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

Consensus statements

A multidisciplinary core committee created the outline and framework of the four papers making up the 3rd edition of the ECFS Standards for the care of people with CF.

The core committee identified and invited authors to write certain sections of each paper, according to their expertise. The paper sections provide background and context, and were written using an evidence-based hierarchy and referencing existing ECFS guidance.

For each section, authors were asked to provide two or three statements to highlight key messages, to act as a focus for policymakers. These statements were reviewed using a modified Delphi methodology. Participants from a range of backgrounds were asked to review the statements and state if they agreed or disagreed (yes/no/cannot answer). If they disagreed, they were asked to explain why and provide an alternative version. Consensus was achieved when ≥80% of contributors agreed with a statement (participants who ticked "cannot answer" were not included in the calculation, as stakeholders previously commented that a "cannot answer" response was not the same as a "disagree").

For each paper, all statements reached consensus in the first round of consultation. Click the title of each paper to find more details about the statements and the Delphi process to reach agreement.

A timely and accurate diagnosis

doi: 10.1016/j.jcf.2023.09.008

#	Statement
1	Individuals presenting with a positive newborn screen, clinical features consistent with CF, or a positive family history (affected sibling), require efficient and accurate diagnostic confirmation.
2	A sweat chloride concentration ≥ 60 mmol/L and/or two CF-causing variants in trans confirm a CF diagnosis.
3	Results supporting a diagnosis of CF should be promptly reported back to the patient and/or their parents/carers, with early CF follow-up arranged. Clear information about the disease and its management should be provided, and genetic counselling offered.
4	A newborn bloodspot screening (NBS) programme for CF should be designed to best address the geography, social health circumstance and ethnicity of the population in that region.
5	The responsible parties for NBS programmes should annually monitor and report their programme's performance using the ECFS-defined key outcome parameters to achieve the ECFS standards (at a minimum).
6	The sweat test remains the diagnostic gold standard for CF and should be performed according to the ECFS standards.
7	In people with CF, the *CFTR* genotype should always be investigated to determine whether variant-specific therapy may be indicated.

#	Statement
8	If the criteria for CF are not met, and clinical consideration of a diagnosis remains, further CFTR functional tests in a specialist diagnostic hub are required.
9	Infants with a CRMS/CFSPID designation should be evaluated and managed according to ECFS standards [4].
10	Individuals with CFTR-RD should be evaluated and managed according to revised ECFS standards.
11	CF carrier testing and screening should only aim to identify CF-causing variants.
12	Raising concerns about future health risks for *CFTR* carriers may be premature until more consistent data are available.

Establishing and maintaining health

doi: 10.1016/j.jcf.2023.12.002

#	Statement
1	Whenever breast feeding is possible, it should be encouraged and supported for infants with CF.
2	Infants with CF presenting with meconium ileus are at risk of both short and long-term nutritional deficits and require early support from the CF team.
3	Support from a specialist CF dietitian is essential.
4	The CF team should encourage healthy feeding behaviours early in life to promote a good relationship with food and a positive body image.
5	Pancreatic enzyme replacement therapy should be initiated if there is clinical evidence of pancreatic insufficiency.
6	Nutritional status should be monitored at each clinic visit.
7	For people on CFTR modulator therapy, special consideration should be given to the need for salt and vitamin supplementation.
8	Physiotherapy advice for airway clearance, including physical activity and exercise, should begin at diagnosis.
9	Physiotherapy for airway clearance should be individualised and provide a framework for people with CF to self-manage.
10	Adolescents should be supported to take increasing responsibility for airway clearance techniques, in preparation for independent adult life.
11	The CF team should regularly evaluate people with CF for rhino-sinus disease.
12	People with CF should avoid tobacco smoke (direct and environmental).
13	People with CF should avoid e-cigarette use (vaping).
14	Regular standardised exercise testing (as per the guidance of the ECFS Exercise Working Group and PhySIIG) should guide the advice and support given by the CF team.
15	CF teams should support people with cystic fibrosis to be physically active and exercise regularly.
16	Access to a multidisciplinary team with CF expertise and to closely associated specialties remains a key requirement for all people with CF.
17	The CF centre should adapt to reflect the improved life expectancy of people with CF.
18	People with CF should be educated by the CF multidisciplinary team and supported (including with telehealth) to help them best manage their health.

#	Statement
19	Remote care provides an opportunity for monitoring and interventions without hospital visits, but further research is needed to determine optimal strategies.
20	Virtual clinics and homecare offer an alternative to traditional structures but should not replace all face-to-face clinic reviews.
21	As new therapies emerge, the role of the CF pharmacist is increasingly important to optimise drug delivery and management.
22	A variety of approaches are available to monitor adherence to therapies and these should be used in an open manner to support people with CF and their families.
23	CF teams should work in partnership with people with CF and parent/caregivers to support adherence to therapies.
24	Starting and stopping therapies should be guided by the best evidence available and decided in partnership with the person with CF.
25	People with CF with eligible *CFTR* gene variants should be offered CFTR modulator therapy.
26	For young children and infants, certain CFTR modulator therapies may not yet be licensed, and options should be considered on an individual basis.
27	When initiating CFTR modulator therapy, people with CF and families should be encouraged to promptly report any significant physical or mental health changes to the CF team.
28	All adverse events experienced on CFTR modulator therapy should be reported to a post market surveillance scheme and the pharmaceutical company.

Recognising and addressing CF health issues

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#	Statement
1	Respiratory function testing and lung imaging should be regularly performed, as per previous guidance [2].
2	Inhaled mucoactive therapies have an important role in the respiratory management of people with CF.
3	CF teams should work in partnership with people with CF and their families to determine the most appropriate regimen for inhaled mucoactive therapies.
4	Airways should be sampled for infection at each clinic visit and with each respiratory exacerbation (expectorated or induced sputum sample preferred; otherwise, oropharyngeal sample).
5	Induced sputum should be performed at least once a year in people with CF who cannot expectorate sputum even if they are asymptomatic.
6	Lower airway sampling via bronchoalveolar lavage should be considered in people with CF who have persistent symptoms despite appropriate culture-directed therapy from expectorated or induced sputum.
7	There is good evidence to support eradication protocols for *Pseudomonas aeruginosa* identified on respiratory culture of any airway sample.
8	A proactive approach to managing CF pathogens other than *Pseudomonas aeruginosa* is reasonable with antibiotic choice determined by local protocols, patient tolerability, and adverse effects.

#	**Statement**
9	The CF team should work closely with the microbiology department to ensure targeted therapy, good antibiotic husbandry and appropriate cross infection strategies.
10	A respiratory sample should be obtained at least annually (sputum, induced sputum or bronchoalveolar lavage) for non-tuberculous mycobacteria detection.
11	A diagnosis of allergic bronchopulmonary aspergillosis (ABPA) should be considered for people with CF who are symptomatic and not responding to antibiotic therapy.
12	Formal annual screening for glucose intolerance should commence from ten years of age for people with CF, although a low threshold for screening should be practised if there is clinical concern.
13	Management of CF diabetes requires multi-disciplinary care, including diabetes specialists, and support for the person with CF through the significant psychological impact of this diagnosis.
14	Therapy for CF diabetes is insulin based and should aim for usual standards of glycaemic control, but not at the expense of high-nutrient dietary support
15	Routine assessment for liver disease is recommended in people with CF, including annual blood tests and regular ultrasonography.
16	Early referral to a gastroenterologist or hepatologist with CF expertise should be initiated for persistent abnormal scan findings, evidence of portal hypertension or persistent transaminitis.
17	Double X-ray absorptiometry (DXA) should be performed on children and adults with CF, who are at risk of low mineral density, for example: low BMI, low FEV_1, history of steroid therapy, history of hypogonadism.
18	Bisphosphonates should be considered for people with CF with significant osteoporosis on DXA scan despite standard therapy (adequate nutrition, physical activity, and calcium/vitamin D supplementation).
19	CF teams should take active measures to minimise the risk for people with CF of renal compromise or stones, including routinely assessing for acute kidney injury and chronic kidney disease, modifying potentially nephrotoxic treatments, and ensuring adequate hydration.
20	Constipation and DIOS are common co morbidities and CF physicians should routinely assess if people with CF are experiencing symptoms suggestive of these conditions.
21	Screening for colorectal cancer in people with CF should commence at an earlier age than the general population.
22	Colonoscopy is the most suitable screening method, other less invasive screening tests (faecal immunochemical testing and CT colonoscopy) need further evaluation.
23	The CF psychologist promotes mental health and quality of life through education, prevention, screening and intervention; helping to facilitate a more holistic approach of treatment for people with CF and their families.
24	People with more advanced CF disease should receive holistic individualised care aimed at balancing quality of life, treatment burden and clinical outcomes.
25	People with advanced CF disease must be assessed regularly for respiratory failure using a combination of clinical evaluation, pulse oximetry and blood gas analysis (including nocturnal and ambulatory assessments).
26	To improve symptoms and quality of life for those with advanced CF lung disease, supplemental oxygen should be offered to people with CF with chronic hypoxaemia and nocturnal non-invasive ventilatory support offered with evidence of chronic hypercarbia.
27	In acute or decompensated chronic respiratory failure, non-invasive ventilatory support should be considered, with appropriate plans for escalation made including consideration of invasive ventilation if appropriate.

#	Statement
28	Lung and/or liver transplant should remain available for people with end-stage CF lung or liver disease.
29	CF teams and transplant centres should collaborate to establish optimal transplant assessment and post-transplant care.
30	For people with CF with advanced disease, planning for end of life care is important.
31	Clear lines of communication are important between the CF and palliative care teams to minimise anxiety and stress for people with CF and their families.

Planning for a longer life

doi.org/10.1016/j.jcf.2024.05.007

#	Statement
1	The CF team should support people with CF through their life journey, recognising key stress points such as changing school, gaining employment and living independently.
2	The CF team should acknowledge and respect the differing values and beliefs of people with CF.
3	The CF team should work with the family to introduce the concept of subfertility to young people with CF in a sensitive manner.
4	The CF team should provide clear, age-appropriate information about assisted reproductive technologies.
5	For women with CF who wish to conceive, the CF team should promote optimisation of pre-conceptional health including lung function, nutritional and metabolic status.
6	Pregnant women with CF should undergo regular monitoring by the CF and obstetric teams including screening for gestational diabetes and re-assessment of chronic supportive care.
7	The decision to continue or discontinue CFTR modulator therapy during and after pregnancy should be made in partnership between the CF team and the person with CF.
8	More research and high-quality evidence are needed to characterise maternal and foetal outcomes following CFTR modulator therapy use during pregnancy and breastfeeding.
9	The CF team should work in close partnership with primary care to ensure that people with CF have appropriate support and screening as they grow older.
10	CF teams should be aware of issues of incontinence (even from an early age) and provide support appropriately.
11	The CF team should evaluate for musculoskeletal problems and postural changes to facilitate early and appropriate management.
12	The CF community should work towards minimising global inequities, through collaboration, agreed standards of care and improved access to therapies.
13	The CF team should be aware of and take measures to tackle the inequalities in health outcomes experienced by people with CF from a less well-resourced backgrounds.

#	**Statement**
14	The CF community should endeavour to act on a micro and macro scale to minimise the impact of providing complex healthcare on the planet, without compromising quality of care.
15	Access to research participation should be equitable.
16	People with CF, from all backgrounds and ages, should be involved in the prioritisation and design of clinical studies, from an early stage.
17	The role of clinical trial networks is to facilitate the delivery of a wide portfolio of commercially sponsored and investigator-led studies, across multiple sites and/or countries.
18	CF registries are instrumental to guide policy and funding, improve quality of care and facilitate research for the benefit of the CF community.