Cystic Fibrosis Trust

# Standards for the nursing management of cystic fibrosis

## Second edition

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Uniting for a life unlimited

# The Cystic Fibrosis Nursing Association (CFNA) Working Group

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Recognition and thanks to previous CFNA Chairs and committee members who have worked on preceding versions.

Cystic Fibrosis Trust

# Contents

# Glossary

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| **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.* |
| **Concordance** | An agreement reached after negotiation between a patient and a healthcare professional that respects the beliefs and wishes of the patient in determining whether, when, and how medicines are to be taken. |
| **CF CNS** | Cystic fibrosis clinical nurse specialist. |
| **CFD** | CF diabetes (formerly CF-related diabetes, or CFRD). |
| **CFNA** | Cystic Fibrosis Nursing Association. |
| **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. |
| **CGM** | Continuous glucose monitoring. |
| **Echogenic bowel** | A condition where a baby's bowel appears brighter than normal on an ultrasound scan. |
| **Genotype** | The genetic makeup of an organism; it refers to the two alleles present at a specific locus in the genome. |
| **Genotyping** | The process of determining the DNA sequence, called a genotype, at positions within the genome of an individual. |
| **MDT** | Multidisciplinary team. |
| ***Meconium ileus*** | Occurs when a newborn's first stool, called meconium, is abnormally thick and sticky, blocking the small intestine. |
| **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. |
| **NMP** | Non-medical prescriber – nurses, physiotherapist, pharmacists and other allied healthcare professionals (AHPs) who have completed an accredited prescribing course and registered their qualification with their regulatory body, are able to prescribe. |
| **Nasal potential difference** | Used to measure the voltage across the nasal epithelium, which results from transepithelial ion transport and reflects in part CFTR function. |
| **OGTT** | Oral glucose tolerance test. |

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| **Phenotype** | The observable characteristics or traits in an individual based on the expression of their genes. |
| **Remote monitoring** | The ability to monitor certain aspects of a person with CF's health from their own home. |
| **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 has been produced by the UK and Ireland Cystic Fibrosis Nursing Association (CFNA). It supersedes the National consensus standards for the nursing management of cystic fibrosis published in 2001. The guideline is intended for nurses who provide care for people with cystic fibrosis (CF). It has been developed by both paediatric and adult nurses to provide an overview of care throughout the person with CF's lifespan. It provides guidance on the role of the CF clinical nurse specialist (CF CNS) within each area of care, and suggestions for audit based upon expert opinion and up-to-date evidence-based practice, but does not cover treatment pathways. Please refer to Cystic Fibrosis Trust's **Standards for the clinical care of children and adults with cystic fibrosis in the UK (2024)**[1] and local clinical guidelines for specific care pathways. It is recognised that multidisciplinary roles within CF teams can often vary across centres/clinics and guidance has been collated using the experience of the committee across multiple centres/clinics throughout the UK and Ireland.

Quotes from people with CF are included throughout to reflect how the role of the CF CNS has positively impacted their care experience.

> **"The CF CNSs are my first point of contact. I feel I can discuss anything with them about my CF care. The CF CNS is a crucial conduit to all the other healthcare professionals at a hospital/clinic. The CF CNSs are key to maintaining my health; I am grateful for the care of the CF CNS, keeping me as well as possible."**
>
> Peter, who has CF

## 1.1 Specialist care and the multidisciplinary team

Cystic fibrosis is one of the most common, life-threatening inherited diseases in the UK, affecting more than 11,300 people (1 in 2,500 live births). Cystic fibrosis is caused by mutations (variants) in the CF gene. Although F508del is the most common variant that causes CF in the UK population, more than 2,000 variants have been identified since the CF gene was discovered in 1989. Cystic fibrosis is a multi-system disease that causes a build-up of mucus in the lungs, digestive system, and other organs. Over time, this mucus blocks and damages the airways, leading to infections and making it hard to breathe. Other disease manifestations can include infertility in men with CF, liver disease, CF diabetes (CFD), and reduced bone mineral density.

Advances in medical and clinical management over recent years, such as the introduction of CFTR modulator therapy, means that people with CF are living longer and healthier lives. Due to the complexity of the condition, people with CF are best cared for under a specialist multidisciplinary team (MDT), made up of doctors, nurse specialists, dietitians, physiotherapists, pharmacists, psychologists, and social workers. These teams may be supported by other staff such as respiratory function technicians and ward nurses skilled in caring for people with CF. The CF MDT works together to provide holistic care for people with CF. Good communication is key for an effective CF MDT; nurses working in areas where there is no opportunity to work within an MDT must communicate with the nearest specialist CF centre, both for their own support and to assure optimal care for the person with CF.

> **"I am now in my early 60s and am very grateful for the level of care I have received from the nursing team."**
>
> Alan, who has CF

## 1.2 The cystic fibrosis clinical nurse specialist

Nursing is a partnership between people with CF, their family/carers, and nurses. The roles of the CF CNS include advocacy, clinical management, education, support and advice, and research.

- **Advocacy:** the CF CNS must act to safeguard the rights of the person with CF and ensure they receive the highest possible level of care by advocating for the person and their family.
- **Clinical management:** the CF CNS should work with the person with CF and the CF MDT in decision-making and monitoring of care. The CF CNS should maintain up-to-date knowledge to ensure that people with CF receive optimum care on an individualised basis. The role of the CF CNS will often involve co-ordination with other services, both in the hospital and community.
- **Support and advice:** the CF CNS can offer expert advice and specialist support to people with CF

and their families, signposting to other services when required.

- **Education for people with CF, families, and carers:** the CF CNS will teach people with CF, families, and carers about the condition and treatments according to individual need. This can support informed decision-making, safe and effective carrying out of treatments, and adherence to treatments.
- **Professional development:** the CF CNS is responsible for maintaining their continued professional development, remaining up-to-date with advances in the treatment and management of CF, and using evidence-based practice to guide clinical care.

The CF CNS often acts as the first point of contact and will co-ordinate many aspects of a person's care, including appointments, letters, prescription requests, accessing other services and signposting.

**The CF CNS should work in line with the CFNA mission statement of inclusivity:**

The CFNA is committed to promoting equality, diversity and inclusion in everything we do. We feel passionately that inequalities or discrimination should not exist and we actively want to play a part in tackling this. We are an organisation where diversity is embraced, celebrated and welcomed. We stand against all forms of discrimination and prejudice, whether on the grounds of ethnic background, sex, gender, sexual orientation, age, religion, disability, or any other aspect of a person's identity. We will always champion tolerance, fairness and equality in all aspects of our work. We recognise that inequalities in society lead to inequalities in health outcomes and this is no different for those affected by CF, as for any health condition. Our nurses respect and embrace diversity and are committed to caring for individuals with CF and their families.

## The role of the cystic fibrosis clinical nurse specialist

# 2. Advanced practice

Nurses have been working at an advanced level within CF care for many years, however in recent years frameworks have been developed to standardise advanced practice across healthcare. The Health Education England **Multi-professional framework for advanced clinical practice in England (2017)**[2] defines advanced clinical practitioners as – healthcare professionals whose practice encompasses a high degree of autonomy and complex decision-making across four pillars: clinical practice, leadership and management, education, and research. According to the Royal College of Nursing (RCN) **Standards for advanced level nursing practice** (2018),[3] advanced practice in nursing requires staff to be trained to MSc level and have expert clinical knowledge and skills. As some advanced practice roles within CF preceded the framework, some nurses may be in advanced practice roles without an MSc.

**The advanced CF CNS practitioner should:**

- Be trained to MSc level.
- Be an expert in the field of CF.
- Share their knowledge through education.
- Head nurse-led clinics.
- Conduct nurse-based research.
- Lead on CF service development.

They may also be an independent prescriber.

**Suggestions for audit:**

- What banding and qualifications do the CF CNSs in your service hold?
- How is CPD maintained?
- What research has been conducted and has it led to changes in practice?
- What service development has been implemented and what impact has it had?
- What are service users' experiences of advanced practitioners in their service?
- Has the journey of a person with CF been impacted by advanced practitioners within your service?

# 3. Non-medical prescribing

Non-medical prescribing (NMP) within CF care is popular as it enables timely prescriptions and a seamless, responsive service, reducing the need for people with CF to travel to hospital or interact with their GP to obtain prescriptions. Furthermore, many CF CNSs are skilled in physical assessment, which enables diagnosis and requisition of radiological investigations in the absence of medical staff. NMP is an additional qualification that can be undertaken when appropriate, based on service need and staffing requirement. For more information, see the **RCN advice guide for non-medical prescribers**.[4]

**The CF CNS should:**

- practice within the NMP competency framework (**prescribing-competency-framework.pdf**)

**Suggestions for audit:**

- How many prescriptions have been completed and for what medications?
- Have there been any prescription errors?
- What time savings have been observed using non-medical prescribers?
- What are service users' experiences of non-medical prescribers in their service?

# 4. Research

Research is paramount to inform evidence-based practice. Opportunities to participate in research may range from observational studies and questionnaires to clinical trials. People with CF and family/carers who wish to take part in research studies must be given the correct information and time to allow them to make an informed decision about participation. People with CF and family/carers should be reassured that their routine care will not be compromised by their participation or non-participation in research. All members of the MDT will be governed by standards of clinical research as defined by the National Institute for Health Research to ensure they are qualified by education, training and experience.

> **"Nurses always let us know of a new trial but don't pressure us into doing them, it's nice to be informed."**
>
> Molly, who has CF

Cystic Fibrosis Trust supports clinical trials in the UK through the Clinical Trials Accelerator Platform (CTAP). They work with pharmaceutical companies (sponsors) to promote the robust clinical trials infrastructure established in the UK through the CTAP programme, encouraging sponsors to open their CF trials in the UK. CTAP also brings together and funds a network of CF centres and trial coordinators to support the delivery of a broad range of CF trials in the UK. They can support healthcare professionals to inform people with CF about clinical trials through their online and print resources.[5] They also list current UK trial opportunities on their Trials Tracker.[6]

**The CF CNS should:**

- Complete and maintain good clinical practice and research training, and follow the study protocol and standard operating procedures.
- Maintain knowledge and understanding of research studies and trials available to people with CF.
- Signpost people with CF and family/carers to opportunities to participate in research.
- Work collaboratively with the research team to ensure that people with CF and family/carers are fully informed of the specific research study or trial being offered.
- Support people with CF and family/carers with their continued participation or their choice to withdraw from a study or trial.
- Maintain competencies to perform delegated tasks, as required.
- Maintain records accordingly in line with Nursing and Midwifery Council (NMC) standards, local policy, clinical research standards and study protocols.

**Suggestions for audit:**

- Is the CF CNS aware of available current research studies?
- Is research documentation complete?
- Are consent forms filed in patients' notes?

# 5. Shared care

People with CF may require care provided across more than one CF clinic or centre; this is often the case when a student goes to university. It is important that people with CF know where their agreed designated/nearest centre is when living away from home, so they can access help if needed.

**The CF CNS should:**

- (at the referring centre or clinic) Advise the person with CF where their agreed designated/nearest centre is when away from home, and if possible provide contact details for them (see **Cystic Fibrosis Trust's list of adult and children's specialist CF centres**).[7]
- (at the referring centre) Co-ordinate a referral to the receiving CF centre. This may involve a separate nursing referral, or phone call to the receiving centre for verbal handover.
- (at the receiving centre) Send the centre's contact information and, if applicable, offer an appointment at their centre.
- Work collaboratively and maintain good communication between the centres and/or clinics.

**Suggestions for audit:**

- What are the number of referrals made for shared care and the reasons for referral?
- What are the number of referrals received for shared care and the reason for referral?
- Is there a clearly documented referral for shared care?

# 6. Diagnosis

A diagnosis of CF can be made at any age, however most people are diagnosed with CF shortly after birth from newborn screening. Since 2003 in Scotland and 2007 in the rest of the UK, all newborn babies have been screened for CF as part of the national newborn screening programme. This is offered when babies are five to eight days old. A 'CF suspected' screening result is referred to a specialist CF centre, as per the national standard. Other signs of CF in newborns include echogenic bowel (antenatally), *meconium ileus*, and rectal prolapse.

Whilst a diagnosis of CF is usually made within the first year of life, some people with CF may not be diagnosed until later in life, even well into adulthood. There are a number of false negatives each year from the screening programme, those that decline screening, and those designated with **Cystic Fibrosis Screen Positive, Inconclusive Diagnosis (CFSPID)**.[8] There are also adults born prior to the introduction of newborn screening who may have gone undiagnosed as children due to variance in the symptoms and severity of CF.

**When clinical evidence leads to suspicion of CF, a diagnosis can be made with:**

- sweat testing
- genotyping
- nasal potential differences.

**The CF CNS should:**

- Actively participate in delivering the diagnosis to a child and their family or adult, aiming to review the person suspected to have CF as soon as practically possible.
- Provide support to the family with the psychosocial impact of the diagnosis, offer opportunities to discuss concerns, ensure ongoing psychosocial support as required, and signpost to relevant information and support resources in their preferred language.
- Provide contact details and information about the CF MDT, ensure timely involvement of the CF MDT, and promote good communication within the team.
- Assess and formulate a clinical management plan with the person with CF and/or their family/carers, including appropriate follow-up arrangements.
- Establish a good working relationship with primary care to ensure the needs of the person with CF are met, providing training and education where required.
- Identify and raise concerns to the CF MDT and refer to multi agencies to ensure the wellbeing of the person with CF.
- Convey knowledge and understanding of the genetic variants of the person with CF and the implications for prognosis and treatment, accessing the CFTR2 database where appropriate.
- Convey knowledge and understanding of current and historical research in the field of CF, and introduce people with CF and their family/carers to the opportunity of research, current studies, and seek to gain informed consent to the UK CF Registry.
- Act as an advocate from diagnosis, striving to overcome social, cultural, and racial injustice and access to healthcare and meeting the needs of the diverse population we serve.
- Educate people with CF and their families/carers about the risk of infection and cross-infection. See **section 15. Infection prevention and control** for further detail.
- Share information with people with CF and their family/carers about the basic genetic implications for the wider family and refer to clinical genetics for counselling where required.

**Suggestions for audit:**

- Did the CF CNS participate in diagnosis?
- Has the person with CF and/or their family/carers met with all members of the CF MDT within the first two weeks of diagnosis?
- Has the person with CF and/or their family/carers been given contact details for the CF MDT?
- Has the person with CF and/or their family/carers been given written information about CF?

# 7. Inpatient care

People with CF who require inpatient admission should be cared for by nursing staff who have a good understanding of CF at a recognised specialist CF centre or network CF clinic. Infection control guidelines should be adhered to in accordance with local policy. A treatment plan will be discussed and decided between the person with CF and/or their family/carers and the CF MDT. Any changes will be documented and discussed with the person with CF and/or their family/carers. The option of home intravenous (IV) therapy can be discussed where appropriate. Upon discharge, changes to treatment should be communicated to all relevant people, such as the primary care team, shared care consultant, schools and nurseries.

The CF CNS often facilitates the admission; co-ordinating the date and logistics of the admission with the person with CF/family/carers and the ward staff.

> **"After spending 27 days in hospital, the nursing team checked in every day making sure I had everything I needed and was okay. No one thing was too small and I felt I could ask them anything during this time."**
>
> Molly, who has CF

**The CF CNS should:**

- Facilitate and co-ordinate the admission.
- Support people with CF and their family/carers during inpatient admission.
- Support the ward nurses with their assessment on admission to the ward.
- Provide ongoing support to ward nurses ensuring they have adequate knowledge and experience to deliver appropriate care, using and managing various access devices.
- Facilitate access to the CF MDT for the duration of the admission.
- Co-ordinate and/or provide appropriate follow-up arrangements upon discharge.

**Suggestions for audit:**

- Do ward staff have access to appropriate education and assessment to ensure continuity in maintaining skills and learning in CF?
- Can ward staff state which recognised specialist CF centre the person with CF is registered with?

# 8. Outpatient care

Outpatient care may encompass virtual or face-to-face appointments to review and maintain patient wellbeing. For detail on virtual consultations and remote monitoring, see section 2.2.1 of the **Cystic Fibrosis Trust Standards for the clinical care of children and adults with CF in the UK (2024)**.[1] At each routine outpatient visit the MDT should be available to assess the physical, physiological and psychosocial needs of the person with CF. The person with CF and/or their family or carers should be given opportunity to discuss their care with each member of the MDT. For measurements to take at outpatient clinics, see **Cystic Fibrosis Trust's Standards for the clinical care of children and adults with cystic fibrosis in the UK (2024)**.[1]

Other care completed in the outpatient setting includes annual reviews, allied health professional-led clinics, commencement of home IV antibiotics, and clinical trials.

People with CF and/or their families/carers should be made aware of which specialist CF centre they are under and how to contact them, particularly when a person receives outpatient care within an agreed designated network CF clinic.

**The CF CNS should:**

- Offer the option of a virtual appointment to people with CF, when appropriate. Virtual appointments should be delivered in line with local policy.
- Support attendance at clinic by establishing barriers to attendance.
- Facilitate appointment reminders where appropriate.
- Implement/facilitate infection control policies in the outpatient setting.
- Facilitate use of equipment to measure lung function, oxygen saturation, weight, height.
- Support people with CF and their families/carers during outpatient appointments.
- Act as an advocate for the person with CF.
- Perform necessary assessments, investigations and sampling.
- Co-ordinate the MDT to facilitate time-efficient appointments.
- Co-ordinate or facilitate nurse-led clinics and appointments.
- In the event of 'failure to attend', aim to establish the reasons and how to facilitate future appointments.

**Suggestions for audit:**

- Does the person with CF have the names and contact numbers of the CF MDT?
- What measurements were taken and documented during the person with CF's outpatient appointment? Do these align with measurements recommended in **Cystic Fibrosis Trust's Standards for the clinical care of children and adults with cystic fibrosis in the UK (2024)**?[1]
- Which members of the CF MDT were available at the appointment?
- What was the duration of visits and the time spent with each member of the MDT?
- Is telemedicine a suitable method of review?

# 9. Community care (home/domiciliary visits)

People with CF will have access to specialist advice, support and direct supervision from the CF CNS in the community as required, enabling the individual to meet their health needs. Home/domiciliary visits will be conducted in line with local policy, patient need, and centre provision. Geographical area may impact ability to provide home visits.

> **"This ensured continuity of care and reduced hospital admissions where possible. Treatment at home gave my son more independence and was better for his mental wellbeing."**
>
> Tracey, whose son Levi passed away in 2023

**The CF CNS should:**

- Facilitate opportunities to discuss community care.
- Formulate a care plan and document in conjunction with the person with CF and/or their family/carers, which should be re-assessed on a regular basis.
- Provide the person with CF and their family/carers with 24-hour contact numbers for support and advice.
- Facilitate safe delivery of effective treatment and care in the community.
- Liaise with community nursing teams/schools/nurseries as required.

**Suggestions for audit:**

- How many home visits occurred in the previous 12 months?
- What were the reasons for the home visit?
- What was the duration of the home visit?
- What was the total travel time incurred for the home visit, compared to the visit duration?

# 10. Transition, adolescence and adulthood

Transition is the process of moving from paediatric to adult care. It can be an unsettling time for people with CF and their families/carers. People with CF should receive individualised care, appropriate to their needs during the transition process. A date/timeframe for transition should be agreed by both the paediatric and adult MDT. The young person with CF and their family/carers should have the opportunity to meet the adult MDT on more than one occasion and view the adult facilities.

> **"Nurses sat in on appointments with paediatrics so we knew them and they knew us, how we work, and how we like things, making the transition to adult care as smooth as it could be."**
>
> Molly, who has CF

**The CF CNS should:**

- Provide a person-centred approach to transition according to individual needs.
- Provide education and resources to aid the transition process.
- Encourage and support collaborative working between the paediatric and adult services.
- Provide an overview and contact details of the adult service.
- Promote autonomy of care.

**Suggested areas for audit:**

- Does the person with CF know the details of their transition, such as the planned date and the adult centre, ahead of time?
- Was a tool used and documentation completed?

# 11. Lifestyle

**People with CF should be encouraged to live a healthy, balanced lifestyle, this includes:**

- Consuming a healthy and balanced diet and staying well hydrated.
- Regular exercise.
- Plenty of sleep.
- Abstinence from smoking/vaping.
- Consumption of no more than 14 units of alcohol per week (men and women).
- Where appropriate, maintaining safe sexual health.

The multidisciplinary team approach in CF care provides the person with CF/their families/carers with team members expert in these fields.

Dietary advice for people with CF should be individualised as this is influenced by factors such as pancreatic status, weight and nutritional status, gastrointestinal issues such as inflammatory bowel disease or secondary co-morbidities such as diabetes, which may require dietary modifications. (CF dietitian).

Exercise is beneficial for people with CF as it not only helps lung health and clearance, but also helps to reduce the risk of other medical conditions, such as high blood pressure, diabetes and even some cancers. It strengthens muscles and bones and reduces risk of fractures; improves balance, reducing the risk of slips, trips and falls; and improves brain function and mood. Physical activity and exercise are important to promote good bone health and reduce the risk of osteopenia and osteoporosis (CF physiotherapist/ exercise specialist).

Good sleep hygiene can help improve sleep quality, which in turn can improve mood, concentration and memory; prevent sleep disorders (such as insomnia); help maintain a healthy weight; and lower the risk of developing health conditions such as diabetes and heart disease.

**The CF CNS should:**

- Provide a person-centred approach to lifestyle advice according to individual needs.
- Facilitate opportunity for people with CF to openly discuss smoking, vaping and recreational drug use.
- Have knowledge of the special dietary requirements of a person with CF and factors influencing this.
- Monitor height, weight and Body Mass Index (BMI) in people with CF.
- Have an awareness (and provide patient education) regarding possible weight gain after starting CFTR modulators.
- Have an awareness of NHS alcohol consumption guidance.
- Ensure people with CF are referred for screening for reduced bone mineral density (BMD) if deemed necessary depending on risk factors, baseline scans, and fracture history.
- Ensure people with CF are referred for annual screening for CF diabetes through an oral glucose tolerance test (OGTT) or continuous glucose monitoring (CGM).
- Be aware of services and information to signpost people with CF to for lifestyle advice.
- Be aware of smoking cessation services and referral processes.
- Monitor vaping use (including what is being vaped), and offer support to reduce usage.
- Be aware of support services for those using recreational drugs.
- Have knowledge of sleep hygiene recommendations.
- Provide opportunity to discuss and review sexual health, with knowledge of where to signpost for expert advice.

**Suggestions for audit:**

- What evidence is there of chart documentation of weight, BMI and pancreatic sufficiency?
- What is uptake of annual OGTT/CGM among people with CF? What is the percentage of people with CFD in the service?
- What information has been provided to people with CF starting CFTR modulators about possible side effects, including insomnia and weight gain?
- Has there been opportunity to discuss sexual health?

# 12. Family planning, pregnancy, perimenopause and menopause

People with CF should have the opportunity to discuss and receive appropriate information and support about fertility, contraception, family planning, and pregnancy according to their individual health and psychosocial needs. People with CF and their partners should be supported through any fertility treatment, pregnancy, and becoming parents.

## 12.1 Fertility and contraception

**The CF CNS should:**

- Support conversations around:
  - Male and female fertility (including impact of CFTR modulator therapy on fertility in women) and, when appropriate, contraception during paediatric care. Conversations should continue throughout and beyond transition.
  - Family planning and the impact of pregnancy on women with CF.
  - The likelihood of a couple having a baby with CF.
- Provide education, information resources, and support on male and female fertility, contraception and family planning.
- Facilitate CF carrier testing for partners of people with CF (which requires a blood sample), or refer to an appropriate service to facilitate this.
- Facilitate or co-ordinate a referral to clinical genetics for genetic counselling, if required.
- Support referral to appropriate departments for fertility testing and treatment.
- Support people with CF through adoption and surrogacy processes.

## 12.2 Pregnancy

**The CF CNS should:**

- Support medication reviews and make adjustments as appropriate, particularly the use of CFTR modulator therapy. Non-medical (independent) prescribers with experience and competency can conduct the review.
- Facilitate joint CF and obstetrics clinics, and promote systems to allow a co-ordinated approach.
- Support diabetes care throughout pregnancy and co-ordinate care with specialist obstetric diabetes services.
- Facilitate access to the full CF MDT for pregnant women with CF.
- Support pregnant women with CF in complex situations.
- Provide education, information resources, and support around breastfeeding with CF.
- Provide education, information resources, and support around being a parent with CF.

## 12.3 Perimenopause and menopause

Recent evidence suggests that women with CF develop perimenopause and menopause symptoms two to three years earlier than people in the general population (The Cystic Fibrosis Reproductive and Sexual Health Collaborative (CFReSHC)).[9] Symptoms can sometimes be overlooked alongside their CF, therefore it is important the CF CNS facilitates opportunity to discuss and explore perimenopause/menopause.

**The CF CNS should:**

- Facilitate and support conversation about perimenopause/menopause.
- Maintain a basic knowledge of perimenopause/menopause symptoms.

- Signpost primary care providers/specialists who can provide expert care and advice.
- Support and empower women with CF throughout their perimenopause/ menopause journey.
- Provide education, information resources, and support around perimenopause/ menopause, including signposting. For example Cystic Fibrosis Trust's **Menopause and CF: Let's talk about it!** webpage.

**Suggestions for audit:**

- Has literature on fertility, family planning and contraception been provided to the person with CF?
- Did the person with CF, and their family/ carers have the opportunity to discuss the process and impact for them?
- Has perimenopause/menopause been discussed with women with CF? Do staff feel confident that they have basic knowledge of perimenopause/menopause symptoms?

# 13. Transplant

Conversations around lung and other solid organ transplantation referral can be daunting for people with CF. The CF CNS will offer support and information around transplantation appropriate to the individual physiological, psychological and social needs of the person with CF. The process of referral for transplantation should be approached in conjunction with discussion of end-of-life care. If the person with CF makes an informed decision not to be referred for transplant assessment, the CF CNS will provide appropriate support and consider end-of-life care planning. The person with CF will be shown respect in their decision not to pursue transplantation as an option, if this is their wish.

> **"Support from nursing staff was key when considering and agreeing upon the decision to sign up for transplantation. The continued support during the assessment process and subsequent time on the waiting list was much welcomed."**
>
> Gareth, who has CF

**The CF CNS should:**

- Support the person with CF and family/ carers in discussions leading up to transplant referral, documenting their physical and emotional needs, and signposting to other services where necessary.
- Provide information about the transplant process, including timelines.
- Support the co-ordination of investigations required for transplant referral.
- Facilitate communication with the transplant team where necessary.

**Suggestions for audit:**

- What was the CNS's input in completed referrals to transplant team?
- Is there transplant in CF literature available for the person with CF and their family/ carers?
- Is there a documented assessment of the needs of the person with CF, including their emotional needs?
- Did the person with CF and their family/ carers have the opportunity to discuss the transplant process?

# 14. End-of-life care and symptom management

People with CF at the end stage of disease will receive support, symptom control and care appropriate to their individual needs to allow them to die with dignity. People with CF and their families/carers will be fully informed about the choices available to them (within the parameters of resources available). Good communication is paramount when planning all aspects of end-of-life care. The goal is to ensure that the wishes of a person with CF and their family are respected.[10]

An Advanced Care Plan (ACP) is a formal document done voluntarily by person with CF and their family/carers so that final wishes are known and supported by the MDT and family.[11] The person with CF and their family/carers will have the opportunity to choose home, hospital, or hospice-based end of life care. This arrangement will be flexible with changing needs. Regular assessment and monitoring of symptoms will be carried out and management planned accordingly, including anticipatory prescribing for symptom management. The person with CF and their family/carers should be informed of changes and have the opportunity to explore their feelings. Bereavement support often begins before death and continues afterwards.[10]

**The CF CNS should:**

- Engage in advance care planning discussions with the person with CF and their family/carers, along with other appropriate members of the CF MDT.
- Co-ordinate support between the ward team, community team and the family.
- Signpost and co-ordinate appropriate support services, such as for emotional or financial support.
- Collaborate with the palliative care and pain teams, forming links for advice and support (memory boxes, keepsakes etc).

**Suggestions for audit:**

- Is there a documented ACP for the person with CF?
- Are people with CF given the opportunity to explore end-of-life discussions and/or planning at annual review?

# 15. Infection prevention and control

Effective infection prevention and control (IPC) in CF care is paramount. The CF CNS should have knowledge of IPC in CF to ensure that people with CF have minimal possible exposure to the potential acquisition of infection.

> **"Infection control has always been key, especially for totally implantable venous access devices, midlines etc. The CF nursing team go above and beyond, which helps put the mind of the patient to ease."**
>
> Gareth, who has CF

**The CF CNS should:**

- Provide ongoing evidence-based and up-to-date information on IPC to people with CF at a level to meet their individual need, and document this provision.
- Educate people with CF on how to minimise infection transmission within their daily life and ensure they can demonstrate this.
- Provide people with CF and their family/carers with information resources relating to IPC in CF (if required).
- Facilitate infection control practices in clinic and during home/domiciliary visits.
- Ensure the person with CF has contact numbers for support if they require further information.
- Ensure people with CF and their family/carers have the understanding and ability to correctly disinfect their equipment, such as nebulisers or home spirometers.
- Ensure people with CF and their family/carers are provided with information about their infection/colonisation and the antibiotics required to suppress or eradicate it, including possible side effects.
- Understand all potential routes of transmission of infection in people with CF in healthcare and community settings, including:
  - airborne
  - blood-borne
  - faecal
  - oral
  - environmental, including stagnant water, warm-water systems
  - pets and animals.
- Support the availability of the necessary equipment and materials to ensure good IPC.
- Identify all potential risks of infection for people with CF, such as with:
  - the inpatient environment
  - the outpatient clinic
  - enteral feeding via percutaneous endoscopic gastrostomy (PEG) or jejunostomy
  - IV antibiotic therapy or the insertion of any IV device
  - respiratory support
  - nebuliser therapy
  - care of indwelling venous devices in hospital and at home.
- Operate within local IPC guidelines.

**Suggestions for audit:**

- Are arrangements in place for microbiological surveillance to identify control issues and use of particular antibiotics?
- What proportion of people with CF have chronic *Pseudomonas* infection?
- Is data collected on lung function, rate of decline, BMI? Are all deaths reviewed? How does data compare to other centres or UK CF Registry data?
- What are the number and resolution time of complaints about IPC?
- Does the service have and adhere to a departmental risk register?

# 16. Adherence/concordance

Good adherence to treatment is directly related to a longer, better quality of life for people with CF. People with CF and their families/carers will have access to education and support from the CF MDT, to empower them to take control of their CF and treatment regimes.

**The CF CNS should:**

- Have an understanding of CF medications and potential barriers to adherence, including time constraints of school, work, denial of need, lack of understanding, frustration, accessibility, embarrassment, forgetfulness, and hopelessness.
- Support people with CF and their families/carers to identify barriers to adherence and think of solutions to overcome these.
- Encourage and co-ordinate attendance at clinics to facilitate assessment of clinical response to treatments.
- Support people with CF and their family/carers to understand and manage their CF.
- Facilitate access to the CF MDT for support as needed.
- Identify markers for non-adherence, such as missed appointments, reduced response to medications, and missing repeat prescriptions from the GP.
- Encourage and facilitate people with CF and their family/carers to access psychosocial support and advice regarding adherence when required.
- Be easily contactable in between clinics for advice surrounding all aspects of CF care that may interfere with adherence to treatments.

**Suggestions for audit:**

- Are barriers to adherence documented in notes?
- Are adherence monitoring systems in place?

# 17. Safeguarding

Safeguarding aims to protect the health, wellbeing and human rights of people with CF, enabling them to live free from harm, abuse and neglect. Safeguarding practice for both adults, children and young people should reflect best practice nationally. A person with CF should be treated as an individual, with dignity and respect to ensure they feel safe in services, and empowered to make informed choices.

The ethnic background, language, religious beliefs, sexuality, and gender of the person with CF should be taken into account when planning care. Reasonable adjustments should be made as needed.

**The six safeguarding principles are:**[12, 13]

- **Empowerment** – patients should be supported and encouraged to make their own decisions and informed consent.
- **Prevention** – to take action before harm occurs.
- **Proportionality** – to make the least intrusive response appropriate to the risk presented.
- **Protection** – to offer support and representation for those in greatest need.
- **Partnership** – to look for local solutions through services working within their communities. Communities have a part to play in preventing, detecting and reporting neglect and abuse.
- **Accountability** – for accountability and transparency in safeguarding practice.

Protecting your own wellbeing when responding to safeguarding concerns is important to ensure that staff can manage the emotional and work challenges that they might encounter. Teams should support each other and signpost to appropriate local resources to maintain the wellbeing of their staff. In some cases, such as medical neglect, the CF CNS will lead safeguarding plans, liaising and advising the social care team.

**The CF CNS should:**

- Complete approved safeguarding training and development commensurate with their roles and responsibilities.
- Act in line with the values and principles set out in the NMC **Code of professional standards of practice and behaviour for nurses, midwives and nursing associates**.[14]
- Recognise when immediate action is necessary to protect the safety of children, young people and adults at risk and be aware of local/national policies and procedures. Where ongoing risk is recognised, the CF CNS will adhere to agreed multi-agency plans.
- Ensure the principles and duties of safeguarding adults and children are holistically, consistently and conscientiously applied to the wellbeing of all people with CF, families and carers.
- Maintain accurate records to document their involvement in safeguarding issues which are clear, accessible and comprehensive.
- Where appropriate, lead/co-ordinate safeguarding cases/framework.
- Support colleagues during safeguarding processes and be aware of your own wellbeing.

**Suggestions for audit:**

- Do staff have appropriate safeguarding training?
- Are safeguarding concerns documented in clinical records?
- Are staff aware of local support to protect their wellbeing during safeguarding processes?

**Note:** The following sections cover specific treatments, therapies and care. Unlike other complications and comorbidities, CFD is included in this guidance due to the additional care and education involved, often provided by the CF CNS.

# 18. Nebuliser therapy

People with CF may require nebulised medication on a short-term basis, during exacerbations, or on a long-term basis. People with CF receiving nebulised medication should be provided with education to understand the importance of the therapy, on safe and effective administration and how to care for and store equipment.

**The CF CNS should:**

- Together with the person with CF, their family/carers, and/or physiotherapist, assess and formulate an appropriate plan of action to ensure safe practice.
- Provide appropriate nebuliser and compressor delivery system and ancillaries (this may also be done by the physiotherapist).
- Provide contact names and numbers for advice and problems associated with nebuliser and compressor systems.
- Advise on frequency and method for maintaining compressor and ancillaries.

**Suggestions areas for audit:**

- Is there a documented test dose assessment?
- Are there arrangements to assess the knowledge of people with CF, for example does the person with CF and/or their family/carers know the frequency of nebulised medication and how this fits into their current medication routine?

# 19. Central venous access devices

People with CF often require central venous access devices (CVAD) for administering IV antibiotics. CVAD care should be provided by suitably trained team members to ensure safety and best practice. CVAD complications should be addressed, and documented appropriately, with referral to other services where appropriate.

**The CF CNS should:**

- Support and advise people with CF during the CVAD decision-making process.
- Facilitate and support referral to appropriate services for CVAD insertion.
- Support and advise people with CF on ongoing CVAD management and care.
- Teach people with CF, families, carers and relevant staff about CVAD management and care.
- Complete ongoing assessments of CVAD care.
- Provide information to people with CF on maintenance requirements and who to contact should complications arise.

**Suggestions for audit:**

- Is there a CF CNS available with experience of CVAD therapy?
- Is there a training and assessment process available to staff for caring for CVAD?
- Can staff state complications of CVAD and problem solve?
- Is there a record of all those trained to access and manage CVAD?
- Can the person with CF and/or their family/carers state who to contact for advice with regard to the CVAD?
- Can the person with CF and/or their family/carers state complications of CVAD?

# 20. Self-administration of intravenous therapy

People with CF may require regular IV antibiotics to help prevent or treat exacerbations. People with CF and/or families/carers should be provided with appropriate education to safely and competently administer prescribed IV medication within their homes. The rationale for implementing home IV therapy should be discussed with the person with CF and their family/carers, using a risk assessment to determine their suitability.

> **"Nurses are great with self-administration of home IVs. Letting me do my IVs at home helps a lot, and dropping in on their way to work to do blood levels makes life a lot easier."**
>
> Molly, who has CF

**The CF CNS should:**

- Together with the person with CF and their family/carers, formulate an appropriate plan of action to ensure safe practice in the administration of home IV antibiotics.
- Provide adequate teaching and supervision to ensure individual needs are met and assessed according to set criteria.
- Provide appropriate administration guidelines, medication and equipment.
- Provide contact details for 24-hour emergency advice and questions.
- Provide information and training on the possibility and recognition of anaphylaxis.
- Assess ongoing need for IV therapy and competence of the person with CF or their family/carers.

**Suggestions for audit:**

- Is there evidence of formal education for people with CF?
- Are there arrangements for assessing the knowledge of people with CF and their family/carers, such as if they know who to contact in an emergency?

# 21. Enteral tube feeding

People with CF may require enteral feeding if nutritional status is deteriorating or failing to improve. People with CF and their family/carers should be provided with education and training to enable them to manage their enteral tube and administer their feed safely, ensuring minimal disruption to their lifestyle.

**The CF CNS should:**

- Together with the CF dietitian, assess the person with CF and/or their family/carers for ability to manage and administer feeds.
- Provide an appropriate education plan, including the care of enteral feeding tubes and the safe administration of feeds.
- Monitor the person with CF for any complications associated with enteral feeding and tube care.
- Support the provision of equipment, including instructions of sourcing ongoing equipment.
- Provide appropriate contact information should any queries arise.

**Suggestions for audit:**

- Is there a documented care plan and competency assessment?
- Is there evidence of formal education for people with CF?

# 22. Oxygen therapy

People with CF may require oxygen on a short-term basis, during exacerbations, or long-term. People with CF receiving oxygen should be provided with education on the safe and effective administration and storage of oxygen. The person with CF will be assessed to determine flow rate and hours of usage (arterial blood gases and oxygen saturation measurements). Assessment of suitability may require full MDT input.

**The CF CNS should:**

- Together with the person with CF and their family/carers, and/or appropriate MDT member, formulate an appropriate plan of action to ensure safe administration of oxygen.
- Co-ordinate oxygen care and delivery. Where available, collaboration with specialist oxygen teams/nurses is recommended.
- Provide education regarding flow rate and hours of usage.
- Provide contact details for emergency advice.
- Continually assess the need for oxygen therapy.

**Suggestions for audit:**

- Is there a documented assessment and oxygen care plan?
- Are there arrangements to assess the knowledge of the person with CF, such as do they know their correct flow rate and who to contact in an emergency?

# 23. Non-invasive ventilation

Non-invasive ventilation (NIV) refers to the mechanical augmentation of minute ventilation.

It is a flexible form of ventilation, which can be used continuously, at night, or intermittently for specific treatments in the day, as indicated by an individual's clinical status. Historically, NIV was first used for people with CF to treat and manage severe life-threatening respiratory failure as a bridge to lung transplantation. The outcome was that oxygenation was improved without worsening hypercapnia and symptoms related to hypercapnia were alleviated.

Increasingly, NIV is used beyond this specific indication to support ventilation in people with CF who are not necessarily listed for transplantation. NIV as an adjunct to airway clearance has become recognised as a treatment option for people with CF and is considered in individuals with more severe lung disease, during infective exacerbations, or in people with CF having difficulty with expectoration of secretions.

This intervention is largely considered, implemented and monitored by CF physiotherapists, if expertise available. The role of NIV for airway clearance can allow the concept to be introduced for those that may go on to require nocturnal or intermittent ventilatory support.

NIV in an acute setting may be initiated by clinical staff depending on local practice. An introduction to NIV in this presentation may induce anxiety and psychological distress for the person with CF and this should be recognised. NIV can be difficult to tolerate, it may also indicate an acute or chronic change in condition for which the person with CF and their family/carers may need support and the opportunity for discussion.

**The CF CNS should:**

- Provide psychological support to the person with CF and their family/carers (although the CF CNS may not directly be involved with the introduction and monitoring of NIV and the equipment required).

**Suggestions for audit:**

- Does the CF CNS have knowledge of NIV?
- What NIV training has been provided to staff?

# 24. Cystic fibrosis diabetes care

People with CF may develop cystic fibrosis diabetes (CFD). People with CFD and their families/carers should be educated and empowered to manage CFD, including appropriate dietary advice, how to prevent associated complications, obtaining appropriate medicines and monitoring equipment.

**The CF CNS should:**

- Educate the person with CFD and their family/carers on CFD, including management of blood glucose levels and the prevention of long-term complications.
- Work collaboratively with the CF dietitian and diabetes specialist teams (where available).
- Support the provision of equipment, including instructions on sourcing ongoing equipment.
- Provide contact details for emergency advice.
- Work within **Cystic Fibrosis Trust's Management of cystic fibrosis diabetes guideline (2022)**.[15]

**Suggestions for audit:**

- Is there a documented CFD educational assessment and care plan?
- What CFD training/education has been provided to staff?
- Do staff feel confident with CFD care?

# 25. Resources and references

## References


1. Cystic Fibrosis Trust. Standards for the clinical care of children and adults with cystic fibrosis in the UK (3rd ed). 2024. Available at: **cysticfibrosis.org.uk/about-us/resources-for-cf-professionals/consensus-documents**

2. Health Education England. Multi-professional framework for advanced clinical practice in England. 2017. Available at: **hee.nhs.uk/sites/default/files/documents/multi-professionalframeworkforadvancedclinicalpracticeinengland.pdf**

3. Royal College of Nursing. **Standards for Advanced Level Nursing Practice**. 2018. Available at: **rcn.org.uk/Professional-Development/Advanced-Practice-Standards**

4. Royal College of Nursing (RCN). Non-medical prescribers. Available at: **rcn.org.uk/Get-Help/RCN-advice/non-medical-prescribers**

5. Cystic Fibrosis Trust. Clinical Trials. Available at: **cysticfibrosis.org.uk/research/clinical-trials**

6. Cystic Fibrosis Trust. Trials Tracker. Available at: **cysticfibrosis.org.uk/trialstracker**

7. Cystic Fibrosis Trust. Specialist cystic fibrosis care. Available at: **cysticfibrosis.org.uk/what-is-cystic-fibrosis/cystic-fibrosis-care/specialist-cystic-fibrosis-care**

8. Cystic Fibrosis Trust. CFSPID. Available at: **cysticfibrosis.org.uk/what-is-cystic-fibrosis/diagnosis/cfspid**

9. The Cystic Fibrosis Reproductive and Sexual Health Collaborative (CFReSHC). The CF Reproductive and Sexual Health Guide. Available at: **cfreshc.org/**

10. Sands D, Repetto T, Dupont LJ, Korzeniewska-Eksterowicz A, Catastini P, and Madge S. End of life care for patients with cystic fibrosis. J Cyst Fibros. 2011(10): Suppl 2:S37-44. Available at: **sciencedirect.com/science/article/pii/S1569199311600076**

11. Cathcart, F, Wood, J, and Madge, S. Improving end-of-life care for adults with cystic fibrosis: an improvement project. *BMJ Open Quality*, 2020 (9). Available at: **bmjopenquality.bmj.com/content/9/3/e000861.long**

12. Department of Health. Adult safeguarding: statement of government policy. 2011. Available at: **gov.uk/government/publications/adult-safeguarding-statement-of-government-policy**

13. Social Care Institute for Excellence (SCIE). What are the six principles of safeguarding? Available at: **scie.org.uk/safeguarding/adults/introduction/six-principles/**

14. Nursing and Midwifery council (NMC).The Code Professional standards of practice and behaviour for nurses, midwives and nursing associates. 2015. Available at: **nmc.org.uk/standards/code/**

15. Cystic Fibrosis Trust. Management of cystic fibrosis diabetes (2nd ed). 2022. Available at: **cysticfibrosis.org.uk/about-us/resources-for-cf-professionals/consensus-documents**

## Other useful information and resources


Age UK. Getting a good night's sleep. Available from: **ageuk.org.uk/information-advice/health-wellbeing/mind-body/getting-a-good-nights-sleep/**

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**

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**

Cystic Fibrosis Trust. Consensus documents. Available at: **cysticfibrosis.org.uk/about-us/resources-for-cf-professionals/consensus-documents**

Cystic Fibrosis Trust. Publications. Available at: **cysticfibrosis.org.uk/the-work-we-do/information-resources/publications**

Cystic Fibrosis Trust. UK CF Registry. Available at: **cysticfibrosis.org.uk/about-us/uk-cf-registry**

Edenborough FP, Borgo G, Knoop C, Lannefors L, Mackenzie WE, Madge S, Morton AM, Oxley HC, Touw DJ, Benham M, Johannesson M; and European Cystic Fibrosis Society. Guidelines for the management of pregnancy in women with cystic fibrosis. J Cyst Fibros. 2008; 7 Suppl 1:S2-32. Available at: **cysticfibrosisjournal.com/article/S1569-1993(07)00129-4/fulltext**

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**

Fagan C, Sharpe C  Sewell A, and Field J. Electronic prescribing: A service improvement project.  J Cyst Fibros. 2023(22): S177-S177. Available at: **cysticfibrosisjournal.com/article/S1569-1993(23)00739-7/abstract**

Manchester Foundation Trust (MFT) Research and Development Directorate. **researchdirectorate.org.uk**

Medical Research Council (MRC). **mrc.ukri.org**

Mental Health Foundation. How to sleep better. Available from: **mentalhealth.org.uk/explore-mental-health/publications/how-sleep-better**

National Health Service. Alcohol units. Available at: **nhs.uk/live-well/alcohol-advice/calculating-alcohol-units/**

National Health Service. Benefits of exercise. Available at: **nhs.uk/live-well/exercise/exercise-health-benefits/**

National Health Service. Sleep problems. Available at: **nhs.uk/every-mind-matters/mental-health-issues/sleep/**

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

National Institute for Health and Care Excellence (NICE). Transition from children's to adults' services for young people using health or social care services [NG43]. 2016. Available at: **nice.org.uk/guidance/ng43**

National Institute for Health Research (NIHR). **nihr.ac.uk**

National Institute for Health Research (NIHR). Good Clinical Practice (GCP). Available at: nihr.ac.uk/career-development/clinical-research-courses-and-support/good-clinical-practice

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

Suntharalingam J, Wilkinson T, Annandale J, et al. British Thoracic Society quality standards for home oxygen use in adults. BMJ Open Resp Res. 2017;4:e000223. Available at: brit-thoracic.org.uk/quality-improvement/quality-standards/home-oxygen/

Witt J, Lewis S, McKirdy F, Brownlee K, and Clarke S. Challenges and innovations in cystic fibrosis clinical teams: insights from the Cystic Fibrosis Trust staffing tool. J Cyst Fibros. 2024(23): S125-S125.

World Medical Association (WMA). Declaration of Helsinki – Ethical principles for medical research involving human subjects. 2013. Available at: wma.net/policies-post/wma-declaration-of-helsinki-ethical-principles-for-medical-research-involving-human-subjects/

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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