GIG CYMRU NHS WALES
Cyd-bwyllgor Comisiynu
Joint Commissioning Committee

Cystic Fibrosis Adults and Young People

Service Specification: SS193

Policy Proposal

June 2024
Version: 1.0

Document information	
Document purpose	Service Specification
Document name	Cystic Fibrosis – Adults and Young People
Author	NHS Wales Joint Commissioning Committee
Publication date	January 2012
Commissioning Team	Cardiac
Target audience	Chief Executives, Medical Directors, Directors of Finance, Nurse Directors Director Adult and Young People CF Services, Adult and Young People CF Service Managers, CF Specialist teams
Description	NHS Wales propose to routinely commission this specialised service in accordance with the criteria described in this policy
Document No	SS193
Review Date	2024

GIG CYMRU NHS WALES | Cyd-bwyllgor Comisiynu Joint Commissioning Committee

Contents

Statement

NHS Wales Joint Commissioning Committee (NWJCC) will commission the service of Cystic Fibrosis (CF) for Adults and Young People (aged 16 years of age and over) in accordance with the criteria outlined in this specification.

In creating this document NWJCC has reviewed the requirements and standards of care that are expected to deliver this service.

Welsh Language

NWJCC is committed to treating the English and Welsh languages on the basis of equality, and endeavour to ensure commissioned services meet the requirements of the legislative framework for Welsh Language, including the Welsh Language Act (1993), the Welsh Language (Wales) Measure 2011 and the Welsh Language Standards (No.7) Regulations 2018.

Where a service is provided in a private facility or in a hospital outside of Wales, the provisions of the Welsh language standards do not directly apply but in recognition of its importance to the patient experience, the referring health board should ensure that wherever possible patients have access to their preferred language.

In order to facilitate this, NWJCC is committed to working closely with providers to ensure that in the absence of a Welsh speaker, written information will be offered and people have access to either a translator or 'Language-line' if requested. Where possible, links to local teams should be maintained during the period of care.

Decarbonisation

NWJCC is committed to taking assertive action to reducing the carbon footprint through mindful commissioning activities. Where possible and taking into account each individual patient's needs, services are provided closer to home, including via digital and virtual access, with a delivery chain for service provision and associated capital that reflects the NWJCC commitment.

Disclaimer

NWJCC assumes that healthcare professionals will use their clinical judgment, knowledge and expertise when deciding whether it is appropriate to apply this document.

This document may not be clinically appropriate for use in all situations and does not override the responsibility of healthcare professionals to make decisions appropriate to

the circumstances of the individual patient, in consultation with the patient and/or their carer or guardian, or Local Authority.

NWJCC disclaims any responsibility for damages arising out of the use or non-use of this policy.

1. Introduction

This document has been developed as the Service Specification for the planning and delivery of Cystic Fibrosis (CF) services for adults and young people (aged 16 years and over) for people resident in Wales. This service will only be commissioned by the NHS Wales Joint Commissioning Committee (NWJCC) and applies to residents of all seven Health Boards in Wales.

1.1 Background

Cystic fibrosis (CF) is a multi-system genetic disorder affecting the lungs, pancreas, liver and intestines. It results from mutations affecting a gene that encodes for a chloride channel called the CF transmembrane conductance regulator (CFTR), which is essential for the regulation of salt and water movements across cell membranes (Cystic Fibrosis Trust's Standards for the clinical care of children and adults with cystic fibrosis in the UK). Absent or reduced function of CFTR results in thickened secretions in the lungs, digestive system and other organs.

The UK Cystic Fibrosis Registry Annual Data Report 2019 reports that 10,655 people in the UK have CF in 2018, with 193 people newly diagnosed that year.

Diagnosis is primarily made during newborn screening. The Annual Data Report 2018 19shows the median age at diagnosis is 2 months, and one in every 2500 babies born in the UK has CF.

Cystic fibrosis can have a significant impact on life expectancy and quality of life. The Annual Data Report 2019 shows the current median age at death is 31 years and the median predicted survival is 49 years. More than 60% of people on the UK cystic fibrosis registry are aged over 16 years.

Lung function is often reduced in CF, and lung infections are a cause of significant morbidity. For example, 44.2% of adults with CF have chronic Pseudomonas aeruginosa infection and 19.9% have chronic Staphylococcus aureus infection. Such chronic infection may need long-term use of antibiotics.

In about 85% of cases the pancreatic exocrine ducts become sufficiently blocked to cause maldigestion and intestinal malabsorption (Cystic Fibrosis Trust's Standards for the clinical care of children and adults with cystic fibrosis in the UK). Infants may fail to thrive and older children and adults may become under-nourished

1.2 Aims and Objectives

The aim of this service specification is to define the requirements and standards of care essential for delivering CF services.

The service should aim to deliver care that will improve life expectancy and enhance quality of life by providing high quality care at the point of need, with optimised treatment and management, led by a specialist team trained to provide multidisciplinary specialist CF care.

The objectives of this service specification are to:

- Ensure minimum standards of care are met.
- Ensure equitable access to CF services.
- Identify centres that are able to provide specialist CF services for people with CF and resident in Wales.
- Improve outcomes for people accessing CF services.
- Make a timely diagnosis with appropriate counselling and psychological support to people with CF and their family or carer.
- Provide high quality proactive and preventative treatment and care to ensure optimal lung function, nutritional status and management of complications associated with CF.
- Ensure a safe, cost effective, high quality service for the recipients of the services commissioned.
- Facilitate autonomy and transition to adult care and encouraging independent care.
- Support people 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.
- Ensure effective communication between people with CF and service providers.
- Provide a personal service, sensitive to physical, psychological and emotional needs of people with and their families or carers.

1.3 Relationship with other documents

This document should be read in conjunction with the following documents:

- **NHS Wales**
  - All Wales Policy: Making Decisions in Individual Patient Funding requests (IPFR).
  - COVID-19 rapid guideline: cystic fibrosis. NICE Guideline (NG170), October 2020
  - Cystic Fibrosis Modulator Therapies, Policy Position Statement, (PP198). NWJCC, January 2021

- The CF Trust: Standards for the Clinical Care of Children and Adults with Cystic Fibrosis in the UK (2011)
- Cystic Fibrosis: diagnosis and Management, NICE Guideline (NG78), October 2017.
- Cystic Fibrosis – Adults, NHS England Service Specification, A01/S/a
- Transition from children's to adults' services for young people using health or social care services. NICE Guideline (NG43), February 2016

2. Service Delivery

The NHS Wales Joint Commissioning Committee will commission a Cystic Fibrosis (CF) service for adults and young people (aged 16 years of age and over) in Wales in-line with the criteria identified in this specification.

2.1 Service description

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).

This service specification does not include general healthcare services such as general practice services, dental, or ophthalmologic services required by individuals. However, close liaison is vital between CF services and general services, and the CF service must have processes in place to ensure that effective communication takes place.

The service provider will demonstrate that they are meeting, or with the support of their commissioners are working towards meeting the requirements for CF 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 the commissioners.

Care will be directed by the specialist centres.

Specialist Centre Responsibilities

- The Specialist Centre is responsible for providing the care plan for all patients. This includes responsibility for determining and prescribing high cost drugs (such as inhaled mucoactives and antibiotics, anti-fungals and the more recent CFTR modulator therapies[1]), in accordance with the national commissioning policy.

- The Centre will:
  - provide urgent care needs and advice 24 hours a day, 7 days a week
  - have systems in place to enable appropriate cover for annual and study leave and any long term absences
  - have telephone advice available, with contact numbers given to patients to enable them to obtain advice from the specialist team at any time
  - be fully operational and able to take referrals at all times

[1] WHSSC, Policy Position Statement , PP198 Cystic Fibrosis Modulator Therapies

- have policies and procedures in place to protect patients from the risk of cross infection both as in-patients and out-patients (Cystic Fibrosis Standards of Care and subsequent updates to Infection Control Standards)
- have a Service Director/Lead CF Consultant and a dedicated Service Manager (see section 2.4)

Provision of Care

General

- All staff working within the CF Service have an obligation to undertake continuing education and training to ensure knowledge and skills remain up to date.
- Core members of the CF team should be members of and regularly contribute to their relevant specialist interest group.
- Each professional group are required to meet the minimum competencies defined within section 3 of the CF Trust Standards for the Clinical Care of Children and Adults with Cystic Fibrosis in the UK (2011) and the defined care pathway. Care will be delivered by a multidisciplinary team of trained, experienced, specialist healthcare professions who routinely care for a critical mass of CF patients at a specialist centre.
- The levels of staffing should be within the recommendations set out in section 3 of the Cystic Fibrosis Trust document Standards for the Clinical Care of Children and Adults with Cystic Fibrosis in the UK (2011).
- A full annual review should be undertaken by the specialist centre, 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 should developed by a consultant and agreed with the patients as a result of the annual review.

Outpatient care

- A dedicated outpatient facility should be available for routine outpatient care.
- A separate outpatient facility should be available for special infection CF patients unable to attend 'normal CF clinics'; allowing for timely review at the point of need.
- Video-conferencing/telephone consultation should be available and use of these facilities encouraged for patient review.
- Individuals should be seen by the multidisciplinary team in a CF clinic (or via video-conferencing) every 3 – 6 months dependant on their individual requirements. Problems that are more active or chronic disease severity may require more frequent reviews by the multidisciplinary team.
- The outpatient clinic should be multidisciplinary and patients should be reviewed by any of the specialist team dependant on their individual requirements.
- CF patients and their families should have access to appropriate counselling and psychosocial support. The service should ensure that facilities are available to

support the best quality of care between home and hospital. Patients can be seen routinely in an outpatient facility or via video-conferencing but there needs to be provision for urgent review and for providing the first dose of an antibiotic course either in an outpatient setting or a day-case facility or ward.

- The facilities should take into consideration the need for infection control 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 routine clinic review, annual reviews, treatment, day case, exercise and physiotherapy etc. This should include ensuring that CF patients are not kept waiting in communal areas and that they remain segregated from each other at all times so as to minimise the risk of cross infection.
- Outreach satellite clinics/videoconferencing clinics should be provided for adults and young people with geographical constraints that make attendance at the specialist centre more difficult. The full multidisciplinary team should be available to provide appropriate review of patients when required.

Inpatient Care

- Inpatient facilities should 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).
- Inpatient care should be available within 24 hours for an urgent admission. Inpatient capacity should be available to ensure elective and urgent admissions can be managed appropriately.
- There should be no more than a 7-day delay between the proposed admission date and the start of the routine/ planned course of treatment.
- The in-patient service should have an infection control policy in place which demonstrates compliance with the Cystic Fibrosis Trust Standards of Clinical Care of Children and Adults with Cystic Fibrosis and subsequent updates to Infection Control Standards.
- Provision of dedicated inpatient facilities should include single rooms with en-suite facilities for every CF patient to minimise the risk of cross infection to enable them to continue life as normally as possible.
- Nurses on the inpatient wards require specific expertise, and should be committed to the CF service, with regular input and training from the specialist CF nurses. Patients need to be admitted to a ward staffed by CF specialists or admitted to wards that are familiar with the care and management of individuals with CF and have developed the required expertise.

- In-patients should:
  - 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 7 days a week
- have provision for appropriate vascular access available at all times
- have an assessment of their exercise capacity and access facilities and support to continue their exercise programme (as appropriate), taking into account the need to prevent cross-infection
- have access to a choice of food including high-energy healthy eating options and access to high-energy mid-meal snacks and drinks, and healthy option supplementary drinks.
- have access to the internet to continue with their education or work and connect with their family/carer support.

Home Care

The life-long multi-system nature of CF means that a complex regimen of home treatment is often recommended. Many patients and families require regular and consistent outreach from the multidisciplinary team in this care.

Home care includes:

- Support in the community by the specialist CF multidisciplinary team.
- Multidisciplinary support through virtual contact platforms.
- Open access to nursing care in the community, this may be the CF specialist nurse from the CF centre or local community nurses who have had specific training, experience and supervision in CF.
- Patients undertaking home IV antibiotic therapy should have a formal assessment of suitability. This should include formal training and an assessment of competency of the patient and/or their carer's in administering the IVs as well as the suitability of the home environment. There should also be a 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.
- Psychosocial support.
- Liaison with school or college for patients still in education.
- Support through times of change in an individual's health including the introduction of treatment for diabetes or home oxygen therapy and end of life care.
- Where clinically appropriate, home treatment should be encouraged.

IV Antibiotic Service

- The service should have the ability to commence IV antibiotics on any day of the week. An urgent course of treatment should be implemented within a maximum of 24 hours of the clinical decision being made.
- There should 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.

Access to Equipment

- The service should ensure that there is access to the provision of high quality spirometry for all appropriate patients with separate spirometry equipment for each microbiological organism available to patients. This should be either available through the use of cloud based devices to enable patients to be reviewed via video conferencing or made available to the Home Care Team and satellite clinics.
- Patients who need home oxygen therapy should receive timely assessment and prescription of oxygen according to the National Home Oxygen Service.
- Patients should have access to a range of clinically appropriate airway clearance devices.
- There should be a comprehensive nebuliser service, which aims to provide devices that deliver drugs in a fast and efficient manner. The service also needs to provide a range of mechanical devices required to provide intermittent positive pressure breathing and non-invasive ventilation where needed.
- Patients should have access to blood sugar monitors or continuous glucose monitoring systems.
- In-patients should have access to enteral feeds, feeding pumps, nasogastric (NG) tubes, percutaneous endoscopic gastrostomy (PEG) tubes and gastrostomy buttons. Primary care should provide NG tubes, feeds and feeding pumps and giving sets for enteral feeding through an approved/agreed contractor or local community nursing service.

Access to Diagnostics

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

- A Laboratory that meets the Laboratory Standards for Processing Microbiological samples from people with Cystic Fibrosis and that routinely cultures for recognised CF pathogens such as Burkholderia cepacia complex and atypical mycobacteria, and a biochemistry laboratory for processing of aminoglycoside levels including tobramycin.
- A Laboratory that performs specialist biochemical analysis such as faecal elastase and complies with the Association for Clinical Biochemists guidelines on performance of sweat tests.

- A specialist radiology service including contrast GI studies for bowel obstruction, ventilation perfusion scans, CT Thorax, angiography, specialist liver scans, DXA bone scans and interventional services.
- Specialist lung function laboratory that can test patients as well as provide support and training for those staff performing spirometry in the clinical setting.
- Facilities to undertake bronchoscopy.

Staffing

- All multi-disciplinary CF, staff commissioned to provide specialist CF care within the adult and young people CF service should not be expected to provide cover to other areas within the hospital setting without agreement from the service director, service manager or commissioning service.
- The service will ensure that processes are in place to ensure adequate workforce planning.
- The service will demonstrate that an appraisal process is in place for all staff.
- Regular audit of services should be performed. Specific audits may be requested by the commissioner. Participation in research studies should be encouraged.
- The service should participate in and demonstrate quality improvements projects.
- Each professional group is required to meet the minimum competencies defined within section 3 of the 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 should be achieved:
    - 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 multidisciplinary teams shall be members of and regularly contribute to their relevant specialist interest group.
    - Attendance at National/International specialist conferences should be demonstrable. It is recognised that not all staff will be able to attend every meeting every year; therefore, the service should be able to demonstrate that there are internal mechanisms for feedback to the multidisciplinary team.
    - Each member of each professional group should demonstrate Continuing Professional Development (CPD) in CF.
    - Specialist centres need to provide cover for annual leave, study leave and long term absence (e.g. long-term sickness or maternity leave).

The Specialist Centre should have a:

- Service Director/Lead CF Consultant
    - The Service Director/Lead CF Consultant is responsible for all aspects of the clinical delivery of the service, ensuring standards are met and the service is continually developed to meet the current and future needs of the adult and young people CF population in Wales

  - The Service Director/Lead CF Consultant of the adult and young people service must have appropriate experience working as a consultant in an accredited CF centre.
  - The Service Director/Lead CF Consultant should be able to demonstrate active participation and attendance at National/international meetings and have a record of accomplishment in teaching, audit and research.
  - The Service Director/Lead CF Consultant should engage in the management of the service as a whole, ensuring leadership of the multi-disciplinary team and clinical governance of the service.

- CF Service Manager
  - The CF service manager should have dedicated time and managerial responsibility for the delivery of an efficient, effective service in close liaison with the service director to ensure standards are met and service continually developed.

Multi-disciplinary Specialist Team

Medical Staffing

- Specialist Consultants should have had training in a recognised CF centre. They should be able to demonstrate active participation and attendance at National/International meetings and have a record of accomplishment in teaching, audit and research.
- Middle grade medical support should in most circumstances comprise speciality doctors within the CF service.

Cystic Fibrosis Clinical Nurse Specialists

- CF Nurse Specialists should meet the standards identified in the CF Trust document 'National Consensus Standards for Nursing Management of Cystic Fibrosis' May 2001. Nurse specialists shall be members of the UK Cystic Fibrosis Nursing Association and should work within a CF multidisciplinary team.

Specialist Physiotherapists

- Specialist CF Physiotherapists should meet the standards identified in the Association of Chartered Physiotherapists in Cystic Fibrosis (ACPCF) document[2] 'Physiotherapy National Standards of Care for people with Cystic Fibrosis 2017'. They shall be members of the ACPCF special interest group.

Specialist Dieticians

[2] Standards of Care and Good Clinical Practice for the Physiotherapy Management of Cystic Fibrosis

- Specialist CF Dieticians should meet the standards defined in 'Nutritional Management of Cystic Fibrosis' (April 2016) and shall be members of the UK Cystic Fibrosis Nutrition Group.

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 Worker

- 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).

Specialist 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.

Administrative Staff

- The adult and young people CF service should have dedicated secretarial provision to ensure, timely, appropriate and extensive administrative services to the full clinical team and MDT. In addition, dedicated data entry clerk provision is important for the timely input of annual review data into the UK CF registry.

Other aspects of Adult and Young People CF Specialist Care

CF Related diabetes (CFRD)

- Management of CF related diabetes should be in accordance with the document 'Management of Cystic Fibrosis Related Diabetes Mellitus (2004). In particular, there will be joint management, including joint outpatient and inpatient review, between the CF multi-disciplinary team and diabetes specialists experienced in the management of CF related diabetes (CFRD).
- The provider must have a documented protocol, which describes how CFRD will be identified, and the provider will undertake an annual audit, which demonstrates compliance with the protocol.

Transitional Care

- Transition should be planned with the patient and their parent or carer with due regard to patient choice. There should be an underlying assumption that transition is natural and expected. Transition planning should be developmentally appropriate.
- All parents or carers should be made aware as early as possible that transition into adult and young people services will take place.

- Arrangements for transition to adult and young people services should commence from around the age of 14 but the age for transfer should be agreed between the patient, the paediatric and adult centres and determined by the individuals' maturity and preparedness.
- All CF services should have a coordinated and documented pathway for transition from children's to adults' services that includes plans for managing all cystic-fibrosis related aspects of care.
- A joint first outpatient review in clinic within the adult and young people service should take place for formal handover; this should include a detailed clinical handover.
- Transition documentation and referral should be completed and forwarded to the adult and young people service prior to first review in the clinic setting.
- Specialist adult and young people CF centres should demonstrate that they are actively engaging in the transition process for each child via an annual report to commissioners of the experience of patients who transitioned during the year.
- The adult and young people CF service MDT and clinicians should attend relevant pre-arranged joint transition clinics in paediatric centres.

Surgery

- The decision to undertake surgery for patients with CF should be made jointly between the relevant surgeon, the CF clinicians and the patient or carer.
- The CF team, in collaboration with other relevant clinicians, should manage admissions for acute abdominal pain.
- Where possible any surgical procedure 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 guidelines relating to adults and young people with CF, which has been developed in collaboration with the CF service. The 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.

Solid Organ Transplantation

- When the possibility of transplantation is appropriate, this should be discussed with the patient and family (where appropriate) as early as possible.
- Information regarding transplantation should be available to all CF patients.
- Referral to the transplant centre for further assessment (if appropriate) should be made as soon as potential candidacy has been assessed.

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

Palliative Care

The specialist CF Centre needs to 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 multidisciplinary team with a specific interest in palliative care
- joint patient review by the CF clinical team and palliative care clinician with cohesive joint management plans in place for end of life care and/or pain management
- access to a CF psychologist for the patient and their 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 should 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 should 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 should be evidenced.
- Strong links should be made with the pre-natal screening services to ensure that patients with a CF diagnosis have rapid access to specialist advice and support when considering pregnancy.
- The service should specify a pathway for CF males to have fertility assessed and a referral pathway for infertility treatment

Core Service Standards

- The model of care should 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 and satellite clinics attended by the full CF MDT.
- Inpatient, day care, outpatient, diagnostic and homecare services should be co-ordinated to ensure continuity of care for the patient.
- Patients and their families should be seen in an age appropriate, comfortable environment, ensuring privacy, dignity and protection from cross infection.

- Patients and their families should 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 should be encouraged to participate in the planning of their care.
- Patients and their families should be made aware of how to contact their clinical team and CF support groups.
- Within the timescales, complete and accurate data should be submitted to the UKCF Registry (subject to patient consent).

2.2 Interdependencies with other services or providers

There is no requirement for co-location with other services.

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

- Endocrinology
- Diabetology
- Palliative care
- Clinical genetics
- Liver services
- Psychiatry
- Renal services
- Gastroenterology
- Gastro-Intestinal Surgery
- Hepatology
- Rheumatology
- ENT
- Vascular Services
- Thoracic Surgery
- Transplantation Services
- Obstetrics and Gynaecology
- Fertility Services.

If not available at a CF Centre, processes need to be in place to demonstrate clear pathways, which include out of hours/Emergency Care.

2.3 Exclusion Criteria

Patients aged under 16 years are excluded from the service.

2.4 Acceptance Criteria

The service outlined in this specification is for patients ordinarily resident in Wales, or otherwise the commissioning responsibility of the NHS in Wales. This excludes patients who whilst resident in Wales, are registered with a GP practice in England, but includes patients resident in England who are registered with a GP Practice in Wales.

2.5 Patient Pathway (Annex i)

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

For adult and young people (16 years of age and over), the majority of referrals will come from paediatric CF services with appropriate transition arrangements. However, referrals may also come from other adult and young people CF services, general physicians, fertility services or General Practitioners.

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

For the purposes of this specification, a CF 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 diagnostic sweat test
        - two known disease causing CF gene variants
        - evidence of functional epithelial ion transport abnormality

2.6 Service provider/Designated Centre

Adult and Young People Cystic Fibrosis Services for Welsh patients are provided by:

The All Wales Cystic Fibrosis Centre based at:

University Hospital Llandough
Penlan Road
Llandough
CF64 2XX

Patients also have access to:

Bristol Royal Infirmary
Marlborough Street

Bristol
BS2 8HW

Royal Brompton Hospital
Sydney Street
Chelsea
London
SW3 6NP

Adult and Young People in North Wales and Powys have access to:

Liverpool Heart and Chest Hospital.
Thomas Drive
Liverpool
L14 3PE

West Midlands Adult Cystic Fibrosis Centre at:

Birmingham Heartlands Hospital
Bordesley Green East
Birmingham
B9 5SS

Manchester Adult Cystic Fibrosis Centre at:

Wythenshawe Hospital
Manchester Adult CF Centre
South Moor Road
Manchester
M23 9LT

Royal Stoke University Hospital
Newcastle Road
Stoke-on-Trent
ST4 6QG

2.7 Exceptions

If the patient does not meet the criteria for treatment as outlined in this policy, an Individual Patient Funding Request (IPFR) can be submitted for consideration in line with the All Wales Policy: Making Decisions on Individual Patient Funding Requests. The request will then be considered by the All Wales IPFR Panel.

If the patient wishes to be referred to a provider outside of the agreed pathway, an IPFR should be submitted.

Further information on making IPFR requests can be found at: Individual Patient Funding Requests

3. Quality and Patient Safety

The provider must work to written quality standards and provide monitoring information to the lead commissioner. The quality management systems must be externally audited and accredited.

3.1 Quality Indicators (Standards)

Locally agreed

The <<service>> should aim to deliver the following:

- The number of patients at the centre should 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 not be less than 50 for a specialist centre.
- 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 should be in line with the recommendations set out in section 3 of the Standards for the Clinical Care of Children and Adults with Cystic Fibrosis in the UK (2011).
- The service should ensure that the facilities are available to support the best quality CF service allowing seamless care between home and hospital.
- The facilities should 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 routine clinic review, annual reviews, treatment, day case etc.
- The service should 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.

3.2 National Standards

The following standards are regarded as core standards:

- A specialist centre should have a Director who is responsible for the service and in addition, the specialist service should have a designated service manager.
- The CF multidisciplinary team (minimum of a doctor, nurse specialist, physiotherapist and dietician) should be available for care of inpatients and outpatients.
- Policies and procedures should be in place to protect patients from the risk of cross infection, both as inpatients and outpatients.
- Microbiological analysis of respiratory samples and age appropriate lung function should be carried out at all outpatient visits.

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

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

- Standards for the Clinical Care of Children and Adults with Cystic Fibrosis in the UK (2011)
- Overview | Cystic fibrosis: diagnosis and management | Guidance | NICE (2017)
- Nutritional Management of Cystic Fibrosis. April 2002
- Physiotherapy National Standards of Care for people with Cystic Fibrosis (2017).
- Standards of care for patients with cystic fibrosis: A European consensus (2005)
- Pharmacist Standards in Cystic Fibrosis Care (2011)
- Quality statements | Cystic fibrosis | Quality standards | NICE (2018)

3.3 Other quality requirements

- the provider will have a recognised system to demonstrate service quality and standards
- the service will have detailed clinical protocols setting out nationally (and local where appropriate) recognised good practice for each treatment site
- the quality system and its treatment protocols will be subject to regular clinical and management audit
- the provider is required to undertake regular patient surveys and develop and implement an action plan based on findings
- the service will participate as and when required in Multidisciplinary Peer Review

4. Performance Monitoring and Information Requirement

4.1 Performance Monitoring

NWJCC will be responsible for commissioning services in line with this policy. This will include agreeing appropriate information and procedures to monitor the performance of organisations.

For the services defined in this policy the following approach will be adopted:

- Service providers to evidence quality and performance controls
- Service providers to evidence compliance with standards of care

NWJCC will conduct performance and quality reviews on an annual basis

4.2 Key Performance Indicators

The providers will be expected to monitor against the full list of Quality Indicators derived from the service description components described in Section 2.1 and against the NICE Quality Indicators Quality statements | Cystic fibrosis | Quality standards | NICE

The provider will be expected to benchmark with other similar centres, including use of UKCF Registry data when available.

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 provider is expected to specifically monitor against the following quality measures:

Quality Measure	Method of Reporting
Number and percentage of patients with chronic pseudomonas aeruginosa infection	UK CF Registry Data
Number and percentage of patients receiving inhaled antibiotics	CF Service reporting dataset
Adherence to standards of care to prevent cross infection	UK CF Registry Data

The service should aim to reduce their prevalence of chronic pseudomonas aeruginosa infection	CF Service reporting dataset
Body Mass Index (BMI) (Median BMI of centre cohort)	UK CF Registry data CF Service reporting dataset
Monitoring of lung function Forced Expiratory Volume (FEV_1) in one second and rate of decline in FEV_1 (Number of patients and percentage with FEV_1 >65%)	UK CF Registry data CF Service reporting dataset
Number and percentage of patients offered mucoactive therapies	UK CF Registry data CF Service reporting dataset
Number of patients who have had a post-annual review management plan and discussion	CF Service reporting dataset
Timely initiation of treatment for infective exacerbations. (number and percentage of patients breaching standards of care for timing of admission)	CF Service reporting dataset
Number and percentage of patients who have been offered a clinical psychology review within last 12 months	CF Service reporting dataset
Number and percentage of patients admitted to a ward with specialist CF staff	CF Service reporting dataset
Number and percentage of patients receiving satellite clinic review for some aspects of CF care closer to home and percentage of patients receiving virtual clinic review	CF Service reporting dataset
Systematic measurement of patient experience and satisfaction	Self-report demonstrating systemic engagement and feedback on actions
Median Survival of national population	CF Registry data

4.3 Date of Review

This document is scheduled for review before 2023 where we will check if any new evidence is available.

If an update is carried out the policy will remain extant until the revised policy is published.

5. Equality Impact and Assessment

The Equality Impact Assessment (EQIA) process has been developed to help promote fair and equal treatment in the delivery of health services. It aims to enable NHS Wales Joint Commissioning Committee to identify and eliminate detrimental treatment caused by the adverse impact of health service policies upon groups and individuals for reasons of race, gender re-assignment, disability, sex, sexual orientation, age, religion and belief, marriage and civil partnership, pregnancy and maternity and language (Welsh).

This policy has been subjected to an Equality Impact Assessment.

The Assessment demonstrates the policy is robust and there is no potential for discrimination or adverse impact. All opportunities to promote equality have been taken.

6. Putting Things Right

6.1 Raising a Concern

Whilst every effort has been made to ensure that decisions made under this policy are robust and appropriate for the patient group, it is acknowledged that there may be occasions when the patient or their representative are not happy with decisions made or the treatment provided.

The patient or their representative should be guided by the clinician, or the member of NHS staff with whom the concern is raised, to the appropriate arrangements for management of their concern.

If a patient or their representative is unhappy with the care provided during the treatment or the clinical decision to withdraw treatment provided under this policy, the patient and/or their representative should be guided to the LHB for NHS Putting Things Right. For services provided outside NHS Wales the patient or their representative should be guided to the NHS Trust Concerns Procedure, with a copy of the concern being sent to NWJCC.

6.2 Individual Patient Funding Request (IPFR)

If the patient does not meet the criteria for treatment as outlined in this policy, an Individual Patient Funding Request (IPFR) can be submitted for consideration in line with the All Wales Policy: Making Decisions on Individual Patient Funding Requests. The request will then be considered by the All Wales IPFR Panel.

If an IPFR is declined by the Panel, a patient and/or their NHS clinician has the right to request information about how the decision was reached. If the patient and their NHS clinician feel the process has not been followed in accordance with this policy, arrangements can be made for an independent review of the process to be undertaken by the patient's Local Health Board. The ground for the review, which are detailed in the All Wales Policy: Making Decisions on Individual Patient Funding Requests (IPFR), must be clearly stated.

If the patient wishes to be referred to a provider outside of the agreed pathway, and IPFR should be submitted.

Further information on making IPFR requests can be found at: Individual Patient Funding Requests

Annex ii Codes

Code Category	Code	Description
ICD-10	E840	Cystic fibrosis with pulmonary manifestations
	E841	Cystic fibrosis with intestinal manifestations
	E848	Cystic fibrosis with other manifestations
	E849	Cystic fibrosis, unspecified
	P75X	Meconium ileus in cystic fibrosis

Annex iii Abbreviations and Glossary

Abbreviations

AWMSG All Wales Medicines Strategy Group

IPFR Individual Patient Funding Request

NWJCC – NHS Wales Joint Commissioning Committee

MDT Multidisciplinary Team

Glossary

Individual Patient Funding Request (IPFR)

An IPFR is a request to NHS Wales Joint Commissioning Committee (NWJCC) to fund an intervention, device or treatment for patients that fall outside the range of services and treatments routinely provided across Wales.

NHS Wales Joint Commissioning Committee (NWJCC)

NWJCC is a joint committee of the seven local health boards in Wales. The purpose of NWJCC is to ensure that the population of Wales has fair and equitable access to the full range of Tertiary Services. NWJCC ensures that services within our portfolio are commissioned from providers that have the appropriate experience and expertise. They ensure that these providers are able to provide a robust, high quality and sustainable services, which are safe for patients and are cost effective for NHS Wales.

Cystic Fibrosis Trust

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

Cystic Fibrosis Registry

The UK Cystic Fibrosis Registry is a secure centralised database, sponsored and managed by the Cystic Fibrosis Trust. It records health data on consenting people with CF in England, Wales, Scotland and Northern Ireland.