Cystic Fibrosis Trust

Standards for the clinical care of children and adults with cystic fibrosis in the UK

Third edition

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Uniting for a life unlimited

Cystic Fibrosis Trust

Contents

Glossary

Adherence	The extent to which an individual's behaviour coincides with health-related advice or recommendations given by a healthcare provider. *This term will be used throughout the document instead of compliance and concordance, as it is most commonly used and understood*.
Bcc	*Burkholderia cepacia* complex.
CF community	People with CF, their families and professional caregivers.
CFD	CF diabetes (formerly CF-related diabetes, or CFRD).
CF professionals	Members of the CF MDT and other CF specialist professionals that look after people with CF.
CFTR	CF transmembrane conductance regulator protein.
CFTR modulator therapy	Treatments which are designed to correct the malfunctioning protein made by the CFTR gene, and are effective in people with specific CF-causing variants.
CFSPID	CF screen positive, inconclusive diagnosis.
Health inequalities/ disparities	The unfair and avoidable differences in health status seen between groups or populations. These are closely linked with social, economic and/or environmental disadvantages. Health and illness follow a social gradient: the lower the socioeconomic position, the worse the health.
DIOS	Distal intestinal obstruction syndrome.
MDT	Multidisciplinary team.
Medicines reconciliation	The process of identifying an accurate list of a person with CF's current medicines, including over-the-counter and complementary medicines, and comparing this list to the current list in use, recognising any discrepancies, and documenting any changes. It also takes into account the current health of the person with CF and any active or long-standing issues.
Remote monitoring	The ability to monitor certain aspects of a person with CF's health from their own home.
Social determinants of health	The non-medical factors such as income, social support, early childhood development, education, employment, housing and gender that can positively or negatively influence health outcomes. They are the conditions in which people are born, grow, work, live, and age.
Telehealth	Remote monitoring of people with CF in their own homes, to anticipate exacerbations early and build their self-care competencies. It is a form of technology-enabled care service, like telemedicine (providing health services remotely).
Variant	Changes (or mutations) to a gene. CF is caused by CF-causing variants to the CFTR gene.
Virtual consultation	Virtual consultations are real-time consultations between a person with CF and a healthcare professional over video or telephone, as opposed to face-to-face. They should be as effective as a face-to-face consultation.

1. Introduction

This document is intended to supersede the 2011 version of this guideline. Much has changed in the last 13 or so years. There have been significant improvements in life with cystic fibrosis (CF): the national roll-out of newborn screening for CF across the UK was completed in 2007, and access to CFTR modulators has become wider. After the initial licence for ivacaftor was given in 2012, and others, including elexacaftor/tezacaftor/ivacaftor in 2020, it is anticipated that a child born with CF today would live into late adult life. More people with CF are engaging in education and employment, have careers, and are considering experiencing parenthood. The improvements have resulted from the partnership between people with CF, their families and carers, and CF teams. However, it must be acknowledged that there are still gaps in knowledge and that clinical care continues to evolve. There is further to go until a life unlimited can be truly achieved for all people with CF, especially for those unable to take modulators.

1.1 What is cystic fibrosis (CF)?

CF is one of the most common life-limiting, recessively inherited diseases in the UK, affecting more than 11,100 people (1 in 2,500 live births). It results from variants affecting a gene that encodes a chloride channel called the CF transmembrane conductance regulator (CFTR). This channel is essential for the regulation of salt and water movements across cell membranes. Absence or reduced function of CFTR results in thickened secretions in organs with epithelial cell lining. Hence, CF is a multi-system disease, although it mainly affects the lungs, digestive system and vas deferens.

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

In about 85% of people with CF, the pancreatic exocrine ducts become blocked to cause maldigestion and intestinal malabsorption (exocrine pancreatic insufficiency). Infants may fail to thrive, and older children and adults may become undernourished. About 15% of babies with CF are born with a bowel blockage (meconium ileus). Many children and adults struggle with abdominal pain and constipation and can be at risk of bowel blockages due to distal intestinal obstruction syndrome (DIOS). Appetite can be adversely affected, and, in some people with CF, an underlying increase in metabolic demands can lead to a need for a higher energy intake.

There are many other disease manifestations: nearly all men with CF are infertile; a high proportion of older people with CF will develop CF diabetes (CFD); chronic liver disease and portal hypertension may develop; joints can be affected (CF arthropathy); bones can be affected with age by reduced bone mineral density (BMD); and nasal polyps and sinusitis are not uncommon. Psychosocial problems that are often associated with living with a long-term health condition may also be present. The manifestations of CF vary depending upon a variety of factors, including genotype and treatments.

1.2 Demographics

There are 11,148 people with CF in the UK, with 63% aged 16 years or older. Currently, CF is seen mostly in white populations. However, CF is increasingly being diagnosed in people from other ethnic backgrounds; in the UK, mainly in people from Asian (Indian subcontinent) or Middle Eastern backgrounds. The carrier rate of a CF gene variant in the UK is 1 in 25, with an incidence of approximately 1 in 2,500 live births, although this figure varies by region. Different variants (mutations) are more common in different regions and ethnic groups, with future work needed to better understand these varying genotypes.

The newborn screening programme for CF became UK-wide in October 2007. Those that are diagnosed with CF through newborn screening are usually under four weeks old. People born before October 2007 (or abroad), or those missed by newborn screening, may still be diagnosed with CF following the development of symptoms.

Life expectancy has improved significantly over recent decades due to a number of factors, including Specialist CF Centre care with a multidisciplinary team (MDT), better nutritional support, improved treatment of respiratory disease, the transformational impact of CFTR modulator therapies, and a more proactive and individualised approach to CF care. For a baby born with CF

today, survival into late adult life is expected. Individual outcomes, however, still vary widely. The healthcare needs of the changing demographic of people with CF, with an increasing aging population, will need to be reflected in the models of care, service provision, and clinical expertise.

For further detail, see the **UK CF Registry 2022 Annual Data Report**.[1]

1.3 Why are Standards of Care needed?

- All people with CF in the UK should have equal access to the highest level of multidisciplinary specialist care that is adequately resourced and encompasses the latest evidence-based therapies.
- The aim is to improve quality of life and extend life expectancy.
- The Standards can be used for benchmarking and outlining objective measures of service provision to inform commissioning of CF services.
- It is hoped that the Standards will also act as an aid for CF Centres undertaking Quality Improvement (QI) programmes.

1.4 Who is this document for?

- Clinicians, allied health professionals, and other healthcare professionals working with people with CF across the UK.
- Commissioners and others in the NHS and elsewhere who are responsible for the provision of care for people with CF.
- Parents/carers of children with CF, young people and adults with CF, and their families, to help them understand what level of care they should expect and how they can play an active role in their care.

1.5 How was the document written?

This is the third edition of the Standards for the Clinical Care of Children and Adults with Cystic Fibrosis and replaces the 2011 version. This update was produced to reflect changes in the current understanding of CF and the practice of CF care in the UK. These consensus standards were compiled by a working group of CF professionals (paediatrics and adult) based in regions around the UK, including specialist physicians, physiotherapists, pharmacists, social workers, psychologists, nurses, and dietitians; clinical quality improvement specialists; and members of the CF community (see **Appendix 1** for working group member details). The first draft of this update was produced based on the expertise and experience of working group members and was reviewed by a secondary peer review group of professionals in the CF MDT. The second draft was reviewed by members of the Cystic Fibrosis Trust Clinical Advisory Group (a group of professionals from across the CF MDT). Comments from each round of reviews were discussed and resolved among the working group. Feedback from focus groups with members of the CF community was incorporated into the final draft.

This is a guideline document that draws consensus on best practice. Recommendations aim to provide a realistic and pragmatic approach to an area where randomised controlled trials cannot always provide evidence. Additional data was collected specifically to inform and develop updated recommendations for staffing (see **Appendix 2** for details). References are not offered to support recommendations in this document, aside from relevant Cystic Fibrosis Trust, European Cystic Fibrosis Society (ECFS), US Cystic Fibrosis Foundation, and National Institute for Health and Care Excellence (NICE) consensus documents and guidelines. Useful information and support resources are signposted where possible. Other key guidance for the care of people with CF includes the ECFS Standards for the care of people with CF[2-5] and **NICE guideline NG78**.[6]

2. Models of CF care

2.1 Models of care

All people with CF must have their care delivered under the direct supervision of a recognised Specialist CF Centre throughout their lives. Concentrating care for people with CF in Specialist adult and paediatric CF Centres ensures that the MDT will see sufficient numbers of people with CF to be able to maintain expertise so that they treat people with CF effectively and recognise the more unusual manifestations to delay the onset of the multi-system complications associated with the condition. Centre care also maximises access to clinical trials.

People with CF and their families (in the case of children's services) should be involved in service planning and delivery. The logistics of delivering care may differ for children and adults (for detail on the frequency of review, see section 5.2.1). Where care is delivered in a Network, or where certain elements of care are delivered outside the main CF Centre, Service Level Agreements (SLAs) and Standard Operating Procedures (SOPs) should be agreed by both parties involved in care delivery.

2.1.1 Paediatric model

- Children will either receive their care from a Specialist CF Centre, or receive shared care within an agreed designated Network CF Clinic (for detail on Network CF Clinics, see section 2.4).
- Shared care must be delivered as part of an agreed designated Network with an SLA and SOPs, as agreed between the Specialist CF Centre and the Network CF Clinics.
- Parents of children with CF should be aware of their options and know they can choose for their children to have all their care at a Specialist CF Centre if they wish.

2.1.2 Adult model

- Due to the increasing complexity of CF in adulthood, all care should be delivered by a Specialist CF Centre. Only in exceptional circumstances should adults with CF receive CF care from non-specialists. With the rapidly evolving needs of this population, innovative models of care may be developed, such as specialist outreach clinics, so that some specialist CF care can be delivered closer to home.

2.2 Person-centred CF care

The care delivered to people with CF must take into consideration a number of different factors related to the clinical needs of each individual, as well as a range of non-clinical factors. The care plan for each individual will be reached through a shared decision-making process, with partnership between the person with CF, their parents or carers (in the case of children and young people), and their clinical team. The person's individual perspectives and preferences influence their choices in balancing the burdens and benefits of treatments and interventions; therefore, these should be elicited and explored.

The care plan needs to take into account the variation of clinical phenotypes of CF and its complications on the health of an individual person with CF; the severity and stage of the disease; and current uncertainties in CF care due to some people being unable to benefit from recent treatment advances. CF teams should continually monitor, review, and re-evaluate care to assess individual responses, in partnership with people with CF and their families/carers.

It is crucial that the delivery of CF care also takes into account the wider conditions of people's lives, such as their living environment, financial resources, and other psychosocial factors that may impact on health and wellbeing, and to recognise and take steps to reduce health inequalities and provide accessible care for everyone. Patient experience surveys or feedback forms can provide insights into whether people with CF and their families/carers feel that their preferences are considered and that they are involved in decisions about their CF care.

Recommendations

1. **Provide equitable access to care**, taking into consideration factors including neurodiversity, learning disability, gender, sexual orientation, ethnicity, and language needs.
2. **Provide support for factors that impact on burden of care**, such as number of siblings with CF, other health needs within a family, and the needs of caregivers.
3. **Ensure that care is collaborative with people with CF and their families/carers**, taking into consideration individual preferences and providing information

for informed decision-making, whilst also recognising where intervention is required, for example, for safeguarding issues, or if there is reason to believe the person lacks mental capacity.

4. **Ensure that care is collaborative with other services**; CF services must collaborate with other health and care providers to deliver joined-up care that centres the needs of people with CF and their families/carers.
5. **Use innovation to optimise the flexible delivery of care whilst ensuring that standards are met for all**, taking into consideration the frequency of visits, in-person vs telehealth (video and telephone clinics), home-based care (such as home intravenous (IV) therapy), outreach care, and flexible clinic timings for education and employment flexibility.

2.2.1 Virtual consultations and remote monitoring

The use of these innovations plays an important role in access to care. However, if people with CF or their families/carers do not want to or cannot access virtual consultations or remote monitoring, they should have access to equivalent face-to-face services.

Virtual consultations are real-time consultations between a person with CF and a healthcare professional over video or telephone, as opposed to face-to-face. They should be as effective as a face-to-face consultation. Remote monitoring is the ability to monitor certain aspects of the health of a person with CF from their own home. Guidance for healthcare professionals on virtual consultations and remote monitoring is available, for example on the **FutureNHS website**.[7]

The needs of most people with CF are likely to be met by a 'blended model' of virtual and face-to-face appointments. A bespoke approach should be considered on a case-by-case basis (for detail on person-centred CF care, see the start of **section 2.2**). Although many aspects of a consultation can be achieved virtually, it is important that people with CF have clear routes of communication with their CF team and access to urgent face-to-face review if required.

Key elements of core virtual consultation:

- Virtual consultations should take place at an appropriate time for the person with CF.
- Children with CF should be present, unless otherwise agreed with their parents or carers.
- Presence of core MDT members.
- Centres should provide the facilities and options for virtual consultations of the same quality as face-to-face consultations:
  - Technology: uninterrupted connectivity and good audio and video quality (on both sides).
  - Facilities: a private safe space, as would be provided in face-to-face contact, enabling confidential discussions.
- Access to appropriate clinical information:
  - Spirometry (with the provision and training in use of appropriate equipment so the test may be performed adequately)
  - Weight (with the provision of appropriate equipment)
  - Height, until growth is expected to cease (with adequate training to perform measurements)
  - Assessment of adherence for electronic data capture
  - Glucose monitoring (with the provision of appropriate equipment)
  - Microbiological surveillance (with postal sampling system).
- Other remote monitoring services include:
  - Blood monitoring
  - Additional monitoring, according to local or individual specific priorities
  - Assessments of psychological wellbeing.

2.3 Specialist CF Centre care

High-quality care for the complex multi-system symptoms and complications of CF can only be provided by an MDT of trained, experienced, specialist healthcare professionals who routinely see a critical mass of people with CF at a Specialist CF Centre, which treats either children or adults.

Factors such as new treatments and the digitalisation of some elements of care have already impacted CF care delivery and workload:

- While inpatient workload has reduced due to the transformational impact of modulators, workload in other areas of CF care has increased, particularly time spent prescribing and delivering virtual care and remote monitoring.
- Workforce planning will need to evolve to meet the changing needs of people with CF. Analysis of current staffing levels and of patient-reported experience measures (PREMs) highlights deficiencies in the provision of clinical psychology, social work, and pharmacy services within CF MDTs.
- There is an urgent need to develop services for an increasing number of adults with CF and to plan services for an ageing population of people with CF with increased disease complexity, complications, and co-morbid conditions. Emerging issues now include increasing rates of pregnancy, treatment of infertility, management of obesity, liver disease, and the long-term complications of CFD, as well as screening, diagnosis, and management of cancer.
- The number of children with CF is expected to remain relatively stable, with key areas of work focusing on the initiation and management of CFTR modulators, preventative advice, support for young people with mental health issues and social deprivation, as well as transition to adulthood, higher education, and employment.
- This remains a time of change and transition for CF. Therefore, the guidance in this chapter should be reviewed every three years as the landscape, data, and evidence base evolve.

This guidance focuses on Specialist CF Centre staffing only. The workforce requirements of a Network or local outreach clinic should be decided based on local priorities and adhering to the principles of Network care (for detail on Network CF Clinics, see section 2.4).

The following criteria should be met for designation as a Specialist CF Centre:

- In the context of many people with CF experiencing improved health outcomes, and smaller numbers presenting with advanced disease, Centres must be of sufficient size to ensure that the MDT will continue to have a diverse range of experiences to maintain its CF expertise and skills. To reflect this, the working group considered that the minimum size for designation as a Specialist CF Centre should be 100 children or 200 adults, including any shared care or outreach clinic patients whose care is overseen by the Specialist CF Centre but may be delivered locally.
  - However, it is acknowledged that the adult CF population is growing rapidly and current adult Specialist CF Centres with fewer than 200 people with CF will build towards the recommended minimum number.
  - It is also recognised that in exceptional circumstances, the geographical location of a Specialist CF Centre may mean that the number of people with CF is less than what is recommended.
  - Smaller Centres need to have formal arrangements in place with other, larger, Centres, in order to maintain clinical expertise through educational peer support, complex case discussions and shared learning.
- The Centre must have a core MDT of trained and experienced CF specialist healthcare professionals:
  - Specialist consultant paediatricians or adult physicians
  - Medical trainee(s) and speciality, associate specialist and specialist (SAS) grades
  - Clinical nurse specialists
  - Physiotherapists
  - Dietitians
  - Clinical psychologists
  - Social workers
  - Pharmacists
  - Secretarial support
  - Database coordinator.

Note: There is flexibility in terms of the exact staffing model used, and it is acknowledged that there are new and emerging roles in some CF Centres (for detail on new and emerging roles, see section 3.10). Such emerging roles are not replacements for the core MDT, but allow flexible development of services and skill mix in line with local needs and priorities.

- The Centre must have an MDT of appropriate number for the size of the patient population to ensure they can meet the needs of individual people with CF, with built-in resilience to cover staff absences.
- The Centre should be able to attract new staff, enabling them to enter the profession and gain expertise in CF care, in order to support succession planning and ensure continuity of specialist care for the Centre's population.
- The Centre must have access to other medical and surgical specialists when required, particularly: Gastroenterology and Hepatology, Diabetes and Endocrinology, Ear Nose and Throat (ENT), Cardiothoracic and General Surgery, Specialist Anaesthesia and Pain Control, Rheumatology, Obstetrics and Gynaecology, Psychiatry, Intensive Care, Interventional Radiology, Palliative Medicine, Microbiology, Genetics, and Primary Care.

To develop staffing guidance, the working group considered that the core services of a Specialist CF Centre comprise three distinct areas of activity, outlined below, each with its own set of tasks and staffing needs. Whilst some tasks require a specific MDT member, others can be devolved, and it is acknowledged that expanding roles, such as non-medical prescribing, as well as new roles, such as CF practitioners, are emerging as innovative approaches.

1. **Patient-facing clinical activity (attributable to individual people with CF and their families/carers)**
   - Clinics (outpatient and virtual) – monitoring and annual review of people with CF.
   - Acute care – seeing people with CF with acute issues and dealing with emergencies.
   - Inpatient care and day cases – ward rounds, seeing inpatients and day cases, starting IVs, and complex case management.
   - Home visits and home-based care – home visits, home physiotherapy, and home IVs.
   - Patient assessments – carrying out investigations, tests, and assessments (including mental health assessments and social determinants of health) and remote monitoring (including equipment training and troubleshooting with people with CF and their families/carers).
   - Interventions with people with CF – sessions with a psychologist or social worker, physiotherapist and dietetics-led interventions, MDT interventions, preventative work, adherence support, group support, and supporting transition from paediatric to adult care.
   - Ad hoc support – phone calls with people with CF, telephone advice, and answering text or email queries from people with CF and their families/carers.

2. **Non-patient-facing clinical activity**
   - Multidisciplinary working – MDT meetings, MDT reviews, joint meetings with other specialties, complex case discussions, and liaison with other services (such as hospice, community mental health, etc).
   - Patient administration – planning clinics, booking appointments, reviewing notes, updating records, making referrals, prescribing, care coordination, advocacy, and UK CF Registry work.
   - Travel – any travel required to deliver patient-facing activities.

3. **Non-clinical work and supporting professional activities**
   - General administrative tasks and meetings – organising workload, checking non-patient-related emails, recruitment, creating rotas, business planning, and staff and team meetings.
   - Clinical governance – managing complaints, infection control, and risk assessments.
   - Research and clinical trials – planning and conducting research or clinical trials and disseminating research or trial findings.
   - Service development – audit, QI work, service improvement projects, and patient and public involvement.
   - Personal development – continuing professional development (CPD).

- Staff development – training, appraisals, supervision, teaching, and mentoring.
- National and international work – special interest groups, working groups and committees, and guideline development.

2.3.1 Staffing guidance

The guidance below describes the current established model of care and outlines the required staffing levels for core MDT staff within a designated Specialist CF Centre.

2.3.2 Considerations for workforce planning in Specialist CF Centres

- Staffing numbers should be appropriate for the Centre's size and reflect the model of care being used, taking into account the time spent by staff from the Specialist CF Centre seeing people with CF in a local hospital or outreach clinic, if applicable.
- At a minimum, the CF MDT should include all roles required to meet designation for a Specialist CF Centre. This must include psychosocial and pharmacy staff.
- Sufficient individuals must be in post to provide cover. In particular, services must ensure:
  - Adequate cover is available for annual leave, sickness absence, parental leave etc.
  - Adequate uplift is included for non-clinical work, including professional development, governance, research, audit, QI work, and national and international work (MDT staff are expected to demonstrate a commitment to ongoing CPD through membership of their relevant special interest group, as well as attendance at relevant national and international conferences).
  - Adequate time is available for training and supervision of new and junior staff, to support succession planning and enable continuous high-quality care.
- Local models of care will be adapted by the local CF MDT to deliver best care, taking into account factors including patient choice, differences in population needs, and service delivery arrangements. Some models may require an increase in staffing, for example if a service delivers more home-based care across a large geographical area, if the Centre serves a population with high levels of deprivation, or if the geographical area served by the Centre has a lower-than-average eligibility for highly effective modulators, for example in ethnically diverse populations.
- CF time should be protected, and staff should not routinely be required to cross-cover other departments or specialties during CF time.
- Teams must have, and be able to maintain, experience in managing the complex problems and rarer complications that may arise in people with CF.
- The full MDT should be available for all routine clinics and the team should be resourced to deliver all aspects of CF care, including monitoring of people with CF.
- Teams must have the ability to provide advice for urgent needs on a 24-hour basis, 7 days per week, and sufficient capacity in clinics for outpatients to be seen urgently (within 24 hours). This may be in clinic, a day case unit, or a ward visit.
- There will be some overlap between areas of activity – for example, staff may complete non-clinical work while on call for acute care or while being available for but not actively involved in clinics.
- Inadequate levels of staffing may be indicated by poorer outcomes for people with CF, feedback from people with CF about difficulties accessing staff, sickness levels in staff, or not meeting guidance around frequency of patient reviews.

The development of staffing guidance in this chapter was informed by existing UK CF workforce data, insights from a survey of the UK CF workforce, patient experience feedback, as well as national and international guidelines, combined with the experience and expertise of working group members. For detail on the evidence and process to develop staffing guidance, see **Appendix 2**.

Table 1: To enable delivery of the areas of CF care activity outlined above, a designated Specialist CF Centre for 100 children should have a core MDT of 9.25 overall whole-time equivalent (WTE). This resource should be used flexibly to meet the needs of the population with an example of the breakdown being:

	WTE for 100 children with CF
Overall WTE	**9.25**
Medical staff*	1.5
Nurse	1.75
Physiotherapy	2.0
Dietitian	1.0
Psychology	0.75
Social work	0.75
Pharmacy	0.75
Administrative staff	0.75

*This refers to consultants and doctors with CF expertise.

Table 2: To enable delivery of the areas of CF care activity outlined above, a designated Specialist CF Centre for 200 adults should have a core MDT of 18.5 overall WTE. This resource should be used flexibly to meet the needs of the population with an example of the breakdown being:

	WTE for 200 adults with CF
Overall WTE	**18.5**
Medical staff*	3.0
Nurse	3.5
Physiotherapy	4.0
Dietitian	2.0
Psychology	1.5
Social work	1.5
Pharmacy	1.5
Administrative staff	1.5

*This refers to consultants and doctors with CF expertise.

2.4 Network CF Clinics

It is recognised that different forms of Network care exist around the UK. These have arisen historically and have been designed to meet local needs. For Network care to be successful, it is crucial that certain criteria are fulfilled in all Centres, as set out in the principles of Network care below.

The advantages of Network care include the delivery of care closer to home; facilitating closer monitoring as necessary, especially for those in remote areas; the ability to involve other non-CF healthcare professionals; and more joined-up working with other agencies, including education and social care. It aligns with the integrated healthcare model.

Whilst Network care is a model adopted in the paediatric setting, the principles for optimal Network care outlined below are applicable across the life journey of the person with CF.

Principles of Network care:

- Key principle: partnership working between people with CF, their families/carers, the Specialist CF Centre, and the Network CF Clinic.
- The Network team should be involved from the very start of the patient journey (such as diagnosis after newborn screening).
- People attending a Network CF Clinic should receive quality of care that is at least equal to the Specialist CF Centre.
- People attending a Network CF Clinic should have access to an MDT with CF expertise, with dedicated time for the CF service, led by a paediatrician or adult physician with training and experience in CF.
- People attending a Network CF Clinic should have access to MDT members with expertise in CF (for example, psychology and social work) from the Specialist CF Centre, if not available locally.
- People attending a Network CF Clinic should have access to new therapies in the same manner as those attending the Specialist CF Centre.
- People attending a Network CF Clinic should have opportunity to participate in research and clinical trials.
- People attending a Network CF Clinic should have access to outpatient and inpatient facilities and routine laboratory tests in the same manner as those attending the Specialist CF Centre.

- People attending a Network CF Clinic should see the Specialist CF Centre team in person at least once a year for annual review and more often if required – for example, for specific reviews, investigations, or preparation for transition to adult care. This will depend upon the severity of disease and level of expertise at the Network CF Clinic. People may attend the Specialist CF Centre, or Specialist CF Centre teams may travel to local Network CF Clinics. Close communication should exist between the Specialist CF Centre and Network CF Clinic teams.
- Optimal communication is facilitated by:
  - Specific members of the Specialist CF Centre team being designated for each Network CF Clinic as the first point of contact. These members of the Specialist CF Centre team will be the MDT members who do the annual reviews and know the Network CF Clinic patients.
  - Shared patient reviews and, if possible, shared patient records.
  - Network CF Clinic MDT members linking regularly with Specialist CF Centre counterparts to share knowledge and expertise.
  - The whole Network meeting at least twice a year (face-to-face once a year) to monitor quality, for service evaluation, to support CPD, and to ensure best practice.
  - SLAs between the Specialist CF Centre and the Network CF Clinic, setting out the key elements of the service required, which should be reviewed annually.
  - SOPs for the delivery of care, co-produced within the Network.
  - Formal agreements in place for provision at another Network site if an essential component of the CF service cannot be provided by a site within the Network CF Clinic – for example, portacath insertion or liver services.

3. Multidisciplinary CF care

3.1 Principles

- Specialist multidisciplinary care is essential in the management of children and adults with CF. Essential team members are listed in **section 2.3**, with further detail on individual roles in **sections 3.3-3.9**. Agenda for Change banding must adequately reflect levels of responsibility, taking into account issues of recruitment, retention, and career progression.
- All people with CF must have access to specialist advice and care from their CF Centres at all times.
- CF Centres must have access to other specialists (**section 2.3**) who are familiar with the complications of CF. These services should develop their experience in managing CF-related complications in close liaison with the core CF MDT. Referral pathways with SLAs must be established.
- Where appropriate, joint clinics should be established, particularly for CFD and antenatal clinics.
- If no suitable candidates with the stipulated experience are available, gaining expertise in post is acceptable with a period of mentoring from a senior colleague.
- All CF MDT members should:
  - have capacity to maintain CPD in CF, which would include attendance at national or international respiratory and CF meetings
  - have a track record in teaching, audit, and/or research, and take part in audits and QI carried out on behalf of the CF service
  - support research in all areas of CF, either through developing individual projects or through participating in research carried out by the MDT
  - be a member of relevant UK CF professional bodies, such as the UK CF Medical Association for specialist doctors, and consider membership to relevant international bodies, such as ECFS special interest groups
  - be responsible for service evaluation and service development, ensuring up-to-date, evidenced-based clinical practice
  - contribute to education and training of other healthcare professionals, including primary care where required
  - have access to health and wellbeing programmes, as well as self-care support.

3.2 The role of the general practitioner (GP)

- Good communication between the Specialist CF Centre, the general practitioner (GP), the person with CF, and their family/carers is essential. The Specialist CF Centre must support people with CF to register with a GP and collaborate with the GP to ensure they are able to adequately support people with CF.
- The GP is responsible for prescribing much of the routine therapy recommended by the Specialist CF Centre. The GP must be adequately informed about the medication recommended, particularly when it may be unfamiliar or used for an unlicensed indication.
- The GP should provide adequate amounts of medication – a minimum of one month at a time, but ideally longer for long-term prescriptions if advised by the specialist team. Some local pharmacies and hospitals coordinate an ordering and delivery service.
- The GP will provide routine immunisation, including any alterations for people with CF, and will arrange for annual influenza immunisation every autumn for people with CF and their family members and carers.
- The GP will invite people with CF to participate in NHS population screening programmes.
- The GP is responsible for non-CF health-related issues.
- The GP may be requested to work in partnership with the CF team, and with palliative care or hospice teams, when appropriate.

3.3 Clinical specialists and consultants

Senior clinical specialists working with people with CF fall into one of the following categories:

1. Consultant in a paediatric Network CF Clinic.
2. Consultant in a Specialist CF Centre.
3. Lead clinical specialist or Centre director in a Specialist CF Centre, either paediatric or adult.

General information

- Doctors must be registered with the General Medical Council.
- Doctors should be a member of the UK CF Medical Association.

3.3.1 Consultants in a paediatric Network CF Clinic

General information

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

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

3.3.2 Consultant in a Specialist CF Centre

General information

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

1. Training in paediatric CF care during a two-to three-year period. They will have a Certificate of Completion of Training (CCT) in paediatrics with National GRID or Specialist Interest in Paediatric Respiratory Medicine which will have provided training in paediatric CF care.
2. Training in adult CF care as part of higher specialist training as a respiratory physician. Basic respiratory training may often not include CF other than for a few weeks.
3. For a specialist CF post, it is expected that the trainee has at least one year (and preferably more) full-time in CF. This may be part of a Cystic Fibrosis Trust post-Certificate of Completion of Training fellowship, or whilst spending two to three years working for an MD or PhD in CF-related research. There may need to be a period of mentoring from a senior colleague.

3.3.3 Lead clinician or Centre director in a Specialist CF Centre

General information

In addition to fulfilling the criteria for a clinical specialist in a Specialist CF Centre, a clinical lead would be expected to:

- Have at least three years' experience working in a CF clinical specialist MDT role in a Specialist CF Centre.
- Have training or experience in management.
- Be fully engaged with the management of the NHS organisation in which they work, in order to be closely involved in the interactions and negotiations with commissioners in planning and contracting CF care.
- Have leadership skills to direct the CF MDT.

3.4 Clinical nurse specialist

General information

- Clinical nurse specialists must be registered with the Nursing and Midwifery Council.
- They should be a member of the UK CF Nursing Association.
- Those working with children must have undergone specific paediatric training.
- They must have specialist knowledge and be experienced in the care of children and/or adults with CF.

Role

- Provide advocacy and psychosocial support, particularly at important times such as the notification of a screening result and diagnosis, admission to hospital, courses of IV antibiotics (inclusive of IV access), starting new treatments, a secondary diagnosis (for example CFD), transition, dealing with reproductive issues, pre- and postnatal care, transplant, and dealing with palliative care issues.
- The clinical nurse specialist may develop additional skills and extended roles in a variety of other areas, such as CFD, midline insertion, and independent and supplementary prescribing.

- Provide home-based care support, particularly for home IV antibiotic therapy.
- Provide education about CF to others, including nurseries, schools, places of higher education, and workplaces.
- Act as a link between the person with CF and their family and primary care, community services, and hospitals.
- Act as a resource for training and education for other professionals involved in CF care.

For further detail, see the Cystic Fibrosis Trust **National Consensus Standards for the Nursing Management of Cystic Fibrosis (2001)**.[8] *Please note, this consensus document is currently being updated and a new version will be available soon.*

3.5 Physiotherapist

General information

- Physiotherapists must be registered with the Health and Care Professions Council (HCPC).
- They should be a member of the Association of Chartered Physiotherapists in CF.
- They must have specialist knowledge and be experienced in the care of children and/or adults with CF.

Role

- Be responsible for assessing issues on admission to and discharge from hospital, at outpatient appointments, and at annual review, including airway clearance, inhaled therapy, musculoskeletal (MSK) issues, exercise, urinary incontinence, and non-invasive ventilation, where necessary.
- Ensure access to appropriate treatment based on assessment findings.
- Review and adapt treatment approaches as requirements change.
- Work with other members of the MDT to assess, monitor, and improve adherence to prescribed medications.
- Maintain community contact to respond to changing needs, support with equipment supply and/or monitor microbiology status through regular sputum sampling.
- Be a resource on physiotherapy assessments, treatments, and interventions, for the training, education, development, and support of others involved in CF care.

For further detail, see the Cystic Fibrosis Trust **Standards of Care and Good Clinical Practice for the Physiotherapy Management of Cystic Fibrosis (2020)**.[9] *Please note, this consensus document is currently being updated and a new version will be available soon.*

3.6 Dietitian

General information

- Dietitians must be registered with the HCPC.
- They should be a member of the British Dietetic Association CF Specialist Group.
- They must have specialist knowledge and be experienced in the care of children and/or adults with CF.

Role

- Be responsible for providing full nutritional advice and assessment, including anthropometric measurements, nutritional support (including supplementation and dietary management), pancreatic enzyme replacement therapy (PERT) and gastrointestinal (GI) support, enteral tube feeding, CFD, and pregnancy, as appropriate to both in- and outpatients.
- Follow clinical dietetic practice that is evidence-based and reflects current research, clinical guidelines and consensus views.
- Be a resource on nutrition for the training, education, development and support of others involved in CF care.

For further detail, see the Cystic Fibrosis Trust **Nutritional Management of Cystic Fibrosis (2016)**.[10]

3.7 Clinical psychologist

Note

- Clinical psychologists are part of a wider category of specialised, doctorate-level trained practitioner psychologists, **as defined by the HCPC**.[11]
- Certain other practitioner psychologists, such as counselling psychologists, may be employed by the NHS to provide CF psychology services if they meet the person specification for a psychologist post and can deliver the full range of roles required in the job description.
- The title 'clinical psychologist' will be used throughout this guideline document.

General information

- Psychology services must be provided by a clinical psychologist embedded within the CF team. This work is best conducted when supervised or overseen by a consultant clinical psychologist.
- Clinical psychologists must be registered with the HCPC.
- Clinical psychologists should be members of the UK Psychosocial Professions in CF Group (UKPPCF).
- The psychology team should be adequately staffed and have a clear operational process that ensures that all referrals (for both in- and outpatients) can be assessed for urgency, and that urgent referrals can be responded to within one week.

Role

- Undertake psychological screening, as part of annual review, which allows an overview of emotional wellbeing, mental health (including but not limited to anxiety and depression), adjustment, coping, health-related behaviours, health management issues, family functioning, and the early identification of a wide range of psychological difficulties.
- Take a preventative approach to mental health issues for people with CF and their families/carers, for example through help to build coping skills, empowerment, and promotion of emotional wellbeing.
- Provide evidence-based psychological therapies.
- Provide support for newly diagnosed people with CF and their families/carers (at any age of diagnosis).
- Work jointly with other MDT members to help develop coordinated and psychologically informed approaches for people with CF with complex health problems and presentations.
- 'Gatekeeping' screening and onward referral of people with CF to mental health services and/or other relevant agencies (such as liaison psychiatry, community mental health services, specialist neurodiversity assessment services), and expert ongoing liaison with these services as appropriate.
- Liaise with Network CF Clinics, social services, and other community agencies for psychosocial input.
- Advise the CF MDT on management of significant risk issues when required and coordinate rapid access to on-call child and adult psychiatry services in cases of urgent psychiatric assessment for mental health concerns or risks (all CF Centres and clinics should be aware of their Trust's emergency psychiatric access policy, which in paediatric Centres should include access for family members).
- Provide consultation, support, and training to other CF professionals and supervision to those providing psychosocial interventions, to provide a 'stepped care' model, where the level of intervention and professional involved is matched to the severity of need.
- Help the CF team develop good models of self-care through advising on team processes to prevent, monitor, and manage staff stress, and manage staff wellbeing particularly at times of high stress, such as following a death or serious incident.
- Lead on development of psychological aspects of the CF service, for example through membership of working groups, team meetings and away days, and through service evaluation and QI initiatives.

For further detail, see the Cystic Fibrosis Trust **Guidelines for UK clinical psychology services in cystic fibrosis (2024)**.[12]

3.8 Social worker

General information

- Social workers should have at least three years' post-qualification experience working with vulnerable children or adults.
- They must be registered with their respective regulatory professional body.
- They should be members of the UKPPCF.

Role

- Understand the potential day-to-day and longer-term psychosocial impacts of living with chronic and progressive health conditions on the lives of people with CF and their families/carers.
- Contribute to the annual review of people with CF to assess physical and psychological wellbeing, cognitive capacity, ability to carry out activities of daily living, social functioning, physical environment, family functioning, and assessment of families and carers.
- Provide expertise in applying social work models and approaches, and contribute to multi-agency plans, procedures, assessments, and reviews for people with CF.

- Through ongoing assessments, identify and build on the strengths of a person with CF and empower them to manage their own lives and bring about positive change for and by themselves.
- Identify and provide early intervention to manage health inequalities that are due to adverse social factors, such as lack of economic or psychosocial support, and discrimination or abuse.
- Apply advocacy skills to ensure that people with CF understand their rights in accordance with relevant statutory legal frameworks and guidance for children and adults, can express their views and wishes, and have their voice heard.
- Ensure that stage-appropriate plans are offered and developed in a person-centred and timely way, such as at diagnosis, transition, transplant, and end-of-life care.
- Promote individuals' inclusion and participation in society, enable them to engage in meaningful and fulfilling activities, and provide support to strengthen their relationships with their families, significant others, and networks.
- Support people with CF and their families/carers to navigate and access statutory and voluntary services in line with their cultural, social, communication and personal needs.
- Work in partnership and share information with the MDT and internal and external agencies (such as social services, mental health organisations, education providers, employers, housing providers, substance misuse services, charities, care navigators, etc) in order to improve the life of the person with CF.
- Understand the financial needs of the person with CF and their families/carers, inform them of their welfare rights, and connect them to a variety of economic resources, including welfare benefits, charitable funds, and specialist financial advice and support.
- Identify social risks for the person with CF, such as food insecurity, poor housing and digital exclusion, and connect them to the necessary resources to address their social needs.
- Help people with CF stay safe and well, balance managing risk alongside being supportive, and apply the principles of proportionality in deciding if and when to intervene in their lives, whilst adhering to children and adult safeguarding legislation and local policy and procedures.
- Seek ways to advocate and influence policy at local and national levels along with UKPPCF consultations.

Further detail will provided in the upcoming Cystic Fibrosis Trust UK Social Work Services in Cystic Fibrosis guideline, which will be available soon.

3.9 Pharmacist

General information

- Pharmacists must be registered with the General Pharmaceutical Council.
- They should be members of the UK CF Pharmacy Group.
- The lead pharmacist should work in a highly specialist role with advanced practice, clinical leadership, and be actively prescribing, often working at regional and national level to influence service developments and policy.

Role

- Provide a prescription monitoring and medication review service for both inpatients and outpatients, which includes providing education to people with CF and their families/carers.
- Provide a full review at annual review and disseminate information to the GP and community pharmacist.
- Assist in optimising adherence to medicines by identifying intentional and unintentional non-adherence, and support and empower people with CF to improve their adherence.
- Pharmacy service provision should allow for access to an out-of-hours on-call pharmacist who can provide advice, information, and urgent medication where appropriate.
- Assist in the management of formularies, development and provision of clinical guidelines and treatment protocols.
- Provide advice on the legal and ethical responsibility of using medicines, including storage and unlicensed or off-label medicines.
- Advise on the procurement of difficult-to-source medications and aid in the resolution of any medication supply problems across secondary and primary care.
- Be involved in financial reporting on CF medication use.

For further detail, see the Cystic Fibrosis Trust **Pharmacy Standards in Cystic Fibrosis Care in the UK (2022)**.[13]

3.10 New and emerging roles

There are several new and emerging roles in the transforming field of CF, helping to provide holistic care across an extended scope of practice to support the broader CF MDT. These include CF practitioner roles, exercise practitioners, youth workers, and occupational therapists.

4. Principles of CF care

4.1 CFTR modulator therapies

- CF teams should continually review the availability of CFTR modulators for people with CF, taking into account any licensing and/or commissioning updates that may allow use in new age groups or CFTR variants. Access to a record of patients' genetics may be a useful aid in regular review of eligibility.
- People with CF should be offered the most effective, but tolerable, CFTR modulator, should more than one be available to them. The choice of which modulator, or whether they will take one, should be a shared decision between the person with CF, or their parents/carers depending on age, and the CF team.
- Education should be provided on the potential adverse drug reactions associated with CFTR modulators before initiating therapy and tolerability should be assessed at every contact with the CF team.
- CF teams, with input from their clinical psychologist, need to be aware of the potential for apparent direct psychological, psychiatric, and/or cognitive impacts of modulators, including a possible causal relationship between Kaftrio® and depression, and follow guidance to monitor any such impacts.
- People with CF taking modulators should be monitored in line with the recommendations of the manufacturer as detailed in the relevant Summary of Product Characteristics, including liver function testing and eye examination as appropriate.
- Early introduction of modulators can lead to pancreatic function changes. This should be monitored, and fat-soluble vitamin levels and enzymatic supplements should be reviewed.
- Education should be provided on the optimal way to take CFTR modulators in order to gain maximum benefits. The potential for drug-drug and drug-food interactions should be assessed before initiating treatment with CFTR modulators and reassessed whenever a new treatment is prescribed. Dose adjustments of CFTR modulators should be made in line with the recommendations of the manufacturer when use of an interacting medication cannot be avoided. Pharmacists are best placed to advise on drug interactions, dose adjustments, and alternative options. People with CF affected by this should be monitored closely for changes in CFTR modulator efficacy and adverse drug reactions. It is advisable that written information is sent to GPs so they are aware of potential drug-drug interactions between CFTR modulators and commonly used medications in primary care.
- Effectiveness of CFTR modulators should be monitored at every contact with the CF team.
- Non-effectiveness of CFTR modulators should be investigated in detail. This can include reassessing for drug-drug interactions, confirming adherence to treatment, measuring sweat chloride if indicated, and reviewing genetic analysis of CFTR variants.
- If any adverse drug reactions are present, they should be assessed by the CF team as soon as possible. All options should be considered to maintain treatment with a CFTR modulator if possible. This may include switching to a different modulator, if applicable, or a dose reduction. This will be dependent on the severity of the adverse reaction and it must be a shared decision between the person with CF, or their parents or carers depending on age, and the CF team, with a robust plan for future monitoring of the adverse reaction in place. Adverse drug reactions should be reported to the Medicines and Healthcare products Regulatory Agency (MHRA) via the Yellow Card scheme.
- Females of child-bearing age with CF should be educated on the link between increased fertility and CFTR modulators and either signposted to or provided with contraception advice, should they wish.
- CF teams should keep up to date with current recommendations for the use of CFTR modulators in pregnancy and breastfeeding. A detailed discussion should take place between the person with CF and the CF team outlining the risks and benefits of CFTR modulators in such situations, to allow the person with CF to make an informed decision of whether to continue or temporarily withhold CFTR modulators. If it is agreed that CFTR modulators are to be withheld, the person with CF should be monitored closely for possible deterioration in health.

For further detail about variant-specific therapy including modulators, see the ECFS **Standards of Care for CFTR Variant-specific Therapy (Including Modulators) for People with Cystic Fibrosis**.[14]

4.2 Medicines optimisation

Medicines optimisation is "a patient-centred approach to safe and effective medicines use, to ensure people obtain the best possible outcomes from their medicines". Important elements include taking medicines correctly, avoiding taking unnecessary medicines, reducing wastage and improving medicines safety. This involves cooperation between the person with CF and healthcare professionals to design, implement and monitor a therapeutic plan to produce the best possible healthcare outcomes. Although the pharmacist is a key member of the team assisting in medicines optimisation, this is done in partnership with the CF MDT.

The 2022 Cystic Fibrosis Trust **Pharmacy Standards in Cystic Fibrosis Care in the UK**[13] details requirements for pharmacy and support services, the role of the pharmacist, and strategies for optimising medicines. These standards describe the support services required to provide an effective service (clinical pharmacy technicians, dispensing, medicines information, procurement, distribution and homecare services).

4.2.1 Shared decision-making

- The person with CF should be placed at the heart of everything and advocate for shared decision-making to be embedded within MDT practice.
- People with CF, or their parents/carers, should be encouraged to share their perspectives and preferences about medicines with the CF team.

4.2.2 Prescribing new treatment

- Recommended medicines should be based on the best available evidence and address specific needs of individuals. Local and national formularies can be used to help guide these decisions.
- When a new medication is prescribed, the CF team should ensure that the person with CF, or their parents/carers, receives adequate education on intended benefits and how to optimally take the medicine to maximise benefits. They should also discuss timepoints for reviewing treatment effectiveness. Written information can help this process.
- Where possible, support children with CF and their families to move to tablet/capsule preparations, using available resources, such as the **KidzMed Programme**.[15]
- Any medication changes should be clearly and promptly communicated to the GP in order to maintain continuity of care.
- An up-to-date and accurate record of allergies and adverse drug reactions should be maintained and documented so that it is accessible to all members of the CF team and others involved in the care of the person with CF.
- A summary of common adverse drug reactions and potential ways to address or minimise these should be provided when a new medication is started.
- All potential adverse drug reactions should be reported in line with the recommendations of the MHRA Yellow Card scheme to ensure robust pharmacovigilance.
- The potential for drug-drug interactions should be assessed whenever any new medicines are considered.
- Individual considerations, including age, organ dysfunction, pregnancy, breastfeeding and immunocompetence, should be taken into account when medicines are prescribed or whenever such circumstances change.

4.2.3 Antibiotic prescribing and stewardship

- Antibiotics should only be used when clinically indicated. They should be started promptly and prescribed at the optimal dose and duration to give the best clinical outcome while minimising harm and potential for resistance. For further detail, see the Cystic Fibrosis Trust **Antibiotic Treatment for Cystic Fibrosis (2009)**.[16] *Please note, this consensus document is currently being updated and a new version will be available soon.*
- People with CF, or their parents/carers, should be given adequate education about antibiotic choice and the importance of completing the prescribed course to reduce antimicrobial resistance.
- Therapeutic drug monitoring should be used when available, to optimise dosing and/or reduce toxicity.
- Inhaled antibiotics delivered through continuous flow devices should be administered with an expiratory filter valve set and filter pads to reduce unnecessary exposure for other household members.
- CF teams should consider producing an antimicrobial formulary to assist in such decision-making.

- The need for long-term antibiotics should be continually reviewed.
- While antimicrobial sensitivity testing is controversial in CF, it can be a useful guide in targeting treatment against a particular pathogen. This is relevant where empiric treatment has not achieved an adequate response or antimicrobial resistance is suspected.
- CF teams should consider implementing other antimicrobial stewardship strategies, such as developing an MDT with involvement from microbiology and/or infectious disease consultants to assist in antimicrobial selection in the context of antimicrobial resistance.

4.2.4 Review of existing prescribed medication

- Ongoing appropriateness of all medicines should be reviewed at every interaction. This will include, but not be limited to, reviewing effectiveness of treatment, dosage, formulations and potential interactions.
- Assessment of adherence, including objective measures and a discussion around self-management of adherence to treatment and any barriers to adherence. Provide support to improve adherence, particularly in key life stages where more intervention is required (for detail on life stages of CF, see section 5.6).
- Consider stopping any treatments that are not necessary, in order to minimise treatment burden.
- All people with CF should have, at minimum, an annual review of their medication by the MDT, ideally by a pharmacist.
- Medicines reconciliation should take place during inpatient admissions and at annual review as a minimum. This should include a review of prescribed and non-prescribed medication (including herbal medicines and additional over-the-counter nutritional supplementation), and medicines supplied by homecare companies and any other specialities within secondary care.

4.2.5 Medicines management (medicine supply, stock, home delivery, and wastage)

CF teams should:

- Work with the person with CF, or their family/carers, to ensure that there is a clear understanding of the routes of supply required for their medicines.
- Resolve medication supply issues and liaise with primary care and homecare delivery companies to ensure an ongoing supply of medication and suggest alternative medication where necessary.
- Encourage safe inhaler disposal, either through recycling projects or through incineration.
- Avoid unnecessary stock accumulation, which is key in sustainable healthcare (for detail on CF care and sustainability, see section 4.3).

4.3 CF care and sustainability

The NHS has committed to a carbon net zero target by 2040.

Recommendations for the delivery of CF care:

- Make sustainable choices of treatment, for example in the choice of inhalers.
- Reduce waste, especially in medicines, and avoid the accumulation of unnecessary stocks.
- Reduce the impact of travel through the utilisation of technology, for example more virtual clinics.
- Reduce paper use, for example through the use of electronic health records.
- Empower people with CF to make sustainable choices, if appropriate, through access to information.

4.4 Infection control

- It is very important to prevent respiratory infections in people with CF. Whilst most bacteria are contracted from the environment, there is evidence of patient-to-patient spread of bacteria such as *Burkholderia cepacia* complex (Bcc), *P. aeruginosa*, *Mycobacterium abscessus*, and methicillin-resistant *S. aureus* (MRSA).
- Regular monitoring for cross-infection and epidemiological surveillance should take place by molecular typing.
- There must be local policies and clear operating procedures so that all people with CF are isolated from each other. Particular care must be taken over those infected with *B. cenocepacia*, non-tuberculous mycobacteria (NTM), *P. aeruginosa* and MRSA.
- Every person with CF admitted as an inpatient will be in their own room with en-suite facilities, to minimise the risk of cross-infection and to give them as much privacy as possible. Hospital facilities must maintain a high standard of cleanliness.

- Processes should be in place to avoid people with CF being in contact with each other in waiting areas, such as in CF clinics, wards, pharmacy, radiology etc.
- A high standard of hygiene should be practised by staff at all times, in particular hand washing; alcohol gels or other suitable preparations must be available in every room. All equipment, including stethoscopes, spirometers, and infant weighing scales, must be cleaned between each patient.

For further detail on infection control, see the Cystic Fibrosis Trust consensus documents:

- **Methicillin-resistant *Staphylococcus aureus* (MRSA) (2008)**[17]
- ***Pseudomonas aeruginosa* infection in People with Cystic Fibrosis. Suggestions for Prevention and Infection Control (2004)**[18]
- **The *Burkholderia cepacia* complex. Suggestions for Prevention and Infection Control (2004)**[19]
- ***Mycobacterium abscessus*: Recommendations for Infection Prevention and Control (2017)**.[20]

Please note, a new guideline on infection prevention and control in CF is in production and will replace these consensus documents once published.

4.5 Respiratory care

There are several important principles of respiratory care.

4.5.1 Monitoring of disease

- Respiratory samples should be obtained from people with CF who do not expectorate sputum, with cough swabs, cough plates, oropharyngeal culture, laryngeal or nasopharyngeal aspirate, induced sputum following hypertonic saline, bronchoalveolar lavage and bronchoscopy brush specimens.
- Airways should be sampled for infection at each clinic visit and with each respiratory exacerbation (expectorated or induced sputum sample preferred; otherwise, oropharyngeal sample). Induced sputum should be performed at least once a year in people with CF who cannot expectorate sputum, even if they are asymptomatic.
- Samples should be sent to a suitable microbiology laboratory. For further detail, see the Cystic Fibrosis Trust **Laboratory Standards for Processing Microbiological Samples from People with Cystic Fibrosis (2022)**.[21]
- Lung function should be monitored regularly with spirometry (including forced expiratory volume in one second (FEV_1) and forced vital capacity (FVC)) in adults and in children and young people with CF who can do this. Oxygen saturation measurements should be monitored with pulse oximetry.
- There should be regular (annual) monitoring with a chest radiograph. Chest CT scans should be carried out when appropriate and not routinely.
- There should be monitoring for complications, particularly allergic bronchopulmonary aspergillosis (ABPA), such as via serology, sputum, and radiology.

4.5.2 Therapies

4.5.2.1 General principles

Respiratory health is maintained with a combination of preventative and specific therapies. Non-respiratory factors also affect respiratory health.

- All routine childhood immunisations, annual influenza and COVID vaccines, are recommended as per the Green Book.[22]
- Smoking and vaping (the use of e-cigarettes) must be strongly discouraged for family members and carers from the time of diagnosis, as well as for people with CF. They should be offered help to engage in smoking cessation programmes.
- People with CF with persistent symptoms, such as decline in lung function and frequent pulmonary exacerbations despite appropriate therapy, should be evaluated for other causes, complications (including ABPA and NTM infection), and contributing factors, and assessed for levels of adherence.

4.5.2.2 Specific therapies

These include antibiotics for acute exacerbations and chronic infection, and use of airway clearance techniques and mucolytics to improve airways clearance.

Antibiotics and antivirals

For further detail, see the Cystic Fibrosis Trust **Antibiotic Treatment for Cystic Fibrosis (2009)**.[16] *Please note, this consensus document is currently being updated and a new version will be available soon.*

For acute exacerbations (oral and IV):

- There should be prompt recognition followed by early and aggressive treatment of lung exacerbations.
- Antiviral agents, for example for influenza and COVID, are indicated.
- Higher doses and longer antibiotic courses (both oral and IV) may be required compared to people without CF. However, treatment should be targeted to response and, in a changing population with CF, dose and duration should reflect individual need and tolerance.
- Eradication regimens should be implemented for first and subsequent new *P. aeruginosa* infections. This should take place promptly (within one week) from when the microbiology result is available. This also applies to Bcc and MRSA.
- Dual therapy using a combination of two IV antibiotics of different classes is recommended for IV antibiotic courses in people with CF infected with *P. aeruginosa*.
- IV gentamicin should be avoided; IV tobramycin and amikacin should be available on hospital formularies.
- Hospital laboratories must have provision to measure aminoglycoside blood levels, which, when prescribed, must be monitored regularly.

For prophylaxis and treatment of chronic infection:

- Antibiotic prophylaxis against respiratory *S. aureus* infection with flucloxacillin should be offered to children with CF from the point of diagnosis up to between the ages of three to six years, following a discussion of the uncertainties and possible adverse effects with their parents or carers.
- As chronic infection with *P. aeruginosa* is an important predictor of survival, treatment of chronic *P. aeruginosa* lung infections should be with inhaled antibiotics.
- Long-term azithromycin may be used in those with chronic infection.

Airway clearance and mucolytics

- People with CF should gain competency in airway clearance techniques (chest physiotherapy). For further detail, see the Cystic Fibrosis Trust **Standards of Care and Good Clinical Practice for the Physiotherapy Management of Cystic Fibrosis (2020)**.[9] *Please note, this consensus document is currently being updated and a new version will be available soon.*
- Other mucoactive respiratory therapies, for example dornase alfa, hypertonic saline and mannitol, should be used when appropriate.
- Initiation of dornase alfa should be considered in all people with CF from six years of age and started in all those not eligible for CFTR modulators.

Inhaled therapies must be delivered through an appropriate device. For further detail, see the Cystic Fibrosis Trust **Standards of Care and Good Clinical Practice for the Physiotherapy Management of Cystic Fibrosis (2020)**[9] and the ECFS **Inhaled Medication and Inhalation Devices for Lung Disease in Patients with Cystic Fibrosis: A European Consensus (2009)**.[23]

4.5.3 Respiratory complications and co-morbidities

- ABPA needs to be considered, especially when there is a lack of response to standard antibiotics. Monitoring of ABPA markers should also include a baseline at annual review.
- Pneumothorax may require initial urgent management in an emergency department, but the person with CF should be immediately referred on to a Specialist CF Centre; management is influenced by future impact on lung transplant surgery.
- Haemoptysis should be treated in a Specialist CF Centre when significant. It can often be managed conservatively but may require an expert bronchial artery embolisation procedure.
- For respiratory failure, assess the need for long-term or nocturnal oxygen therapy. If CO_2 elevation has developed, consider the appropriateness of instituting non-invasive ventilation.

4.6 Nutritional and gastroenterological care

There are several important principles of nutritional care.

4.6.1 Monitoring nutrition and gastroenterology status

- The main aim is to achieve a healthy nutritional status.
- Exocrine pancreatic status should be established at diagnosis by clinical assessment and confirmed by faecal elastase measurement.

People with CF who have exocrine pancreatic sufficiency may need re-checking when clinically indicated.

- Early introduction of CFTR modulators can lead to pancreatic function changes. This should be monitored, and fat-soluble vitamin levels and enzymatic supplements reviewed.
- Growth of infants and children should be measured regularly, including weight, height, and head circumference in infants up to two years old, and plotted on appropriate charts. Body mass index (BMI) should be monitored in all people with CF (and for children from two years of age, plotted on appropriate centile charts). In addition, alternative measures, such as body composition with dual energy X-ray absorptiometry (DEXA) scan, bioelectrical impedance, or hand grip strength, should also be considered.
- If growth is faltering or weight loss identified, a diagnostic review should be made. This should include analysis of dietary intake, malabsorption, and glucose metabolism.
- People with CF should receive support with weight management (losing, gaining, maintaining), regardless of whether they are on a CFTR modulator or not. People with CF may struggle with adjusting behaviours, for example after starting CFTR modulators or when their health status changes. For some people with CF, support may involve joint working between a psychologist, a physiotherapist, and a dietitian, focusing on a healthy lifestyle and supporting people with body dissatisfaction and disordered eating.
- At annual review, comprehensive nutritional assessments should be completed (for detail, see section 5.3.1).

For further detail, see the Cystic Fibrosis Trust **Nutritional Management of Cystic Fibrosis (2016)**.[10]

4.6.2 Therapies

- Nutritional status is independently linked to survival, so prompt intervention to normalise nutritional status is essential, especially in the first year of life. Generally, people with CF with optimal growth have better lung function and fewer infections. Advice from a dietitian is critical. For further detail, see the Cystic Fibrosis Trust **Nutritional Management of Cystic Fibrosis (2016)**.[10]
- PERT and fat-soluble vitamin supplements should be prescribed to people with CF with exocrine pancreatic insufficiency. Amounts given should be tailored to each individual – some may not need supplementation with all fat-soluble vitamins, as long as they are appropriately monitored.
- Nutritional support may be required for some people with CF. This can be through oral nutritional supplements or alternatively via enteral tube feeding.
- Access to a gastroenterologist with experience of CF is important for managing GI symptoms and complications such as malabsorption, severe gastro-oesophageal reflux, or DIOS not responding to standard therapies.

4.6.3 Complications

- Meconium ileus is the presenting feature in 10–15% of newborns with CF and is usually manifest before results of newborn screening are known. All babies with this condition must have CF excluded with genetic testing and a sweat test.
- DIOS will usually be managed by the CF specialist with input from the dietitian, however, when resistant to therapy, will need referral to a gastroenterologist with CF experience.
- Gastro-oesophageal reflux is common and should be excluded when people with CF are not responding to standard therapy, particularly when presenting with poor weight gain or intractable wheezing. In adults, cough, wheeze, and deterioration in lung function may be the presenting symptoms.
- Psychological difficulties related to body, weight, and shape, and/or disordered eating behaviours, will need specialist input from a dietitian and psychologist. In cases where these difficulties meet criteria for an eating disorder, then referral to, and ongoing liaison with, specialist eating disorders services will be needed.
- For recurrent acute pancreatitis (the majority of people with CF with this are pancreatic sufficient), referral to a gastroenterologist is appropriate.
- There is an increased risk of GI cancers in CF. For detail on bowel screening, see section 4.7.9.

4.7 Management of other CF manifestations and CF-related complications

There are a number of other problems associated with CF due to the multi-system nature of the condition.

4.7.1 Impaired glucose metabolism and CF diabetes (CFD)

- Impaired glucose metabolism can be accompanied by deteriorating clinical status, with a fall in lung function and weight for up to five years before diagnosis of CFD. CFD is associated with a worsening of health status, so early identification is critical. CFD results from pancreatic dysfunction related to CF and is distinct from Type 1 and Type 2 diabetes, but people with CF may be affected by any of these. Women with CF are commonly affected by gestational diabetes during pregnancy. Review of the diagnosis of CFD is important as glucose impairment may be transient. The diagnosis may be confirmed by the use of random blood glucose profiles or continuous glucose monitoring (CGM).
- It is recommended that all people with CF, including those who are pancreatic sufficient, are screened annually for CFD from 10 years of age, using an oral glucose tolerance test or CGM. For further detail, see the Cystic Fibrosis Trust **Management of Cystic Fibrosis Diabetes (2022)**.[24]
- Treatment of CFD should follow the Cystic Fibrosis Trust **Management of Cystic Fibrosis Diabetes (2022)**.[24]
- Joint management with a diabetes specialist experienced in CFD is important, as its management may be different to the management of diabetes in someone without CF.
- People with CF need specialist CF dietetic management as it is different from that given to people with diabetes without CF.
- All people with CFD should aim for optimal control of diabetes to reduce the chances of long-term complications.

4.7.2 Liver disease

- Annual screening for liver disease should be carried out in all people with CF.
- This will include regular examination for hepatosplenomegaly, annual blood liver function and clotting tests, and liver ultrasound for abnormal architecture and signs of splenomegaly starting from three years of age and subsequently when indicated. Routine repeat ultrasounds may not be necessary in adults with previous normal scans. Other means of imaging, such as fibroscans or MRI scans, may be helpful.
- People with CF should be referred to a hepatologist with CF experience for management of significant liver disease and its complications (for example, portal hypertension and oesophageal varices).
- Liver transplantation is an option for end-stage liver disease.
- Dietitians should regularly review any person with CF who has advancing liver disease.

4.7.3 Reduced bone mineral density (BMD)

- Screening for reduced BMD with a DEXA scan should be considered, depending on risk factors, baseline scans, and fracture history. For further detail, see the ECFS **European Cystic Fibrosis Bone Mineralisation Guidelines (2011)**.[25]
- Attention should be paid to nutritional status and intake, corticosteroid intake, vitamin D levels, and levels of physical activity (particularly weight-bearing exercise).
- Significant abnormalities, especially in children, should lead to referral to a bone specialist, endocrinologist or rheumatologist.

4.7.4 CF arthropathy and other musculoskeletal (MSK) issues

- Most MSK joint pain is non-inflammatory (non-CF arthropathy) and can be assessed and treated without rheumatology referral. When inflammatory arthritis (CF arthropathy or unknown) is suspected, then a rheumatology referral should be made for accurate diagnosis and management.
- There should be MSK expertise to manage postural issues and other common MSK complications of CF within the CF physiotherapy team, or a clear pathway of access to a physiotherapist with training and expertise in CF.

4.7.5 Nasal polyposis and sinusitis

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

4.7.6 Urinary incontinence

- A member of the CF team, such as a clinical nurse specialist or physiotherapist, should enquire about urinary incontinence for all people with CF. If present, physiotherapists should advise and manage or make a referral when necessary.

4.7.7 Renal complications

- Renal impairment can occur as a result of drug therapies, especially the frequent use of aminoglycosides – gentamicin in particular – as well as other complications, including CFD. Renal function (eGFR) and blood pressure should be monitored on a regular basis.
- Monitoring of renal function and blood pressure are of particular importance in people with CF who have undergone lung transplantation, as renal failure is a common complication of immunosuppressive therapy.
- Renal stones are more common in CF than in the general population, in particular among those with exocrine pancreatic insufficiency. People with CF with renal stones should be referred to a urologist with a specialist interest in stone management.

4.7.8 Hearing

- Hearing problems are more common in people with CF than in the general population. People with CF who are at risk of hearing impairment, including due to exposure to frequent courses of IV aminoglycosides, should be referred for an audiology assessment.
- Mitochondrial screening for risk of deafness with aminoglycosides should be offered to all people with CF, ideally immediately following a diagnosis of CF.

4.7.9 Bowel screening

- Due to an increased risk of colorectal cancer within an ageing population of people with CF, screening should be offered. At present, there are no UK bowel screening guidelines for people with CF. The Cystic Fibrosis Foundation **Cystic Fibrosis Colorectal Cancer Screening Consensus Recommendations (2018)**[26] should be followed. These recommend screening from the age of 40 years (or 30 years for those in receipt of a solid organ transplant), repeated every three to five years. For colonoscopies, appropriate CF bowel preparation should be followed.
- Faecal Immunochemical Test (FIT) kits are not recommended.

4.7.10 Psychosocial challenges of living with CF

- Psychological and behavioural problems are common and varied, and include issues with adherence to therapies, symptoms of anxiety and depression (which have been found to be two to three times higher in people with CF than would be expected in the general population), difficulties with body image and identity, eating difficulties, procedural distress, school problems, problems coping with distressing physical symptoms, concerns over family planning, adjusting to changes in morbidity and mortality and/or commencing new treatments, and end-of-life and transplant issues. The clinical psychologist can assess and offer support and psychological therapies for these difficulties (for detail on the clinical psychologist role, see **section 3.7**).
- The psychological impact of CF on children may change as they age, and CF teams should ensure children have a developmentally and age-appropriate knowledge and understanding of CF and how to take care of their physical and mental health.
- Access to psychosocial support for people with CF and their families/carers at key life stages (such as diagnosis, transition, pregnancy and parenthood, and end of life) is essential.
- The psychosocial impacts of new treatments (in particular CFTR modulators), both for those who are eligible and also for those who are not eligible (or are unable to take the treatment for any reason), need to be considered and monitored (for detail on CFTR modulator therapies, see **section 4.1**). For CFTR modulators, this should include both apparent direct psychological, psychiatric, and/or cognitive impacts, and more indirect difficulties with psychological adjustment (such as adjusting to wellness, changing illness perceptions and identities, etc).
- The **Your life and CF survey**[27] between March and April 2023 revealed the extent of additional costs that living with CF incurs. Financial strain and poverty are associated with worse physical and mental health outcomes. People with CF and their families/carers should have access to social workers, who have an important role in identifying social deprivation (and key elements of food insecurity, poor housing and digital exclusion) and connecting people with CF and their families/carers with appropriate sources of support.

- CF teams, particularly social workers, must be aware of challenges and offer strategies to manage CF-related issues in educational and workplace settings, assisting with decision-making about education and career options, signposting to programmes and initiatives to support people with CF into training and employment, and implementing reasonable adjustments to enable the person with CF to remain in or return to work and education.
- People with CF should be screened annually for potential psychosocial problems by a psychologist and social worker with experience in CF.
- All people with CF must have access to clinical psychology and social work services within the CF team. All referrals (for both in- and outpatients) should be assessed for urgency; urgent referrals should be responded to within one week.

5. Delivery of care

5.1. Making the diagnosis of CF

Where a clinical diagnosis of CF is confirmed, every effort should be taken to establish the genotype.

5.1.1 Diagnosis through newborn screening

In the majority of cases, the CF diagnosis will be made through the **newborn screening programme**.[28]

- When CF is suspected, the screening laboratory refers the child to the relevant Specialist CF Centre, which will depend on where the child is born and resides. Pathways must be in place for a referral from the newborn screening laboratory to the relevant Specialist CF Centre (within 24 hours). Pathways must be robust so that cases are not missed and all positive results are processed appropriately.
- Specialist CF Centres and their Networks must have a care pathway in place for contacting the parents in person and making the diagnosis in a timely fashion. Consideration must be given to reducing the waiting time and stress for the parents. A sweat test and clinical assessment should be performed no later than the day after informing the parents that CF is suspected, and five days after a positive screening result. This information should be given by someone who has comprehensive knowledge of CF, who will preferably be accompanied by a healthcare professional who already knows the family (such as a health visitor). This should ideally be face-to-face; if done by telephone, a structured call format should be used.
- Diagnosis should be confirmed by a sweat test and/or genetic variant analysis. Sweat tests must comply with the Guidelines for the Performance of the Sweat Test for the Investigation of Cystic Fibrosis in the UK (2014), available on the **Association for Laboratory Medicine website**.[29] The sweat test result should be given to the parents by the CF consultant as soon as it is available, preferably on the same day. If only one parent is available, a friend or relative should be invited to be present.
- While most children with a positive result from screening have a clear diagnosis of CF, there are cases when the diagnosis remains in doubt, with a discrepancy between genotype, phenotype and sweat test results. This may result in a CF screen positive, inconclusive diagnosis (CFSPID) designation. The management of babies and children with this designation is beyond the scope of this document. For further detail, see the ECFS **updated guidance on the management of children with CRMS/CFSPID (2020)**.[30]

For further information, see:

- ECFS **European Best Practice Guidelines for Cystic Fibrosis Neonatal Screening (2009)**[31]
- ECFS **Consensus on the Use and Interpretation of Cystic Fibrosis Mutation Analysis in Clinical Practice (2008)**[32]
- ECFS **A European Consensus for the Evaluation and Management of Infants With an Equivocal Diagnosis Following Newborn Screening for Cystic Fibrosis (2009)**[33]
- ECFS **Guidelines on the Early Management of Infants Diagnosed With Cystic Fibrosis Following Newborn Screening (2010)**.[34]

5.1.2 Diagnosis through clinical features

Some children and adults will be diagnosed following clinical suspicion of CF (symptomatic diagnosis) or through genetic testing, for example due to family history.

- People in whom CF is suspected should be referred to a Specialist CF Centre. A diagnostic pathway should be in place for older children and adults.
- A sweat test and genotyping should be arranged for everyone with clinical features suggestive of CF (irrespective of newborn screening test result) and results interpreted in line with guidelines to allow a diagnosis of CF, CFTR-related disorder, or CF unlikely.
- Extended genetic testing should be made available, and additional CFTR functional tests (such as nasal potential difference and intestinal current measurement) could be considered where sweat test and genotype results lead to an inconclusive diagnosis.
- Those presenting with CFTR-related disorders, such as congenital bilateral absence of the vas deferens (CBAVD), should be referred to a CF Centre for further evaluation and management as appropriate.

- Where a clinical diagnosis of CF is confirmed, every effort should be made to establish the genotype.
- Genetic counselling should be offered following diagnosis at any age of CF, CFSPID, or CFTR-related disorder.
- Infants presenting with meconium ileus should be treated as if they have CF until proven otherwise, with an early referral to the CF team. Awareness of the link between meconium ileus and CF should be raised with paediatric surgical teams.
- A detailed explanation and baseline investigation, similar to that offered to infants detected through newborn screening, should be given to all those newly diagnosed. Literature should be age-appropriate.

5.1.3 Following the diagnosis

- Within the first three to four months of a diagnosis, all people with CF should have the following investigations:
  - airway culture (cough swab, sputum)
  - assessment of pancreatic function, such as faecal elastase
  - extended genotype if only one of the common variants has been identified.
- Education for the parents/carers of the child with CF, or the person with CF if diagnosed later in life, should be provided by the full MDT within seven days. Contact details, including telephone numbers for the CF team, should be provided.
- Siblings of children with CF should be offered tests to exclude CF.
- The families of people with CF should be offered referral to the regional genetics service for counselling for future pregnancies. Siblings without CF will be offered carrier testing when they are old enough to make the choice.
- For adults with CF, consent is necessary to share the diagnosis.
- Access to appropriate information should be made available, including details of Cystic Fibrosis Trust.
- Psychosocial difficulties should be screened for and support offered to the individual and/or their family as appropriate following confirmation of the diagnosis.
- Consent to register on the UK CF Registry should be discussed.
- Testing for mitochondrial variants for risk of deafness with aminoglycosides should be offered following a diagnosis of CF.

5.2 Outpatients

Principles of infection control must be strictly maintained in clinic. For further detail on infection control, see section 4.4.

5.2.1 Frequency

- People with CF should be reviewed regularly, with a frequency appropriate to their individual needs.
- Adults and children with CF must be reviewed face-to-face at least once a year (including annual review) by the full Specialist CF Centre MDT, which may take place at either the Network CF Clinic or Specialist CF Centre hospital.
- Progress of children with CF should be discussed between the Specialist CF Centre and Network CF Clinic more than once a year. This may take different forms in different Networks, depending on the level of expertise at the Network CF Clinic and the severity of the individual's disease.
- It may be appropriate for a child with CF to be seen more often at the Specialist CF Centre.
- Newly diagnosed infants should be seen every one to two weeks during the initial learning period, which may include home visits by the community team. After this, they should be seen up to monthly for the first year.
- CF Centres should act promptly upon requests for follow-up from people with CF.

5.2.2 Procedures

The following should be carried out at every in-person clinic visit:

- Measurement of growth in children and weight or BMI in adults (for further detail, see section 4.6.1).
- Regular monitoring of lung function with spirometry (from five to six years of age), and oxygen saturation measurements with pulse oximetry.
- Culture of respiratory secretions (for further detail, see section 4.5.1).
- Blood pressure measurement, where appropriate.

For remote clinic consultations:

- Regular monitoring of lung function with spirometry (from five to six years of age), ensuring measurements are quality-assured by respiratory physiologists or other appropriately trained staff.
- Oxygen saturation measurements with pulse oximetry, when necessary.
- Weight. The same set of scales should be used for consistency.
- Height (for children with CF). Consider what measures can be put in place for reproducible height measurements using the same method. Consider supplying a suitable tape measure or stadiometer, and instructions for home use. Advice on obtaining accurate height and weight measurements has been published by the British Dietetic Association.[35]
- Microbiological surveillance for CF-related pathogens. Community/outreach resources might usefully quality-assure sampling; otherwise, postal services, private providers, or GP services might provide bespoke solutions for transporting specimens for laboratory processing.
- Uploading of intermittent and CGM reading.

5.2.3 Consultations

People with CF will see the members of the multidisciplinary CF team, who should be present at every clinic.

- **Doctor:** People with CF may not necessarily see a consultant at every visit, but the consultation is under their supervision, and should be discussed with the consultant either in the clinic or at a multidisciplinary meeting.
- **Clinical nurse specialist:** The nurse has a pivotal role in the clinic and should be available to see people with CF.
- **Physiotherapist:** Review of current respiratory status and physiotherapy-related treatments must be available to people with CF at every clinic visit. The physiotherapist will usually collect the microbiology samples.
- **Dietitian:** Nutritional advice and education must be available at every clinic visit.
- **Clinical psychologist:** People with CF and their families should have access to a clinical psychologist.
- **Social worker:** People with CF and their families should have access to advice from a social worker, particularly during times of clinical deterioration and life change, as adjustments may need to be made to work patterns, education, and employment.
- **CFD specialist nurse:** People with CFD or impaired glucose tolerance should have access to expert advice.
- **Pharmacist:** Advice relating to medicines must be available at every clinic visit through the pharmacist.

5.3 Annual review

- The annual review is a detailed assessment of every aspect of the person with CF's condition and therapies, to assess changes over the last year, identify where treatments can be improved, and produce a management programme for the following year.
- An annual review is a process, and does not necessarily need to happen all in one day, but will always include some in-person contact.
- A report should be written once all results are available, and sent to the GP, Network consultant, and person with CF or their family/carers.
- The report should be discussed with the person with CF or their family/carers and the treatment plan agreed.
- Data from the review is entered onto the UK CF Registry.

5.3.1 Consultations

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

- Collection of UK CF Registry data.
- Specific mention when appropriate of puberty, fertility, transition, plans for pregnancy, and transplantation.
- Physiotherapy review of airway clearance techniques, exercise, and inhaled medication regimens. They should also discuss any MSK issues and enquire about urinary incontinence. Home nebulisers and other home-based care equipment should be brought in for annual service when appropriate.
- Comprehensive nutritional assessment, incorporating anthropometric measurements and dietary and clinical considerations. Clinical considerations include a GI review (symptoms, complications and PERT management), glycaemic control, screening and bone health.

- Screening of the person with CF and their family/carers by a clinical psychologist and by a social worker, to identify and address any psychosocial issues related to living with chronic illness.
- Pharmacist review and discussion of all medication taken (both prescribed and non-prescribed), discussion around adherence, and review of the adequacy of supply arrangement.

5.3.2 Investigations

Centres may do additional investigations, but those listed below are the minimum expected:

- Lung function for people with CF over the age of six years.
- Exercise testing, when clinically indicated.
- Oxygen saturation measured by pulse oximetry. Assessment of blood gases may be measured in some adults with CF when clinically indicated.
- Respiratory sample such as sputum (may be induced) or a cough swab for microbiology. Lower respiratory tract samples should be used for mycobacterial sampling.
- Chest radiograph.
- Ultrasound of liver and spleen in children aged three years and above and as needed. Routine repeat ultrasounds may not be necessary in adults with CF with previous normal scans.
- Screening for reduced BMD with DEXA scan should be considered depending on risk factors, baseline scans, and fracture history.
- Assessment of glucose metabolism. Everyone with CF should have yearly screening for CFD from the age of 10 years. A variety of tools exist to identify abnormal glucose, including oral glucose tolerance tests (ideally with intermediate sampling), CGM, and serial blood glucose monitoring. Children under 10 years should be screened for CFD if there is concern about weight or height gain, decline in lung function, or any symptoms of hyperglycaemia.
- Blood taken for the following (which may vary according to local policy and include additional tests): full blood count, clotting studies, electrolytes and renal function, liver function tests, HbA1C, iron studies, aspergillus markers, and vitamins A, D, and E (ideally vitamin E:cholesterol ratio).

In addition to the above, other relevant investigations may include multiple breath washout (MBW) tests and fibroscans.

5.4 Inpatients

5.4.1 Principles

The majority of admissions are for IV antibiotics, either for a chest exacerbation or as part of routine management. However there are a number of other common reasons, including: education of the person with CF or their family/carers at time of new diagnosis or at other times; any deterioration in clinical condition that fails to respond to outpatient measures, including poor weight gain or weight loss; treatment of DIOS; liver disease; management of uncontrolled CFD; haemoptysis or pneumothorax; psychosocial support; elective procedures, such as insertion of a totally implantable venous access device, or gastrostomy, ENT or dental surgery; ante- or postnatal care; and palliation of symptoms, respite, or end-of-life care.

Inpatient care is a fundamental part of the management of CF, and beds in a ward suitable for CF care should always be available for an emergency admission, as well as capacity to ensure elective and urgent admissions can be managed appropriately.

- An urgent course of treatment should be implemented within a maximum of 24 hours of the decision being made. There should not be a delay of longer than seven working days of the proposed admission date for a non-urgent course of treatment.
- Principles of infection control must be strictly maintained in the hospital. For detail on infection control, see **section 4.4**.
- Facilities must exist for a parent or carer of a child with CF to stay with them in hospital.
- Local Authorities have a duty to provide suitable education for children of compulsory school age who cannot attend school due to illness. This education might be provided in a number of ways, which include hospital schools. However, children with CF should not be together in a hospital school room, so facilities must also exist for schooling at the bedside of the person with CF. In January 2013, the Department for Education (DfE) updated guidance on **arranging education for children who cannot attend school because of health needs**.[36] There is also statutory guidance document produced in 2015 on **supporting pupils at school with medical conditions**.[37]
- Reasonable adjustments should be made for anyone with communication needs.
- There should be access to appropriate play and/or recreation, with facilities for studying.

- Physiotherapists should be responsible for a full assessment (including airway clearance, MSK issues, exercise, urinary incontinence and respiratory support, including non-invasive ventilation when necessary) on admission. They should also be available to administer or supervise physiotherapy treatment twice daily (or more if necessary), including weekends; this may be with assistance from a physiotherapist or independently if the physiotherapist has previously assessed that to be appropriate.
- People with CF should have dietetic input as required during inpatient admission. Provision should be made for inpatients to have a choice of high-quality food and fluids to meet individual requirements.
- People with CF should be seen by a consultant with specialist CF knowledge at least twice a week. The consultant will be updated daily by the rest of the team.
- People with CF should have access to a clinical nurse specialist.
- People with CF should be reviewed by the pharmacist on admission for medicines reconciliation and inpatient prescription, at discharge for discharge medications, and also as needed during the week, with service provision of on-call advice on weekends and out of hours when necessary.
- People with CF should have access to a clinical psychologist within the CF team when on the ward.
- People with CF should have access to a social worker for advice and support with any financial, social, or emotional concerns.
- For people with CF without a totally implantable venous access device, there should be provision at all times for appropriate vascular access. This will usually be a peripherally inserted central catheter or peripheral long line, but there may be circumstances when a short IV cannula is used. Attention must be paid to procedural distress and an experienced doctor, nurse, or suitably qualified practitioner should perform the procedures. Procedural distress may be helped by play specialists and/or psychology support. Sedation may also be required.

Discharge planning is essential, especially if the person with CF is finishing the course of IV antibiotics at home. A summary must be sent to the GP, shared care consultant, and person with CF or their family/carers at discharge. The person with CF should be given a follow-up appointment at the time of discharge.

5.4.2 Investigations

For respiratory exacerbations:

- Lung function tests (spirometry, if feasible), oxygen saturation and blood gases (in adults with CF), and overnight continuous monitoring where appropriate.
- Monitoring of clinically relevant blood tests.
- Blood glucose monitoring. As well as for those with CFD, this is often carried out in older people with CF for the first few days of an admission.
- Aminoglycoside levels. Particular attention must be paid to this, especially if the person with CF is dehydrated (for example, due to DIOS or CFD) or has renal impairment or liver disease.
- A sputum sample or cough swab must be sent to microbiology within 24 hours of admission and repeated during admission if appropriate. A respiratory sample for virology, such as via nasopharyngeal aspirate, is sometimes indicated.
- Weight on admission and as advised and agreed between the person with CF and dietitian throughout admission. Additional weight investigations are, at minimum, weekly and upon discharge.
- A chest radiograph is only performed if clinically indicated, for example to exclude pneumothorax. They are not performed to check long line position.

Not all inpatient admissions are related to respiratory exacerbations, and other clinical investigations should be performed relevant to the suspected underlying clinical problem. For example, this might include abdominal imaging for suspected DIOS.

5.5 CF care delivery at home

5.5.1 Principles

- This innovation encompasses a range of activities related to CF care provided at home, from routine clinical care to the CF virtual ward, which delivers high-intensity hospital-level care to the person with CF in their home with input from the specialist CF team.
- Most Specialist CF Centres in the UK offer a home-based care service. The clinical nurse specialist usually provides this service, but in some places, other members of the MDT, such as CF practitioners, physiotherapists, dietitians and social workers, are also involved.

- A comprehensive service may support engagement with and adherence to treatment, reduce hospital admissions, and help people with CF and their families/carers balance healthcare needs alongside other competing demands and priorities, such as education, employment, and parenting. It can also lead to more timely interventions, especially when there are hospital capacity constraints.

A home-based care service can support many aspects of clinical and social care, including:

- Providing psychosocial support to people with CF and their families/carers, including advising on and addressing non-medical issues that support health and wellbeing. This could include tackling social risks – such as poor housing, isolation, and barriers to employment, education and patient engagement – as well as supporting with behaviour change and linking into community resources.
- Health education.
- Clinical assessments at home, including weight, glucose monitoring, lung function, and oxygen saturation.
- Home IV antibiotic courses (for detail, see section 5.5.2), including drug level monitoring and blood tests.
- Flushing of totally implantable venous access devices.
- Optimising airway clearance techniques and encouraging exercise programs.
- Collecting respiratory samples.
- Discussing personal issues, such as urinary incontinence.
- Supporting home enteral feeding.
- Pregnancy and postnatal support.
- Support when starting a new treatment, such as insulin or home oxygen.
- Liaison with the local authority regarding aids and adaptations in the home.
- Support to people with CF waiting for a transplant.
- End-of-life and bereavement support for family members.
- Liaison and communication with GPs and other local healthcare providers.
- Liaison with nurseries, schools, colleges, and workplaces.

5.5.2 Home intravenous (IV) antibiotics

- Home IV antibiotics may be suitable for some people with CF. CF teams need to evaluate their appropriateness, for example where non-adherence or home conditions are an issue. They should be sensitive to the needs of families/carers; for some families, admitting a person with CF to hospital is important for respite, to prevent home becoming hospital, to ease employment pressures, to prevent isolation of some adults, or because of the needs of other family members.
- When a person with CF is receiving IV antibiotics at home, close monitoring is required to assess progress and ensure aminoglycoside levels (if required) are within range.
- Home delivery of pre-prepared IV antibiotics should be considered and when appropriate offered to people with CF.
- Appropriate training to administer IV antibiotics must be given to the person with CF or their family/carers, and written competency checks recorded.

The risks of anaphylaxis and options available should be discussed – for example, first doses being given in hospital, possible use of EpiPens, and education for people with CF and their families/carers.

5.6 Life stages of CF

There is an increasing expectation that people with CF can live long, fulfilling lives largely unlimited by CF. It is important for CF teams to explore the beliefs of those with CF and their families/carers around future life expectations, hopes, and goals and to promote a 'start well, live well, and age well' approach.

As well as needing to acquire the more practical skills associated with independent living, such as cooking, cleaning, shopping, budgeting and money management skills – all of which are important prerequisites for a successful transition to adulthood and independent living – people with CF also need to consider how they will manage their health needs alongside other significant transitions, including entering employment, higher education, and living independently. It is important for CF teams to be able to signpost people with CF and their families/carers to services, to help facilitate reasonable adjustments that can help people with CF to achieve their goals.

5.6.1 Infancy, early years and childhood

- CF teams need to be aware of how a child's cognitive, social, and emotional development will impact on their engagement with CF and their care.
- Particular milestones where specific support might be needed from the CF team may include:
  - Weaning.
  - Starting nursery and primary school, where there may be anxieties about exposure to infections and/or CF treatments being managed outside the home for the first time. CF teams may need to work with families and these settings to support individual healthcare plans.
  - Transitioning to secondary school, where there may be anxieties about increasing independence with managing treatment routines, integrating CF into a person's identity, and/or disclosure of CF to new peers.

5.6.2 Adolescence and the transition to adulthood

- CF teams should consider how living with CF may impact on some of the key developmental tasks of adolescence, such as developing one's own identity (including sexuality and body image) and forming peer and intimate relationships. CF teams should support young people as appropriate as they consider how to disclose and manage CF in new environments and relationships.
- Consideration should be given by the CF team to issues that may become more pertinent in adolescence, such as sexual and reproductive health and psychological wellbeing. For detail on sexual and reproductive health, see **section 5.6.5**.
- The CF team should promote an awareness of a healthy lifestyle and the potential consequences of high-risk behaviour in the context of CF (for example, illicit drug use, smoking, vaping, excess alcohol, and unprotected sex).
- Adherence may be particularly challenging, and CF teams should be aware of the importance of manageable routines and the obstacles to adherence which may present in adolescence.
- Young people with CF need to be supported by professionals to develop self-care, empowerment, and patient leadership skills in their CF care, in line with national guidance, such as the **NHS Long Term Plan**.[38] For detail on person-centred CF care, see **section 2.2**.
- The CF team should support the young person with CF and their families/carers with the process of transferring CF care responsibilities to the young person in a structured and managed way, considering individual family functioning.
- The CF team should support and discuss plans for higher education, employment, and independent living. The CF team can work with the young person with CF to consider strategies for balancing healthcare needs alongside these transitions; assess and identify any gaps in living skills; and offer support and advice on a range of issues, including accessing suitable accommodation, financial entitlements, and paying for prescriptions, and can liaise with education providers and employers if required.

5.6.3 Transition to adult care

- Refer to **NICE Quality Standard 2016 QS140**,[39] as well as the recommendations from the **report on transition from children's to adults' services by the National Confidential Enquiry into Patient Outcome and Death (NCEPOD)**.[40] Transition includes both the gradual transition to independence in healthcare management and the transfer of care from a paediatric to an adult centre.
- When old enough, all children with CF will have their care transferred to adult services. It is essential that the young person with CF and their family/cares are involved in the early planning of this. Discussions should be started with the child with CF and their family/carers from when the child is 12 years old.
- The transition and transfer of young people with CF with additional needs (such as learning disability, neurodiversity, looked-after children, and those with complex comorbidities) requires a coordinated and integrated approach between CF teams, the young person with CF, their family/carers, and any other multi-agency providers, such as other healthcare providers, education providers, Child and Adolescent Mental Health Services, and social care services.
- Collaboration between the adult and paediatric teams is essential for successful transition and transfer of care. There needs to be a formally agreed protocol for transition, with identified coordinators from both teams and a full written MDT handover, including any information the young person with CF would like to share.
- Information about the process and the choice of adult CF Centre (when more than one Centre exists locally) must be given to the young person with CF and their family/carers.

- During the transition period, young people with CF should be offered the opportunity to attend part of their consultation without their family/carers present.
- The young person with CF and their family/carers should have the opportunity to formally meet the adult team on more than one occasion (usually in joint clinics) and view the adult facilities.
- The upper age limit by which transfer to adult services should have taken place is the 18th birthday of the person with CF.
- Transfer of care between CF Centres is equally important and requires close liaison between both services with full MDT communication.

5.6.4 CF in adulthood

CF teams should work to support people with CF to:

- Continue to develop self-management strategies, focusing on independence, autonomy, and decision-making skills, such as living well with CF alongside competing demands of education, work and parenthood, and supporting them to get reasonable adjustments in line with national guidance, such as the **NHS Long Term Plan**.[38]
- Attain goals that are important to them in education, career development, travel and/or moving geographically. This includes working with other adult Specialist CF Centres to facilitate Centre transition or to co-ordinate care, for example when a person with CF relocates periodically for education. For detail on the role of the social worker, see **section 3.8**.
- Access information and make informed decisions regarding sexual health, fertility, family planning, pregnancy and parenthood. For detail, see **section 5.6.5**.

5.6.5 Sexual health, fertility, family planning, pregnancy and parenthood

- The CF team should be aware of the potential impacts that having CF may have on sexual wellbeing and functioning, including those related to physical symptoms (such as breathlessness, fatigue, pain), treatment (such as the need for oxygen), and/or psychological factors (such as body image concerns, low self-esteem, low mood, and anxiety).
- The CF team should be ready to discuss these impacts and offer support and appropriate signposting to suitable information resources and services, such as **CF Reproductive and Sexual Health Collaborative (CFReSHC)**,[41] for information about sexual health, fertility, and contraception.

5.6.5.1 Female fertility

- Developmentally appropriate conversations about fertility issues should be had with all people with CF, throughout childhood and teenage years.
- The CF team should consider proactive discussion with all people with CF of childbearing age about fertility and their interest in pregnancy or becoming a parent.
- In particular, the CF team should check that people with CF are aware of the increased fertility on CFTR modulator therapies.
- The CF team should discuss contraception and signpost people with CF to suitable sources of information and services.

5.6.5.2 Male fertility

- Developmentally appropriate conversations about fertility issues should be had with all people with CF, throughout childhood and teenage years.
- All males should be offered semen analysis when appropriate.
- Males should be educated that they cannot assume they are infertile unless formally tested, and should take appropriate contraceptive (and safe sex) measures.
- There are options for males with CF to father babies, and referral for genetic counselling and assisted conception choices should be offered to males and their partners.

5.6.5.3 Pre-conception health and genetic counselling

- Where possible, all individuals with CF should be encouraged to discuss potential pregnancy (or partner's pregnancy) with their CF team before attempting to become pregnant, including genetic counselling and psychosocial issues. Pre-conception health status (such as BMI, diabetic control, vitamins, and haemoptysis risk) should be optimised, with more regular reviews if needed.
- Medication review should be carried out in conjunction with the pharmacist, and adjustments to treatments made where appropriate.

- People with CF should be counselled regarding the potential effects of becoming a parent on their health, and supported to explore how the family might cope with any future changes to health status.
- Open conversations need to be had with people with CF around any potential risks of pregnancy to the individual and to the unborn child. It may be helpful to involve obstetricians in these conversations and to offer support, including from the psychologist.
- Shared decision-making tools around becoming a parent with CF are available. CF teams should consider signposting people with CF to these resources – for example, the Cystic Fibrosis Trust **guide for those thinking of starting a family (2016)**.[42] Other options for becoming a parent, such as adoption, fostering, and surrogacy, should also be discussed, and sources of support should be signposted as appropriate.

5.6.5.4 Pregnancy

- With the advent of CFTR modulators, the number of people with CF becoming pregnant has increased year upon year.
- Approximately half of the pregnancies in people with CF are unplanned. It is important that CF teams can offer support to pregnant people with CF about all options available to them and provide signposting to additional support services, including counselling.
- Many guidelines and data on pregnancy in people with CF predate CFTR modulators, such as the **ECFS Guidelines for the Management of Pregnancy in Women with Cystic Fibrosis (2008)**.[43]
- CF teams should counsel pregnant people with CF on unknown risks to their foetus and tangible maternal benefits of continuing medications, including CFTR modulator therapies, through pregnancy.
- Normal changes that occur during pregnancy may adversely affect the pregnant person with CF. Thresholds of medical contraindication to pregnancy have not been established.
- Screening for diabetes should be offered at weeks 12 and 28 of pregnancy if the pregnant person with CF is not known to have diabetes.
- Diabetic control should be optimised, as poorly controlled CFD may contribute to adverse outcomes similar to diabetes in any pregnancy. For further detail about pregnancy and CFD, see section 6.2 in the Cystic Fibrosis Trust **Management of Cystic Fibrosis Diabetes (2022)**.[24]
- CF teams should encourage good dental care and offer exercise advice during pregnancy.

Coordination of care

- Where possible, the CF service should develop a relationship with obstetricians with expertise in CF (with support from suitably trained anaesthetists), who understand the issues associated with pregnancy and CF for initial discussions to plan the appropriate care pathway for each person with CF.
- Good communication is essential between the person with CF, the CF MDT, and obstetric teams during pregnancy. Consider monthly clinical review by the CF team during the second trimester of pregnancy, increasing to weekly or fortnightly during the third trimester.

Postnatal care

- Ongoing good communication between obstetric, paediatric, and CF teams and the parent with CF is of paramount importance.
- Close follow-up is necessary by the CF team to optimise the health of the mother postnatally. Particular consideration should be given to the formulation of a peripartum plan around management of constipation, airways clearance, and pelvic floor care.
- Clinical teams should consider potential CFTR modulator-related adverse events for the newborn related to *in-utero* exposure to CFTR modulators (animal reproductive models suggest no toxicity at normal human doses, but very limited human data is available to inform drug-associated risks during pregnancy), such as:
  - risk of cataracts for ivacaftor-containing products, based on juvenile animal models (newborn infant physical examination within 72 hours includes red reflex and is repeated at a six-week check by the GP, with referral to ophthalmology if concerned)
  - risk of hepatotoxicity, based on the known metabolism of the CFTR modulator.
- Postpartum care should include CF MDT review at four and eight weeks (more frequently if required). This should include contraception discussions, assessment of physical and mental wellbeing, and checking that adequate social support is available.

Breastfeeding

- CF teams should support people with CF with their decision-making regarding breastfeeding while on medications for CF, including CFTR modulators, in conjunction with the pharmacist.

- CFTR modulators have maternal benefits on nutritional status and lung health during the postpartum period, but have been shown in animal models and in humans to be present in breast milk.
- CF teams should liaise with obstetric and paediatric teams regarding possible CFTR-modulator-related adverse events for newborn exposure via breast milk, such as:
  - risk of cataracts for ivacaftor-containing products, based on juvenile animal models (newborn infant physical examination within 72 hours includes red reflex and is repeated at a six-week check by the GP, and an annual eye screen for the duration of breastfeeding, with referral to ophthalmology if concerned)
  - risk of hepatotoxicity, based on the known metabolism of CFTR modulators (suggest three-monthly whilst breastfeeding).

CF newborn screening

- Babies born to people with CF should receive the heel prick test at the standard time (between days five and eight).
- CF teams should speak with individuals who remained on CFTR modulator therapy through pregnancy about the potential for false negative CF screening of their infant.

5.6.5.5 Being a parent with CF

- The CF team should be alert to the additional needs of parents with CF, such as baby-changing facilities in clinics and trying to be flexible with clinic arrangements.
- New parents with CF may find it difficult to prioritise their own healthcare needs and regular monitoring, and support around adherence may be particularly important during this stage.
- The CF MDT should be alert to the potential needs of the dependents of people with CF under their care. Support needs may escalate at certain time points, such as admission to hospital, referral for transplantation, or end-of-life care.

5.6.6 Growing older with CF

With ongoing therapy advancements, the CF population in the UK is expanding and increasing in age and complexity. CF teams should work to adapt their service to help provide holistic care for this increasing and ageing population with CF.

- Multi-morbidity and coordinating care:
  - People with CF should be encouraged to lead their care (for detail on person-centred CF care, see **section 2.2**), but the CF MDT will have an increasingly important role in liaising with multiple allied specialties to deliver coordinated care for people with CF with comorbidities such as:
    - Cardiovascular disease
    - Diabetes (for retinal screening)
    - Hearing problems
    - Cancer (for engagement in screening programmes, such as for GI and cervical cancer)
    - Renal diseases
    - Bone diseases
    - Frailty
- Menopause:
  - It is important for CF teams to liaise with primary care practitioners to support women with CF during the menopause. For information about menopause and CF, see the Cystic Fibrosis Trust blog post: **Menopause and CF: Let's Talk About It!**[44]
- Retirement:
  - With the increasing life expectancy of people with CF, many will live to and beyond pensionable age and should be supported to access appropriate advice regarding financial planning and other considerations of this life stage. For further information on pensions, see **Citizens Advice**.[45]

5.7 Transplantation

5.7.1 Lung transplantation: pre-transplant considerations

- Lung transplantation offers the possibility of improved quality of life and survival for some people with advanced CF lung disease.
- All CF Centres should have a working relationship with at least one National Commissioning Group (NCG) designated lung transplant centre. Good communication is paramount to good care, and there should be clear lines of communication between the two services. Best practice is provided by regular joint MDT meetings between the two services.
- Clear indications for referral should be developed by each Specialist CF Centre in conjunction with the transplant unit.
- The care of people with CF should include consideration of the possible future need for lung transplantation, particularly with

respect to the management, avoidance, and mitigation of possible contraindications to future transplantation.

- Formal assessment of suitability and decisions about timing of placement on the waiting list should be carried out by the transplant centre.
- Early discussion and referral can help to optimise access and outcomes.
- The ongoing psychosocial needs of people with CF being considered for transplantation should be reviewed by the CF team, including support around deciding whether to pursue a transplant or not, and for those where transplant may not be possible or desired.
- Due to the uncertainty of transplant outcomes, end-of-life care should be discussed with the person with CF and their family/carers as part of the preparation for transplant.

For further detail, see the Cystic Fibrosis Foundation **consensus guidelines for lung transplant referral for individuals with CF (2019)**[46] and the International Society for Heart and Lung Transplantation **consensus document for the selection of lung transplant candidates (2021)**.[47]

5.7.2 Lung transplantation: post-transplant considerations

People with CF who have undergone lung transplant still have CF, with its systemic complications, and should be managed with collaborative working between the CF MDT and transplant teams.

- There should be a clear agreement, either on a person-by-person basis, or on an inter-service level, on which conditions/areas of care are the primary responsibility of which team. Due to the complexity of post-transplant immunosuppression, all lung complications should be managed and/or directed by the lung transplant centre. Responsibilities for other areas of care (for example, but not limited to, diabetes, low BMD, mental health, liver issues, GI complications, and renal issues) may be the responsibility of either the CF Centre or the lung transplant centre, but must be the responsibility of one or the other, agreed in advance, and clearly communicated to all those involved in care.
- Good communication is paramount to good care, and there should be clear lines of communication between the two services and the person with CF. Best practice is provided by regular joint MDT meetings between the two services.

5.7.3 Other solid organ transplantation

Other solid organ transplants, in particular liver, kidney, and pancreas, may be required by some people with CF. The CF MDT should have a working relationship with one of the NCG designated centres for liver/pancreas and renal transplantation.

5.8 Palliative and end-of-life care

- All Centres should have a palliative care team readily available to help and advise on management issues. Ideally, staff (especially in adult CF Centres) should have received some training in end-of-life care.
- All people with CF should have the opportunity to discuss end-of-life care with their physician and other members of the MDT, and should be offered advance care planning. End-of-life care and advance care planning should be patient-focused, and a care plan should be individualised according to the wishes of the person with CF. Individualised care plans should be reviewed upon deterioration of health or otherwise initiated by the person with CF. There needs to be clear documentation of end-of-life discussions and decisions with the person with CF and their family/carers. They should be asked where their preferred place of care would be at the end of their life (home, hospital, or hospice). For further detail about advance care planning, see the Cystic Fibrosis Trust **Advance Care Planning for People with Cystic Fibrosis (2017)**.[48]
- There should be recognition of physical, psychosocial, and spiritual concerns for all people with CF and their families/carers.
- Advance care planning should be undertaken with full involvement of the family.
- Balancing active treatment with adequate palliation of symptoms, especially for those awaiting transplant, can be a major challenge and may require support from the whole MDT and the palliative care team.
- When a person with CF has been identified as entering the last days or hours of life, this should be clearly communicated to the person with CF and their family/carers. There should be access to bereavement support for families/carers. If not available at the CF Centre, the CF MDT should signpost or help them access this support as appropriate. For information and support resources, see the Cystic Fibrosis Trust webpage on **bereavement and CF**.[49]

- Following the death of a person with CF, there should be a review of how well end-of-life care was managed, whether the person's priorities for care were achieved, and the effect of the death on the family, other people with CF, and the MDT. The CF Centre should undertake QI measures following this.
- Liaison with primary care is essential throughout the process.

6. Data, audits and quality monitoring

- Participation in audit and service evaluation is necessary to maintain high clinical standards and to ensure equity of care. Issues or challenges that impact on service delivery should be identified and acted on early.
- Specialist CF Centre staff should have protected time to contribute to audit, service evaluation, and QI.
- It is recommended that, at a minimum, Specialist CF Centres and, where relevant, Network CF Clinics:
  - contribute to the UK CF Registry and review the annual Registry reports
  - regularly gather and review patient experience feedback – for example, through the Cystic Fibrosis Trust PREMs surveys
  - contribute to the Cystic Fibrosis Trust staffing tool and review their annual summary.
- CF Centres should regularly discuss and review any action plans developed from the above data. Where services are linked through a Network, it is recommended that Specialist CF Centre and Network CF Clinic staff discuss and review data insights and actions plans together as a Network.

6.1 UK CF Registry

- The UK CF Registry is a key resource in service delivery in the UK, with the primary objective of raising standards of care.
- It is a secure centralised database, sponsored and managed by Cystic Fibrosis Trust, which records health data on consenting people with CF in England, Wales, Scotland, and Northern Ireland.
- CF Centres should encourage people with CF to consent and participate.
- In order for the UK CF Registry to be used effectively, the coverage should be complete (currently 99%) and the data collection should be detailed and accurate.
- Specialised CF Centre staff should have time and expertise to complete the data entry, review the reports, and disseminate the findings.
- The web-based software enables the collection of a standardised dataset, including patient-reported outcomes, across all paediatric and adult Specialist CF Centres and associated Network CF Clinics throughout the UK.
- The data can be accessed both regionally and locally. CF centres can access a dashboard and extracts from the UK CF Registry platform directly, to map and drive improvements in outcomes.
- A **UK CF Registry data report**[50] is published annually by Cystic Fibrosis Trust, and includes UK-wide as well as Centre-level analyses. It allows services to review their key outcomes and use of some medications, alongside UK averages.
- The UK CF Registry provides longitudinal data on the changing demographics of the CF population, the occurrence of complications (such as diabetes, liver disease, and cancer), and additional issues (such as increasing rates of pregnancy and parenthood). This data informs service planning for commissioners and service providers.
- Data collection via the UK CF Registry is of growing importance for clinical trials and effectiveness studies (such as CF STORM) and post-market authorisation of new medications for CF.

6.2 Other recommended audits and outcomes

- Aside from contributions to the UK CF Registry, Specialist CF Centres and Network CF Clinics in the UK should also monitor patient experiences (such as satisfaction with CF care, infection control, access and support from MDT staff, and hospital and home-based care), clinical activity, and resourcing levels (MDT staffing configurations, WTEs available for CF, banding of roles, and vacancies).
- **NICE Quality Standard QS168**[51] offers a service improvement Excel template to support an initial assessment of performance against NICE Quality Standard statements for CF.
- Services are also encouraged to participate in the three-yearly Cystic Fibrosis Trust PREMs survey, as well as the annual UK CF staffing tool, to systematically capture information on patient experiences and staffing levels in CF care. Data from these initiatives enable longitudinal monitoring of patient experiences and the CF workforce across the UK, as well as providing contributing CF Centres with bespoke data summaries to facilitate discussions about patient experiences and resourcing.

- The Cystic Fibrosis Trust QI team[52] also offer advice and support with data exploration and QI work.
- Alternatively, or in addition, Specialist CF Centres and Network CF Clinics may choose to undertake or participate in external peer review, internal audits, or activity monitoring, as well as other service evaluations, to explore quality and equity of CF care.

6.3 Quality considerations

This section outlines questions that should be considered when reviewing and reflecting on clinical practice and quality of care in CF services. These may be used for internal reflection and self-assessment, or during external service evaluations, such as peer review.

Staffing and resourcing

- How does the Specialist CF Centre ensure there is an MDT of trained and experienced CF specialist health professionals available and that staffing levels are appropriate for the Centre's size and population?
- How do staff maintain and evidence their CPD relevant to CF?
- What models to support staff with self-care, such as to manage stress, are in place? How can staff access health and wellbeing programmes?
- How does the service ensure that people with CF have access to specialist advice and care from their CF MDT at all times (as outlined in section 3.1)?
- What processes are in place to ensure all people with CF requiring hospital admission are placed in individual rooms with en-suite facilities?

Processes and pathways

- What SLAs and SOPs are in place for delivery of care by all outreach clinics and paediatric Network CF Clinics? How are these kept up to date and agreed with the Specialist CF Centre?
- Do the Centre and any paediatric Network CF Clinics (if relevant) send respiratory samples to a microbiology laboratory fulfilling the Cystic Fibrosis Trust Laboratory Standards for Processing Microbiological Samples from People with Cystic Fibrosis?[21] What is the process for checking clinic respiratory microbiology results?
- What arrangements are in place to minimise the risk of cross-infection in clinics, inpatient facilities and when delivering care at home?
- What is the provision for supporting care at home and in the community?
- What arrangements are in place to offer a blended model of care using new technologies, such as virtual clinics and remote monitoring?
- What arrangements are in place for medicines optimisation for people with CF?
- What arrangements are in place for joint care with a CFD specialist?
- What referral pathways exist in the Centre for referral to other medical and surgical disciplines, including hepatology, gastroenterology, obstetrics, ENT, and palliative care?
- What arrangements are in place for transplant advice and post-transplant care?
- What is the pathway for transition from paediatric to adult care? How do transition processes align with recommendations from the NCEPOD[40] into transition?
- How does the service monitor adherence and risk-taking behaviours?
- What arrangements are in place to ensure data is entered onto the UK CF Registry? What proportion of people with CF are entered?
- What arrangements are in place to ensure access to clinical trials and research for people with CF?

Service delivery

- How does the service ensure that all people with CF are seen at least once a year face-to-face by the full Specialist CF centre MDT?
- What proportion of the clinic population has had an annual review? Do all members of the CF MDT input into annual reviews?
- What evidence is there that all people with CF discuss the outcome of their annual review with a member of the CF MDT? How is written feedback from annual reviews offered to people with CF?
- What arrangements are in place for annual screening for psychosocial issues?
- What arrangements are in place to ensure all inpatients see a CF consultant a minimum of twice per week? What arrangements are in place for inpatient physiotherapy?
- What arrangements are in place for people with CF to receive home IV antibiotics? How does the service ensure all people with CF or their families/carers administering home IV antibiotics have undergone training and competency assessment?

Therapies and outcomes

- How, and how regularly, is eligibility for CFTR modulators assessed for all people with CF? What arrangements are in place to support shared decision-making about modulator therapies between the CF team and people with CF or their families/carers?
- How does the service monitor BMI? What data are available about BMI at the Centre?
- How does the service monitor lung function? What data are available about lung function at the Centre?
- What arrangements are in place in the CF Centre for the surveillance of transmissible organisms?
- What treatment protocols are in place to eradicate infections in all people with CF with a new infection, such as *P. aeruginosa*, MRSA or Bcc?
- What treatment protocols are in place for people with CF with chronic *P. aeruginosa*?

Patient experiences

- What arrangements are in place to elicit the preferences of people with CF and their families/carers and make shared decisions?
- What evidence is there of person-centred care (such as from patient experience surveys)?
- What arrangements are in place to capture the views of and experiences with the service from people with CF? What evidence exists of user satisfaction with care received (such as from routine feedback or patient experience surveys)?
- How are service users involved in service planning and delivery?

7. References

1 Cystic Fibrosis Trust. UK Cystic Fibrosis Registry. 2022 annual data report. 2023. Available at: **cysticfibrosis.org.uk/sites/default/files/2023-12/CFT_2022_Annual_Data_Report_Dec2023.pdf**

2 Castellani C, Simmonds NJ, Barben J, Addy C, Bevan A, Burgel PR et al. Standards for the care of people with cystic fibrosis (CF): A timely and accurate diagnosis. J Cyst Fibros. 2023 Nov;22(6):963–968. Available at: **doi.org/10.1016/j.jcf.2023.09.008**

3 Southern KW, Addy C, Bell SC, Bevan A, Borawska U, Brown C et al. Standards for the care of people with cystic fibrosis; establishing and maintaining health. J Cyst Fibros. 2024 Jan;23(1):12–28. Available at: **doi.org/10.1016/j.jcf.2023.12.002**

4 Burgel PR, Southern KW, Addy C, Battezzati A, Berry C, Bouchara JP et al. Standards for the care of people with cystic fibrosis (CF); recognising and addressing CF health issues. J Cyst Fibros. 2024 Mar;23(2):1569–1993. Available at: **doi.org/10.1016/j.jcf.2024.01.005**

5 Gramegna A, Addy C, Allen L, Bakkeheim E, Brown C et al. Standards for the care of people with cystic fibrosis (CF); Planning for a longer life. J Cyst Fibros. 2024 May;23(3):375-387. Available at: **doi.org/10.1016/j.jcf.2024.05.007**

6 National Institute for Health and Care Excellence (NICE). Cystic fibrosis: diagnosis and management. NICE guideline NG78. 2017. Available at: **nice.org.uk/guidance/ng78**

7 National Health Service. FutureNHS. Available at: **future.nhs.uk/**

8 Cystic Fibrosis Trust. National consensus standards for the nursing management of cystic fibrosis. 2001. Available at: **cysticfibrosis.org.uk/sites/default/files/2020-12/Nursing%20management%20of%20CF.pdf**

9 Cystic Fibrosis Trust. Standards of care and good clinical practice for the physiotherapy management of cystic fibrosis (4th ed). 2020. Available at: **cysticfibrosis.org.uk/sites/default/files/2020-12/Standards%20of%20Care%20and%20Good%20Clinical%20Practice%20for%20the%20Physiotherapy%20Management%20of%20Cystic%20Fibrosis%20Fourth%20edition%20December%202020.pdf**

10 Cystic Fibrosis Trust. Nutritional management of cystic fibrosis (2nd ed). 2016. Available at: **cysticfibrosis.org.uk/sites/default/files/2020-12/Nutritional%20Management%20of%20cystic%20fibrosis%20Sep%2016.pdf**

11 Health & Care Professions Council. The standards of proficiency for practitioner psychologists. 2023. Available at: **hcpc-uk.org/standards/standards-of-proficiency/practitioner-psychologists/**

12 Cystic Fibrosis Trust. Guidelines for UK clinical psychology services in cystic fibrosis. 2024. Available at: **cysticfibrosis.org.uk/sites/default/files/2024-07/CF%20Trust%20Psychology%20Guidelines%20V5.pdf**

13 Cystic Fibrosis Trust. Pharmacy standards in cystic fibrosis care in the UK (3rd ed). 2022. Available at: **cysticfibrosis.org.uk/sites/default/files/2022-12/CF%20Trust%20Pharmacy%20Standards%20of%20Care%20V5.pdf**

14 Southern KW, Castellani C, Lammertyn E, Smyth A, VanDevanter D, van Koningsbruggen-Rietschel S et al. Standards of care for CFTR variant-specific therapy (including modulators) for people with cystic fibrosis. J Cyst Fibros. 2022 Oct;22(1):17–30. Available at: **doi.org/10.1016/j.jcf.2022.10.002**

15 Royal College of Paediatrics and Child Health. KidzMed programme. 2022. Available at: **learning.rcpch.ac.uk/external-resources/kidzmed-programme/**

16 Cystic Fibrosis Trust. Antibiotic treatment for cystic fibrosis (3rd ed). 2009. Available at: **cysticfibrosis.org.uk/sites/default/files/2020-11/Anitbiotic%20Treatment.pdf**

17 Cystic Fibrosis Trust. Methicillin-resistant *Staphylococcus aureus* (MRSA). 2008. Available at: **cysticfibrosis.org.uk/sites/default/files/2020-12/MRSA.pdf**

18 Cystic Fibrosis Trust. *Pseudomonas aeruginosa* infection in people with cystic fibrosis. Suggestions for prevention and infection control (2nd ed). 2004. Available at: **cysticfibrosis.org.uk/sites/default/files/2020-12/Pseudomonas%20aeruginosa.pdf**

19 Cystic Fibrosis Trust. The *Burkholderia cepacia* complex. Suggestions for prevention and infection control (2nd ed). 2004. Available at: **cysticfibrosis.org.uk/sites/default/files/2020-12/Burkholderia%20cepacia.pdf**

20 Cystic Fibrosis Trust. *Mycobacterium abscessus*: Recommendations for infection prevention and control. 2017. Available at: **cysticfibrosis.org.uk/sites/default/files/2020-12/NTM%20guidelines%20Mar%202018.pdf**

21 Cystic Fibrosis Trust. Laboratory standards for processing microbiological samples from people with cystic fibrosis (2nd ed). 2022. Available at: **cysticfibrosis.org.uk/sites/default/files/2023-01/CF%20Lab%20Standards%20FINAL.pdf**

22 UK Health Security Agency. Immunisation against infectious disease. 2013 (updated 2020). Available at: **gov.uk/government/collections/immunisation-against-infectious-disease-the-green-book#the-green-book**

23 Heijerman H, Westerman E, Conway S, Touw D, and Döring G; consensus working group. Inhaled medication and inhalation devices for lung disease in patients with cystic fibrosis: A European consensus. J Cyst Fibros. 2009 Sep;8(5):295–315. Available at: **doi.org/10.1016/j.jcf.2009.04.005**

24 Cystic Fibrosis Trust. Management of cystic fibrosis diabetes (2nd ed). 2022. Available at: **cysticfibrosis.org.uk/sites/default/files/2022-12/CF%20Trust%20Diabetes%20Consensus%20FINAL.pdf**

25 Sermet-Gaudelus I, Bianchi ML, Garabédian M, Aris RM, Morton A, Hardin DS et al. European cystic fibrosis bone mineralisation guidelines. J Cyst Fibros. 2011 Jun;10 Suppl 2:S16–23. Available at: **doi.org/10.1016/S1569-1993(11)60004-0**

26 Hadjiliadis D, Khoruts A, Zauber AG, Hempstead SE, Maisonneuve P, Lowenfels AB; Cystic Fibrosis Colorectal Cancer Screening Task Force. Cystic fibrosis colorectal cancer screening consensus recommendations. Gastroenterology. 2018 Feb;154(3):736–745. Available at: **doi.org/10.1053/j.gastro.2017.12.012**

27 Cystic Fibrosis Trust. Your life and CF. 2023. Available at: **cysticfibrosis.org.uk/sites/default/files/2023-06/Your%20Life%20and%20CF%20Report%202023.pdf**

28 National Health Service England. NHS newborn blood spot (NBS) screening programme. 2023. Available at: **gov.uk/government/collections/nhs-newborn-blood-spot-nbs-screening-programme**

29 Association for Laboratory Medicine. Our Resources. 2024. Available at: **labmed.org.uk**

30 Barben J, Castellani C, Munck A, Davies JC, de Winter-de Groot KM, Gartner S et al; European CF Society Neonatal Screening Working Group (ECFS NSWG). Updated guidance on the management of children with cystic fibrosis transmembrane conductance regulator-related metabolic syndrome/cystic fibrosis screen positive, inconclusive diagnosis (CRMS/CFSPID). J Cyst Fibros. 2021 Sep;20(5):810–819. Available at: **doi.org/10.1016/j.jcf.2020.11.006**

31 Castellani C, Southern KW, Brownlee K, Dankert Roelse J, Duff A, Farrell M et al. European best practice guidelines for cystic fibrosis neonatal screening. J Cyst Fibros. 2009 May;8(3):153–173. Available at: doi.org/10.1016/j.jcf.2009.01.004

32 Castellani C, Cuppens H, Macek M Jr, Cassiman JJ, Kerem E, Durie P et al. Consensus on the use and interpretation of cystic fibrosis mutation analysis in clinical practice. J Cyst Fibros. 2008 May;7(3):179–196. Available at: doi.org/10.1016/j.jcf.2008.03.009

33 Mayell SJ, Munck A, Craig JV, Sermet I, Brownlee KG, Schwarz MJ et al; European Cystic Fibrosis Society Neonatal Screening Working Group. A European consensus for the evaluation and management of infants with an equivocal diagnosis following newborn screening for cystic fibrosis. J Cyst Fibros. 2009 Jan;8(1):71–78. Available at: doi.org/10.1016/j.jcf.2008.09.005

34 Sermet-Gaudelus I, Mayell SJ, Southern KW; European Cystic Fibrosis Society (ECFS), Neonatal Screening Working Group. Guidelines on the early management of infants diagnosed with cystic fibrosis following newborn screening. J Cyst Fibros. 2010 Sep;9(5):323–329. Available at: doi.org/10.1016/j.jcf.2010.04.008

35 British Dietetic Association Paediatric Specialist Group. COVID-19 – Guidelines for remote dietetic consultations. 2020. Available at: bda.uk.com/static/7bf38706-cf65-4bbe-97eec8f59928cbf9/BDA-PSG-Guidelines-for-Remote-Dietetic-Consultations-Final-2420.pdf

36 Department for Education. Arranging education for children who cannot attend school because of health needs. 2023. Available at: assets.publishing.service.gov.uk/media/657995f0254aaa000d050bff/Arranging_education_for_children_who_cannot_attend_school_because_of_health_needs.pdf

37 Department for Education. Supporting pupils at school with medical conditions. 2015. Available at: assets.publishing.service.gov.uk/government/uploads/system/uploads/attachment_data/file/803956/supporting-pupils-at-school-with-medical-conditions.pdf

38 National Health Service. The NHS Long Term Plan. 2019. Available at: longtermplan.nhs.uk/publication/nhs-long-term-plan/

39 National Institute for Health and Care Excellence (NICE). Transition from children's to adults' services (QS140). 2016 (updated 2023). Available at: nice.org.uk/guidance/qs140

40 National Confidential Enquiry into Patient Outcome and Death (NCEPOD). Transition from child into adult healthcare. 2023. Available at: ncepod.org.uk/2023transition.html

41 The Cystic Fibrosis Reproductive and Sexual Health Collaborative (CFReSHC). 2020. Available at: cfreshc.org/

42 Cystic Fibrosis Trust. Thinking of starting a family? A guide for adults with cystic fibrosis and their partners. 2016. Available at: cysticfibrosis.org.uk/sites/default/files/2020-11/Starting%20a%20family%20pack%202016.pdf

43 Edenborough FP, Borgo G, Knoop C, Lannefors L, Mackenzie WE, Madge S et al; European Cystic Fibrosis Society. Guidelines for the management of pregnancy in women with cystic fibrosis. J Cyst Fibros. 2008 Jan;7 Suppl 1:S2–32. Available at: doi.org/10.1016/j.jcf.2007.10.001

44 Cystic Fibrosis Trust. Menopause and CF: Let's talk about it! 2022. Available at: cysticfibrosis.org.uk/news/menopause-and-cf-lets-talk-about-it

45 Citizens Advice. Pensions. 2024. Available at: citizensadvice.org.uk/debt-and-money/pensions/

46 Ramos KJ, Smith PJ, McKone EF, Pilewski JM, Lucy A, Hempstead SE et al; CF Lung Transplant Referral Guidelines Committee. Lung transplant referral for individuals with cystic fibrosis: Cystic Fibrosis Foundation consensus guidelines. J Cyst Fibros. 2019 May;18(3):321–333. Available at: doi.org/10.1016/j.jcf.2019.03.002

47 Leard LE, Holm AM, Valapour M, Glanville AR, Attawar S, Aversa M et al. Consensus document for the selection of lung transplant candidates: An update from the International Society for Heart and Lung Transplantation. J Heart Lung Transplant. 2021 Nov;40(11):1349–1379. Available at: doi.org/10.1016/j.healun.2021.07.005

48 Cystic Fibrosis Trust. Advance care planning for people with cystic fibrosis. 2017. Available at: cysticfibrosis.org.uk/sites/default/files/2020-12/Advance%20Care%20Plan%20guideline%20for%20healthcare%20professionals%20v2.pdf

49 Cystic Fibrosis Trust. Bereavement and cystic fibrosis. Available at: cysticfibrosis.org.uk/life-with-cystic-fibrosis/difficult-conversations/bereavement

50 Cystic Fibrosis Trust. UK Cystic Fibrosis Registry: Reporting and resources. Available at: cysticfibrosis.org.uk/about-us/uk-cf-registry/reporting-and-resources

51 National Institute for Health and Care Excellence (NICE). Cystic fibrosis (QS168). 2018. Available at: nice.org.uk/guidance/qs168

52 Cystic Fibrosis Trust. Quality Improvement. Available at: cysticfibrosis.org.uk/the-work-we-do/quality-improvement

53 Standards of Care Working Group. Results of a survey exploring how CF workload is evolving for MDT staff in the UK. 2023. Unpublished.

54 Cystic Fibrosis Trust. UK Patient Reported Experience Measures (PREMs) Survey: Children's CF Services Report. 2023. Available at: cysticfibrosis.org.uk/sites/default/files/2023-09/PREMs_Paediatric_Report_2023.pdf

55 Cystic Fibrosis Trust. UK Patient Reported Experience Measures Survey (PREMs): Adult Services Report. 2022. Available at: cysticfibrosis.org.uk/sites/default/files/2022-06/PREMs_Adult_Report_UK_2022_FINAL.pdf

56 Cystic Fibrosis Trust. UK Cystic Fibrosis Service Resourcing 2020 to 2022. 2023. Available at: cysticfibrosis.org.uk/sites/default/files/2023-03/CF_Staffing_Report_UK_2023_FINAL.pdf

8. Appendices

Appendix 1: The UK Cystic Fibrosis Trust Standards of Care Working Group

Chairs

Professor Andrew Jones, Consultant Physician in Respiratory Medicine and Cystic Fibrosis, Manchester University Hospitals NHS Foundation Trust

Dr Maya Desai, Consultant Paediatrician in Respiratory Medicine and Cystic Fibrosis, Birmingham Women's and Children's NHS Foundation Trust

Members

Dr Caroline Elston, Consultant Physician in Adult Cystic Fibrosis and Respiratory Medicine and Site Medical Director Denmark Hill, King's College Hospital NHS Foundation Trust

Fiona Dowdall, Cystic Fibrosis Specialist Social Worker, Manchester University Hospitals NHS Foundation Trust

Dr Iram Haq, Consultant Paediatrician in Respiratory Medicine and Cystic Fibrosis, Great North Children's Hospital, Newcastle Hospitals NHS Foundation Trust

Isabelle Rosamond, Cystic Fibrosis Community Representative

Jacqueline Lowdon, Clinical Specialist Paediatric Dietitian in Cystic Fibrosis, Leeds Teaching Hospitals NHS Trust

Dr Jamie Duckers, Consultant Physician in Respiratory Medicine and Cystic Fibrosis, Cardiff and Vale University Health Board

Jade Conroy, Cystic Fibrosis Community Representative

Dr Jana Witt, Clinical Quality Improvement Manager, Cystic Fibrosis Trust

Dr Keith Brownlee, Director of Medical Affairs, Cystic Fibrosis Trust

Kerry-Lee Watson, Clinical Lead Cystic Fibrosis Specialist Dietitian, King's College Hospital NHS Foundation Trust

Michael Dooney, Cystic Fibrosis Specialist Pharmacist, Blackpool Teaching Hospitals NHS Foundation Trust

Professor Martin Walshaw, Consultant Physician in Respiratory Medicine and Cystic Fibrosis and Centre Director, Liverpool Heart and Chest Hospital NHS Foundation Trust

Dr Rachel Massey-Chase, Principal Clinical Psychologist in Cystic Fibrosis, King's College Hospital NHS Foundation Trust

Rachel Neilan, Cystic Fibrosis Community Representative

Rob Dixon, Cystic Fibrosis Community Representative

Samantha Henman, Lead Nurse for Cystic Fibrosis & CF Service Co-Lead, Royal Papworth Hospital NHS Foundation Trust

Dr Simon Langton-Hewer, Consultant Paediatrician in Respiratory Medicine and Cystic Fibrosis, University Hospitals Bristol NHS Foundation Trust

Dr Stephen J. Bourke, Consultant Physician in Respiratory Medicine and Cystic Fibrosis, Newcastle Hospitals NHS Foundation Trust

Dr Su Madge, Consultant Nurse and Cystic Fibrosis Centre Director, Royal Brompton & Harefield Hospitals

Tracey Daniels, Non-medical Clinical Lead, Cystic Fibrosis Specialist Physiotherapist, York and Scarborough Teaching Hospitals

Appendix 2: Evidence and process to develop staffing guidance

An open consultation with CF MDT staff invited by email via their respective CF professional groups (such as the CF Medical Association, CF Nursing Association, UKPPCF, etc) during September 2023 sought to identify activities and tasks that form part of CF care but are not routinely monitored or reported in NHS statistics. The consultation was open for one month and gathered 451 free text responses via Google Jamboard from CF MDT professionals. From the open consultation, a follow-up survey conducted with members of the CF workforce in the UK in October/November 2023 sought to understand how CF workload was evolving.[53] The survey was disseminated by email through the Chairs of CF professional groups and gathered 151 responses from CF staff across numerous services in the UK.

The results indicate that there has been a shift, rather than a reduction, in workload since the introduction of modulators:

- Inpatient workload was reported to have reduced in line with evidence from NHS statistics and the UK CF Registry, which show reduced rates of admissions and IV days.
- However, workload in other areas had reportedly increased – specifically, prescribing, virtual care, patient assessments, managing complexity, wellbeing support, and fertility and pregnancy care.
- Current time spent on patient-facing activities was perceived as adequate by most staff in the survey, though many felt they needed more time for personal and service development.
- Many staff also noted that they were increasingly being asked to cross-cover other specialties.

Insights from the Cystic Fibrosis Trust PREMs surveys:[54,55]

1. Access to doctors, nurses, physiotherapists and dietitians is perceived to be good, and the majority of people with CF are satisfied with the care and support they receive.
2. However, there are some persistent issues with access to psychosocial staff (clinical psychologists and social workers) and pharmacists in CF MDTs, with some people with CF reporting that they had to seek support privately due to a lack of access via their CF MDT.

Insights from the Cystic Fibrosis Trust annual staffing tool:[56]

1. There is variation in the composition of CF MDTs, and some services lack dedicated clinical psychology and social work support that is embedded within the MDT.
2. There are persistent issues with recruitment and retention, particularly with – but not limited to – psychosocial roles, with several services reporting long-term vacancies (more than six months) every year.
3. Many services also report that they are not satisfied with their current staffing levels.

The committee was concerned about the future, with a rising number of adults with CF and increasing complexity and comorbidity in an ageing population of people with CF. It was recommended that Specialist CF Centres, particularly for adults, need to start planning for a growing population now to be ready for the future.

Based on the minimum recommended sizes for paediatric and adult Centres, the committee considered the weekly sessions required to deliver a Specialist CF Centre service and agreed the required staffing levels for both service types. The tables below show how guidance was developed.

Paediatric care

Model for 100 patients

	Weekly sessions				Weekly WTE	Weekly WTE, incl. 20% uplift*
	Clinics	Inpatient work	Other work	TOTAL sessions		
Medical staff	5	5	2	12	1.2	**1.5**
Nurse	5	2	7	14	1.4	**1.75**
Physiotherapist	5	7	4	16	1.6	**2.0**
Dietitian	5	1	2	8	0.8	**1.0**
Psychology	5	0.5	0.5	6	0.6	**0.75**
Social work	5	0.5	0.5	6	0.6	**0.75**
Pharmacy	5	0.5	0.5	6	0.6	**0.75**
Administrator	0	0	6	6	0.6	**0.75**
				TOTAL WTE per 100 patients:	**9.25**	

*for development, training, leave, sickness etc.

Adult care

Model for 200 patients

	Weekly sessions				Weekly WTE	Weekly WTE, incl. 20% uplift*
	Clinics	Inpatient work	Other work	TOTAL sessions		
Medical staff	10	10	4	24	2.4	**3.0**
Nurses	10	4	14	28	2.8	**3.5**
Physiotherapists	10	14	8	32	3.2	**4.0**
Dietitians	10	2	4	16	1.6	**2.0**
Psychology	10	1	1	12	1.2	**1.5**
Social work	10	1	1	12	1.2	**1.5**
Pharmacy	10	1	1	12	1.2	**1.5**
Administrators	0	0	12	12	1.2	**1.5**
				TOTAL WTE per 200 patients:	**18.5**	

*for development, training, leave, sickness etc.

Cystic Fibrosis Trust

Cystic Fibrosis Trust is the charity uniting people to stop cystic fibrosis. Our community will improve care, speak out, support each other and fund vital research as we race towards effective treatments for all.

We won't stop until everyone can live without the limits of cystic fibrosis.

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We welcome your feedback on our resources. You can also ask for this resource in large print or as a text file.

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