Cystic Fibrosis our focus

Standards for the Clinical Care of Children and Adults with cystic fibrosis in the UK

Second edition. December 2011

Fighting for a Life Unlimited

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Summary of principles and standards of cystic fibrosis care

A. The diagnosis of CF should not be delayed but must be handled sensitively, and be followed by education of the parents / carers and patient (if old enough).

- Pathways must be in place for a referral within 24 hours from the newborn screening laboratory to the relevant Specialist CF Centre which must be robust so that cases are not missed and all positive results processed appropriately.
- Specialist CF Centres and their networks must have a newborn screening care pathway in place for contacting the parents in person and making the diagnosis within 5 working days. Consideration must be given to reducing the waiting time and stress for the parents; the sweat test and clinical assessment should be performed no later than the day after informing the parents that CF is suspected.
- Diagnosis should be confirmed by a sweat test and genetic mutation analysis on a separate sample.
- Infants presenting with meconium ileus should be treated as if they have CF until proven otherwise, with an early referral to the CF team.
- A diagnostic pathway should be in place for older children and adults. All patients (outside the newborn period) diagnosed symptomatically must be referred to a Specialist CF Centre for ongoing care.
- Education of the parents / carers and/or older patients should be provided by the full multidisciplinary team within 7 days, including written information and direction to reliable websites.

B. All patients must be under the direct supervision with regular follow-up from an adequately resourced designated Specialist CF Centre, sometimes in partnership with a Network CF Clinic.

Specialist CF Centre

- A Specialist CF Centre has a minimum of 100 adults or children. In exceptional circumstances, the geographical location of a Specialist CF Centre may mean that the number of patients is less, although it should not be less than 50.
- Parents should be aware of their options and know they are able to choose full care from a Specialist CF Centre if they wish. Due to the increasing complexity of CF in adulthood, full care should be delivered by a Specialist CF Centre. In exceptional circumstances adults may receive some care through an Outreach Clinic.
- The centre must participate in the Cystic Fibrosis Trust / British Thoracic Society and British Paediatric Respiratory Society programme of peer review.
- The centre should have access to diagnostic facilities (e.g. sweat testing, lung function, bronchoscopy, and radiology) and microbiology services fulfilling the 2010 CF Trust laboratory standards for processing microbiological samples.
- All CF patients must have access to their CF centres for routine and emergency care and advice.
- Patients should be reviewed regularly with a frequency appropriate to their individual needs, but routine appointments for a stable patient should be every 2-3 months depending on the severity of their disease. Newly diagnosed infants should be seen more frequently (initially weekly).
- There must be sufficient capacity in clinics for outpatients to be seen urgently with sufficient space to ensure optimal infection control.
- There must be sufficient inpatient beds so that patients do not wait unnecessarily for an admission. Beds in a ward suitable for CF care (adhering to cross-infection policies) should always be available for an emergency admission, and there needs to be capacity to ensure elective and urgent admissions can be managed appropriately.
- An urgent course of treatment should be implemented within a maximum of 24 hours of the decision being made, and a non-urgent admission within 7 working days of the planned date.
- Hospital ward nursing staff must have sufficient knowledge and experience to provide CF care.
- Patients should be seen by a consultant with specialist CF knowledge at least twice a week when a hospital inpatient; they will be kept updated every day by the rest of the team.

Network CF Clinic

- Care delivered by a Network CF Clinic should be to the same standard as that delivered by the Specialist CF Centre.
- If under the care of a Network CF Clinic, patients must be seen at the Specialist Centre for annual review, (unless the Network CF Clinic can provide all recommended clinical reviews and investigations, in which case it may be done jointly by both teams in the local centre).
- Patients must be seen at least twice a year by the full Specialist Centre multidisciplinary team (MDT), which may take place at either the Network or Specialist hospital. It may be appropriate for the patient to be seen more often at the Specialist Centre, depending on the severity of the disease and level of expertise at the Network CF Clinic. Patients in network care who have an unstable or deteriorating condition should be reviewed promptly by the specialist CF team.
- Patients and their families should have a clear understanding of how the network functions and who is responsible for their care.

- There must be clear lines of communication between the Network team and the Specialist CF Centre at all levels.
- Shared care must be delivered as part of an agreed designated Care Network with a Service Level Agreement and Standard Operating Procedures as laid down by the Specialist CF Centre. There must be regular communication between the consultants and multidisciplinary team of the Specialist and Network CF Clinics.

C. Specialist multidisciplinary care must be delivered by a team of trained and experienced CF specialist health professionals with staffing levels appropriate to the size of the patient population.This is done as a partnership with the patient's parents/carers and/or the patient when old enough.

- The MDT will consist of: specialist consultant paediatricians or adult physicians; medical support from trainee(s); clinical nurse specialists; physiotherapists; dietitians; clinical psychologists; social workers; pharmacists; secretarial support; and database coordinator.
- Continuity of care is essential. All patients should have a named consultant even though they may be looked after by a team of consultants.
- There must be a commitment from all Specialist and Network MDT members to ongoing Continual Professional Development (CPD), demonstrated by membership of their relevant Special Interest Group, as well as attendance at relevant national and international conferences.
- There should be access to other medical and surgical specialists when appropriate, and particularly, Gastroenterology and Hepatology, Diabetes and Endocrinology, Ear Nose and Throat, Cardiothoracic and General Surgery, Specialist Anaesthesia & Pain Control, Rheumatology, Obstetrics & Gynaecology, Psychiatry, Intensive Care, and Interventional Radiology.
- All patients should have an annual review with a report written by a consultant who should discuss it with the patient/carers, and the treatment plan agreed (written copy given to patient/carers). Data should be entered onto the CF Registry as part of national audit of outcomes.

D. Measures must be in place to prevent cross-infection from other patients.

- There must be policies that involve segregating patients so that all CF patients are isolated from each other.
- Patients should not be in close contact with each other in waiting areas, e.g. CF clinics, pharmacy, radiology etc.
- Patients should not share rooms including bathrooms, toilets and social areas whilst inpatients in hospital. Preferably they should be in a cubicle with en-suite facilities.
- Arrangements must be in place for patients infected with Burkholderia cepacia complex and MRSA, for example separate clinics and appropriate inpatient segregation.

E. Treatment of airway infections are critical, so antibiotics are a key part of CF therapy – for prophylaxis, eradication therapy, long-term treatment of chronic infection, and treatment of acute exacerbations.

- All patients must have frequent (every clinic visit) microbiological surveillance of respiratory secretions (e.g. cough swab, sputum culture, induced sputum).
- All patients must have regular monitoring of lung function with spirometry (from 5-6 years of age), and oxygen saturation measurements with pulse oximetry.
- Younger children should receive oral antibiotic prophylaxis for Staphylococcus aureus; this may be considered for adults as well. Whilst this is common practice in the UK, it is not considered best practice in the USA and is still controversial.
- Attempted eradication should be carried out for first and subsequent new Pseudomonas aeruginosa infections. This should take place promptly (within 1 week) from when the microbiology result is available. This also applies to Burkholderia cepacia complex and MRSA.
- Long-term treatment of chronic Pseudomonas aeruginosa lung infections with inhaled antibiotics should be prescribed.
- Inhaled therapies must be delivered through an appropriate device.
- Acute chest exacerbations must be treated promptly with adequate doses of oral, nebulised and/or intravenous antibiotics. See Flume PA, Mogayzel PJ Jr, Robinson KA, et al; Clinical Practice Guidelines for Pulmonary Therapies Committee. Cystic fibrosis pulmonary guidelines: treatment of pulmonary exacerbations. Am J Respir Crit Care Med 2009;180:802-8.
- Higher doses and longer antibiotic courses (both oral and intravenous) are required compared to people without CF; this includes dual therapy for intravenous courses (βlactam-based antibiotic with an aminoglycoside).
- Intravenous gentamicin should be avoided so tobramycin and amikacin need to be on the hospital formularies for use in CF patients. Hospital laboratories must have the ability to measure tobramycin or amikacin blood levels, which must be measured regularly.
- CF teams must evaluate the appropriateness of

homecare intravenous treatment, for example where non-adherence or home conditions are an issue. Appropriate training to administer IV antibiotics must be given to the patient / carers and written competency checks recorded.

F. Chest physiotherapy with airway clearance techniques are a lifelong mainstay part of treatment.

- All patients should be reviewed by specialist physiotherapists in clinic and at annual review.
- Patients should receive physiotherapy treatment twice daily (including weekends), or more if necessary, when an inpatient in hospital. This may be carried out with assistance from a physiotherapist or independently if the physiotherapist has previously assessed that to be appropriate.
- Treatment with inhaled dornase alfa or hypertonic saline should be considered as an adjunct to airway clearance.

G. Nutritional support is crucial for all patients.

- Pancreatic status should be established at diagnosis through clinical assessment and measurement of faecal elastase. Pancreatic sufficient patients may need re-checking when clinically indicated. Patients with pancreatic insufficiency should receive pancreatic enzyme replacement therapy and fat-soluble vitamins; and pancreatic sufficient patients may also require fat-soluble vitamin supplementation.
- Prompt intervention to normalise nutritional status is essential, especially in the first year of life.
- All pancreatic insufficient patients should be reviewed by a specialist dietitian in every clinic to ensure optimal energy intake and dose of pancreatic enzyme therapy. Pancreatic sufficient patients will be seen if necessary.
- Growth of infants and children should be measured in every clinic, including weight, height and in infants head circumference (and plotted on appropriate charts). BMI should be monitored in all children and adults.
- If growth is faltering or weight loss identified a diagnostic review should be made. This should include assessment of dietary intake, malabsorption and glucose tolerance.
- All CF patients should be seen at annual review by a dietitian.
- Nutritional supplements may be necessary for some people and are usually given by mouth, and less often nasogastrically / nasojejunally or via a gastrostomy.
- Inpatients should have specialist CF dietetic input at least twice a week, and more frequently if appropriate.

H. Other manifestations of CF as well as complications must be recognised promptly and in some cases should be screened for, particularly liver disease and impaired glucose metabolism.

- The CF team must have experience in managing the complex problems and rarer complications that may arise in patients with CF.
- Annual screening for liver disease (with biannual ultrasound) should be carried out on all patients aged 5 years and above. Routine repeat ultrasounds may not be necessary in adults with previous normal scans.
- Early identification of impaired glucose metabolism and CF-related diabetes (CFRD) is critical; it is recommended that patients are screened annually for CFRD from 12 years and above using an oral glucose tolerance test. The diagnosis may be confirmed by the use of random blood glucose profiles or continuous glucose monitoring. Insulin therapy should be started early and is the mainstay of treatment
- It is important boys are informed about fertility issues at an appropriate age (usually 10-12 years old).
- Screening for reduced Bone Mineral Density (BMD) with dual energy x-ray absorptiometry (DXA scan) should be initiated from 10 years of age with serial measurements every 1-3 years.

I. Psychosocial support is often required and should be available.

- All patients must have access to clinical psychology and social work services (which are part of the CF team); the timing will depend on the urgency of the particular situation.
- Access to psychological support at the time of late diagnosis (e.g. adults) is essential.
- Patients should be screened annually for potential psychosocial problems by a psychologist or social worker with experience in CF.

J. Transition to adult care should be planned and managed appropriately.

- A joint paediatric and adult CF team approach is necessary for successful transition with identified coordinators from both teams and a formally agreed protocol.
- There needs to be a plan agreed by the CF team and patient / carers for transition, which is usually completed by the 17th birthday (and certainly by the 18th).
- The adolescent and carers should have the opportunity to formally meet the adult team on more than one occasion (usually in joint clinics), and view the adult facilities.

K. Transplantation, palliative and end of life care must be planned and managed appropriately.

- All CF centres should have a working relationship with one of the National Commissioning Group (NCG) designated lung transplant centres, with clear indications for referral agreed.
- Formal arrangements must be in place for the appropriate multidisciplinary continuing care of CF patients after transplantation. This may be based in the transplant centre (with regular support from the CF centre), or in the CF centre (with close liaison with the transplant centre).
- All centres should have a palliative care team readily available to help and advise on management issues.
- All people with CF should have the opportunity to discuss end of life care with their physician and other members of the MDT. Issues around advanced care planning should be patient focused and the care pathway for each patient individualised according to patients wishes; this must be clearly documented.

1. Introduction

1.1 What is cystic fibrosis?

Cystic fibrosis (CF) is the most common, life-limiting, recessively inherited disease in the UK, affecting about 9,000 people (1 in 2,500 live births). It results from mutations affecting a gene that encodes for a chloride channel called the cystic fibrosis transmembrane conductance regulator (CFTR), which is essential for the regulation of salt and water movements across cell membranes. Absent or reduced function of CFTR results in thickened secretions in organs with epithelial cell lining hence it is multi-system, although mainly affects the lungs, digestive system and vas deferens.

The airways become clogged with thick sticky mucus, which impairs the clearance of microorganisms. This leads to recurrent infection, inflammation, bronchial damage, bronchiectasis and eventually death from respiratory failure. Patients are often infected with Staphylococcus aureus and Pseudomonas aeruginosa but also by a number of other organisms, some of which are resistant to many antibiotics.

In about 85% of cases the pancreatic exocrine ducts become sufficiently blocked to cause maldigestion and intestinal malabsorption (pancreatic insufficiency). Infants may fail to thrive and older children and adults may become under-nourished. About 15% of CF babies are born with a bowel blockage (meconium ileus) and some older patients develop recurrent blockages due to distal intestinal obstruction syndrome. Appetite is often adversely affected which is a problem as there is an underlying increase in metabolic demands leading to a need for an increased energy intake.

There are a number of other complications: most males are infertile; a high proportion of older patients will develop CF-related diabetes requiring multiple daily insulin injections; chronic liver disease and portal hypertension may develop; joints can be affected (CF-arthropathy) and with age bones can be affected by reduced bone mineral density; nasal polyps and sinusitis are not uncommon. Behavioural and psychological problems that are often associated with any severe long-term medical condition may also be present.

1.2 Demographics

Cystic fibrosis mainly affects Caucasian populations. There are a number of cases in the UK amongst families from Asia (Indian subcontinent) and the Middle East; it is rare in people of Afro-Caribbean origin and other ethnic groups. The carrier rate of a CF gene mutation in the UK is 1 in 25 with an incidence of 1 in 2,500 live births. It is reported that the population in the UK is 9,027 patients. See CF Registry annual data report 2009, available at www.cftrust.org.uk/aboutcf/publications/cfregistryreports. The proportion and number who are adults has increased so over half (56%) are now 16 years

or older [CF Registry annual data report 2009]. Since October 2007, the whole of the UK has had newborn screening so that the sweat test confirmed diagnosis is now made at around 4 weeks of age. People born before that date (or abroad) may still be diagnosed following development of symptoms.

Median survival is currently 34.4 years [CF Registry annual data report 2009] and has been predicted to be at least 50 years for children born in 2000 [Dodge ERJ 2007]. This has improved dramatically over recent decades due to a number of factors, including specialist centre care, better nutritional support, and improved treatment of lung complications with aggressive use of antibiotics. However, the median age at death is currently 27 years [CF Registry annual data report 2009] and most people with CF who die each year are young adults, and occasionally some are children (only 3 in 2009) [CF Registry annual data report 2009]. Dodge JA, Lewis PA, Stanton M, Wilsher J. Cystic fibrosis mortality and survival in the UK: 1947-2003. Eur Respir J 2007;29:522-6.

1.3 Why standards of care are needed

- The CF community wishes to ensure all patients have equal access to the highest level of multidisciplinary specialist care that is adequately resourced and encompasses the latest evidence-based therapies.
- The aim is to improve quality of life and extend life expectancy.
- The standards can be used for benchmarking and form the basis for peer review and designation of CF services. This will aid the commissioning of CF services.
- It is hoped that these standards will also act as an aid for CF centres undertaking Quality Improvement programmes.

1.4 Who is this document for?

- Clinicians and other allied health professionals.
- Commissioners and others in the NHS and elsewhere who are responsible for the provision of care for people with CF.
- Parents and carers of children with CF; older children and adults with CF; and their families, to help them understand what level of care they should expect.

1.5 How this document was written

This is the 2nd edition of the Standards of Care document that was published in 2001. A revision was started by the CF Trust Standards of Care Committee in 2007, and this version formed the template for the current document. At the request of the CF Trust, Dr Ian Balfour-Lynn and Dr Susan Madge (members of the 2001 & 2007 Standards of Care Committee) wrote the 1st draft, collated comments and incorporated them into subsequent drafts. The 1st draft was reviewed by members of the CF Trust Peer Review Project Steering Group, and the 2nd draft by members of the CF Trust Medical Advisory Committee and by Directors of all the CF centres in the UK, and Chairs of all the CF Nursing and Allied Health Professional groups. Finally, the 3rd draft was ratified by the CF Trust Medical Advisory Committee who resolved any remaining issues. The 4th draft was then approved by the British Thoracic Society Specialist Advisory Group. It is therefore a consensus document that draws on best practice, and is not intended to be an evidence-based clinical guideline, hence is not referenced throughout the text. However all relevant CF Trust, British Thoracic Society, European CF Society, and US Cystic Fibrosis Foundation consensus documents and guidelines are referenced with their online links; multiple references are included in these documents. Additionally the CF Trust clinical care pathway www.cfcarepathway.com, the Department of Health Definition 10 of the National Specialised Services Definition Set 3rd edition (2010), and the Service Specifications document produced by West Midlands and South East Coast Specialised Commissioning teams are referenced. Thus the omission of references to each statement does not weaken our recommendations but is a more realistic and pragmatic approach to an area where randomized controlled trials can not always provide evidence.

Many helpful comments were received to improve the document at different stages of development. However it should be noted that whilst this is a consensus document, not all reviewers agreed with all recommendations. Thanks are due to:

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1.6 Other sources

- CF Trust Clinical Care Pathway

www.cfcarepathway.com

- CF Trust publications (factsheets, consensus documents, Registry)

www.cftrust.org.uk/aboutcf/publications/

- NHS Map of Medicine for Cystic Fibrosis

http://healthguides.mapofmedicine.com/choices/map/cystic_fibrosis1.html

- European CF Society – Standards of care for patients with cystic fibrosis: A European consensus

www.elsevier.com/framework_products/promis_misc/2005.pdf

2. Models of cf care

2.1 Models of care

Concentrating care for CF patients in specialist adult and paediatric CF centres ensures that the multidisciplinary team will see sufficient numbers of patients to be able to maintain expertise so that they treat CF patients effectively, recognise the more unusual manifestations and delay the onset of the multi-system complications associated with the condition.

Different models of care have been outlined in detail on the CF Trust clinical care pathway www.cfcarepathway.com under the heading 'Model of care for people with CF in the UK'; also available at www.cftrust.org.uk/aboutcf/cfcare/Model_of_CFCare.pdf.

- All patients with CF must have their care delivered under the direct supervision of a recognised Specialist CF Centre for treatment throughout their lives.
- The logistics of delivering care may differ for children and adults:
- Children will either receive full care from a Specialist CF Centre, or shared care within an agreed designated network.
- Parents should be aware of their options and know they are able to choose full care from a Specialist CF Centre if they wish.
- Due to the increasing complexity of CF in adulthood, full care should be delivered by a Specialist CF Centre. In exceptional circumstances (e.g. long travelling distances) geography may dictate that adults can receive care through a Network CF Clinic provided by specialists with appropriate training and dedicated time working as part of the Specialist CF Centre team.
- Care delivered by a Network CF Clinic should be to the same standard as that delivered by the Specialist CF Centre.
- If under the care of a Network CF Clinic, patients must be seen at the Specialist Centre for annual review (unless the Network CF Clinic can provide all recommended clinical reviews and investigations, in which case it may be done jointly by both teams in the local centre).
- Patients must be seen at least twice a year (including annual review) by the full Specialist Centre multidisciplinary team (MDT), which may take place at either the Network or Specialist hospital. It may be appropriate for the patient to be seen more often at the Specialist Centre, depending on the level of expertise at the Network CF Clinic, and the severity of their disease.
- Shared care must be delivered as part of an agreed designated Network with a Service Level Agreement

and Standard Operating Procedures as laid down by the Specialist CF Centre.

The importance of Specialist CF Centre care cannot be over-emphasised. The need for Specialist CF Centres has been recognised by the Royal College of Paediatrics and Child Health, the Royal College of Physicians of London, the British Thoracic Society, and the British Paediatric Respiratory Society. It is also recognised by the Department of Health in Definition 10 of the National Specialised Services Definition Set (2010) www.specialisedservices.nhs.uk/library/26/Cystic_Fibrosis_Services_all_ages.pdf. The US Cystic Fibrosis Foundation and the European Cystic Fibrosis Society also strongly endorse the principle and importance of Specialist CF Centre care.

Audit and outcomes

- Are all patients seen at least once a year by a team from the Specialist CF Centre for annual review?
- Are all patients seen at least twice a year by the full Specialist Centre MDT?
- Are Service Level Agreements in place for all Network CF Clinics?

2.2 Specialist CF Centre care

The level of expertise required to treat the complex multi-system symptoms and complications in CF can only be acquired by a multidisciplinary team of trained, experienced, specialist health professionals who routinely see a critical mass of patients at a Specialist CF Centre.

A Specialist CF Centre treats either children or adults. It has a minimum of 100 adults or children. It is recommended that when numbers reach 250 and are set to continue to rise, the development of alternative Specialist Centres should be considered. It is recognised that in exceptional circumstances, the geographical location of a Specialist CF Centre may mean that the number of patients is less than 100, although it should not be less than 50.

Criteria for designation as a Specialist CF Centre have been outlined by the West Midlands Specialised Commissioning Team – 'Designating cystic fibrosis services'. The following criteria must be met:

- A sufficient number of patients (see above).
- A core multidisciplinary team of trained and experienced CF specialist health professionals, which must be of appropriate number for the size of the patient population:
- Specialist consultant paediatricians or adult physicians
- Medical support from trainee(s)
- Clinical nurse specialists
- Physiotherapists
- Dietitians
- Clinical psychologists
- Social workers
- Pharmacists
- Secretarial support
- Database coordinator
- Recommended staffing levels have been agreed by the chairs of all the CF special interest groups.
- Staffing levels do not necessarily change incrementally with patient numbers above 250 (ie, 500 patients do not automatically require double the staff of 250 patients). This does depend on the specialty though, for example it is likely physiotherapy numbers may need to increase as opposed to the database coordinator.
- Maximum staff requirements may be limited by the number of beds available and clinics held each week, regardless of the total clinic population.
- Staffing numbers should reflect the model of shared care being used, taking into account time spent by staff from the Specialist Centre seeing patients in a local hospital CF clinic.
- It is important that adequate cover is available for annual leave etc.

Adult centres

Whole time equivalents per clinic size

	75 Patients	150 Patients	250 Patients
Consultant 1	0.5	1	1
Consultant 2	0.3	0.5	1
Consultant 3	---	---	0.5
Staff grade / fellow	0.5	1	1
SpR	0.4	0.8	1
Specialist nurse	2	3	5
Physiotherapist	2	4	6
Dietitian	0.5	1	2
Clinical psychologist	0.5	1	2
Social worker	0.5	1	2
Pharmacist	0.5	1	1
Secretary	0.5	1	2
Database coordinator	0.4	0.8	1

Paediatric centres

Whole time equivalents per clinic size

	75 Patients	150 Patients	250 Patients
Consultant 1	0.5	1	1
Consultant 2	0.3	0.5	1
Consultant 3	---	---	0.5
Staff grade / fellow	0.5	1	1
SpR	0.3	0.5	1
Specialist nurse	2	3	4
Physiotherapist	2	3	4
Dietitian	0.5	1	1.5
Clinical psychologist	0.5	1	1.5
Social worker	0.5	1	1
Pharmacist	0.5	1	1
Secretary	0.5	1	2
Database coordinator	0.4	0.8	1

- The team must have experience in managing the complex problems and rarer complications that may arise in patients with CF.
- A commitment from the MDT to ongoing CPD, demonstrated by membership of their relevant Special Interest Group, as well as attendance at relevant national and international conferences.
- Access to other medical and surgical specialists when appropriate particularly, Gastroenterology and Hepatology, Diabetes and Endocrinology, Ear Nose and Throat, Cardiothoracic and General Surgery, Specialist Anaesthesia & Pain Control, Rheumatology, Obstetrics & Gynaecology, Psychiatry, Intensive Care, and Interventional Radiology.
- Access to diagnostic facilities (e.g. sweat testing www.acb.org.uk/docs/sweat.pdf, lung function, bronchoscopy, and radiology).
- Access to microbiology services fulfilling the criteria set out in 'Laboratory Standards for Processing Microbiological Samples from People with Cystic Fibrosis' (CF Trust, 2010) www.cftrust.org.uk/aboutcf/publications/consensusdoc/CD_Laboratory_Standards_(for_web)_4_Oct_2010.pdf.
- Ability to provide 24-hour 7 days per week advice for urgent patient needs.
- Sufficient capacity in clinics for outpatients to be seen urgently (within 24 hours). This may be in clinic, day case unit or a ward visit.
- Sufficient inpatient beds so that patients do not wait for an admission (see section 5.4.1).
- Hospital ward nursing staff with sufficient knowledge and experience in CF care.
- All CF facilities must allow for adequate patient segregation.
- Participation in the UK CF Registry.
- Participation in the Cystic Fibrosis Trust / British Thoracic Society and British Paediatric Respiratory Society programme of peer review.
- Active involvement in audit and research.
- Active involvement in education and training.
- Written protocols for the delivery of care.
- Ability to demonstrate evidence that views of the service users (patients and families / carers) have been considered and influenced service delivery.

Audit and outcomes

- Is there a multidisciplinary team of trained and experienced CF specialist health professionals in the Specialist Centre and are the staffing levels appropriate for the clinic size?
- Is there evidence that the staff maintain their CPD relevant to CF?
- Does the centre have referral pathways in place for other medical / surgical disciplines?
- Does the centre send respiratory samples to a microbiology laboratory fulfilling the 2010 CF Trust laboratory standards for processing microbiological samples?
- What clinical audits in CF have been carried out in the last 3 years?
- What peer-reviewed papers have been published in the last 5 years?
- Are there local guidelines for CF care?

2.3 Network CF Clinics

It is recognised that different forms of shared and network care exist around the UK. These have arisen historically and been designed to meet local needs. Whilst these are mostly successful, it is crucial that certain criteria are fulfilled in all centres.

- There must be a Service Level Agreement between the Specialist CF Centre and the Network CF Clinic setting out the key elements of the service required.
- There must be clearly defined protocols (Standard Operating Procedures) for the delivery of care as established by and agreed with the Specialist CF Centre.
- All patients attending a Network CF Clinic must be seen by the multidisciplinary team from the Specialist CF Centre at least twice a year, and maybe more depending on the severity of the disease and level of expertise at the Network CF Clinic.

Criteria for designation as a Network CF Clinic are as follows:

- A Network CF Clinic is led by a consultant with the criteria as per section 3.3.1.

- There needs to be a core local multidisciplinary team, comprising an appropriate number of named nurse specialists, dietitians and physiotherapists, who liaise and work with the multidisciplinary team at the Specialist CF Centre, and who are members of the appropriate Special Interest Group.
- There must be regular communication between the consultants and multidisciplinary team of the Specialist Centre and Network CF Clinic.
- Serious clinical problems should be communicated by telephone or email (with appropriate patient confidentiality maintained) to the Specialist CF Centre within 24 hours.
- All significant episodes of care as set out in the Service Level agreement will be communicated to the MDT at the Specialist CF Centre within 5 working days.
- Routine letters should be sent out within 7 days.
- Conversely, any change in treatment recommended by the Specialist CF Centre should be communicated to the local team within 10 days (or within 5 days for an acute problem).
- There must be a formal system in place (organised by the Specialist CF Centre) to support CPD for the Network Clinic team. These meetings should occur at least annually and include the active participation of Network Clinic teams.
- Sufficient capacity for outpatients to be seen urgently (within 24 hours). This may be in a clinic, day case unit or as a ward visit.
- Sufficient inpatient beds so that patients do not wait for an admission (see section 5.4.1).
- All CF facilities must allow for adequate patient segregation.
- Access to microbiology services fulfilling the criteria set out in 'Laboratory Standards for Processing Microbiological Samples from People with Cystic Fibrosis' (CF Trust, 2010) www.cftrust.org.uk/aboutcf/publications/consensusdoc/CD_Laboratory_Standards_(for_web)_4_Oct_2010.pdf.
- Participation in the CF Registry in conjunction with the Specialist CF Centre.

Audit and outcomes

- Is there a multidisciplinary team of trained and experienced CF specialist health professionals in the Network CF Clinic?
- Is there evidence that the staff maintain their CPD relevant to CF?
- Is there a Standard Operating Procedure for the delivery of care by the Network CF Clinic agreed with the Specialist CF Centre?
- Does the Network CF Clinic send respiratory samples to a microbiology laboratory fulfilling the 2010 CF Trust laboratory standards for processing microbiological samples?

3.1 Multidisciplinary cf Care

3.1 Principles

- Specialist multidisciplinary care is essential in the management of children and adults with CF. Essential team members are listed in section 2.2, with further details in sections 3.3-3.9.
- All CF patients must have access to specialist advice and care from their CF centres at all times.
- Access to the CF centres (routine or urgent) should follow the CF Trust clinical care pathway – www.cfcarepathway.com.
- Continuity of care is essential. All patients should have a named consultant even though the CF centre may work with a team of consultants. The patient may not necessarily see their consultant on each visit, but it is important that they see the consultant at least once a year, usually after annual review. Additionally, the patient should have access to him / her at times of particular concern.
- CF centres must have access to other specialists (section 2.2) who are familiar with the complications of CF. Referral pathways with Service Level Agreements must be established.
- Where appropriate joint clinics should be established, particularly for CF-related diabetes.

Audit and outcomes

- Do all patients have a named consultant, and how often the patients seen by them?
- Are there joint clinics held with a CF diabetes specialist?

3.2 The role of the general practitioner (GP)

- The GP is responsible for prescribing much of the routine therapy recommended by the Specialist CF Centre. The Specialist CF Centre must ensure that the GP is adequately informed about the medication recommended particularly when it may be unfamiliar or used out of product license. It is reasonable to expect the GP to provide adequate amounts of medication – a minimum of one month at a time but ideally this would be longer for chronic medications e.g. pancreatic enzymes, vitamins etc. Some local pharmacies and hospitals coordinate an ordering and delivery service.
- The GP will ensure that patients are fully immunised and arrange for annual influenza immunisation every autumn. Please see European CF Society – Immunisation in the current management of cystic fibrosis patients www.journals.elsevierhealth.com/periodicals/jcf/ article/S1569-1993(05)00032-9/fulltext.
- The GP will be responsible for non-CF health-related issues.
- The GP may be asked to be involved with certain referrals, for example fertility and pregnancy issues, and genetic counselling.
- The GP (as well as hospital consultant) will have responsibility for certification of illness for patients.
- The GP may be requested to work in partnership with the CF homecare team, particularly in the management of end of life issues.

3.3 Medical consultant

Senior doctors working with CF patients fall into one of the following categories:

1. Consultant in a paediatric Network CF Clinic.

2. Consultant in a Specialist CF Centre.

3. Lead clinician / centre director in a Specialist CF Centre. They will usually work either with children or adults.

3.3.1 Consultant in a paediatric Network CF Clinic

A Network CF Clinic is usually led by a consultant with the following criteria:

- Training that will have included CF care; this may be as a general paediatrician with a respiratory interest, or full time national grid respiratory training. It is likely this criterion will now apply to new consultants taking up such a post.
- Specialist interest and clinical experience in CF; there may be older experienced paediatricians in this category who have worked with CF children for many years.
- Job plan with adequate programmed activities in CF.
- Clearly defined arrangements for cover during annual leave and absence.
- Arrangements for attending clinics with the Specialist CF Centre team.
- Capacity to maintain CPD in CF which would include attendance at national or international respiratory / CF meetings.

3.3.2 Consultant in a Specialist CF Centre

A consultant who works in a Specialist CF Centre will be expected to fulfil the following criteria:

- Training in paediatric CF care during a 2-3 year period as a higher specialist National Grid Respiratory trainee; they will have a Certificate of Completion of Training (CCT) in paediatrics with a respiratory interest. It would now be unusual to be appointed to work in a

Specialist CF Centre with primary training in paediatric gastroenterology as most specialist CF posts are part of a respiratory post.

- Training in adult CF care as part of higher specialist training as a respiratory physician. Basic respiratory training may often not include CF other than for a few weeks, but for a specialist CF post it is expected that the trainee has at least 1 year (and preferably more) full time in CF. This may be part of a CF Trust post-CCT fellowship, or whilst spending 2-3 years working for an MD or PhD in CF-related research. There may need to be a period of mentoring from a senior colleague.
- Job plan with adequate programmed activities in CF.
- Capacity to maintain CPD in CF which would include attendance at national or international respiratory / CF meetings.
- A track record in teaching, audit and/or research.

3.3.3 Lead clinician/centre director in a Specialist CF Centre

In addition to fulfilling the criteria for a consultant in a Specialist CF Centre, a lead clinician would be expected to have the following:

- At least three years' experience working as a consultant in an accredited Specialist CF Centre.
- Training or experience in management.
- They should be fully engaged with the management of the NHS organisation in which they work in order to be closely involved in the interactions and negotiations with commissioners in planning and contracting CF care.
- Leadership skills to direct the multidisciplinary CF team.

3.4 Clinical nurse specialist

General information

- Must be registered with the Nurses and Midwives Council.
- Must be a member of the UK CF Nursing Association.
- Those working with children must have undergone specific paediatric training.
- Must have specialist knowledge and be experienced in the care of children or adults with CF.
- Must maintain their CPD through the attendance of courses and conferences.

Role

- Provide advocacy and psychosocial support, particularly at important times such as the notification of a screening result and diagnosis, first admission to hospital, first course of intravenous antibiotics, a secondary diagnosis (e.g. CFRD), transition, reproductive issues, pre- and postnatal care, transplant and end of life issues.
- Provide home care support particularly for home intravenous antibiotic therapy.
- Provide education to others about CF including nurseries, schools, places of higher education and work places.
- Act as a link between the patient and family, primary care/community services and hospital.
- Act as a resource for training and education for other professionals involved in CF care.
- See CF Trust consensus document 'National Consensus Standards for the Nursing Management of Cystic Fibrosis', 2001, available at www.cftrust.org.uk/aboutcf/publications/ consensusdoc/C_3300Nursing.pdf.

Research and audit

- Contribute to research in all areas of CF, either through developing individual projects or participating in research carried out by the MDT.
- Take part in audit carried out on behalf of the CF service.

3.5 Physiotherapist

General information

- Must be registered with the Health Professions Council.
- Must have a minimum of 3 years' post-graduate experience working with patients with CF.
- Should be a member of the Association of Chartered Physiotherapists in Cystic Fibrosis (ACPCF).
- Must work to the Standards of Respiratory Care ACPCF (2009) and Standards of care and good clinical practice for the physiotherapy management of cystic fibrosis (2011).
- Must maintain their CPD through attendance at meetings such as national and international CF conferences and ACPCF study days.

Role

- Be responsible for a full assessment (including airway clearance, posture, exercise, urinary incontinence and non-invasive ventilation where necessary) on admission to and discharge from hospital, at every outpatient appointment and at annual review.
- Maintain community contact when necessary.
- Be responsible for service evaluation.
- Be responsible for service development, ensuring up-to-date, evidenced-based clinical practice.
- See CF Trust consensus document 'Standards of care and good clinical practice for the physiotherapy management of cystic fibrosis', 2011, available at www.cftrust.org.uk/aboutcf/ publications/ consensusdoc/Physio_standards_of_care.pdf.

Research and audit

- Contribute to research in all areas of CF either through developing individual projects or participating in research carried out by the multidisciplinary team.
- Take part in audit carried out on behalf of the CF service.

3.6 Dietitian

General information

- Must be registered with the Health Professions Council.
- Must be a member of the UK Dietitians CF Interest Group.
- Have specialist knowledge and be experienced in the care of children and/or adults with CF.
- Must maintain CPD through attendance at study days and meetings such as national and international CF conferences.

Role

- To be responsible for providing full nutritional advice and assessment including nutritional supplementation, PERT, enteral tube feeding, CFRD etc as appropriate to both in- and outpatients.
- Clinical dietetic practice should be evidence-based and reflect current research, clinical guidelines and consensus views.
- Be a resource on nutrition for the training, education, development and support for others involved in CF care.
- See CF Trust consensus document 'Nutritional management of cystic fibrosis', 2002, available at www.cftrust.org.uk/aboutcf/publications/consensusdoc/C_3500Nutritional_Management.pdf.

Research and audit

- Participate in dietetic and multi-professional CF audit and research.
- Be responsible for auditing and evaluating the service they provide ensuring improvement where necessary.

3.7 Clinical psychologist

General information

- Psychology services must be provided by a clinical psychologist, however at larger Centres this should be provided by a consultant clinical psychologist.
- Must be registered with the Health Professions Council.
- Must be a member of the UK Psychosocial Professions in CF Group (UKPPCF).
- Must maintain CPD through attendance at study days and meetings such as national and international CF conferences.

Role

- Undertake psychological review as part of annual review including an assessment of behaviour, emotions and family functioning.
- Provide psychological therapies including cognitive, behavioural, and family therapies.
- Respond to referral of inpatient for specialist psychological input within 1 week.
- Provide a psychology service in parallel with CF clinics by responding to referrals within 2 weeks.
- Provide support for newly diagnosed patients.
- 'Gatekeeping' for the onward referral of patients to mental health services and/or other relevant agencies (e.g. liaison psychiatry, community psychology services).
- Liaise with Network Clinics, social services and other community agencies for psychosocial input.
- Coordinate rapid access to on-call child and adult psychiatry services in cases of urgent psychiatric assessment for mental health concerns or risk of self-harm (all CF centres and clinics should be aware of their Trust's emergency psychiatric access policy).
- Provide consultation, support, training and supervision to other CF team professionals providing psychosocial interventions.

Research and audit

- Contribute to research in all areas of CF either through developing individual projects or participating in research carried out by the multidisciplinary team.
- Take part in audit carried out on behalf of the CF service.

3.8 Social worker

General information

- Desirable to have 3 years' post-qualification experience.
- Must be registered with the General Council of Social Care.
- Must maintain CPD through attendance at study days and meetings such as national and international CF conferences.
- Must be a member of the UK Psychosocial Professions in CF Group (UKPPCF).

Role

- Maintain up-to-date knowledge on all significant welfare and benefit changes and understand and apply relevant and current legislation to support patients.
- Have knowledge of chronic illness and how this impacts on patients and their families both day to day and long-term. Increase understanding amongst local and national government departments regarding these hidden consequences of CF.

- Act as a gatekeeper and liaison for the onward referral of patients to social services, housing services and other relevant agencies.
- Follow child protection procedures and ensure effective information sharing and referral and liaison to home authority team where appropriate.
- Liaise with schools / colleges / universities to access suitable support, e.g. arrangements for getting work to student, home access to laptop computers, time extensions for coursework and special arrangements for exams.
- Provide information and advice on employment rights and arrange access to Disability Employment Advisors if appropriate.
- Advocate on behalf of individuals and educate government and local authority agencies regarding the possible impact of CF on access to appropriate benefits and suitable housing.
- Provide support to patients' carers as needed and provide bereavement support / end of life support to patients and families.

Research and audit

- Contribute to research in all areas of CF either through developing individual projects or participating in research carried out by the multidisciplinary team.
- Take part in audit carried out on behalf of the CF service.

3.9 Pharmacist

General information

- Must be registered with the Royal Pharmaceutical Society of Great Britain.
- Must be a member of the Cystic Fibrosis Pharmacists Group.
- Must maintain CPD through appropriate study and attendance at relevant study days, national and international conferences.

Role

- Provide a prescription monitoring and medication review service, to include education and counselling to patients and carers.
- Provide a full review at annual review and disseminate information to GP and community pharmacist.
- Pharmacy service provision should allow for access to an out of hours on call pharmacist who can provide advice, information and urgent medication where appropriate.
- Assist in the management of formularies, development and provision of clinical guidelines and treatment protocols.
- Contribute to education and training of other healthcare professionals, including primary care where required.
- Provide advice on the legal and ethical responsibility of using medicines, including storage, unlicensed / off-label medicines.
- Advise on the procurement of difficult to source medications and aid in the resolution of any medication supply problems across secondary and primary care.

Research and audit

- Provide horizon scanning and critical evaluation of recent studies on new and existing therapies.
- Collaborate with CF research and development.
- Audit treatment guidelines and new therapies.
- Be involved in financial reporting on CF medication use.
- See 'Pharmacy Standards in CF Care' (Cystic Fibrosis Pharmacists Steering Group, 2011), available at www.cftrust.org.uk/aboutcf/publications/other_articles.

4. Principles of cf care

4.1 Infection control

- It is very important to prevent bacterial infections in people with CF. Whilst most bacteria are contracted from the environment, there is evidence of patient to patient spread of bacteria such as Burkholderia cepacia complex, Pseudomonas aeruginosa, and MRSA.
- Regular monitoring for cross-infection and epidemiological surveillance should take place by molecular typing.
- There must be local policies and clear operating procedures that involve segregating patients so that all CF patients are isolated from each other.
- Patients should not share rooms including bathrooms and toilets whilst inpatients in hospital.
- Hospital facilities must maintain a high standard of cleanliness.
- Patients should not be in contact with each other in waiting areas, e.g. CF clinics, wards, pharmacy, radiology etc.
- A high standard of hygiene should be practised by staff, in particular hand washing; alcohol gels or other suitable preparations must be available in every room. All equipment including stethoscopes, spirometers, infant weighing scales etc. must be cleaned between each patient.
- Particular care must be taken over patients infected with Burkholderia cepacia complex and MRSA. These patients should always come to separate, segregated clinics, and be admitted to separate wards where possible.
- People with CF, their families, and professionals recognise that there are psychosocial implications of segregation. It is important to acknowledge that limiting friendships between people with CF may lead to feelings of isolation and loneliness, and may contribute to difficulties in adjusting to the many challenges involved in living with CF.
- See CF Trust consensus documents:
  - Methicillin-resistant Staphylococcus aureus (MRSA), 2008. www.cftrust.org.uk/aboutcf/publications/consensusdoc/MRSA_1st_Edition_Final_web.pdf
  - Pseudomonas aeruginosa infection in people with cystic fibrosis. Suggestions for prevention and infection control. 2nd edition, 2004. www.cftrust.org.uk/aboutcf/publications/consensusdoc/C_Pseudomonas_aeruginosa_Nov_04.pdf
  - The Burkholderia cepacia complex. Suggestions for prevention and infection control. 2nd edition, 2004. www.cftrust.org.uk/aboutcf/publications/consensusdoc/C_Burkholderia_cepacia_Sep_2004.pdf

Audit and outcomes

- What arrangements are in place to minimise the risk of cross-infection in clinics and inpatient facilities?
- Is there evidence of cross-infection in the unit?
- What proportion of patients is infected with Burkholderia cepacia complex and MRSA, and what is the annual rate of new acquisition of these organisms?

4.2 Respiratory care

There are a number of important principles of respiratory care:

4.2.1 Monitoring of disease

- Frequent (at every clinic visit) microbiological surveillance of respiratory secretions (e.g. cough swab, sputum culture, and induced sputum) should be undertaken. This is principally for standard CF-associated infections such as Staphylococcus aureus and Pseudomonas aeruginosa, but should also include methicillin-resistant Staphylococcus aureus (MRSA), Burkholderia cepacia complex, Stenotrophomonas maltophilia, Achromobacter xylosoxidans and Aspergillus spp. Non- tuberculous mycobacteria should be tested for annually and more often if appropriate (including before starting long-term azithromycin). Samples should be sent to a suitable microbiology laboratory; see CF Trust consensus document 'Laboratory Standards for Processing Microbiological Samples from People with Cystic Fibrosis', 2010, www.cftrust.org.uk/aboutcf/publications/consensusdoc/CD_Laboratory_Standards_(for_web)_4_Oct_2010.pdf.
- Regular monitoring of lung function with spirometry (from 5-6 years of age), and oxygen saturation measurements with pulse oximetry.
- Regular (annual) monitoring with a chest radiograph. Chest CT scans should be carried out when appropriate and not routinely. The scan should be undertaken in an appropriate centre to minimise radiation exposure and with a suitable protocol to detect bronchiectasis.
- Monitoring for complications, particularly allergic bronchopulmonary aspergillosis (serology, sputum, radiology).
- Please see: European CF Society – Early intervention and prevention of lung disease in cystic fibrosis: a European consensus www.elsevier.com/framework_products/promis_misc/2004.pdf.

4.2.2 Therapies

- Younger children should receive oral antibiotic prophylaxis for Staphylococcus aureus; this may be considered for adults as well. Whilst this is common practice in the UK, it is not considered best practice in the USA and is still controversial.

- Prompt recognition followed by early and aggressive treatment of lung exacerbations.
- Higher doses and longer antibiotic courses (both oral and intravenous) are required compared to people without CF.
- Dual therapy is recommended for intravenous antibiotic courses (ß-lactam-based antibiotic with an aminoglycoside).
- Intravenous gentamicin should be avoided so tobramycin and amikacin need to be on the hospital formularies for use in CF patients. Hospital laboratories must have the ability to measure tobramycin or amikacin blood levels, which must be monitored regularly.
- See CF Trust consensus document 'Antibiotic treatment for cystic fibrosis', 3rd edition, 2009. Available at www.cftrust.org.uk/aboutcf/publications/consensusdoc/Antibiotic_treatment_for_Cystic_Fibrosis.pdf.
- Chronic infection with Pseudomonas aeruginosa is an important predictor of survival. Eradication regimens should be implemented for first and subsequent new Pseudomonas aeruginosa infections. This should take place promptly (within 1 week) from when the microbiology result is available. This also applies to Burkholderia cepacia complex and MRSA.
- Treatment of chronic Pseudomonas aeruginosa lung infections with inhaled antibiotics.
- Regular use of airway clearance techniques (chest physiotherapy). See CF Trust consensus document 'Standards of Care and Good Clinical Practice for the Physiotherapy Management of Cystic Fibrosis', 2011, available at www.cftrust.org.uk/aboutcf/publications/consensusdoc/ Physio_standards_of_care.pdf.
- Use of other respiratory therapies, for example Dornase alfa, hypertonic saline, long-term azithromycin when appropriate. See Flume PA, O'Sullivan BP, Robinson KA, et al.; Cystic Fibrosis Foundation, Pulmonary Therapies Committee. Cystic fibrosis pulmonary guidelines: chronic medications for maintenance of lung health. Am J Respir Crit Care Med 2007;176:957-69.
- Inhaled therapies must be delivered through an appropriate device.
- Patients with persistent symptoms despite appropriate therapy may require further investigations (e.g. chest imaging, pH study, bronchoscopy etc.).
- Smoking must be strongly discouraged in parents/carers from the time of diagnosis, as well as in all patients. Help should be offered for them to engage in smoking cessation programmes.
- Please see: European CF Society – Inhaled medication and inhalation devices for lung disease in patients with cystic fibrosis: A European consensus. www.ecfs.eu/files/webfm/webfiles/File/documents/JCF_article.pdf

4.2.3 Complications

- Please refer to CF Trust clinical care pathway www.cfcarepathway.com.
- Allergic bronchopulmonary aspergillosis (ABPA) needs to be considered, especially when there is a lack of response to standard antibiotics. Monitoring of ABPA markers should take place during admissions for exacerbations, with a baseline at annual review.
- Pneumothorax should be treated in a Specialist CF Centre; management is influenced by future impact on lung transplant surgery. See British Thoracic Society guideline on pneumothorax, 2003: www.britthoracic.org.uk/Portals/0/Clinical%20Information/Pleural%20Disease/Guidelines/ PleuralDiseaseSpontaneous.pdf.
- Haemoptysis should be treated in a Specialist CF Centre when significant. It can often be managed conservatively but may require an expert bronchial embolisation procedure. See Flume PA, Mogayzel PJ Jr, Robinson KA, et al; Clinical Practice Guidelines for Pulmonary Therapies Committee; Cystic Fibrosis Foundation Pulmonary Therapies Committee. Cystic fibrosis pulmonary guidelines: pulmonary complications: hemoptysis and pneumothorax. Am J Respir Crit Care Med 2010;182:298-306.
- Respiratory failure – assess the need for long-term or nocturnal oxygen therapy. If CO2 elevation has developed consider the appropriateness of instituting non-invasive ventilation – this is of proven benefit in helping patients with symptom control, and to help them survive to lung transplantation.

Audit and outcomes

- What proportion of patients is infected with chronic Pseudomonas aeruginosa, MRSA, and Burholderia cepacia complex, and what is the annual rate of new acquisition of these organisms?
- What is the median lung function (FEV1 and FVC) of the whole clinic and patients at transition to adult services? These should be monitored longitudinally to ensure improvement in overall care.
- What is the process of checking clinic respiratory sample microbiology results?
- Is eradication therapy carried out for first Pseudomonas aeruginosa infection?
- What proportion of patients with chronic Pseudomonas aeruginosa infection is on long-term inhaled antibiotics?
- Are tobramycin level results available within 24 hours?
- What proportion of patients aged 6 years and above with FEV1 <70% has been prescribed Pulmozyme (or who have failed a therapeutic trial)?

4.3 Nutritional and gastroenterological care

There are a number of important principles of nutritional care:

4.3.1 Monitoring nutrition and gastroenterology status

- Pancreatic status should be established at diagnosis by clinical assessment and confirmed by faecal elastase measurement. Pancreatic sufficient patients may need re-checking when clinically indicated (including annual fat-soluble vitamin levels).
- Growth of infants and children should be measured regularly, including weight, height and in infants head circumference (and plotted on appropriate charts). BMI should be monitored in all patients (and for children plotted on appropriate centile charts).
- If growth is faltering or weight loss identified a diagnostic review should be made. This should include analysis of dietary intake, malabsorption and glucose metabolism.
- At annual review measurements of nutritional status should be made (see section 5.3).
- Please see European CF Society – Nutrition in patients with cystic fibrosis: a European consensus, available at www.elsevier.com/framework_products/promis_misc/2002.pdf.

4.3.2 Therapies

- Nutritional status is independently linked to survival so prompt intervention to normalise nutritional status is essential, especially in the first year of life. Generally patients with better growth have better lung function and fewer infections. Advice from a specialist CF dietitian is critical. See CF Trust consensus document 'Nutritional management of cystic fibrosis', 2002, available at www.cftrust.org.uk/aboutcf/publications/consensusdoc/C_3500Nutritional_Management.pdf.
- Pancreatic replacement therapy and fat-soluble vitamin supplements should be prescribed to patients with pancreatic insufficiency. Amounts given should be tailored to each individual. Pancreatic sufficient patients may also require fat-soluble vitamin supplementation.
- Nutritional supplements may be necessary for some people and are usually taken by mouth. In some patients nutritional support may be needed and is usually given nasogastrically or via a gastrostomy. Occasionally tube feeds may be given via a nasojejunal tube or jejunostomy.
- Access to a gastroenterologist with experience of CF is important for managing complications such as malabsorption, severe gastro-oesophageal reflux or distal intestinal obstruction syndrome not responding to standard therapies.

4.3.3 Complications

- Please refer to CF Trust clinical care pathway www.cfcarepathway.com.
- Meconium ileus is the presenting feature in 10-15% of newborns with CF, and is usually manifest before results of newborn screening are known. All babies with this condition must have CF excluded.
- Distal intestinal obstruction syndrome will usually be managed by the CF specialist with input from the dietitian, however when resistant to therapy will need referral to a gastroenterologist with CF experience.
- Gastro-oesophageal reflux is common and should be excluded when patients are not responding to standard therapy, particularly when presenting with poor weight gain, or intractable wheezing. In adults cough, wheeze and deterioration in lung function may be the presenting symptoms.
- Eating difficulties can occur at any age, and will need specialist input from a dietitian and psychologist.
- Recurrent acute pancreatitis – the majority of these patients are pancreatic sufficient, referral to a gastroenterologist is appropriate.

Audit and outcomes

- What is the median BMI of the clinic population; and patients at transition to adult services? These should be monitored longitudinally to ensure improvement in overall care
- What proportion of the clinic population has weight <10th centile (children) or BMI <25th centile (children) or BMI <19 (adults).
- What proportion of the clinic is enterally tube fed?

4.4 Management of other CF manifestations and CF-related complications

- There are a number of other problems associated with cystic fibrosis due to the multi-system nature of the condition. See CF Trust clinical care pathway www.cfcarepathway.com for more details.

4.4.1 Impaired glucose metabolism and CF-related diabetes (CFRD)

- Impaired glucose metabolism and particularly overt CFRD is a major determinant of severe lung disease and reduced survival, so early identification is critical. Review of the diagnosis is important as glucose impairment may be transient. The diagnosis may be confirmed by the use of random blood glucose profiles or continuous glucose monitoring (CGMS).
- It is recommended that patients are screened annually for CFRD from 12 years and above using an oral glucose tolerance test. See CF Trust consensus document 'Management of Cystic Fibrosis-related Diabetes Mellitus', 2004, available at www.cftrust.org.uk/aboutcf/publications/ consensusdoc/diabetes.pdf.

- Insulin therapy should be started early and is the mainstay of treatment.
- Joint management with a diabetes specialist is important, but they need to be experienced in CFRD as its management is different from diabetes in someone without CF.
- Patients need specialist CF dietetic management as it is different from that given to diabetic patients without CF.

Audit and outcomes

- What regimen is in place for CFRD screening?
- What arrangements are in place for joint care with a CF diabetes specialist?
- What is the median HbA1c of the patients with confirmed CFRD taken at annual review?

4.4.2 Liver disease

- Annual screening for liver disease should be carried out on all patients aged 5 years and above.
  - This will include regular examination for hepatosplenomegaly, annual blood liver function and clotting tests, and alternate year ultrasound liver for abnormal architecture and signs of splenomegaly in children. Routine repeat ultrasounds may not be necessary in adults with previous normal scans.
- CF clinicians should be able to manage early liver disease and know when to start ursodeoxycholic acid.
- Patients should be referred to a hepatologist with CF experience for management of significant liver disease and its complications (for example portal hypertension, oesophageal varices).
- Liver transplantation is an option for end-stage liver disease.

Audit and outcomes

- How often do patients have a liver ultrasound?
- What is the pathway for referral to a hepatologist?

4.4.3 Male fertility

- It is important all males are informed about fertility issues at an appropriate age (usually 10-12 years old). Clear records should be taken to ensure that understanding is achieved before transfer to adult services.
- All adult males should be offered a sperm test when appropriate.
- Males should be educated that they can not assume they are infertile unless formally tested, and should take appropriate contraceptive (and safe sex) measures.
- Referral for genetic counselling (including their partners) and treatment using intracytoplasmic sperm injection should be offered.

Audit and outcomes

- Is it recorded whether fertility has been discussed before transition to adult services?
- Is there evidence that the service can advise patients appropriately and refer them for treatment of male infertility?

4.4.4 Reduced bone mineral density (BMD)

- Screening for reduced BMD with dual energy x-ray absorptiometry (DXA scan) should be initiated from 10 years of age with serial measurements every 1-3 years. See CF Trust consensus document 'Bone mineralization in cystic fibrosis', 2007, available at www.cftrust.org.uk/aboutcf/publications/consensusdoc/Bone-Mineral-Booklet.pdf, with the addendum www.cftrust.org.uk/aboutcf/publications/consensusdoc/Addendum_for_bone_document.pdf.
- Attention should be paid to nutritional status, corticosteroid intake and levels of physical activity (particularly weight-bearing exercise).
- Significant abnormalities should lead to referral to a bone specialist or endocrinologist (especially in children).

Audit and outcomes

- What proportion of patients aged 10 years and above has had a DXA scan in the last 3 years?

4.4.5 CF arthropathy

- Mild CF arthropathy can be managed by a CF specialist but referral pathways to a rheumatologist with CF experience should be in place for those with significant joint disease.

4.4.6 Nasal polyposis and sinusitis

- Mild ENT disease can be managed by a CF specialist, but referral pathways to an ENT specialist with CF experience should be in place for those with significant disease.

4.4.7 Stress incontinence

- A female member of the CF team (e.g. nurse specialist, physiotherapist) should enquire about stress incontinence for girls aged 11 years and above, and make a referral when necessary.

4.4.8 Puberty

- Puberty is occasionally delayed and its progress needs monitoring. Patients with significant delayed puberty may wish to be referred to a paediatric endocrinologist.

4.4.9 Renal complications

- Renal impairment can occur as a result of drug therapies, particularly the frequent use of aminoglycosides and gentamicin in particular. Renal function should be monitored on a regular basis.
- Monitoring of renal function and blood pressure are of particular importance in patients who have undergone

lung transplantation, as renal failure is a common complication of immunosuppressive therapy.

Audit and outcomes

- What measures are taken to monitor renal function in patients?
- Which intravenous aminoglycosides are used?
- What proportion of patients has had episodes of renal failure in the last 5 years?

4.4.10 Hearing

- Hearing problems may arise as a consequence of frequent courses of intravenous aminoglycosides. Screening is not currently advocated but audiological assessment may be necessary in some patients.

4.4.11 Psychological and social difficulties

- Psychological and behavioural problems are common and varied, including issues with eating difficulties, needle aversion / phobia, adherence to therapies, school problems, anxiety disorders, depression, concerns over infertility, and end of life / transplant issues.
- All patients must have access to clinical psychology and social work services; timing recommendations will depend on the urgency of the particular situation.
- Access to psychological support at the time of late diagnosis (e.g. adults) is essential.
- Patients should be screened annually for potential psychosocial problems by a psychologist or social worker with experience in CF.
- At times of crisis or deterioration in a patient's condition, the social worker can support patients to make adjustments in their lifestyle and facilitate changes in their working / employment / financial life allowing them to maximise treatment and optimise health.
- The CF team must have access to the psychosocial team for advice / consultation about patient care.
- All CF team members to be aware of and to follow safeguarding children / adults procedures where necessary.

Audit and outcomes

- What access do the patients have to clinical psychology and social work services?
- Does screening for psychosocial issues take place annually?

5. Delivery of care

5.1. Making the diagnosis

Please refer to CF Trust clinical care pathway www.cfcarepathway.com for details.

5.1.1 Diagnosis through newborn screening

In the majority of cases, the CF diagnosis will be made through the newborn screening programme. Details are available at http://newbornbloodspot.screening.nhs.uk/nat_std_cf_protocol; the website includes the protocols, information leaflets, communication guidelines and forms, training materials, advice for commissioners, and the laboratory handbook. It also includes the policies and standards for newborn blood spot screening in the UK (April 2005).

- When CF is suspected, the screening laboratory refers the child to the relevant Specialist CF Centre which will depend on where the child is born and resides. Pathways must be in place for a referral from the newborn screening laboratory to the relevant Specialist CF Centre (within 24 hours) which must be robust so that cases are not missed and all positive results processed appropriately.
- Specialist CF Centres and their networks must have a care pathway in place for contacting the parents in person and making the diagnosis in a timely fashion. Consideration must be given to reducing the waiting time and stress for the parents. The sweat test should be performed no later than the day after informing the parents that CF is suspected. This information should be given by someone who has comprehensive knowledge of CF, and preferably accompanied by a health professional who already knows the family (e.g. health visitor, GP).
- Diagnosis should be confirmed by a sweat test and/or genetic mutation analysis. Sweat tests must comply with the 2003 national guidelines written by the Association of Clinical Biochemists (being updated at time of printing) – www.acb.org.uk/docs/sweat.pdf. The sweat test result should be given to the parents by the CF consultant on the same day the test is performed. If only one parent is available, a friend or relative should be invited to be present.
- Whilst the majority of children who are picked up by screening have classic or typical CF, there are cases when the diagnosis remains in doubt, with a discrepancy between genotype, phenotype and sweat test results. These children may have non-classic or atypical CF, or CFTR dysfunction (sometimes called CFTR-related disorder or CFTR metabolic syndrome), and these children must remain under the care of a CF specialist.

Please see:

- European CF Society - European best practice guidelines for cystic fibrosis neonatal screening.

www.ecfs.eu/files/webfm/webfiles/File/documents/Castellani_2009_Journal-of-Cystic-Fibrosis.pdf

- European CF Society Consensus on the use and interpretation of cystic fibrosis mutation analysis in clinical practice. www.elsevier.com/framework_products/promis_misc/JCF7.pdf
- European CF Society - A European consensus for the evaluation and management of infants with an equivocal diagnosis following newborn screening for cystic fibrosis. www.elsevier.com/framework_products/promis_misc/JCF8.pdf

Audit and outcomes

- What is the pathway for notifying the parents of the need for a sweat test after receiving notification from the screening laboratory?
- Are sweat tests conducted according to the national standard?
- How many sweat tests are performed each year in the centre?

5.1.2 Diagnosis through clinical features

Some children and adults will be diagnosed following clinical suspicion of CF (symptomatic diagnosis). Some of the children will have been born before newborn screening was universal in the UK (October 2007), some will have been born abroad, and a small number will have been missed by the screening process.

- A diagnostic pathway should be in place for older children and adults. All the patients must be referred to a Specialist CF Centre for ongoing care.
- Infants presenting with meconium ileus should be treated as if they have CF until proven otherwise, with an early referral to the CF team.
- A similar detailed explanation and baseline investigation as offered to infants detected through newborn screening should be given to all newly diagnosed patients. Literature should be age-appropriate.

5.1.3 Following the diagnosis

- After diagnosis all patients should have the following investigations:
  - airway culture (cough swab, sputum);
  - bloods to include blood count, serum electrolytes, liver function tests and vitamin levels;
  - assessment of pancreatic function e.g. faecal elastase;
  - extended genotype if only one of the common mutations has been identified.
- Education of the parents / carers and/or older patients should be provided by the full multidisciplinary team within 7 days.
- Contact details, including telephone numbers for the CF team should be provided.
- General practitioner(s) must be informed on the day of diagnosis.
- The families of patients with CF should be offered referral to the regional genetic service for counselling re future pregnancies. Siblings will be offered carrier testing when they are old enough to make the choice. For adult patients, consent is necessary to reveal the diagnosis.
- Units should work to raise awareness of missed or late diagnoses which are not uncommon, and usually reflect an incorrect prior diagnosis or an atypical phenotype. A late diagnosis can result in difficulties in psychological and social adjustment, and the person may benefit from support and intervention by the psychologist and CF social worker. Adults in particular face practical problems such as obtaining life or travel insurance, mortgages etc.
- Siblings of children with CF should have a sweat test to exclude cystic fibrosis.
- Access to appropriate information should be made available including details of the Cystic Fibrosis Trust.
- The patient should be consented to be registered on the national CF Registry (Port CF).
- Please see Sermet-Gaudelus I, Mayell SJ, Southern KW; European Cystic Fibrosis Society (ECFS), Neonatal Screening Working Group. Guidelines on the early management of infants diagnosed with cystic fibrosis following newborn screening. J Cyst Fibrosis 2010;9:323-9.

5.2 Outpatients

Please refer to CF Trust clinical care pathway www.cfcarepathway.com for details. Principles of infection control must be strictly maintained in clinic – see section 3.5.

5.2.1 Frequency

- Patients should be reviewed regularly with a frequency appropriate to their individual needs, but must be seen at least twice a year by the full Specialist Centre multidisciplinary team (MDT), which may take place at either the Network or Specialist hospital.
- Routine appointments for a stable patient should be every 2-3 months depending on severity of disease. It is expected that they will be seen more frequently if there are ongoing clinical problems.
- Newly diagnosed infants should be seen every 1-2 weeks during the initial learning period, which may include home visits by the community team.
- Patients with atypical CF e.g. CBAVD without respiratory disease should be reviewed at least annually.

5.2.2 Procedures

The following should be carried out at every clinic visit, wherever that takes place:

- Measurement of growth in children and weight / BMI in adults (see section 3.3.1).
- Regular monitoring of lung function with spirometry (from 5-6 years of age), and oxygen saturation measurements with pulse oximetry.
- Culture of respiratory secretions (see section 3.2.1).
- Urine is tested for glucose if the patient has lost weight or if they are receiving oral corticosteroids.
- Blood pressure is measured in those on regular oral corticosteroids and post-transplant patients.
- When appropriate, flushing of totally implantable venous access devices. These will be flushed at other times as well since they are usually flushed every 4-8 weeks.

5.2.3 Consultations

Patients will see the members of the multidisciplinary CF team.

- Doctor. Patients may not necessarily see a consultant at every visit, but the consultation is under their supervision, and should be discussed with the consultant either in the clinic or at a multidisciplinary meeting. Patients should have the opportunity to request which doctor they wish to see, although must understand that this may not always be possible. They should however see a consultant at least at every other clinic visit, and more frequently depending on the severity of their disease. A letter must be written (and sent within 10 days) to the GP and shared care consultant, with a copy to the patient / carer.
- CF nurse specialist. The nurse has a pivotal role in the clinic and should see all the patients, often for general advice and support.
- Physiotherapist. All patients should have a review of current respiratory status and physical therapy every visit. The physiotherapist will usually collect the microbiology samples.
- Dietitian. Nutritional advice and education should be available at every clinic visit. All pancreatic insufficient patients should be reviewed by a specialist dietitian in every clinic and pancreatic sufficient patients will be seen if necessary.
- Clinical psychologist. Families and patients should have access to a clinical psychologist.
- Social worker. Patients should have access to a social worker.
- Pharmacist. Patients should have access to the CF specialist pharmacist if required in addition to the usual pharmacy support services.

Audit and outcomes

- Are full care patients seen 2-3 monthly in their Specialist CF Centre?
- Are shared care patients seen a minimum of twice a year by the full Specialist Centre MDT?
- What arrangements are in place for patient segregation?
- Are all members of the MDT present in every clinic?
- Is there evidence of user involvement / satisfaction in service delivery?

5.3 Annual review

Please refer to CF Trust clinical care pathway www.cfcarepathway.com for details. The annual review is a detailed assessment of every aspect of the patient's condition and therapies, to assess changes over the last year, identify where treatments can be improved, and produce a management programme for the following year.

- A report should be written by a consultant once all results are available, and sent to the GP, shared care consultant and patient / carers.
- The report should be discussed with the patient / carers and the treatment plan agreed.
- Data from the review is entered on to the UK CF Registry (Port CF).

5.3.1 Consultations

In addition to the consultations carried out in standard clinics, the following are included:

- Collection of Registry data.
- Specific mention when appropriate of puberty, fertility, transition, plans for pregnancy, transplantation.
- Physiotherapy review of airway clearance techniques, exercise and inhaled medication regimens.

They should also check posture and enquire about stress incontinence. Home nebulisers should be brought in for annual service when appropriate.

- Assessment of nutritional status and evidence of malabsorption, which may include a food diary, analysis of pancreatic function and enzyme use, nutritional support and vitamin supplementation, and diabetic therapy. Height and weight, growth velocity and BMI charts should be filled in (where appropriate).
- The patients / families should see a clinical psychologist who will screen for potential problems and use validated screening tools when necessary.
- There should be access to the social worker particularly during times of clinical deterioration as adjustments may need to be made to work patterns and types of employment.
- Specialist pharmacist review and discussion of all medication taken, both prescribed and non- prescribed and review adequacy of supply arrangements.

5.3.2 Investigations

- Centres may do additional investigations, but these listed below are the minimum expected:
- Lung function (which may include plethysmography) for patients over six years.
- Exercise testing when clinically indicated.
- Oxygen saturation measured by pulse oximetry. Arterial blood gases may be measured in some adults when clinically indicated.
- Respiratory sample (sputum or cough swab) for microbiology to include non-tuberculous mycobacteria.
- Chest radiograph.
- Ultrasound liver and spleen in children aged 5 years and above on alternate years (or annually if abnormal). Routine repeat ultrasounds may not be necessary in adults with previous normal scans.
- Bone densitometry (DXA scans) from 10 years of age every 1-3 years, or annually if abnormal.
- Assessment of glucose metabolism – oral glucose tolerance test from aged 12 years (unless CFRD already diagnosed). In those with impaired glucose metabolism, HbA1c (glycosylated Hb) will be measured.
- Blood is taken for the following (which may vary according to local policy and include additional tests): full blood count, clotting studies, electrolytes and renal function, liver function tests, HbA1c, vitamins A, D and E, iron studies, and aspergillus markers.

For purposes of quality improvement, it is useful to check what proportion of annual review recommendations has been implemented.

Audit and outcomes

- What proportion of the clinic population has had an annual review?
- Do all patients discuss the outcome of the annual review with a consultant?
- What proportion of patients is entered on to the CF Registry?

5.4 Inpatients

5.4.1 Principles

The majority of admissions are for intravenous (IV) antibiotics, either for a chest exacerbation or as part of routine management. However there are a number of other reasons, including: education of the family / patient at time of new diagnosis or at other times; any deterioration in clinical condition that fails to respond to outpatient measures including poor weight gain or weight loss; treatment of

DIOS; management of uncontrolled CF-related diabetes; psychosocial support; elective procedures e.g. insertion of a totally implantable venous access device or gastrostomy, ENT or dental surgery; ante- or postnatal care; respite or end of life care.

- Inpatient care is a fundamental part of the management of CF, and beds in a ward suitable for CF care should always be available for an emergency admission, as well as capacity to ensure elective and urgent admissions can be managed appropriately. See CF Trust clinical care pathway www.cfcarepathway.com.
- An urgent course of treatment should be implemented within a maximum of 24 hours of the decision being made. There should not be a delay of longer than 7 working days of the proposed admission date for a non-urgent course of treatment.
- Principles of infection control must be strictly maintained in the hospital – see section 4.1.
- Facilities must exist for a parent / carer to stay with their child in hospital.
- Local Authorities have a duty to provide suitable education for children of compulsory school age who cannot attend school due to illness. This education might be provided in a number of ways which include hospital schools. However children with CF should not be together in a hospital school room so facilities must also exist for schooling at the patient's bedside. In November 2001 the Department for Education and Skills (DfES), jointly with the Department of Health, published statutory guidance called Access to education for children and young people with medical needs, DfES 0732 / 2001, DoH 2001 / 019 which sets out the national minimum standards of education for children who cannot attend school because of illness or injury.
- There should be access to appropriate play and/or recreation, with facilities for studying.
- Physiotherapists should be responsible for a full assessment (including airway clearance, posture, exercise, urinary incontinence and non-invasive ventilation when necessary) on admission. They should also be available to administer or supervise physiotherapy treatment twice daily (or more if necessary), including weekends. This may be with assistance from a physiotherapist or independently if the physiotherapist has previously assessed that to be appropriate.
- Patients should have specialist CF dietetic input at least twice a week, and more frequently if appropriate. Provision should be made for inpatients to have a choice of high quality food including high energy options and access to high energy mid meal snacks and drinks.
- Patients should be seen by a consultant with specialist CF knowledge at least twice a week. The consultant will be updated daily by the rest of the team.
- Patients should have access to a cystic fibrosis nurse specialist.

- Patients should be reviewed by the CF specialist

pharmacist on a daily basis during the week, with service provision of on call advice on weekends and out of hours when necessary.

- Patients should have access to a clinical psychologist within the CF team when on the ward.
- Patients should have access to a social worker who can liaise with employers to minimise stress caused by time off work or with tutors at school / college / universities to minimise disruption to studies.
- For those patients without a totally implantable venous access device, there should be provision at all times for appropriate vascular access. This will usually be a PICC (percutaneously-inserted central catheter) or peripheral long line, but there may be circumstances when a short IV cannula is used. Attention must be paid to procedural distress and an experienced doctor or nurse should perform the procedures. Sedation may be required e.g. oral or sublingual midazolam, self-administered Entonox (nitrous oxide).
- Discharge planning is essential, especially if the patient is finishing the course of IV antibiotics at home. A summary must be sent to the GP, shared care consultant and patient / carers at discharge. The patient should be given a follow-up clinic appointment at the time of discharge.

5.4.2 Investigations

- Lung function tests (spirometry) twice weekly. Oxygen saturation which may include overnight continuous monitoring.
- Admission bloods. These are performed at admission but in children are often taken at the same time as the first aminoglycoside level unless they are needed immediately.
- Blood glucose monitoring. As well as those with CFRD, this is usually carried out in older patients for the first few days of an admission.
- Aminoglycoside levels – particular attention must be paid to this, especially if the patient is dehydrated (e.g. DIOS, CFRD) or has renal impairment or liver disease.
- Sputum/cough swab must be sent to microbiology within 24 hours of admission and repeated during admission. Nasopharyngeal aspirate for viral immunofluorescence is sometimes indicated (usually in infants <1 year old).
- Weight – twice weekly.
- Chest radiograph is only performed if clinically indicated e.g. to exclude pneumothorax. They are not performed to check long line position.

Audit and outcomes

- What arrangements are in place for infection control?
- What is the median waiting time for admission for a chest exacerbation?
- Do all inpatients see a CF consultant a minimum of twice per week?
- How often is an inpatient seen by a physiotherapist on a weekend?
- What arrangements are there for providing additional nutritional requirements during an admission?

5.5 Homecare

5.5.1 Principles

- Most Specialist CF Services in the UK offer a home-based care service. The CF clinical nurse specialist usually provides this service, but in some places CF specialist physiotherapists, dietitians and social workers are also involved.
- A comprehensive service may improve adherence to treatment, reduce hospital admissions, and help patients / parents cope with the psychosocial and practical day-to-day demands of CF.

A homecare service can support many aspects of clinical and social care including:

- Psychosocial support for patients / family.
- Health education.
- Clinical assessments at home including weight, lung function and oxygen saturation.
- Home intravenous antibiotic courses (see below), including drug level monitoring and blood tests.
- Flushing of totally implantable venous access devices.
- Optimising airway clearance techniques and encouraging exercise programs.
- Collecting respiratory samples.
- Discussing personal issues such as urinary incontinence.
- Supporting home enteral feeding.
- Pregnancy and postnatal support.
- Support at times of deterioration in health e.g. starting insulin, home oxygen etc.
- Liaison with local authority re. aids and adaptations in the home.
- Support to patients waiting for a transplant.
- End of life and bereavement support for family members.
- Liaison and communication with GP / local health care providers.
- Liaison with nurseries, schools, colleges and work places.

5.5.2 Home intravenous antibiotics

- Home IV antibiotics are not suitable for all patients. CF teams need to evaluate their appropriateness, for example where non-adherence or home conditions are an issue. They should be sensitive to the needs of the carers; for some families admitting a patient to hospital is important for respite, to prevent home becoming hospital, to ease employment pressures, prevent

isolation of some adults, or because of the needs of other family members.

- When a patient is receiving IV antibiotics at home, close monitoring is required to assess progress and ensure aminoglycoside levels are safe.
- Home care delivery of pre-prepared home IV antibiotics should be considered and when appropriate offered to patients.
- Appropriate training to administer IV antibiotics must be given to the patient / carers and written competency checks recorded.

Audit and outcomes

- What is the provision for supporting care at home?
- What proportion of patients receives home IV antibiotics?
- Have all patients / carers administering home IV antibiotics undergone competency assessment?

5.6 Services for adolescents / young people

- Young people need to be supported by professionals in learning to exercise autonomy and developing confidence in using healthcare services.
- During the transition period, they should be offered the opportunity to attend part of their consultation without their parents present.
- Consideration of lifestyle, sexual health and psychological issues should be made, as well as transition to higher education or starting work.
- See National Service Framework for Children Young People and Maternity Services: Core Standards, 2007 www.dh.gov.uk/en/Publicationsandstatistics/Publications/PublicationsPolicyAnd Guidance/Browsable/DH_4094329.

Audit and outcomes

- What provisions are in place for adolescent patients?

5.7 Transition to adult care

- When old enough all children with CF will have their care transferred to adult services. It is therefore essential that the patient and family are involved in the planning at an early stage to ensure a smooth and managed transition.
- Close liaison between the paediatric and adult teams over routine CF management reduces alterations to their patients' treatments at the time of transition.
- A joint paediatric and adult CF team approach is necessary for successful transition with identified coordinators from both teams. There needs to be a formally agreed protocol for transition.
- Discussions should be started with the child and carers when the child is 11 years old.
- Information about the process and about the choice of adult centre (when more than one centre exists locally) must be given to the adolescent and their parent.
- The adolescent and carers should have the opportunity to formally meet the adult team on more than one occasion (usually in joint clinics), and view the adult facilities.
- Written information about each patient must be given to the adult team.
- The upper age limit by which transition should have taken place is the 18th birthday.
- There should be an opportunity to discuss accessing college / university, including suitable halls of residence, or employment. Help may be required from a social worker in applying for funding to cover prescription charges for the first time.
- The process of transition does not end after the transition clinic. Follow up during the changeover period must be handled carefully between the paediatric and adult CF teams, with an agreement as to which team takes responsibility.

Audit and outcomes

- Is there a pathway in place for transition?

5.8 Pregnancy

- Women with poor lung function (FEV1 <50% predicted) may have increased adverse outcomes during a pregnancy.
- CF-related diabetes may contribute to adverse outcomes similarly to diabetes in any pregnancy.
- Chronic infection with Burkholderia cenocepacia also appears to be an adverse risk factor.
- Where possible, women should be encouraged to discuss pregnancy with their CF team before attempting to become pregnant, including genetic counselling and psychosocial issues. There should be more regular reviews pre-conception to optimise health status.
- Medication review should be carried out in conjunction with the CF specialist pharmacist and adjustments to treatments made where appropriate.
- Pregnant women with CF and their fetuses are at increased risk, so need specialist obstetric care
- working in close collaboration with the CF unit. Where possible, the CF unit should develop a relationship with a single obstetrician (with support from an anaesthetist) who has an understanding of the issues associated with pregnancy and CF. Close monitoring is essential especially in the 3rd trimester, when airway clearance may become problematic.
- Pregnant women should be reviewed frequently with the increasing size of the growing baby.
- Nutrition is a significant challenge during pregnancy in women with cystic fibrosis and regular review should include increased input from the CF specialist dietitian.
- Close postnatal follow-up is necessary in the CF unit

to ensure prenatal respiratory and nutritional health are reached.

- See European CF Society Guidelines for the management of pregnancy in women with cystic fibrosis www.ecfs.eu/ files/webfm/webfiles/File/documents/Pregnancy.pdf.

Audit and outcomes

- Is there an identified obstetrician linked with the adult CF centre and do they see all pregnant women with CF?

5.9 Transplantation

- Lung transplantation offers the possibility of improved quality of life and survival for people with end-stage cystic fibrosis.
- All CF centres should have a working relationship with one of the National Commissioning Group (NCG) designated lung transplant centres.
- Clear indications for referral should be developed by each Specialist CF Centre in conjunction with the transplant unit.
- Formal assessment of suitability and decisions about timing of placement on the waiting list should be carried out by the transplant centre.
- The ongoing psychosocial needs of patients being considered for transplantation should be reviewed by the CF team.
- Formal arrangements must be in place for the appropriate multidisciplinary continuing care of CF patients after transplantation. This may be based in the transplant centre (with regular support from the CF centre), or in the CF centre (with close liaison with the transplant centre).

Audit and outcomes

- Is there a formal link with a National Commissioning Group (NCG) designated lung transplant centre?
- How many patients have been referred for a transplant assessment in the last 3 years?
- What arrangements for post-transplant care are in place?

5.10 Palliative and end of life care

- All centres should have a palliative care team readily available to help and advise on management issues. Ideally, staff (especially in adult centres) should have received some training in end of life care.
- All people with CF should have the opportunity to discuss end of life care with their physician and other members of the MDT. Issues around advanced care planning should be patient focused, and a care pathway (for example the Liverpool Care pathway, www.liv.ac.uk/mcpcil/liverpool-care-pathway/) should be individualised according to the patient's wishes.
- Consideration must be given to patients on the transplant waiting list over the management of the transition from "hoping for transplant" to palliating terminal symptoms.
- There should be recognition of physical, psychosocial and spiritual concerns for all patients.
- This should be undertaken with full involvement of family.
- There should be easy access to psychological therapy within the CF team e.g. intervention for anxiety / depression.
- There needs to be clear documentation of end of life discussions and decisions with the patient and family.
- Patients should be given the choice whether to die at home or in hospital. Sometimes it may be possible to be in a hospice.
- There should be access to bereavement support for families.
- Following a patient's death there should be the opportunity for a multidisciplinary discussion in order to review practice.
- Liaison with primary care is essential throughout the process.

Audit and outcomes

- What access is there to a palliative care team?
- How many deaths have there been at the centre in the last 3 years, and what is the median age of death?

6. Audits and outcomes

6.1 CF Registry

- The UK CF Registry was established by the Cystic Fibrosis Trust in collaboration with the CF community with the primary objective of raising standards of care in the UK.
- Web based software enables the collection of a standardised dataset across all paediatric and adult Specialist CF Centres and associated Network CF Clinics throughout the UK.
- The Registry can be accessed locally and regionally for audit of individual Specialist Centres and clinics (login is at www.portcf.uk/login.asp). The multicentre dataset facilitates national audit, both cross-sectional and longitudinal.
- A report is published annually by the CF Trust and from 2011 centre data is no longer anonymised, available at www.cftrust.org.uk/aboutcf/publications/cfregistryreports.

6.2 Peer review

- The CF Trust established the current programme of reviewing CF Centres and Clinics in 2006. Since 2010 this has been done jointly with the British Thoracic Society and British Paediatric Respiratory Society.
- The aim is to help improve the level of care that Specialist CF Centres and networked clinics in the UK can offer to their patients.
- During a peer review, an independent panel of experts in CF care (including a patient or parent representative) spends a day visiting a CF Centre or Network CF Clinic. They discuss with CF teams how they manage their service and identify any problem areas such as staff shortages.
- Questionnaires are sent to all patients / carers attending the clinic in advance of the visit. A group of them is also interviewed on the day.
- A detailed report is then sent to hospital managers and commissioners, highlighting what is being done well and any areas that need attention.
- The majority of centres have now had at least one review and some centres have already received significant funding increases for their CF service as a result.
- It is expected that all centres are reviewed as part of their designation as a CF service.

6.3 Summary of recommended audits and outcomes

A. The diagnosis of CF should not be delayed but must be handled sensitively, and be followed by education of the parents / carers and patient (if old enough).

1. What is the pathway for notifying the parents of the need for a sweat test after receiving notification from the screening laboratory?

2. Are sweat tests conducted according to the national standard?

3. How many sweat tests are performed each year in the centre?

B. All patients must be under the direct supervision with regular follow-up from an adequately resourced designated Specialist CF Centre, sometimes in partnership with a Network CF Clinic.

4. Are all patients seen at least twice a year by the full Specialist CF Centre multidisciplinary team?

5. Are full care patients seen 2-3 monthly in their Specialist Centre?

6. Does the Specialist Centre / Network CF Clinic send respiratory samples to a microbiology laboratory fulfilling the 2010 CF Trust laboratory standards for processing microbiological samples?

7. What is the median waiting time for admission for a chest exacerbation?

8. Do all inpatients see a CF consultant a minimum of twice per week?

9. Is there evidence of user involvement / satisfaction in service delivery?

10. What proportion of patients is entered on to the CF Registry?

11. Are Service Level Agreements in place for all Network CF Clinics?

12. Are there local guidelines for CF care?

13. Is there a Standard Operating Procedure for the delivery of care by the Network CF Clinic agreed with the Specialist CF Centre?

C. Specialist multidisciplinary care must be delivered by a team of trained and experienced CF specialist health professionals with staffing levels appropriate to the size of the patient population.This is done as a partnership with the patient's parents/carers and/or the patient when old enough.

14. Is there a multidisciplinary team (MDT) of trained and experienced CF specialist health professionals in the Specialist Centre/Network CF Clinic and are the staffing levels appropriate for the clinic size?

15. Do all patients have a named consultant, and how often are the patients seen by them?

16. Are all patients seen at least once a year by a team from the Specialist CF Centre for annual review?

17. What proportion of the clinic population has had an annual review?

18. Do all patients discuss the outcome of the annual review with a consultant?

19. Are all members of the MDT present in every clinic?

20. Does the centre have referral pathways in place for other medical / surgical disciplines?

21. Is there evidence that the staff maintain their CPD relevant to CF?

22. What clinical audits in CF have been carried out in the last 3 years?

23. What peer-reviewed papers have been published in the last 5 years?

D. Measures must be in place to prevent cross-infection from other patients.

24. What arrangements are in place to minimise the risk of cross-infection in clinics and inpatient facilities?

25. What arrangements are in place for patient segregation?

26. Is there evidence of cross-infection in the unit?

27. What proportion of patients is infected with chronic Pseudomonas aeruginosa, MRSA, and Burholderia cepacia complex, and what is the annual rate of new acquisition of these organisms?

E. Treatment of airway infections are critical, so antibiotics are a key part of CF therapy – for prophylaxis, eradication therapy, long-term treatment of chronic infection, and treatment of acute exacerbations.

28. What is the median lung function (FEV1 and FVC) of the whole clinic and patients at transition to adult services? These should be monitored longitudinally to ensure improvement in overall care.

29. What is the process of checking clinic respiratory sample microbiology results?

30. Is eradication therapy carried out for first Pseudomonas aeruginosa infection?

31. What proportion of patients with chronic Pseudomonas aeruginosa infection is on long-term inhaled antibiotics?

32. Which intravenous aminoglycosides are used?

33. Are tobramycin level results available within 24 hours?

34. What proportion of patients aged 6 years and above with FEV1 <70% have been prescribed Pulmozyme (or who have failed a therapeutic trial).

35. What is the provision for supporting care at home?

36. What proportion of patients receives home IV antibiotics?

37. Have all patients/carers administering home IV antibiotics undergone competency assessment?

F. Chest physiotherapy with airway clearance techniques are a lifelong mainstay part of treatment.

38. How often is an inpatient seen by a physiotherapist on a weekend?

G. Nutritional support is crucial for all patients.

39. What arrangements are there for providing additional nutritional requirements during an admission?

40. What is the median BMI of the clinic population and patients at transition to adult services? These should be monitored longitudinally to ensure improvement in overall care

41. What proportion of the clinic population has weight <10th centile (children) or BMI <25th centile (children) or BMI <19 (adults)?

42. What proportion of the clinic is enterally tube fed?

H. Other manifestations of CF as well as complications must be recognised promptly and in some cases should be screened for, particularly liver disease and impaired glucose metabolism.

43. What arrangements are in place for joint care with a CF diabetes specialist?

44. What regimen is in place for CFRD screening?

45. What is the median HbA1c of the patients with confirmed CFRD taken at annual review?

46. How often do patients have a liver ultrasound?

47. What is the pathway for referral to a hepatologist?

48. What proportion of patients aged 10 years and above has had a DXA scan in the last 3 years?

49. What measures are taken to monitor renal function in patients?

50. What proportion of patients has had episodes of renal failure in last 5 years?

51. Is it recorded whether fertility has been discussed before transition to adult services?

52. Is there evidence that the service can advise patients appropriately and refer them for treatment of male infertility?

53. Is there an identified obstetrician linked with the adult CF centre and do they see all pregnant women with CF?

I. Psychosocial support is often required and should be available.

54. What access do the patients have to clinical psychology and social work services?

55. Does screening for psychosocial issues take place annually?

J. Transition to adult care should be planned and managed appropriately.

56. What provisions are in place for adolescent patients?

57. Is there a pathway in place for transition?

K. Transplantation, palliative and end of life care must be planned and managed appropriately.

58. Is there a formal link with a National Commissioning Group (NCG) designated lung transplant centre?

59. How many patients have been referred for a transplant assessment in the last 3 years?

60. What arrangements for post-transplant care are in place?

61. What access is there to a palliative care team?

62. How many deaths have there been at the centre in the last 3 years, and what is the median age of death?

Notes

Notes

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The Cystic Fibrosis Trust is the only UK-wide charity dedicated to fighting for a life unlimited by cystic fibrosis (CF) for everyone affected by the condition. Our mission is to create a world where everyone living with CF will be able to look forward to a long, healthy life.

At the Trust we are:

- Investing in cutting-edge research
- Driving up standards of clinical care
- Providing support and advice to people with CF and their families
- Campaigning hard for the issues that really matter

Fighting for a Life Unlimited