National Institute for Health and Care Excellence

Version 1.0 Pre-consultation

Cystic Fibrosis: diagnosis and management

Review questions

NICE Guideline

Methods, evidence and recommendations

9 March 2017

Draft for Consultation

Commissioned by the National Institute for Health and Care Excellence

Disclaimer

The recommendations in this guideline represent the view of NICE, arrived at after careful consideration of the evidence available. When exercising their judgement, professionals are expected to take this guideline fully into account, alongside the individual needs, preferences and values of their patients or service users. The recommendations in this guideline are not mandatory and the guideline does not override the responsibility of healthcare professionals to make decisions appropriate to the circumstances of the individual patient, in consultation with the patient and/or their carer or guardian.

Local commissioners and/or providers have a responsibility to enable the guideline to be applied when individual health professionals and their patients or service users wish to use it. They should do so in the context of local and national priorities for funding and developing services, and in light of their duties to have due regard to the need to eliminate unlawful discrimination, to advance equality of opportunity and to reduce health inequalities. Nothing in this guideline should be interpreted in a way that would be inconsistent with compliance with those duties.

NICE guidelines cover health and care in England. Decisions on how they apply in other UK countries are made by ministers in the Welsh Government, Scottish Government, and Northern Ireland Executive. All NICE guidance is subject to regular review and may be updated or withdrawn.

Contents

1 Review questions

No.	Number in the workplan	Review question
1	23	In infants, children, young people and adults (including those that have undergone newborn screening) when should cystic fibrosis be suspected?
2	28	What information and support should be given to children, young people and adults with cystic fibrosis?
3	16	What is the effectiveness of different models of care (for example, specialist centre, shared care [delivered by a Network CF Clinic which is part of an agreed designated network with a Specialist CF Centre], community, telehealth and/or home care for people with CF?
4	15	What is the clinical and cost-effectiveness of multidisciplinary teams of various compositions?
5	27	What parts of the transition from children's to adult services are most important for young people with cystic fibrosis and their family members and carers?
6	29	What are the non-lower-respiratory complications of cystic fibrosis in infants, children, young people and adults?
7	22	What is the effectiveness of programmes of exercise in the management of cystic fibrosis?
8	26	What is the clinical and cost effectiveness of nutritional interventions in people with cystic fibrosis?
9	2	In people with cystic fibrosis, what is the most effective regimen of enzyme replacement therapy in the treatment of exocrine pancreatic insufficiency?
10	3	What are the effective strategies for treatment and secondary prevention of distal ileal obstruction syndrome?
11	4	What is the diagnostic accuracy of tests to detect or strategies to detect early and late CF liver disease?
12	5	What is the diagnostic and prognostic value of different strategies to detect CF liver disease and predict progression (including progression to cirrhosis and portal hypertension with (out) oesophageal varices)?
13	1	What is the effectiveness of ursodeoxycholic acid for preventing the development or progression of liver disease in people with cystic fibrosis?
14	24	What is the most effective strategy to monitor for the onset of CF-related diabetes (CFRD)?
15	25	What is the most effective strategy to monitor for the identification of reduced bone mineral density in people with CF?
16	21	What strategies are effective at identifying people with cystic fibrosis for the presence of a psychological and/or behavioural problem?
17	7	What is the value of the following investigative strategies in monitoring the onset of pulmonary disease in people with CF without clinical signs or symptoms of lung disease? • Non-invasive microbiological investigation- induced sputum samples, cough swab, throat swab, and nasopharangeal aspiration • Invasive microbiological investigation- broncheoalveolar lavage • Lung physiological function tests- Cardiopulmonary exercise testing, Spirometry and Lung Clearance Index • Imaging techniques- Chest x-ray and CT scan
18	8	What is the value of the following investigative strategies in monitoring evolving pulmonary disease in people with established lung disease? • Non-invasive microbiological investigation- induced sputum samples, cough

No.	Number in the workplan	Review question
		swab, throat swab, and nasopharangeal aspiration • Invasive microbiological investigation- broncheoalveolar lavage • Lung physiological function tests- Cardiopulmonary exercise testing, Spirometry and Lung Clearance Index • Imaging techniques- Chest x-ray and CT scan • Evolving pulmonary disease defined as decline in lung function (based on FEV1), increased exacerbations and/or infections, (symptom based?) and CT changes.
19	9	What is the added value of imaging and invasive microbiological testing in addition to non-invasive microbiological testing and lung function tests in monitoring the response to treatment following an acute exacerbation? Definition of stablished lung disease: clinical signs and symptoms and/or radiological signs of lung disease.
20	6	What is the effectiveness of airway clearance techniques in people with cystic fibrosis?
21	11	What is the effectiveness of mucoactive or mucolytic agents, including dornase alpha, nebulised sodium chloride (isotonic and hypertonic) and mannitol?
22	14	What is the effectiveness of long-term antimicrobial prophylaxis to prevent pulmonary bacterial colonisation with Staphylococcus aureus in people with CF?
23	12	What is the effectiveness of antimicrobial treatment for acute pulmonary infection or those with an exacerbation in children and adults with cystic fibrosis?
24	13	What is the effectiveness of antimicrobial regimens in suppressing chronic pulmonary infection in children and adults with CF with any of the following pathogens: • Pseudomonas aeruginosa • Burkholderia cepacia complex • Staphylococcus aureus • Aspergillus fumigatus
25	10	What is the effectiveness of immunomodulatory agents in the management of lung disease?
26	17	What is the effectiveness of cohorting on the basis of pathogen status versus not cohorting on the basis of pathogen status in reducing transmission of CF pathogens?
27	18	What is the effectiveness of different models of segregating patient's in reducing transmission of CF pathogens?
28	19	What is the effectiveness of individual protective equipment in reducing transmission of CF pathogens?
29	20	What is the effectiveness of the combination of cohorting, segregating and protective equipment in reducing transmission of CF pathogens?