NICE National Institute for Health and Care Excellence

NICE Pathways

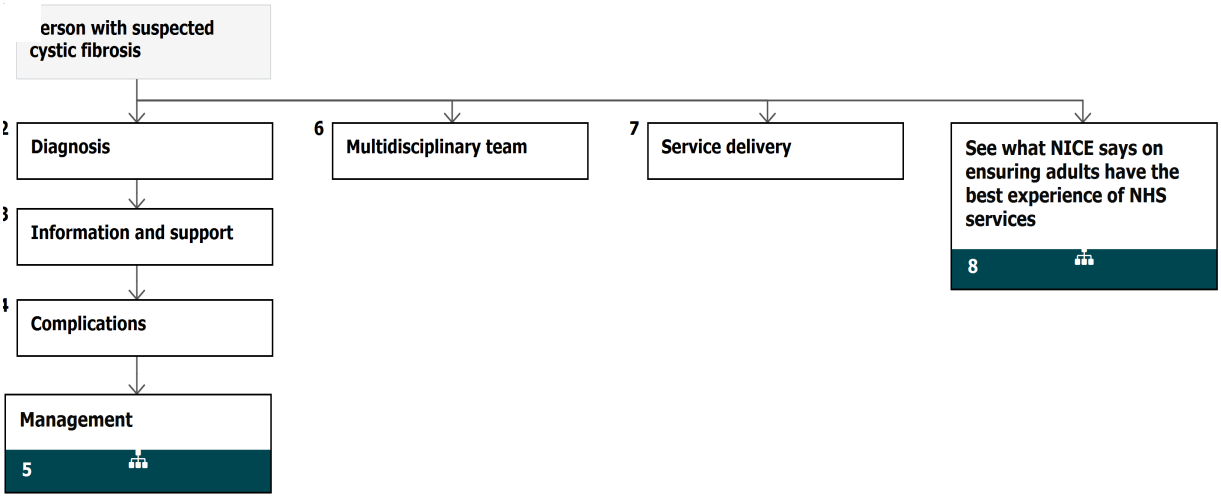
Cystic fibrosis overview

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: 06 September 2018

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



1 Person with suspected cystic fibrosis

No additional information

2 Diagnosis

Be aware that cystic fibrosis can be diagnosed based on:

- positive test results in people with no symptoms, for example infant screening (blood spot immunoreactive trypsin test) followed by sweat and gene tests for confirmation **or**
- clinical manifestations, supported by sweat or gene test results for confirmation **or**
- clinical manifestations alone, in the rare case of people with symptoms who have normal sweat or gene test results.

When and how to assess for cystic fibrosis

Assess for cystic fibrosis and, when clinically appropriate, perform a sweat test (for children and young people) or a cystic fibrosis gene test (for adults) in people with any of the following:

- family history
- congenital intestinal atresia
- meconium ileus
- symptoms and signs that suggest distal intestinal obstruction syndrome
- faltering growth (in infants and young children) (see NICE's recommendations on faltering growth)
- undernutrition
- recurrent and chronic pulmonary disease, such as:
    - recurrent lower respiratory tract infections
    - clinical or radiological evidence of lung disease (in particular bronchiectasis)
    - persistent chest X-ray changes
    - chronic wet or productive cough
- chronic sinus disease
- obstructive azoospermia (in people aged 12 and over)
- acute or chronic pancreatitis (see NICE's recommendations on pancreatitis)
- malabsorption
- rectal prolapse (in under 12s)
- pseudo-Bartter syndrome.

When to refer to a specialist centre

Refer people with suspected cystic fibrosis to a specialist cystic fibrosis centre if:

- they have a positive or equivocal sweat test result
- their assessment suggests they have cystic fibrosis but their test results are normal
- gene testing reveals 1 or more cystic fibrosis mutations.

3 Information and support

Provide people who are newly diagnosed with cystic fibrosis and their family members or carers (as appropriate) with opportunities to discuss their concerns.

Information and support should be provided by healthcare professionals with expertise in cystic fibrosis.

Provide people with suspected or diagnosed cystic fibrosis and their family members or carers (as appropriate) with relevant information that they can understand, and opportunities for discussion on topics such as:

- their diagnosis
- monitoring of their condition
- management choices for their condition
- possible or existing complications or comorbidities
- implications for living independently.

Provide people with cystic fibrosis and their family members or carers (as appropriate) with information about their care pathway.

Give information to people with cystic fibrosis and to family members or carers in ways that are individually appropriate. Avoid jargon and use formats that they prefer, for example:

- face-to-face discussions
- copies of correspondence
- written information (such as leaflets)
- any digital media and reliable internet sources that are available.

When appropriate, provide people with cystic fibrosis and their family members or carers with opportunities for discussion with relevant expert professionals on:

- available resources and support, such as local support and advocacy services
- managing the risks of cross-infection
- implications of the condition for school and education
- career planning
- transition to adult care
- foreign travel
- fertility and contraception
- pregnancy and parenting
- organ transplantation
- end of life care.

Provide people with cystic fibrosis with information about how to contact other people with cystic fibrosis without risking cross-infection (see preventing cross-infection), for example, by directing them to online support groups.

For more information on communication, providing information and shared decision-making in adult NHS services, see NICE's recommendations on patient experience.

Be aware that people with cystic fibrosis and their family members or carers will need emotional support and some may need specialist psychological support (see psychological assessment), in particular:

- at diagnosis
- at times of transition (for example, when starting or changing school, moving from education to work, or changing to living independently for the first time)
- in relation to fertility, including family planning, pregnancy and infertility
- to cope with complications of cystic fibrosis
- when waiting for or having organ transplantation
- when approaching the end of life.

See NICE's recommendations on end of life care for people with life-limiting conditions.

NICE has written information for the public on cystic fibrosis.

4 Complications

Be aware that people with cystic fibrosis are at risk of the following common complications:

- being underweight
- meconium ileus (affects 1 in 7 newborn babies)
- fat-soluble vitamin deficiencies (including vitamins A, D, E and K)
- distal intestinall obstruction syndrome
- muscle pains and arthralgia
- male infertility caused by obstructive azoospermia (almost all males with cystic fibrosis are infertile)
- reduced female fertility
- upper airway complications, including nasal polyps and sinusitis (prevalence increases with age)
- chronic liver disease (the prevalence increases with age until early adulthood)
- urinary stress incontinence
- cystic fibrosis-related diabetes (uncommon in children under 10 years, but the prevalence increases with age and it affects up to 1 in 2 adults)
- reduced bone mineral density (including osteoporosis).

Be aware that people with cystic fibrosis are at risk of the following less common complications:

- cystic fibrosis-related arthritis
- delayed puberty (associated with severe cystic fibrosis)
- renal calculi (incidence increases with age and 1 in 20 adults are affected).

5 Management

See Cystic fibrosis / Managing cystic fibrosis

6 Multidisciplinary team

The specialist cystic fibrosis multidisciplinary team should include at least one of each (depending on the size of the clinic) of the following professionals, who should have specialist expertise in the condition:

- specialist paediatricians or adult physicians
- specialist nurses
- specialist physiotherapists
- specialist dietitians
- specialist pharmacists

- specialist clinical psychologists.

The specialist cystic fibrosis multidisciplinary team should be led by a specialist paediatrician or adult physician.

The specialist cystic fibrosis multidisciplinary team should either include or have access to social workers.

Social workers should provide advice and support to people with cystic fibrosis and their family members or carers (as appropriate), for example on:

- help with adjusting to long-term treatment (such as taking regular medicines)
- education
- employment
- government benefits
- respite care.

Specialist nurses (working with specialist paediatricians or physicians) should coordinate care and facilitate communication between other members of the cystic fibrosis team, and act as advocates for people with cystic fibrosis and their family members or carers (as appropriate). Key clinical roles could include:

- support during and after diagnosis and when starting treatment
- triage
- advanced clinical assessment
- coordinating home intravenous antibiotic services, including intravenous access.

Specialist physiotherapists should assess and advise people with cystic fibrosis at clinic, at inpatient admissions, during pulmonary exacerbations and at their annual review on. Assessment and advice should cover airway clearance, nebuliser use, musculoskeletal disorders, exercise, physical activity and urinary incontinence.

Specialist dietitians should assess and advise people with cystic fibrosis about all aspects of nutrition at outpatient clinic visits, during inpatient admissions and at their annual review (see nutrition).

Specialist pharmacists should advise people with cystic fibrosis on medicines optimisation at outpatient clinic visits, during inpatient admissions, on discharge from hospital and at annual review. They should advise healthcare professionals on all aspects of medicines use and prescribing, and support GPs, community pharmacists and homecare providers to ensure that

people with cystic fibrosis get the medicines they need without interruption. See NICE's recommendations on medicines optimisation.

Specialist clinical psychologists should assess and advise people with cystic fibrosis and their family members or carers (as appropriate) at outpatient clinic visits and (if needed) at other outpatient appointments, during inpatient admissions, and at their annual review (see psychological assessment).

The specialist cystic fibrosis multidisciplinary team should either include or have access to specialist expertise relevant to cystic fibrosis in the following areas:

- microbiology
- pulmonary physiology
- diabetes
- gastroenterology
- hepatology
- rheumatology
- psychiatry
- interventional radiology
- surgery (gastrointestinal, thoracic, and ear, nose and throat)
- obstetrics
- palliative care.

The specialist cystic fibrosis multidisciplinary team should work with the person's GP, providing timely information so that the GP is able to support the person with cystic fibrosis by:

- prescribing cystic fibrosis medicines:
    - in batches of at least 1 month at a time for routine medicines
    - for longer periods if advised by the specialist team
    - following guidance on arrangements for prescriptions of unlicensed medicines
- providing routine annual immunisation, including any alterations for people with cystic fibrosis and flu vaccinations for family members and carers
- managing health problems not related to cystic fibrosis
- certification of illnesses
- working in partnership with cystic fibrosis homecare teams, particularly for end of life care
- providing care for the person's family members or carers.

See NICE's recommendations on end of life care for people with life-limiting conditions.

7 Service delivery

Care for people with cystic fibrosis should be provided by a specialist cystic fibrosis multidisciplinary team based at a specialist cystic fibrosis centre.

Specialist cystic fibrosis centres should:

- plan patient care (including outpatient and inpatient care), taking into account the risk of cross-infection (see preventing cross-infection)
- maintain local and national registers of patients that include information about their clinical condition, treatment and outcomes
- audit practice and outcomes.

When a shared-care model is used for children and young people, it should include:

- formal arrangements between the local paediatric team at the shared-care centre and multidisciplinary team at the specialist cystic fibrosis centre
- direct involvement of specialist cystic fibrosis multidisciplinary team members
- an annual assessment and at least one other review per year by the specialist cystic fibrosis multidisciplinary team, in addition to reviews by the local paediatric team (see annual and routine reviews).

If available and when clinically appropriate, outreach care for adults with cystic fibrosis may be provided by the specialist cystic fibrosis multidisciplinary team at a local hospital.

The specialist cystic fibrosis centre should have a point of contact available at all times (day or night) for urgent enquiries from people with cystic fibrosis and their family members or carers (as appropriate).

Consider telemedicine or home visits for routine monitoring where they are more appropriate than outpatient visits and if the person with cystic fibrosis prefers it.

Make arrangements (including providing equipment and expert support) for people to have intravenous antibiotic therapy at home, when this is appropriate.

8 See what NICE says on ensuring adults have the best experience of NHS services

See Patient experience in adult NHS services

Glossary

CGM

continuous glucose monitoring

CFTR

cystic fibrosis transmembrane conductance regulator

DPI

dry powder for inhalation

DXA

dual energy X-ray absorptiometry

FEF

forced expiratory flow

FEV1

forced expiratory volume in 1 second

FVC

forced vital capacity

MRSA

methicillin-resistant staphylococcus aureus

MSSA

methicillin-sensitive staphylococcus aureus

NPA

nasal pharyngeal aspirate

OGTT

oral glucose tolerance testing

Outreach care

(a model of care in which the specialist multidisciplinary cystic fibrosis team provide outpatient clinics in local hospitals)

PEG

polyethylene glycol and electrolyte

Immunomodulatory dose

(a dose of a drug that is less than the minimum inhibitory dose)

Pulmonary exacerbation

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

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

Shared-care model

(when a local hospital cares for people with cystic fibrosis, with oversight, support and direct involvement from members of a specialist cystic fibrosis multidisciplinary team)

Telemedicine

(providing clinical services remotely, using phone and video messaging to communicate with the patient)

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.