NHS England

SCHEDULE 2 – THE SERVICES

A. Service Specifications

Service Specification No.	A01/S/a
Service	Cystic Fibrosis - Adults
Commissioner Lead	
Provider Lead	
Period	12 Months
Date of Review	

1. Population Needs

1.1 National/local context and evidence base

Cystic fibrosis (CF) is the most common, life-limiting, recessively inherited disease in the UK, affecting about 7,700 people in England (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 it 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.

Cystic fibrosis mainly affects Caucasian populations. It is uncommon 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. Median survival is 41.4 years (CF Registry 2012) and has been predicted to be at least 50 years for children born in 2000. However, the median age at death is currently 29 years and most people with CF who die each year are young adults, and occasionally some are children (3 in 2009).

2. Outcomes

2.1 NHS Outcomes Framework Domains & Indicators

Domain 1	**Preventing people from dying prematurely**
Forced Expiratory Volume in 1 second (FEV1)	Number of patients and % with FEV1 >65% by age group and sex
BMI	Median BMI of centre cohort
Median Survival of National population	UK CF registry data
Domain 2	**Enhancing quality of life for people with long-term conditions**
Annual review and feedback	Number and % of patients who have had a post-annual review management plan with discussion
Accessibility of psychological support	Number and % of patients who have seen a psychologist within the past 12 months
Domain 3	**Helping people to recover from episodes of ill-health or following injury**
Timely initiation of treatment for exacerbation	% patients breaching standards of care for timing of admission.
Mucociliary clearance therapies	Number and % of adults receiving mucociliary clearance therapies
Domain 4	**Ensuring people have a positive experience of care**
Admission to specialist unit/ward	% of patients admitted to a ward with specialist CF staff

Systematically measure patient experience and satisfaction at a frequency driven by patient need	Systematic engagement and feedback on actions taken
Domain 5	**Treating and caring for people in safe environment and protecting them from avoidable harm**
Chronic Pseudomonas Aeruginosa infection (3+ isolates between two annual data sets)	% adults with chronic pseudomonas infection
Pseudomonas (PA) Chronic PA is 3+ isolates between two annual data sets	Number and % of patients with Chronic PA infection on inhaled antibiotics
Data Input	Number of complete annual data sets taken from verified data set expressed as a % of actual patient numbers

3. Scope

3.1 Aims and objectives of service

Aim

The service aims to improve both life expectancy and quality of life for adults with Cystic Fibrosis

Objectives

The service will deliver the aims of improving life expectancy and quality of life for adults with CF by:

- making timely diagnosis with appropriate counselling and psychological support to the patient and their family,
- providing high quality proactive and preventative treatment and care to ensure optimal lung function and nutritional status,
- ensuring a safe, cost effective, high quality service for the recipients of the services commissioned,
- ensuring equity of access to services for the CF population,
- facilitating autonomy and transition to adult care, encouraging independent care,
- supporting the patient in helping them to manage their CF independently in order that they can aspire to a life less hindered by their condition and provide support to their families

where appropriate,
- ensuring effective communication between patients, and the service providers,
- providing a personal service, sensitive to the physical, psychological and emotional needs of the patients and their families.

This specification sets the core elements of the service and standards by which CF services should be provided. Its purpose is not to define who the providers are. It defines the service to be provided which is supported by the new Payment by Results (PbR) currencies and funding streams that are being developed. The specification will be used to define the models of care and agree the providers.

This service specification does not include generic healthcare services such as dental service, general practice services, ophthalmology services etc. required by individuals with CF which will be accessed in the same way as by the non- CF population. However, close liaison is vital between CF services and generic services and the CF service should have processes in place to ensure that communication takes place.

The providers of the service will demonstrate that they are meeting, or with the support of commissioners, are working towards meeting, the requirements for cystic fibrosis care as set out in the CF Trust document "Standards for the Clinical Care of Children and Adults with Cystic Fibrosis in the UK" (2011). Providers not currently meeting the requirements of the CF Trust document will have a plan to do so which has been agreed with commissioners.

3.2 Service description/care pathway

The guiding principle within the service requirements is that all services will be provided in accordance with the CF Trust document "Standards for the Clinical Care of Children and Adults with Cystic Fibrosis in the UK" (2011). Care will be directed by the specialist centres.

All Services

As a minimum:

- every CF specialist centre will have a Director who is responsible for the service,
- every individual will have a named CF consultant in accordance with section 3.1 of the CF Trust document "Standards for the Clinical Care of Children and Adults with Cystic Fibrosis in the UK" (2011),
- the model of care must be governed by assurances of standards of care, access with care at home or close to home (where appropriate), and consistency and equity of access including the provision of home antibiotic services,
- inpatient, day care, outpatient, diagnostic and homecare services will be co- ordinated to ensure continuity of care for the patient,
- patients and their families will be seen in an age appropriate, comfortable environment, ensuring privacy, dignity and protection from cross infection,
- patients and their families will be afforded the right to be fully informed of their condition, and to ensure that information is communicated in an understandable, sympathetic and age appropriate manner,
- patients and their families will be encouraged to participate in the planning of their care,

- patients and their families will be made aware how to contact their clinical teams and cystic fibrosis support groups,
- within the required timescales, complete and accurate data is submitted to the UK CF Registry subject to patient consent.

Specialist Centre Responsibilities

Specialist centres will be responsible for providing the care plan for all patients.
This includes the responsibility for determining when high cost drug (such as Dornase, Tobramycin, Colistimethate sodium and Aztreonam lysine) should be prescribed, in accordance with the national commissioning policy.

All main centres will need a Service Manager with dedicated time and responsibility for the CF service.

Specialist centres must be able to provide cover for annual leave, study leave and long term absence (e.g. long term sickness or maternity leave). Individuals will be seen by the multi-disciplinary team in a CF clinic when clinically indicated. Typically this will be approximately every 8-12 weeks. More active problems will require more frequent reviews by the multi-disciplinary team.

The service must be able to provide for urgent care needs and advice 24 hours a day, 7 days a week. This will include management of emergencies such as haemoptysis, pneumothorax and bowel obstruction (including Distal Intestinal Obstruction Syndrome (DIOS)).

Telephone advice must be available. Clear contact numbers should be given to patients to enable them to obtain advice from the specialist team at any time. During out of hours contact, a process must be in place to ensure a clear line of communication with a CF specialist.

All specialist centres need to be fully operational and in a position to take referrals. Clearly defined links should be in place with community services and hospitals. Centres serving more rural areas should be able to demonstrate an ability to provide an outreach service for adult services where appropriate.

Outreach Care

Outreach care aims to provide specialist centre care using the facilities of a local provider. Outreach care does not require the local provider to have any CF specialist staffing.

Outreach care can be provided for adults where geographical constraints make attendance at the specialist centre difficult. Outreach care will be provided by the specialist CF centre. The full multi-disciplinary team will be present at outreach clinics.

Number of Patients

The number of patients at the centre must be sufficient to support a continuous provision of high quality care for CF patients. The CF Trust standards of care recommend that typically this will be approximately 100 patients and will not be less than 50.

Multi-Disciplinary Approach to Specialist Adult Care

Care will be delivered by a multi-disciplinary team of trained, experienced, specialist healthcare professionals who routinely care for a critical mass of CF patients at a specialist centre. The levels of staffing within multi-disciplinary teams will be in line with the recommendations set out in section 3 of the Cystic Fibrosis Trust document "Standards for Clinical Care of Children and Adults with Cystic Fibrosis in the UK" (2011).

General

All staff working within the CF service have an obligation to undertake continuing education and training to ensure updating of knowledge and skills. Core members of the CF multi-disciplinary teams shall be members of, and regularly contribute to, their relevant specialist interest group. Attendance at National/International specialist conferences will be demonstrable. It is recognised that not all staff will be able to attend every meeting every year; therefore the service will be able to demonstrate that there are internal mechanisms for feedback to the multi-disciplinary team.

Each member of each professional group shall demonstrate Continuing Professional Development in CF.

The service will ensure that processes are in place to ensure adequate workforce planning.

The service will be able to demonstrate that an appraisal process is in place for all staff.

Regular audit of services will be performed. Specific audits may be requested by the commissioner. Participation in research studies will be encouraged.

Each professional group will be required to meet the minimum competencies defined within section 3 of the Cystic Fibrosis Trust "Standards for the Clinical Care of Children and Adults with Cystic Fibrosis in the UK" (2011) and the defined care pathway. In particular the following will be achieved:

Composition of Multi-disciplinary Team

Service Director/Lead CF Consultant

The consultant in charge of the adult service must have appropriate experience working as a consultant in an accredited CF centre. He/she must be able to demonstrate active participation and attendance at national/international meetings and have a track record in teaching, audit and research. He/she must engage in the management of the service as a whole, ensuring leadership of the multi-disciplinary team and clinical governance of the service.

Cystic Fibrosis Nurse Specialists

CF Nurse Specialists will meet the standards identified in the CF Trust document "National Consensus Standards for the Nursing Management of Cystic Fibrosis" May, 2001. Nurse Specialists shall be members of the UK Cystic Fibrosis Nursing Association and must work within a CF multi- disciplinary team.

Physiotherapists

Specialist CF Physiotherapists will meet the standards identified in the Association of Chartered Physiotherapists in Cystic Fibrosis (ACPCF) document “Physiotherapy National Standards of Care for people with Cystic Fibrosis 2011”. They shall be members of the ACPCF special interest group.

Dieticians

Specialist CF Dieticians will meet the standards defined in Nutritional Management of Cystic Fibrosis (April 2002) and shall be members of the UK Cystic Fibrosis Nutrition Group.

Medical Staffing

Specialist Consultants will have had training in a recognised CF Centre. They must be able to demonstrate active participation and attendance at national/international meetings and have a track record in teaching, audit and research.

Middle grade medical support will in most instances comprise a trainee in respiratory medicine but may include a non-career grade with appropriate experience.

Pharmacists

Pharmacists must be registered with the General Pharmaceutical Council Britain and be a member of the Cystic Fibrosis Pharmacists Group. Pharmacists practice will reflect the Pharmacist Standards in Cystic Fibrosis Care 2011

Clinical Psychologists

Clinical Psychologists must be registered with the Health and Care Professions Council and be a member of the UK Psychosocial Professions in CF Group (UKPPCF).

Social Workers

Social workers must be registered with the Health and Care Professions Council and be a member of the UK Psychosocial Professions in CF Group (UKPPCF).

Provision of Care

Annual Review

A full review must be undertaken by the specialist centre once a year, in line with the standards defined in The CF Trust document “Standards for the Clinical Care of Children and Adults with Cystic Fibrosis in the UK” (2011).

A personal care plan must produced by a consultant and agreed with the patient as a result of every annual review undertaken

Outpatient Care

Routine appointments will be every 2 to 3 months when stable and more often if not. The outpatient clinics are multidisciplinary with all patients being reviewed by the doctor and a CF

nurse specialist, physiotherapist and dietician at all routine reviews. There will be access to psycho-social support.

Inpatient Care

Beds in a ward suitable for cystic fibrosis care will be available within 24 hours for an emergency admission, as well as capacity to ensure elective and urgent admissions can be managed appropriately. There will not be a delay of more than one week of the proposed admission date for a routine/planned/elective course of treatment.

Inpatient facilities will meet the standards defined in the Cystic Fibrosis Trust “Standards for the Clinical Care of Children and Adults with Cystic Fibrosis in the UK” (2011).

In particular, inpatients will:

- be entitled to and receive physiotherapy treatment 7 days a week if appropriate,
- have access to a specialist CF dietetic input at least twice a week, and more frequently if clinically appropriate,
- be seen by a consultant at least twice a week, and have access to consultant advice,
- be seen every day by a member of the medical team and have access to a Middle Grade doctor who is formally linked to the CF service,
- have access to a CF nurse specialist,
- have access to education facilities and support for school/college and examinations as appropriate,
- have access to appropriate recreational facilities,
- have provision for appropriate vascular access available at all times,
- have access to facilities for exercise.

Every CF patient will be in their own room, with en-suite facilities to minimise the risk of cross infection and to enable them to continue life as normally as possible.

Nurses on the inpatient wards require specific expertise, and will be committed to the CF service, with regular input and training from the specialist CF nurses. Patients will be admitted to a ward staffed by CF specialists or to wards that are familiar with the care and management of individuals with this condition and have developed the required expertise.

The inpatient service must have an infection control policy in place which demonstrates compliance with section 4.1 of the Cystic Fibrosis Trust “Standards for the Clinical Care of Children and Adults with Cystic Fibrosis in the UK” (2011).

Provision will be made for inpatients to have a choice of food including high energy options and access to high energy mid-meal snacks and drinks. This shall include evenings and weekends.

Intravenous (IV) Antibiotics

The service will have the ability to commence IV antibiotics on any day of the week.

An urgent course of treatment will be implemented within a maximum of 24 hours of the clinical decision being made.

There will not be a delay of longer than one week of the proposed admission date for a routine/elective/planned course of treatment.

Where appropriate, IV antibiotics may be provided at home, following receipt of the initial dose at the specialist CF centre.

Homecare

The life long multi-system nature of cystic fibrosis means that a complex regimen of home treatment is often recommended. Many patients and families require regular and consistent outreach from the multi-disciplinary team in this care. This will include:

- support in the community by the specialist CF multi-disciplinary team,
- open access to nursing care in the community. This may be a CF nurse specialist from the CF service, or local Community nurses who have specific training, experience and supervision in CF,
- patients undertaking home IV antibiotic therapy will have a formal assessment of suitability. This will include formal training and an assessment of competency of the patient and/or their carers in administering the IVs as well as the suitability of the home environment. There will also be planned review and assessment by the prescribing physician to ensure efficacy of each course of home IV antibiotics,
- support for patients receiving overnight enteral feeding,
- care of indwelling vascular access devices, gastrostomies and other stoma,
- physiotherapy input where appropriate,
- psycho-social support,
- liaison with school or college for patients still in education,
- support through times of change in an individual's health including introduction of treatment for diabetes or home oxygen therapy and end of life care,
- where clinically appropriate, home treatment will be encouraged.

Outpatient and Day Case Facilities

The service will ensure that the facilities are available to support the best quality CF service allowing seamless care between the home and hospital. Thus patients can be seen routinely in an outpatient facility but there must be provision for urgent review and providing the first dose of an antibiotic course either in the outpatients or a day case facility or ward.

The facilities must take the need for infection control into consideration and demonstrate compliance with section 4.1 of the Cystic Fibrosis Trust "Standards for the Clinical Care of Children and Adults with Cystic Fibrosis in the UK" (2011) when providing facilities for annual reviews, treatment, day case etc. This will include ensuring that CF patients are not kept waiting in communal waiting areas and that they remain segregated from each other at all times so as to minimise the risk of cross infection.

Equipment

The service will ensure that all relevant equipment is available, maintained and kept up to date in order that patients can receive and make use of appropriate equipment as well as treatment. In particular, the service will ensure that the following equipment is available as required:

The service will ensure that there is access to the provision of high quality spirometry (i.e.

meeting UK/EU standards) for all appropriate patients. Access will be available to the home care team to enable the monitoring of selected patients in the home with oxygen saturation monitors and home spirometry.

Patients who need home oxygen therapy will receive timely assessment and prescription of oxygen according to the National Home Oxygen service.

Individual patients will have access to a range of clinically appropriate airway clearance devices.

There will be a comprehensive nebuliser service, which aims to provide devices that deliver drugs in a fast and efficient manner. The service will also be able to provide a range of mechanical devices required to provide intermittent positive pressure breathing and non-invasive ventilation where needed.

Individual patients will have access to blood sugar monitors and continuous glucose monitoring systems (CGMS). Inpatient access will include enteral feeds, feeding pumps, nasogastric (NG) tubes, percutaneous endoscopic gastrostomy (PEG) tubes and gastrostomy buttons.

Primary Care will provide NG tubes, feeds and feeding pumps and giving sets for enteral feeding through an approved/agreed contractor or local community nursing service.

Diagnostics

The service will have access to all appropriate specialist CF diagnostic services, including:

- a microbiology laboratory that meets the Laboratory Standards for Processing Microbiological Samples from People with Cystic Fibrosis. First edition. September 2010. and that routinely cultures for recognised CF pathogens such as Burkholderia cepacia complex and also performs tobramycin levels,
- specialist radiology services, including contrast GI studies for bowel obstruction, ventilation perfusion scans, computerised tomography (CT) thorax, angiography, specialist liver scans, dual energy X-ray absorptiometry (DXA) bone scans and interventional services,
- a laboratory that performs specialist biochemical analysis such as faecal elastase and complies with the Association for Clinical Biochemists guidelines on performance 2003 of sweat tests,
- specialist lung function laboratory that will test patients as well as provide support and training for those staff performing spirometry in the clinic setting,
- Nasal Potential Difference testing (where required this facility should be available by collaborative arrangements with an appropriately equipped specialist CF Centre),
- facilities to undertake bronchoscopy.

Other specific aspects of Adult CF Specialist Care

Diabetes Care

Management of CF related diabetes will in accordance with the document ‘Management of Cystic Fibrosis-related Diabetes Mellitus (2004)’. In particular:

- There will be joint management between the CF multi-disciplinary team and a diabetes specialist experienced in the management of CF related diabetes (CFRD).
- The provider must have a documented protocol which describes how CFRD will be identified. The provider will undertake an annual audit which demonstrates compliance with the protocol

Transitional Care

Transition from paediatric to adult care is the norm for all patients.

Transition will be planned with the patient and their parent(s)/carer(s) with due regard to patient choice. There should be an underlying assumption that transition is natural and expected. All parents/carers will be made aware as early as possible that transition into adult services will take place.

Arrangements for transition to adult services will commence from the age of 12 years and will be completed by the age of 18, when responsibility for care transfers to the specialist adult cystic fibrosis centre. The specialist paediatric cystic fibrosis centre responsible for the care of the child will be responsible for ensuring that transition arrangements are put in place for each child. It is particularly important that these arrangements are carefully co-ordinated where the patient has had the majority of their care provided at a paediatric network clinic.

Every specialist paediatric CF service will have a formal policy for transition that is agreed with all specialist adult CF services to which their patients transfer.

The specialist paediatric CF centre will ensure the following:

- early discussion with the patient and carers about the process of transition. Options for adult care will be detailed. The age for transition will be flexible but agreed 2-4 years in advance, with the intention to complete before 18th birthday,
- notification to the adult centre of intention to proceed with transition,
- copies of letters and the annual review report are provided to the adult centre at least in the year prior to the anticipated transition clinic,
- there is documented paediatric and adult multi-disciplinary team member liaison, involving all multi-disciplinary team groups,
- there is the opportunity to visit the adult centre, to meet key multi- disciplinary team members and view both inpatient and outpatient facilities. Such a visit could be repeated if requested,
- there is a joint clinic with detailed clinical handover.

Specialist adult CF centres will demonstrate that they are actively engaging in the transition process for each child via an annual audit report to commissioners of the experience of patients who transitioned during the year.

Surgery

The decision to undertake surgery for patients with CF should be made jointly between the relevant surgeon, the CF clinicians and the patient and their parents or carers. Admissions for acute abdominal pain should be managed by the CF team, in collaboration with other relevant clinicians. Where possible, surgical procedures should be undertaken at a hospital which also provides a CF service. If this is not possible, full access to CF specialists should be available to

ensure that the patient's CF needs are fully taken into account, including during any post-operative period of inpatient care. A clear care plan should be developed, with regular contact and review between the relevant parties.

The surgical units should have a protocol or guideline relating to adults with CF which has been developed in collaboration with the CF service. This protocol should specify required standards of cross infection control and dietary/physiotherapy support.

The surgical service should have access to a CF clinician, to ensure communication regarding any surgical procedure, before during and after the procedure.

Transplantation

When the possibility of transplantation is appropriate, it will be discussed with the patient and family (where appropriate) as early as possible. Access to information will be readily available to patients.

Referral to the transplant centre for further assessment, if appropriate, will be made as soon as potential candidacy has been assessed.

Work up for transplantation will be undertaken in line with the guidance, processes and pathways defined by the transplant centre.

Palliative Care

Centres will be able to demonstrate:

- good working relationships with the general palliative care team attached to the hospital/local hospice/local community team and their involvement in all such patients,
- an identified member of the multi-disciplinary team with specific interest in palliative care,
- access to a psychologist for the patient and family,
- clear documentation of End of Life discussions,
- access to bereavement support for families,
- clinical review and debrief following a patient death.

Management of Pregnancy/Fertility Services

The service will define a specific pathway for the planning of a pregnancy for a patient with CF. Ideally, pregnancy shall be planned in this group of patients, with pre-conception counselling and referral to the genetic service as a couple.

Links will be made to the high risk obstetric team and patients will be seen by the CF team at least monthly during pregnancy. Close liaison between the CF team and the specialist obstetric team will need to be evidenced.

Strong links will be made with the pre-natal screening services to ensure that parents with a CF diagnosis have rapid access to specialist advice and support when considering pregnancy or where a diagnosis of CF is suspected in their unborn child, either before termination of pregnancy is considered or before their child is born.

The service will specify a pathway for CF males to have fertility assessed and a referral pathway for infertility treatment.

Infection Control

The service must have policies and procedures in place to protect patients from the risk of cross- infection, both as in- and out-patients (Cystic Fibrosis Trust standards of care and subsequent updates to infection control standards).

Clinical Governance

Clinical Governance will be demonstrated via:

- microbiological surveillance to identify infection control issues and use of particular antibiotics,
- proportion of patients with chronic Pseudomonas infection,
- monitoring of lung function forced expiratory volume in 1 second (FEV1) and rate of decline,
- Body Mass Index,
- reviews of all deaths,
- benchmarking with other similar centres, including use of the UK CF Registry data when available,
- number and resolution times of complaints,
- departmental risk register.

There will be clear succession planning for staffing to ensure continuity of care into the future.

3.3 Population covered

The service outlined in this specification is for patients ordinarily resident in England*; or otherwise the commissioning responsibility of the NHS in England (as defined in *Who Pays?: Establishing the responsible commissioner,* and other Department of Health guidance relating to patients entitled to NHS care or exempt from charges).
* - Note: for the purposes of commissioning health services this EXCLUDES patients who, whilst resident in England are registered with a GP Practice in Wales, but INCLUDES patients resident in Wales who are registered with a GP Practice in England.

The Provider shall provide services (the Service) for patients with cystic fibrosis aged 16 years and over. All eligible patients must be able to access the service.

3.4 Any acceptance and exclusion criteria and thresholds

Referrals can come from a number of sources, following the identification of a patient with suspected CF.

Following referral with suspected CF, the service will be responsible for:

- investigations leading to a rapid and clear diagnosis, where possible appropriate counselling of patients

- early introduction of required treatment.

For adult patients, the majority of referrals will come from paediatric CF services with appropriate transition arrangements. However, referrals may also come from other adult CF services, general physicians or General Practitioners.

Patients already diagnosed with CF may be referred to the CF service when they move from other regions.

Patients aged under 16 years and patients who do not have cystic fibrosis are excluded from the service

For the purposes of this specification, a cystic fibrosis patient is defined as:

- having a confirmed or strongly suspected diagnosis of cystic fibrosis, which includes:
  - a compatible clinical history, supported by one or more of the following:
  - a positive sweat test
  - two known disease forming CF gene mutations
  - evidence of functional epithelial ion transport abnormality

3.5 Interdependencies with other services/providers

There is no requirement for co-location with other services

The service will be able to provide access or referral to specialists within:

- Endocrinology,
- Diabetology,
- Hepatology,
- Gastroenterology,
- Rheumatology,
- ENT,
- Vascular services,
- Thoracic surgery,
- Palliative care,
- Clinical genetics,
- Transplantation services,
- Psychiatry,
- Obstetrics and Gynaecology,
- Renal services,
- Fertility services,
- Gastro-intestinal surgery.

If not available at a CF Adult Centre, processes must be in place to demonstrate clear pathways including Out of Hours/Emergency Care.

4. Applicable Service Standards

4.1 Applicable national standards e.g. NICE

Core Standards

The following standards are regarded as core standards and need to be achieved in order for a contract to be awarded for CF services. Where the remaining standards in this specification are currently met, they must continue to be met.

Where the remaining standards in this specification are not met they will need to be met over time. In such cases, the provider will agree with the commissioner a detailed development plan for achieving them (see section 2.1.5).

Every specialist centre must have a Director responsible for the service

The CF multi-disciplinary team (minimum of doctor, nurse specialist, physiotherapist and dietitian) must be available for care of in- and out-patients.

Policy and procedures must be in place to protect patients from the risk of cross- infection, both as in- and out-patients

Microbiological analysis of respiratory samples and age appropriate lung function must be carried out at all out-patients visits

Within the required timescales, the service will meet the minimum dataset requirements of the UK CF Registry. (subject to patient consent)

4.2 Applicable standards set out in Guidance and/or issued by a competent body (e.g. Royal Colleges)

The services within this specification will be provided with reference to the following publications:

- The CF Trust document “Standards for the Clinical Care of Children and
- Adults with Cystic Fibrosis in the UK” (2011),
- National Consensus Standards for the Nursing Management of Cystic Fibrosis. May 2001,
- Nutritional Management of Cystic Fibrosis. April 2002,
- Association of Chartered Physiotherapists in Cystic Fibrosis document “Physiotherapy
- National Standards of Care for people with Cystic Fibrosis 2011”
- Clinical Care Pathway,
- Department of Health National Definition Set for Cystic Fibrosis (2009),
- Management of Cystic Fibrosis-related Diabetes Mellitus (2004),
- Standards of care for patients with cystic fibrosis,
- Standards of care for patients with cystic fibrosis: A European consensus,
- Pharmacist Standards in Cystic Fibrosis Care 2011.
- Medicines Optimisation: Helping patients to make the most of medicines - Good practice guidance for healthcare professionals in England. http://www.rpharms.com/promoting-pharmacy-pdfs/helping-patients-make-the-most-of-their-medicines.pdf

These standards may change over time and as required, the service specification and service

level agreements will be updated to reflect such changes.

The service will meet and maintain national quality standards and any other national quality requirements that may from time to time be specified including multi-disciplinary Peer Review.

The service will meet the minimum dataset requirements of the UK CF Registry so as to enable all patients in the service to annually be assigned a banding. The bandings are linked to the appropriate national tariff will be used by commissioners for funding purposes. Detailed information on what is and is not included in the national tariff can be found in the Department of Health document “Payment by Results Guidance for 2011-12”. This information may be superseded by subsequent guidance.
(http://www.dh.gov.uk/prod_consum_dh/groups/dh_digitalassets/documents/digitalasset/dh_126157.pdf)

5. Applicable quality requirements and CQUIN goals

5.1 Applicable quality requirements (See Schedule 4 Parts A-D)

5.2 Applicable CQUIN goals (See Schedule 4 Part E)

6. Location of Provider Premises

The Provider’s Premises are located at:

7. Individual Service User Placement

Appendix One

Quality standards specific to the service using the following template:

Quality Requirement	Threshold (trigger for breach)	Method of Measurement	Consequence of breach
Domain 1: Preventing people dying prematurely			
Adults attending a specialist CF centre should have a FEV1 of >65%	< 55%	UK CF Registry data	Internal review with report to commissioners External review for outliers
Median BMI of centres should be greater than 21	Median BMI lower than 21	UK CF Registry data	Internal review with report to commissioners External review for outliers
Domain 2: Enhancing the quality of life of people with long-term conditions			
All adults should have an annual review and management plan discussed with patient	<85%	Self report dashboard	Internal review and submission of improvement plan
All adults should have access to psychological support at annual review	<85%	Self report dashboard	Internal review and submission of improvement plan
Domain 3: Helping people to recover from episodes of ill-health or following injury			
All adults should be offered inhaled mucolytic therapy	<70%	UK CF Registry data	Internal review
There should be no delay in initiating IV antibiotic therapy for pulmonary exacerbation (as defined by national service specification)	Breach percentage >10%	Self report /dashboard/CQIN	Commissioner review
Domain 4: Ensuring that people have a positive experience of care			
All adults requiring inpatient care should be admitted to a ward staffed by CF specialist staff (as defined by national specialist service specification)	<95%	Quality dashboard	Commissioner review
All specialist CF units should systematically measure patient experience and satisfaction at a	Yes/No	Self report demonstrating systematic engagement and feedback on actions	Commissioner review

Quality Requirement	Threshold (trigger for breach)	Method of Measurement	Consequence of breach
frequency driven by patient need		taken	
Domain 5: Treating and caring for people in a safe environment and protecting them from avoidable harm			
Adherence to standards of care to prevent cross infection. Centres should aim to reduce their prevalence of chronic pseudomonas infection	>65%	UK CF Registry	Internal review with report to commissioners Commissioner review for outliers
All adults chronically infected with Pseudomonas Aeruginosa should receive inhaled antibiotic therapy. Percentage of patients with chronic PA on inhaled antibiotics	< 85%	UK CF Registry data	Internal review with report to commissioners Commissioner review for outliers