NICE National Institute for Health and Care Excellence

NICE Pathways

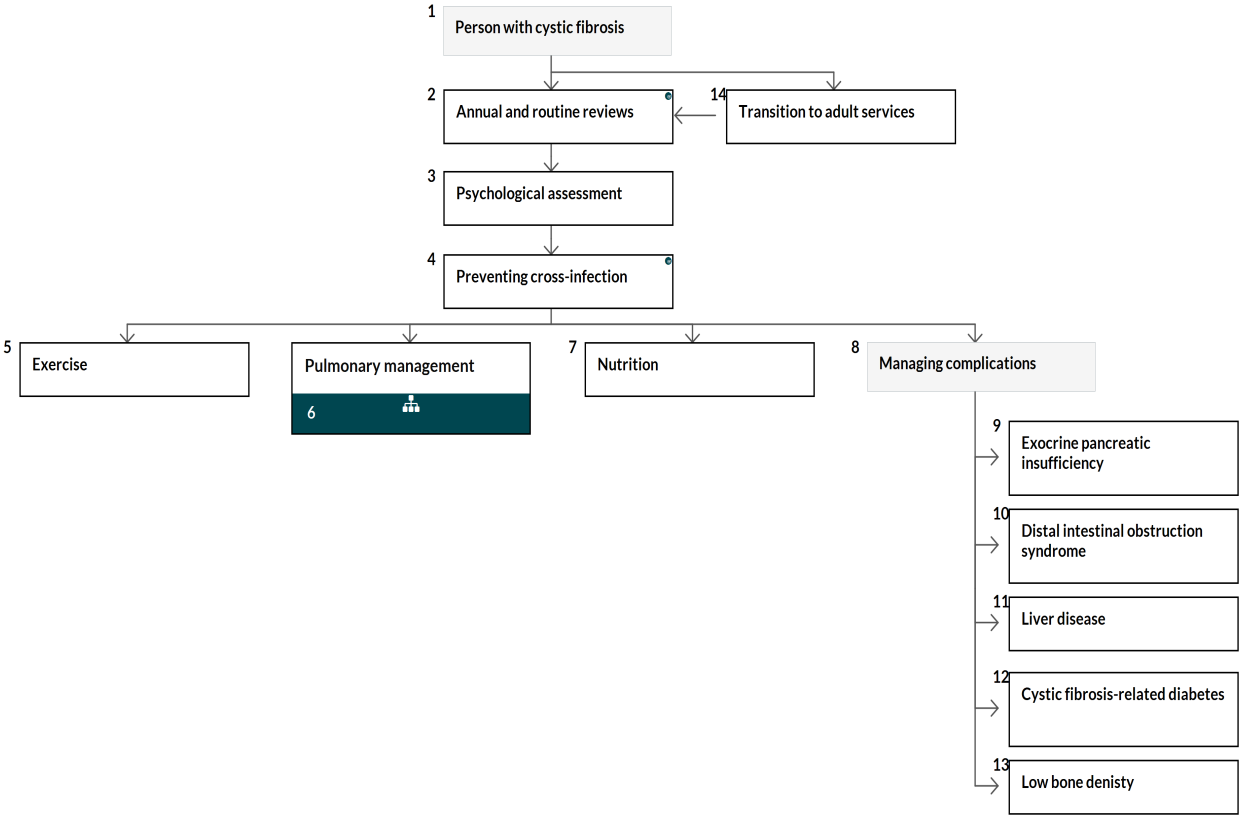
Managing cystic fibrosis

NICE Pathways bring together everything NICE says on a topic in an interactive flowchart. NICE Pathways are interactive and designed to be used online.

They are updated regularly as new NICE guidance is published. To view the latest version of this NICE Pathway see:

http://pathways.nice.org.uk/pathways/cystic-fibrosis
NICE Pathway last updated: 22 October 2021

This document contains a single flowchart and uses numbering to link the boxes to the associated recommendations.



1 Person with cystic fibrosis

No additional information

2 Annual and routine reviews

Be aware that:

- the aim of cystic fibrosis care is to prevent or limit symptoms and complications of the condition
- routine monitoring and annual assessments are crucial in providing effective care.

Annual review

Offer people with cystic fibrosis a comprehensive annual review that includes the following:

- a pulmonary assessment (see pulmonary monitoring)
- an assessment of nutrition and intestinal absorption
- an assessment for liver disease
- testing for cystic fibrosis-related diabetes, from 10 years of age
- an assessment for other potential or existing cystic fibrosis complications
- a psychological assessment
- assessments by a specialist nurse, physiotherapist, pharmacist and social worker (see multidisciplinary team)
- a review of their exercise programme.

Routine reviews

Provide regular routine reviews for people with cystic fibrosis, and do these more frequently immediately after diagnosis and in early life. For example:

- weekly in their first month of life
- every 4 weeks when they are between 1 and 12 months old
- every 6 or 8 weeks when they are between 1 and 5 years old
- every 8 or 12 weeks when they are over 5 years old
- every 3 or 6 months as adults.

Quality standards

The following quality statement is relevant to this part of the interactive flowchart.

1. Annual reviews

3 Psychological assessment

At the annual review, the specialist clinical psychologist should include assessments of:

- general mental health and wellbeing
- quality of life
- any factors that are making treatment adherence difficult
- indicators of emerging psychosocial problems
- behaviours that affect health outcomes.

If a severe mental health condition is identified at any assessment performed by the cystic fibrosis clinical psychologist, refer the person with cystic fibrosis to a mental health practitioner. For guidance on treating mental health conditions, refer to the relevant NICE guideline.

For family members or carers of people with cystic fibrosis, the specialist clinical psychologist should

- assess any cystic-fibrosis-related needs they have
- support their psychological wellbeing
- refer them to mental health practitioners as needed.

4 Preventing cross-infection

See the NICE Pathway on prevention and control of healthcare-associated infections.

To prevent cross-infection among people with cystic fibrosis in outpatient and inpatient care, use microbiological surveillance and a local infection control strategy that includes cohorting.

Inform people with cystic fibrosis, their family members or carers (as appropriate) and staff involved in their care about the risk of cross-infection and how to avoid it.

Each specialist cystic fibrosis clinic should be organised to prevent cross-infection. Separate

people individually during the clinic including by organising:

- the use of communal waiting areas
- attendance at diagnostic, treatment and pharmacy facilities.

Keep people with transmissible or chronic *Pseudomonas aeruginosa* or *Burkholderia cepacia* complex infection separate from people who do not have these infections, for example by using separate outpatient clinics.

Consider keeping people with cystic fibrosis who have intermittent isolation of *Pseudomonas aeruginosa* separate from people who do not have this infection, for example by using separate outpatient clinics. Help people with cystic fibrosis plan their inpatient attendance to avoid contact with each other, for example when they use:

- hospital restaurants, schools and recreation areas
- diagnostic, treatment and pharmacy facilities (see information and support).

During inpatient care, give people with cystic fibrosis individual rooms with en-suite facilities.

Quality standards

The following quality statement is relevant to this part of the interactive flowchart.

2. Preventing cross-infection during hospital admissions

5 Exercise

Advise people with cystic fibrosis and their family members or carers (as appropriate) that regular exercise improves both lung function and overall fitness.

Offer people with cystic fibrosis an individualised exercise programme, taking into account their capability and preferences.

Regularly review exercise programmes to monitor the person's progress and ensure that the programme continues to be appropriate for their needs.

Provide people with cystic fibrosis who are having inpatient care with:

- an assessment of their exercise capacity
- the facilities and support to continue their exercise programme (as appropriate), taking into

- account the need to prevent cross-infection and local infection control guidelines.

6 Pulmonary management

See Cystic fibrosis / Pulmonary management in cystic fibrosis

7 Nutrition

The cystic fibrosis specialist dietitian should offer advice on the benefits of optimal nutrition, and at the annual assessment review the person's:

- total nutritional intake, including energy intake (calories)
- estimated nutritional needs
- pancreatic enzyme replacement therapy, if appropriate.

Encourage people to increase calorie intake by increasing portion size and eating high-energy foods if there is concern about their nutrition (including weight loss and inadequate weight gain).

If increased portion size and high-energy foods are not effective, consider a trial of oral nutritional supplements.

If attempts to increase calorie intake are not effective, consider:

- supplementation with enteral tube feeding **or**
- for adults, a short-term trial of an appetite stimulant[1] (for example up to 3 months).

8 Managing complications

No additional information

9 Exocrine pancreatic insufficiency

Test for exocrine pancreatic insufficiency in people with cystic fibrosis, using a non-invasive technique such as stool elastase estimation. If the test result is normal, repeat if symptoms or signs suggesting malabsorption occur.

Offer oral pancreatic enzyme replacement therapy to people with exocrine pancreatic

[1] At the time of publication (October 2017), appetite stimulants did not have a UK marketing authorisation for use in people with cystic fibrosis for this indication. The prescriber should follow relevant professional guidance, taking full responsibility for the decision. Informed consent should be obtained and documented. See the General Medical Council's Prescribing guidance: prescribing unlicensed medicines for further information.

insufficiency. Adjust the dose as needed to minimise any symptoms or signs of malabsorption.

Consider an acid suppression agent[1] (for example an H2 receptor antagonist or a proton pump inhibitor) for people who have persistent symptoms or signs of malabsorption despite optimal pancreatic enzyme replacement therapy.

10 Distal intestinal obstruction syndrome

Be aware that a variety of conditions can cause acute abdominal pain and resemble distal intestinal obstruction syndrome in people with cystic fibrosis, for example:

- constipation
- appendicitis
- intussusception
- cholecystitis.

Suspect distal intestinal obstruction syndrome in people with cystic fibrosis who have an acute onset of peri-umbilical or right lower quadrant abdominal pain and any of the following:

- a palpable mass in the right lower quadrant
- faecal loading in the right lower quadrant on a plain abdominal X-ray, especially if associated with small intestine air-fluid levels
- clinical features of partial or complete intestinal obstruction, such as vomiting (especially bilious) and abdominal distension.

For people who have an acute onset of peri-umbilical abdominal pain but no other clinical or radiological features of distal intestinal obstruction syndrome, consider further imaging, for example with an:

- abdominal ultrasound scan **or**
- abdominal CT scan.

Manage suspected distal intestinal obstruction syndrome in a specialist cystic fibrosis centre, with supervision from specialists who have expertise in recognising and treating the condition and its complications.

Offer oral or intravenous fluids to ensure adequate hydration (and rehydration if needed) for people with distal intestinal obstruction syndrome.

Consider diatrizoate meglumine and diatrizoate sodium solution (Gastrografin) (orally or via an

[1] At the time of publication (October 2017), acid suppression agents did not have a UK marketing authorisation for use in people with cystic fibrosis for this indication. The prescriber should follow relevant professional guidance, taking full responsibility for the decision. Informed consent should be obtained and documented. See the General Medical Council's Prescribing guidance: prescribing unlicensed medicines for further information.

enteral tube) as first-line treatment for distal intestinal obstruction syndrome.

If diatrizoate meglumine and diatrizoate sodium solution (Gastrografin) is not effective, consider an iso-osmotic PEG solution (macrogols) (orally or via an enteral tube) as a second-line treatment.

Consider surgery as a last resort, if prolonged treatment with a PEG solution is not effective.

To reduce the risk of distal intestinal obstruction syndrome recurring:

- encourage people to drink plenty of fluids
- optimise pancreatic enzyme replacement therapy (see exocrine pancreatic insufficiency [See page 6])
- consider advising regular treatment with a stool-softening agent such as lactulose or a PEG solution.

11 Liver disease

Perform a clinical assessment and liver function blood tests at the annual review for people with cystic fibrosis.

If liver function blood tests are abnormal, perform a liver ultrasound scan and consider ursodeoxycholic acid[1] treatment.

Think about stopping ursodeoxycholic acid if liver function blood tests return to normal and clinical assessment and liver ultrasound scan show no liver disease.

If ursodeoxycholic acid is stopped, monitor for re-emergence of liver disease using clinical assessment and liver function blood tests.

Think about referring people with cystic fibrosis to a liver specialist if the liver function blood test results are persistently abnormal despite treatment with ursodeoxycholic acid.

Refer people with cystic fibrosis to a liver specialist if they have any of the following:

- chronic progressive liver disease, based on clinical assessment, liver function blood tests or the findings on a liver ultrasound scan
- liver failure, based on clinical assessment and liver function tests
- portal hypertension, haematemesis, splenomegaly or findings on a liver ultrasound scan.

[1] At the time of publication (October 2017), ursodeoxycholic acid did not have a UK marketing authorisation for

adults with cystic fibrosis for this indication. The prescriber should check individual brands for licensing in children and young people and follow relevant professional guidance, taking full responsibility for the decision. Informed consent should be obtained and documented. See the General Medical Council's Prescribing guidance: prescribing unlicensed medicines for further information.

12 Cystic fibrosis-related diabetes

Diagnose cystic-fibrosis-related diabetes using one of the following:

- CGM
- serial glucose testing over several days
- OGTT – if OGTT is abnormal perform CGM or serial glucose testing over several days to confirm the diagnosis.

Test for cystic-fibrosis-related diabetes in people with cystic fibrosis annually from 10 years of age.

Test for cystic-fibrosis-related diabetes at the end of the first and second trimesters of pregnancy, using CGM or OGTT.

Test for cystic-fibrosis-related diabetes in people with cystic fibrosis who are taking long-term systemic corticosteroids or receiving enteral tube feeding, using CGM or serial glucose monitoring.

Think about testing for cystic-fibrosis-related diabetes in people who still have any of the following despite optimised cystic fibrosis treatment:

- unexplained weight loss
- a deterioration in lung function as measured by spirometry
- increased frequency of pulmonary exacerbations
- excessive tiredness.

13 Low bone density

Consider DXA bone density scans for people with cystic fibrosis who have factors that put them at high risk of low bone mineral density, such as:

- frequent or long-term oral corticosteroid use
- frequent intravenous antibiotic use
- severe lung disease
- undernutrition
- previous low-impact fractures

- previous transplants
- post menopause.

Seek specialist advice for people with a bone mineral density standard deviation below -2.0 (Z score) or -2.5 (T score).

14 Transition to adult services

Begin discussing the transition process to adult services with young people with cystic fibrosis when they are 12 years old, and with their family members or carers (as appropriate).

All cystic fibrosis services should have a coordinated and documented pathway for transition from children's to adults' services that includes plans for managing all cystic-fibrosis-related aspects of care.

Ask people with cystic fibrosis and their family members or carers (as appropriate) for feedback on the quality of the transition service, taking account of the recommendations on supporting infrastructure in the NICE Pathway on transition from children's to adults' services.

For more guidance on managing transition from children's to adults' services, see the NICE Pathway on transition from children's to adults' services. In particular, see the recommendations on:

- transition planning for young people moving from children's to adults' services
- named worker
- overarching principles.

Glossary

CGM

continuous glucose monitoring

DXA

dual energy X-ray absorptiometry

OGTT

oral glucose tolerance testing

PEG

polyethylene glycol and electrolyte

Pulmonary exacerbations

(sudden or recent worsening of clinical symptoms or signs; frequently caused by an acute pulmonary infection)

Sources

Cystic fibrosis: diagnosis and management (2017) NICE guideline NG78

Your responsibility

Guidelines

The recommendations in this guideline represent the view of NICE, arrived at after careful consideration of the evidence available. When exercising their judgement, professionals and practitioners are expected to take this guideline fully into account, alongside the individual needs, preferences and values of their patients or the people using their service. It is not mandatory to apply the recommendations, and the guideline does not override the responsibility to make decisions appropriate to the circumstances of the individual, in consultation with them

and their families and carers or guardian.

Local commissioners and providers of healthcare have a responsibility to enable the guideline to be applied when individual professionals and people using services 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 complying with those duties.

Commissioners and providers have a responsibility to promote an environmentally sustainable health and care system and should assess and reduce the environmental impact of implementing NICE recommendations wherever possible.

Technology appraisals

The recommendations in this interactive flowchart represent the view of NICE, arrived at after careful consideration of the evidence available. When exercising their judgement, health professionals are expected to take these recommendations fully into account, alongside the individual needs, preferences and values of their patients. The application of the recommendations in this interactive flowchart is at the discretion of health professionals and their individual patients and do 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.

Commissioners and/or providers have a responsibility to provide the funding required to enable the recommendations to be applied when individual health professionals and their patients wish to use it, in accordance with the NHS Constitution. They should do so 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.

Commissioners and providers have a responsibility to promote an environmentally sustainable health and care system and should assess and reduce the environmental impact of implementing NICE recommendations wherever possible.

Medical technologies guidance, diagnostics guidance and interventional procedures guidance

The recommendations in this interactive flowchart represent the view of NICE, arrived at after careful consideration of the evidence available. When exercising their judgement, healthcare professionals are expected to take these recommendations fully into account. However, the interactive flowchart does not override the individual responsibility of healthcare professionals to make decisions appropriate to the circumstances of the individual patient, in consultation with the patient and/or guardian or carer.

Commissioners and/or providers have a responsibility to implement the recommendations, in their local context, in light of their duties to have due regard to the need to eliminate unlawful discrimination, advance equality of opportunity, and foster good relations. Nothing in this interactive flowchart should be interpreted in a way that would be inconsistent with compliance with those duties.

Commissioners and providers have a responsibility to promote an environmentally sustainable health and care system and should assess and reduce the environmental impact of implementing NICE recommendations wherever possible.