NHS England

SCHEDULE 2 – THE SERVICES

A. Service Specifications

Service Specification No.	A01/S/b
Service	Cystic Fibrosis (Children)
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 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 population 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 >85% by age group and sex
BMI centiles	Median BMI centile 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 children 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 and carer 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)	% children 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 by age group
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 children with Cystic Fibrosis

Objectives

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

- Making timely diagnosis (including in response to newborn screening) with appropriate counselling and psychological support to the child and their family.
- Providing high quality proactive and preventative treatment and care to optimise 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 from children's care to adult care and encouraging independent care.
- Supporting parents and families of children with CF, as well as the child.
- Supporting the child in helping them to manage their CF independently in order that they can aspire to a life less hindered by their condition and providing support to their families where appropriate.
- Ensuring effective communication between patients, families 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 out the core elements of the service and standards by which CF services will be provided. Its purpose is not to define who the providers are. It defines the service to be provided and is supported by payment by results (PbR) currencies and funding streams. The specification will be used to define the models of care, agree the providers and establish robust shared care/network care arrangements where appropriate.

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 will 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 of this service specification. Specialist CF Centres not currently meeting the specification will have a plan to do so by April 2014 which has been agreed with commissioners. Network care providers not currently meeting this specification will have a plan to do so by April 2016 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 is directed by a specialist centre.

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 Care 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) will be prescribed, in accordance with the national commissioning policy.

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

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) of centre staff.

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).

Telephone advice must be available. Clear contact numbers will 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. The specialist centre will agree arrangements for 24 hour services with network clinics to ensure equity of access across a network service.

Network Care

Network care providers will typically have fewer numbers of patients than a specialist centre and so may have fewer staff. Care is therefore provided in partnership with the specialist CF centre that co-ordinates the network. Providers of network care for children will meet the requirements detailed in section 2.3 of the CF Trust document “Standards for the Clinical Care of Children and Adults with Cystic Fibrosis in the UK” (2011).

As a minimum the network service will have:

- A local CF multi-disciplinary team meeting the standards detailed in section 2.3 of the CF Trust document “Standards for the Clinical Care of Children and Adults with Cystic Fibrosis in the UK” (2011).
- Regular locally run CF multi-disciplinary team out patient clinics some of which will be joint clinics with the specialist CF centre that co- ordinates the network. Each patient will be seen by the full specialist multi-disciplinary team at least twice /year, either in a joint clinic at the network centre or at the specialist centre.
- Inpatient facilities suitable for routine CF admissions.
- Ward nurses with sufficient CF experience.

Annual reviews will take place at the specialist CF centre (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).

Network care providers must be able to comply with the standards specified below for CF inpatient care.

Network care providers must be able to provide cover for annual leave, study leave and long term absence (e.g. long term sickness or maternity leave).

Outreach Care

Outreach care differs from Shared/Network Care. Outreach care is provided by the specialist centre 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 children where geographical constraint makes attendance at the specialist centre difficult. The full multi-disciplinary team will be present at outreach clinics.

Multi Disciplinary Approach to Specialist Paediatric CF 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 must 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 team will 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 must demonstrate continuing professional

development (CPD) in CF.

The service will have processes in place to ensure adequate workforce planning.
The service will be able to demonstrate that an appraisal process is in place for all staff.

Study days and network meetings will be run by the service for core and wider workforce teams.

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

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

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 Clinical Director/Lead CF Consultant

The consultant in charge of the paediatric service must have a certificate of training (CCT) in paediatrics with accreditation in paediatric respiratory medicine or equivalent in cumulative experience. He/she must also have at least three years experience working as a consultant in an accredited paediatric 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 multidisciplinary team and clinical governance of the service.

Cystic Fibrosis Nurse Specialists

CF Nurse Specialists must meet the standards identified in the CF Trust document "National Consensus Standards for the Nursing Management of Cystic Fibrosis" May, 2001.

Nurse Specialists will be members of the UK Cystic Fibrosis Nursing Association and must work within a CF multidisciplinary team.

All nurse specialists must be registered with the Nurses and Midwives Council and those working with children must have undergone specific paediatric training.

Physiotherapists

Specialist CF Physiotherapists must 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 will be members of the ACPCF special interest group.

Dieticians

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

Medical Staffing

Specialist Consultants must 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. Specialist Consultants will also have CCT in paediatrics with accreditation in paediatric respiratory medicine.

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

Pharmacists

Pharmacists must be registered with the General Pharmaceutical Council and be a member of the Cystic Fibrosis Pharmacists Group. Pharmacists' practice will reflect 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 be produced by a consultant and agreed with the patient as a result of every annual review undertaken.

Outpatient Care

Routine appointments should 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, a physiotherapist and dietician at all routine reviews. There should be access to psycho-social support.

Inpatient Care

Beds in a ward suitable for cystic fibrosis care must be available within 24 hours for an emergency admission, as well as capacity to ensure elective and urgent admissions can be

managed appropriately. There must 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 CF 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 play and recreational facilities 7 days a week,
- have provision for appropriate vascular access available at all times,
- have facilities for sedation for procedures (e.g. line insertion) 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.

Patients must be admitted to wards that are familiar with the care and management of children with this condition and have developed the required expertise.

Nurses on the inpatient wards require specific expertise, and 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.

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 must include evenings and weekends.

IV Antibiotics

The service must 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 must 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 or network care hospital.

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 community services to support them 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 including children's 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 their carers in administering the IVs as well as the suitability of the home environment. There must 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.
- Liaison with nurseries, 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 is 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 to enteral feeds, feeding pumps, nasogastric (NG) tubes, percutaneous endoscopic gastrostomy (PEG) tubes and gastrostomy buttons.

GPs 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, 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.
- Epithelial ion transport testing (where required this facility will be available by collaborative arrangements with an appropriately equipped specialist CF Centre).
- Facilities to undertake bronchoscopy.

Other aspects of Paediatric CF Specialist Care

Diabetes Care

Management of CF related diabetes will be 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 IP and OP facility. 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 must be made jointly between the relevant surgeon, the CF clinicians and the child and their parents or carers. Acute admissions for acute abdominal pain will 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 must be available to ensure that the child's CF needs are fully taken into account, including during any post operative period of inpatient care. A clear care plan must be developed, with regular contact and review between the relevant parties.

The surgical units must have a protocol or guideline relating to children with CF which has been developed in collaboration with the CF service. This protocol will specify required

standards of cross infection control and dietary/physiotherapy support.

General anaesthetic must be undertaken by a paediatric anaesthetist with experience of CF, and conducted within appropriate facilities in accordance with the Royal College of Surgeons' publication regarding quality standards for paediatric surgery; Surgery for Children; Delivering a First Class Service (2008).

The surgical service must 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 child and family as early as possible. Access to information will be readily available to patients and their families.

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 demonstrate:

- Good working relationships with and access to the general palliative care team attached to the hospital/local hospice/local community team and their involvement in all such patients.
- Clear documentation of End of Life discussions.
- Access to bereavement support for families.
- Clinical review and debrief following a patient death.

Infection Control

Policy and procedures must be 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). All children will be admitted to a facility which provides specialist CF care in single rooms with en-suite facilities.

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 (FEV1) and rate of decline.
- Body Mass Index (as a percentile for children).
- 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.

General Paediatric Care

When treating children, the service will additionally follow the standards and criteria outlined in the Specification for Children's Services (attached as Annex 1 to this Specification).

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 paediatric services for patients with cystic fibrosis.
Paediatric services shall be provided for patients up to the age of 18.

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. These will include:

- Antenatal diagnosis of CF.
- A newborn screening result that suggests a high likelihood of CF.
- Clinical suspicion by a general paediatrician, GP or other hospital specialist.
- Neonatal diagnosis of meconium ileus.

Diagnostic services will be provided for patients suspected of having cystic fibrosis.
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/parents.
- Early introduction of required treatment.

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 provide access or referral to specialists within:

- Endocrinology, including diabetes (with an interest in CF related diabetes), with joint clinics available on a regular basis,
- Hepatology,
- Gastroenterology,
- Rheumatology,
- ENT,
- Vascular services,
- Thoracic surgery,
- Palliative care,
- Clinical genetics,
- Transplantation services,
- Psychiatry,
- Paediatric Surgery,
- Gynaecology,
- Renal services,
- Anaesthetic services,
- Gastro-intestinal surgery,
- Pre-natal and new-born screening services.

If not available at a network care 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 multidisciplinary team (minimum of doctor, nurse specialist, physiotherapist and dietician) must be available for care of in- and out-patients. All children will be seen by a specialist physiotherapist and dietitian at each routine clinic visit.

Policy and procedures must be in place to protect patients from the risk of cross-infection, both as in- and out-patients. All children will be admitted to a facility which provides specialist CF care in single rooms with en-suite facilities

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).

Network clinics - There will be regular locally run CF multi-disciplinary team outpatient clinics some of which will be joint clinics with the specialist CF centre that co-ordinates the network. Each patient will be seen by the full specialist multi-disciplinary team at least twice /year, either in a joint clinic at the network centre or at the specialist centre.

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
- Management of Cystic Fibrosis-related Diabetes Mellitus (2004)
- Department of Health National Definition Set for Cystic Fibrosis (2009)
- 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

ANNEX 1 TO SERVICE SPECIFICATION:

PROVISION OF SERVICES TO CHILDREN

Aims and objectives of service

This specification annex applies to all children's services and outlines generic standards and outcomes that would fundamental to all services.

The generic aspects of care:

The Care of Children in Hospital (Health Service Circular 1998/238) requires that:

- Children are admitted to hospital only if the care they require cannot be as well provided at home, in a day clinic or on a day basis in hospital.
- Children requiring admission to hospital are provided with a high standard of medical, nursing and therapeutic care to facilitate speedy recovery and minimize complications and mortality.
- Families with children have easy access to hospital facilities for children without needing to travel significantly further than to other similar amenities.
- Children are discharged from hospital as soon as socially and clinically appropriate and full support provided for subsequent home or day care.
- Good child health care is shared with parents/carers and they are closely involved in the care of their children at all times unless, exceptionally, this is not in the best interest of the child. Accommodation is provided for them to remain with their children overnight if they so wish.

Service description/care pathway

All paediatric specialised services have a component of primary, secondary, tertiary and even quaternary elements.

The efficient and effective delivery of services requires children to receive their care as close to home as possible dependent on the phase of their disease.

Services should therefore be organised and delivered through "integrated pathways of care" (National Service Framework for children, young people and maternity services (Department of Health & Department for Education and Skills, London 2004)

Interdependencies with other services

All services will comply with *Commissioning Safe and Sustainable Specialised Paediatric Services: A Framework of Critical Inter-Dependencies* – Department of Health (DH)

Imaging

All services will be supported by a 3 tier imaging network ('Delivering quality imaging services for children' DH 13732 March 2010). Within the network:

- It will be clearly defined which imaging test or interventional procedure can be performed and reported at each site
- Robust procedures will be in place for image transfer for review by a specialist radiologist, these will be supported by appropriate contractual and information governance arrangements
- Robust arrangements will be in place for patient transfer if more complex imaging or intervention is required
- Common standards, protocols and governance procedures will exist throughout the network.
- All radiologists, and radiographers will have appropriate training, supervision and access to continuing professional development
- All equipment will be optimised for paediatric use and use specific paediatric software.

Specialist Paediatric Anaesthesia

Wherever and whenever children undergo anaesthesia and surgery, their particular needs must be recognised and they should be managed in separate facilities, and looked after by staff with appropriate experience and training.1 All UK anaesthetists undergo training which provides them with the competencies to care for older babies and children with relatively straightforward surgical conditions and without major co-morbidity. However those working in specialist centres must have undergone additional (specialist) training2 and should maintain the competencies so acquired3 *. These competencies include the care of very young/premature babies, the care of babies and children undergoing complex surgery and/or those with major/complex co-morbidity (including those already requiring intensive care support).

As well as providing essential co-dependent service for surgery specialist anaesthesia and sedation services may be required to facilitate radiological procedures and interventions (for example MRI scans and percutaneous nephrostomy) and medical interventions (for example joint injection and intrathecal chemotherapy), and for assistance with vascular access in babies and children with complex needs such as intravenous feeding.

Specialist acute pain services for babies and children are organised within existing departments of paediatric anaesthesia and include the provision of agreed (hospital wide) guidance for acute pain, the safe administration of complex analgesia regimes including epidural analgesia, and the daily input of specialist anaesthetists and acute pain nurses with expertise in paediatrics.

*The Safe and Sustainable reviews of paediatric cardiac and neuro- sciences in England have noted the need for additional training and maintenance of competencies by specialist anaesthetists in both fields of practice.

References

1. Guidelines for the Provision of Anaesthetic Services (GPAS) Paediatric anaesthetic services. RCoA 2010 www.rcoa.ac.uk
2. Certificates of Completion of Training (CCT) in Anaesthesia 2010
3. Continuing Professional Development (CPD) matrix level 3

Specialised Child and Adolescent Mental Health Services (CAMHS)

The age profile of children and young people admitted to specialised CAMHS day/in-patient settings is different to the age profile for paediatric units in that it is predominantly adolescents who are admitted to specialised CAMHS in-patient settings, including over-16s. The average length of stay is longer for admissions to mental health units. Children and young people in specialised CAMHS day/in- patient settings generally participate in a structured programme of education and therapeutic activities during their admission.

Taking account of the differences in patient profiles the principles and standards set out in this specification apply with modifications to the recommendations regarding the following:

- Facilities and environment – essential Quality Network for In-patient CAMHS (QNIC) standards should apply (http://www.rcpsych.ac.uk/quality/quality,accreditationaudit/qnic1.aspx)
- Staffing profiles and training - essential QNIC standards should apply.
- The child/ young person's family are allowed to visit at any time of day taking account of the child / young persons need to participate in therapeutic activities and education as well as any safeguarding concerns.
- Children and young people are offered appropriate education from the point of admission.
- Parents/carers are involved in the child/young persons care except where this is not in the best interests of the child / young person and in the case of young people who have the capacity to make their own decisions is subject to their consent.
- Parents/carers who wish to stay overnight are provided with accessible accommodation unless there are safeguarding concerns or this is not in the best interests of the child/ young person.

Applicable national standards e.g. NICE, Royal College

Children and young people must receive care, treatment and support by staff registered by the Nursing and Midwifery Council on the parts of their register that permit a nurse to work with children (Outcome 14h Essential Standards of Quality and Safety, Care Quality Commission, London 2010)

- There must be at least two Registered Children's Nurses (RCNs) on duty 24 hours a day in all hospital children's departments and wards.

- There must be an Registered Children's Nurse available 24 hours a day to advise on the nursing of children in other departments (this post is included in the staff establishment of 2RCNs in total).

Accommodation, facilities and staffing must be appropriate to the needs of children and separate from those provided for adults. All facilities for children and young people must comply with the Hospital Build Notes HBN 23 Hospital Accommodation for Children and Young People NHS Estates, The Stationary Office 2004.

All staff who work with children and young people must be appropriately trained to provide care, treatment and support for children, including Children's Workforce Development Council Induction standards (Outcome 14b Essential Standards of Quality and Safety, Care Quality Commission, London 2010).

Each hospital who admits inpatients must have appropriate medical cover at all times taking account of guidance from relevant expert or professional bodies (National Minimum Standards for Providers of Independent Healthcare, Department of Health, London 2002)."Facing the Future" Standards, Royal College of Paediatrics and Child Health.

Staff must carry out sufficient levels of activity to maintain their competence in caring for children and young people, including in relation to specific anaesthetic and surgical procedures for children, taking account of guidance from relevant expert or professional bodies (Outcome 14g Essential Standards of Quality and Safety, Care Quality Commission, London 2010).

Providers must have systems in place to gain and review consent from people who use services, and act on them (Outcome 2a Essential Standards of Quality and Safety, Care Quality Commission, London 2010). These must include specific arrangements for seeking valid consent from children while respecting their human rights and confidentiality and ensure that where the person using the service lacks capacity, best interest meetings are held with people who know and understand the person using the service. Staff should be able to show that they know how to take appropriate consent from children, young people and those with learning disabilities (Outcome 2b) (Seeking Consent: working with children Department of Health, London 2001).

Children and young people must only receive a service from a provider who takes steps to prevent abuse and does not tolerate any abusive practice should it occur (Outcome 7 Essential Standards of Quality and Safety, Care Quality Commission, London 2010 defines the standards and evidence required from providers in this regard). Providers minimise the risk and likelihood of abuse occurring by:

- Ensuring that staff and people who use services understand the aspects of the safeguarding processes that are relevant to them
- Ensuring that staff understand the signs of abuse and raise this with the right person when those signs are noticed.
- Ensuring that people who use services are aware of how to raise concerns of abuse.
- Having effective means to monitor and review incidents, concerns and complaints that have the potential to become an abuse or safeguarding concern.
- Having effective means of receiving and acting upon feedback from people who use services and any other person.
- Taking action immediately to ensure that any abuse identified is stopped
- and suspected abuse is addressed by:
    - having clear procedures followed in practice, monitored and reviewed that take account of relevant legislation and guidance for the management of alleged abuse

    - separating the alleged abuser from the person who uses services and others who may be at risk or managing the risk by removing the opportunity for abuse to occur, where this is within the control of the provider
    - reporting the alleged abuse to the appropriate authority
    - reviewing the person's plan of care to ensure that they are properly supported following the alleged abuse incident.
- Using information from safeguarding concerns to identify non-compliance, or any risk of non-compliance, with the regulations and to decide what will be done to return to compliance.
- Working collaboratively with other services, teams, individuals and agencies in relation to all safeguarding matters and has safeguarding policies that link with local authority policies.
- Participates in local safeguarding children boards where required and understand their responsibilities and the responsibilities of others in line with the Children Act 2004.
- Having clear procedures followed in practice, monitored and reviewed in place about the use of restraint and safeguarding.
- Taking into account relevant guidance set out in the Care Quality Commission's Schedule of Applicable Publications
- Ensuring that those working with children must wait for a full CRB disclosure before starting work.
- Training and supervising staff in safeguarding to ensure they can demonstrate the competences listed in Outcome 7E of the Essential Standards of Quality and Safety, Care Quality Commission, London 2010

All children and young people who use services must be:

- Fully informed of their care, treatment and support.
- Able to take part in decision making to the fullest extent that is possible.
- Asked if they agree for their parents or guardians to be involved in decisions they need to make.

(Outcome 4I *Essential Standards of Quality and Safety*, Care Quality Commission, London 2010)

Key Service Outcomes

Evidence is increasing that implementation of the national Quality Criteria for Young People Friendly Services (Department of Health, London 2011) have the potential to greatly improve patient experience, leading to better health outcomes for young people and increasing socially responsible life-long use of the NHS.

Implementation is also expected to contribute to improvements in health inequalities and public health outcomes e.g. reduced teenage pregnancy and STIs, and increased smoking cessation. All providers delivering services to young people should be implementing the good practice guidance which delivers compliance with the quality criteria.

Poorly planned transition from young people's to adult-oriented health services can be associated with increased risk of non adherence to treatment and loss to follow-up, which

can have serious consequences. There are measurable adverse consequences in terms of morbidity and mortality as well as in social and educational outcomes. When children and young people who use paediatric services are moving to access adult services (for example, during transition for those with long term conditions), these should be organised so that:

- All those involved in the care, treatment and support cooperate with the planning and provision to ensure that the services provided continue to be appropriate to the age and needs of the person who uses services.

The National Minimum Standards for Providers of Independent Healthcare, (Department of Health, London 2002) require the following standards:

- **A16.1** Children are seen in a separate out-patient area, or where the hospital does not have a separate outpatient area for children, they are seen promptly.
- **A16.3** Toys and/or books suitable to the child's age are provided.
- **A16.8** There are segregated areas for the reception of children and adolescents into theatre and for recovery, to screen the children and adolescents from adult patients; the segregated areas contain all necessary equipment for the care of children.
- **A16.9** A parent is to be actively encouraged to stay at all times, with accommodation made available for the adult in the child's room or close by.
- **A16.10** The child's family is allowed to visit him/her at any time of the day, except where safeguarding procedures do not allow this
- **A16.13** When a child is in hospital for more than five days, play is managed and supervised by a qualified Hospital Play Specialist.
- **A16.14** Children are required to receive education when in hospital for more than five days; the Local Education Authority has an obligation to meet this need and are contacted if necessary.
- **A18.10** There are written procedures for the assessment of pain in children and the provision of appropriate control.

All hospital settings should meet the Standards for the Care of Critically Ill Children (Paediatric Intensive Care Society, London 2010).

There should be age specific arrangements for meeting Regulation 14 of the Health and Social Care Act 2008 (Regulated Activities) Regulations 2010. These require:

- A choice of suitable and nutritious food and hydration, in sufficient quantities to meet service users' needs
- Food and hydration that met any reasonable requirements arising from a service user's religious or cultural background
- Support, where necessary, for the purposes of enabling service users to eat and drink sufficient amounts for their needs
- For the purposes of this regulation, "food and hydration" includes, where applicable, parenteral nutrition and the administration of dietary supplements where prescribed
- Providers must have access to facilities for infant feeding, including facilities to support breastfeeding (Outcome 5E, of the Essential Standards of Quality and Safety, Care Quality Commission, London 2010)

All paediatric patients should have access to appropriately trained paediatric trained dieticians, physiotherapists, occupational therapists, speech and language therapy, psychology, social work and CAMHS services within nationally defined access standards.

All children and young people should have access to a professional who can undertake an assessment using the Common Assessment Framework and access support from social care, housing, education and other agencies as appropriate.

All registered providers must ensure safe use and management of medicines, by means of the making of appropriate arrangements for the obtaining, recording, handling, using, safe keeping, dispensing, safe administration and disposal of medicines (Outcome 9 Essential Standards of Quality and Safety, Care Quality Commission, London 2010). For children, these should include specific arrangements that:

- They are supported to have a health action plan
- Facilities meet the appropriate requirements of the Disability Discrimination Act 1995

They meet the standards set out in Transition: getting it right for young people. Improving the transition of young people with long-term conditions from children's to adult health services. Department of Health, 2006, London.

Appendix Two

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			
Children (above 5 years) attending a specialist CF centre should have a FEV1 of >85%	< 55%	UK CF Registry data	Internal review with report to commissioners External review for outliers
Median BMI percentile of centres should be 50% or greater	Median BMI percentile less than 50	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 children should have an annual review and management plan discussed with the family	<85%	Self report dashboard	Internal review and submission of improvement plan
All children 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 children over the age of 6 years should be offered inhaled mucolytic therapy	< 70%	UK CF Registry data	Internal review
There should be no delay (as defined by national service specification) in initiating IV antibiotic therapy for pulmonary exacerbation	Breach percentage >10%	Self report /dashboard/CQIN	Commissioner review
Domain 4: Ensuring that people have a positive experience of care			
All children 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 and	Yes/No	Self report demonstrating systematic	Commissioner review

Quality Requirement	Threshold (trigger for breach)	Method of Measurement	Consequence of breach
carer experience and satisfaction at a frequency driven by patient need		engagement and feedback on actions 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 have a low prevalence of chronic pseudomonas infection	>14%	UK CF Registry	Internal review with report to commissioners Commissioner review for outliers
All children chronically infected with Pseudomonas Aeruginosa should receive inhaled antibiotic therapy. Percentage of patients with chronic PA on inhaled antibiotics by age group	< 85%	UK CF Registry data	Internal review with report to commissioners Commissioner review for outliers