smooth transition from paediatric to adult care
- A resource for education and training e.g. lectures, courses, seminars
- An opportunity for clinical research and evaluation of new techniques and equipment.

Physiotherapy Practice

General

Documentation, consent and confidentiality should follow the *Standards of Physiotherapy Practice*⁵. (C, IV)

Professional

- The physiotherapist should have knowledge of the *ACPRC Standards for Respiratory Care*⁴ (C, IV)
- The physiotherapist should have a special interest in CF and must maintain an awareness of evidence based practice in respiratory care
- The physiotherapist should develop his/her role within the CF team as appropriate.

Communication

- Physiotherapists should communicate effectively within the multi-disciplinary framework in order to provide optimum patient care
- Physiotherapists should communicate between specialist, local and community services
- They should be available to offer education and support to patients and their families/carers
- The physiotherapist should attend appropriate meetings and case conferences.

Specific

At diagnosis

- On confirmation of diagnosis the family should be seen by an experienced CF physiotherapist and a regimen of treatment and education commenced in both symptomatic and asymptomatic patients
- Evidence to date suggests that regular physiotherapy may help to maintain pulmonary function in the short term^{6,7} (B, IIa)
- Accurate assessment accompanied by careful consideration of other influential factors such as age, psychosocial needs, cultural background and likely adherence to treatment will facilitate the formulation of an effective physiotherapy regimen
- Patients should be monitored at regular intervals and treatment regimens adapted according to their changing needs. The frequency of visits either to the specialist or local centre will be dictated by health status
- Patients and carers should be instructed in appropriate techniques and taught to recognise deteriorating symptoms.

Outpatients

- Patients should be reviewed at a minimum of 3 month intervals and more frequently in infants, children and patients with complications
- All patients with cystic fibrosis should undergo Annual Review at a Specialist CF Centre or CF Clinic³. (C, IV) This should include a thorough assessment and review by the specialist physiotherapist of the physiotherapy regimen including airway clearance, exercise tolerance, inhalation therapy and any relevant domiciliary equipment.

Domiciliary/Community

- Specific policies should be in place relating to health and safety
- Physiotherapy management in the home should be appropriate and effective
- Equipment provided should be safe and suitable.

Inpatients

- All inpatients should be referred to the physiotherapist and assessed within 24 hours of admission
- Treatment should be monitored and evaluated daily
- Exercise should be continued as appropriate.

Infection Control

- The physiotherapist should be aware of the respiratory pathogens, their transmissibility and the potential for cross-infection⁸. (C, IV)
- The physiotherapist should ensure the risk of cross-infection between patients and from physiotherapy equipment is minimal.

Airway Clearance Techniques (ACT)

- Techniques employed to enhance the clearance of airway secretions include:
 - Active Cycle of Breathing Techniques (ACBT)
 - Autogenic Drainage (AD)
 - Exercise
 - High Frequency Chest Wall Compression
 - High Frequency Oscillation
 - Oscillatory Positive Expiratory Pressure (Flutter/R-C Cornet)
 - Positive Expiratory Pressure
 - Postural Drainage and Percussion.

- Existing evidence to date does not show any one of these techniques to be superior to all others⁹. (A, Ia) Choice of treatment should be dictated by individual requirements
- The therapist should at all times be aware of the treatment related burden that daily therapies place on patients and their families. Unreasonable prescription of time consuming therapies will have a negative effect on adherence to treatment. The therapist should at all times work with the patient and their family/carers, in order to formulate an effective yet manageable treatment programme.

Exercise

- The importance of including exercise as part of the therapeutic regimen is well recognised¹⁰⁻¹² (B, IIa)
- Subjective measures of perceived exertion, breathlessness or fatigue may be useful complementary measures when performing exercise tests or in order to gauge levels of activity during exercise programmes¹¹. (B, IIa)

Exercise Testing

- Exercise testing is useful to assess functional capacity, determine the impact of disease process on everyday function, to monitor progress and allow safe and effective exercise recommendation¹³ (B, IIa)
- Exercise testing in CF on an annual basis is a recommendation of the clinical guidelines working party³ (C, IV)
- The choice of test used will be dependent on the indication for the test, information required, and the facilities available.

Exercise programmes

- Therapeutic benefits gained from exercise programmes in patients with CF include:
 - cardiorespiratory fitness¹⁰ (B IIa)
 - increased ventilatory endurance¹⁴ (B, IIa)
 - decreased breathlessness¹¹ (B, IIa)
 - enhanced sputum clearance¹² (B, IIa)
 - improved body image through increased muscle mass and muscle strength¹⁵ (B, IIa)
 - improved morale¹¹ (B, IIa)
 - enhanced quality of life¹⁶ (B, IIa)
- Exercise programmes should be tailored to the individual taking into consideration, disease severity, level of fitness and the patient's preferences to exercise activities
- Supplementary oxygen during exercise may be necessary to prevent exercise induced desaturation and relieve symptoms of breathlessness. Exercise performance may also be improved¹⁷. (B, IIb)

Management of specific problems

- Complications and the natural progression of the disease process may require modification of treatment regimens or additional therapeutic strategies
- Complications such as liver disease, CF related diabetes, pneumothorax, haemoptysis may occur, particularly as disease severity increases. Modification of exercise regimens and airway clearance techniques are likely to be necessary
- Arthropathy either associated with bronchopulmonary infection or periodic arthritis will require appropriate management in terms of maintaining joint mobility and function
- Pulmonary complications are the greatest cause of post-operative morbidity and mortality. CF patients undergoing surgical procedures will require additional physiotherapy in terms of patient preparation, possible lavage during intubation and intensive post-operative management
- Patients who may potentially benefit from bi-lateral sequential lung or heart-lung transplantation should be referred to a regional transplant centre. Thorough assessment including a full review of the physiotherapy regimen should be undertaken prior to acceptance on to a transplant list. It is essential that patients awaiting transplantation maintain mobility and fitness as far as possible. Post transplant patients should be encouraged to lead active life styles and obtain maximum fitness. Patients should be taught how to review their chest condition and a regimen of airway clearance may still be necessary in some patients
- As the disease progresses, an increasing loss of independence is inevitable. Home care and domiciliary physiotherapy input may be required. In the terminal stages of illness significant modification of treatment will be necessary. Although withdrawal of treatment may eventually be appropriate, contact with a physiotherapist should be maintained to offer support and advice to the patient and family.

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APPENDIX E

Dietetic Management of Cystic Fibrosis. Summary of Recommendations of the UK Cystic Fibrosis Dietitians' Interest Group

- A state registered dietitian at Senior 1 level or above should be responsible for the dietetic care of patients with CF. Care by lower grades of staff should be undertaken with supervision.
- Dietitians responsible for the care of CF should practice in accordance with the National Professional Standards for Dietitians in Healthcare (BDA 1998).
- Clinical dietetic practice in CF should reflect current research, clinical guidelines and consensus views.
- The dietitian will agree with the CF care team relevant measures of nutritional status and guidelines for intervention based on these measures.
- The dietitian will explain the principles of nutritional management to the patient and relevant carers within one week of confirmed diagnosis. These principles include nutritional requirements, provision of requirements, pancreatic enzyme therapy, vitamin therapy. This will be supported by appropriate literature and will be followed up within two months.
- An initial nutritional assessment and appropriate intervention will be carried out within one week of a confirmed diagnosis.
- The dietitian will provide on-going nutritional assessment, advice, and support relevant to the needs of the individual. In particular, this must take account of the numerous situations which may require more intensive dietetic support.
- This will require both in-patient and out-patient work. Some examples of such situations are early infancy, adolescence, pregnancy, transplantation, CF related diabetes, eating disorders, enteral feeding.
- An Annual Review of nutritional status will be carried out and appropriate intervention planned.
- For paediatric patients dietitians should ensure that self-knowledge of nutritional care is developed as a component of transition.
- The dietitian will carry out regular audit of nutritional status and dietetic interventions within the CF clinic population.
- In centres offering shared care the model of care, including the dietetic component, be agreed and resourced ensuring that these recommendations are achieved for all patients.
- The dietitian will act as a resource for the training, education and support of others involved in the care of CF.

British Dietetic Association. National Professional Standards for Dietitians Practicing in Healthcare 1998.

APPENDIX F

Consensus Statement on the Provision of Psychological Services within CF Teams

Prepared by the British Psychosocial Professionals Group (BPPG/CF)

1.0 General Issues for Psychological Therapists

Psychological therapists working with CF patients should

- be registered with their governing professional body (e.g. British Psychological Society, British Association of Counsellors, Royal College of Psychiatry)
- have specialist knowledge of how chronic medical conditions impact on children, adults and their families and of CF itself.

Accepted levels of service provision should be in line with other Professions Allied to Medicine (PAMs) involved in CF management, i.e. 4 sessions per 50 patients on full-care.

2.0 Consensus Standards for Clinical Psychologists Working in CF Teams

Many CF centres and clinics now either employ or have direct access to clinical psychology services. Where this is the case, then the following activities are considered intrinsic to their role.

- Conduct psychological assessments as part of Annual Reviews.
- Run clinical psychology services in parallel with CF clinics.
- Attend CF team meetings.

Furthermore, there is an expectation that the clinical psychologist would

- Act as a 'gatekeeper' for the onward referral of patients to mental health services and/or other relevant agencies (e.g. liaison psychiatry, palliative care teams)
- Provide opinion on cases seen by other members of the CF team
- Support other CF team members in psychological aspects of casework
- Maintain client confidentiality
- Contribute to psychological research, either individually or in conjunction with other CF team members.

Continued overleaf

3.0 Establishing Best Practice Guidelines

During the next 12 months, the BPPG/CF is aiming to establish guidelines for best psychological practice in key areas of CF management. These are: adherence to treatment, QoL, management of procedural distress, management of feeding behaviour difficulties and end-of-life issues.

Further details about the British Psychosocial Professionals Group (BPPG/CF) can be obtained from:

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Notes



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