

# ECFS Standards for the care of people with cystic fibrosis (3<sup>rd</sup> Ed., 2023-2024)

## [A timely and accurate diagnosis](#)

doi: 10.1016/j.jcf.2023.09.008

### **1 Introduction**

### **2 Methods**

### **3 Standards of care for timely and accurate diagnosis**

#### 3.1 Diagnostic definitions and principles

#### 3.2 Screening approaches

#### 3.3 Diagnostic standards

##### 3.3.1 Sweat test standards

##### 3.3.2 Genetic testing standards

##### 3.3.3 Further electrophysiological measures

#### 3.4 Clarifying unclear situations

##### 3.4.1 CRMS/CFSPID designation; definition, evaluation, and management

##### 3.4.2 CFTR-related disorders; definition, evaluation, and management

#### 3.5 Carrier information

##### 3.5.1 Role of counselling

##### 3.5.2 Potential risks associated with carrier status

### **4 Conclusion**

## [Establishing and maintaining health](#)

doi: 10.1016/j.jcf.2023.12.002

### **1 Introduction**

### **2 Eating well**

#### 2.1 Infant feeding

2.2 Supporting good eating; content and behaviours

2.3 Pancreatic enzyme replacement therapy

2.4 Monitoring nutritional progress

### **3 Towards optimal lung health; staying ahead of the curve**

3.1 Physiotherapy for airway clearance

3.1.1 Working with the family from diagnosis through transition to adult life

3.1.2 Adapting airway clearance in older children and adults

3.1.3 Airway clearance approaches for the productive patient

3.1.4 The upper airways; problems and solutions

3.2 Clean air

3.2.1 Cigarette smoking and vaping

3.2.2 Hookah smoking

3.2.3 Passive smoking

### **4 Being active**

4.1 The rationale for physical activity and exercise

4.2 Approaches to measuring and monitoring exercise capability

4.3 Strategies to support and maintain an active lifestyle

### **5 Working with the CF team and other healthcare professionals**

5.1 Expectations and models of care

5.2 Potential for remote care

5.2.1 Remote monitoring

5.2.2 Virtual clinics

5.2.3 Ambulatory care

### **6 Managing medicines**

6.1 Role of the pharmacist in the CF team

6.2 Supporting adherence to therapies

6.3 Starting and stopping medicines

## **7 Variant-specific therapy (CFTR modulator therapy) to correct the underlying defect**

7.1 Progress since interim guidance (January 2023)

7.2 Monitoring for adverse events on CFTR modulator therapy

7.3 Adjusting the dose of CFTR modulator therapy after adverse reactions

7.4 Adjusting the dose of CFTR modulator therapy during pregnancy

## **8 Conclusion**

## **[Recognising and addressing CF health issues](#)**

[doi.org/10.1016/j.jcf.2024.01.005](https://doi.org/10.1016/j.jcf.2024.01.005)

### **1 Introduction**

### **2 Identifying and addressing airway infection and inflammation**

2.1 Monitoring for lung health decline

2.2 Inhaled mucoactive agents

2.3 Surveillance for airway infection

2.4 Approaches to antibiotic therapy

2.4.1 Prophylaxis

2.4.2 Eradication

2.4.3 Suppression

2.4.4 Recognising and treating nontuberculous mycobacterium lung disease

2.5 Addressing pulmonary exacerbations

2.6 Fungal diseases in the CF lung

### **3 Cystic fibrosis specific health issues**

3.1 CF-related diabetes

3.2 CF-related liver disease

3.3 CF bone disease

3.4 Stones and other renal issues

3.5 Intestinal obstruction

3.6 Early identification of cancer

#### **4 Supporting mental and emotional well being**

4.1 The unique risks that people with CF and their families face

4.2 Consider screening tools for mental health issues

4.3 The role of the psychologist

#### **5 When life gets challenging**

5.1 Supporting increased treatment demands

5.1.1 Supporting complex care needs and treatment burden

5.1.2 Maintaining nutritional status and optimising nutritional support

5.2 Supporting breathing

5.2.1 Detecting and treating respiratory failure

5.2.2 Delivering oxygen and non-invasive ventilatory (NIV) support

5.3 Solid organ transplantation

5.4 Planning for end of life

#### **6 Conclusion**

### **[Planning for a longer life](#)**

[doi.org/10.1016/j.jcf.2024.05.007](https://doi.org/10.1016/j.jcf.2024.05.007)

#### **1. Introduction**

1.1 Methods

#### **2. Navigating life**

2.1. Issues around school and further education

2.2. Transitioning to adult life

2.3. Sexual and reproductive health

2.4. Employment

2.5. Insurance options

2.6. Considering retirement

2.7. Respecting values and beliefs

### **3. Planning a family**

3.1. Fertility of people with CF

3.2. Preparing for pregnancy

3.3. Supporting pregnancy

3.4. Raising children and grandchildren

3.5. Variant-specific therapy (CFTR modulator therapy) during and after pregnancy

3.5.1. Impact of variant-specific therapy (VST) on fertility in people with CF

3.5.2. Advice to women with CF considering pregnancy

3.5.3. The potential implications of VST exposure *in utero*

### **4. Growing older with CF**

4.1. The changing CF demographic

4.2. Specific health issues arising in older age for people with CF

4.3. Maintaining health in older age with CF

4.4. Supporting older people with CF emotionally and practically

4.5. Pelvic health

4.6. Menopause

4.7. Posture and musculoskeletal health

4.7.1. MSK pain

4.7.2. Postural problems

4.7.3. CF-associated arthritis

### **5. How inequalities and inequities impact people with CF**

5.1. Addressing global inequities

5.2. CF health inequalities from poverty

### **6. CF and the changing planet**

6.1. How planetary health impacts people with CF

6.1.1. Air quality

6.1.2. Climate

6.2. How healthcare impacts the planet

6.2.1. Mitigation

6.2.2. Reducing admissions and waste

6.2.3. Healthcare teams as advocates

7. Engaging with CF research

7.1. Stakeholder engagement to direct research priorities

7.2. Facilitating involvement in research

7.3. Registries to drive quality improvement

**8. Conclusion**