

COVID-19 rapid guideline: cystic fibrosis

NICE guideline

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Your responsibility

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.

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Overview

The purpose of this guideline is to maximise the safety of patients with cystic fibrosis and make the best use of NHS resources, while protecting staff from infection. It will also enable services to match capacity to patient needs if services become limited because of the COVID-19 pandemic.

This guideline focuses on what you need to stop or start doing during the pandemic. Follow the usual professional guidelines, standards and laws (including those on equalities, safeguarding, communication and mental capacity), as described in [making decisions using NICE guidelines](#).

This guideline is for:

- health and care practitioners
- health and care staff involved in planning and delivering services
- commissioners

The recommendations bring together

- existing national and international guidance and policies
- advice from specialists working in the NHS from across the UK. These include people with expertise and experience of treating patients with cystic fibrosis during the current COVID-19 pandemic.

NICE has developed these recommendations in direct response to the rapidly evolving situation and so could not follow the standard process for guidance development. The guideline has been developed using the [interim process and methods for developing rapid guidelines on COVID-19](#). The recommendations are based on evidence and expert opinion and have been verified as far as possible. We will review and update the recommendations as the knowledge base and expert experience develops.



1 Communicating with patients and minimising risk

- 1.1 Communicate with patients, their families and carers, and support their mental health and wellbeing to help alleviate any anxiety and fear they may have about COVID-19. Signpost to charities (such as the [Cystic Fibrosis Trust](#)), support groups (including NHS Volunteer Responders), and [UK government guidance on the mental health and wellbeing aspects of COVID-19](#).
- 1.2 Be aware that some patients, families or carers may need specialist psychological or social work support in the context of COVID-19.
- 1.3 Minimise face-to-face contact to reduce the risk of infection by:
- using telephone, video or email consultations whenever possible
 - cutting non-essential face-to-face appointments
 - contacting patients via text message, telephone or email
 - using electronic prescriptions rather than paper
 - providing home spirometry and, where appropriate, weighing scales
 - providing facilities to collect samples remotely
 - using different methods to deliver prescriptions and medicines to patients, for example, pharmacy deliveries, postal services, NHS Volunteer Responders, or introducing drive-through pick-up points for medicines.
- However, note that routine childhood vaccinations should continue to take place at the GP surgery.
- 1.4 Tell patients, their families and carers that they should contact their cystic fibrosis team if they think the patient may have COVID-19, to ensure that their symptoms are appropriately assessed. They should do this as soon as they have symptoms. They should also contact the [NHS 111 online coronavirus service](#) or call NHS 111. In an emergency they should call 999 if the patient is seriously ill.

- 1.5 Be aware that symptoms of COVID-19 and pulmonary disease exacerbations may be difficult to differentiate at initial presentation.

Patients not known to have COVID-19

- 1.6 Tell patients, their families and carers to refer to relevant parts of [UK government guidance on shielding and protecting people defined on medical grounds as extremely vulnerable from COVID-19](#), until the risks associated with the COVID-19 pandemic have passed. This includes:
- advice for other members of the household, noting the recommended shielding support for the patient and the need to follow relevant parts of government advice on social distancing (this differs across the UK) stringently. If this is not possible, for example, if they have a caring role for the patient, they may need to adopt shielding measures for themselves
 - advice for visitors who are providing essential care
 - advice on getting assistance with foods and medicines while shielding
 - support for mental wellbeing. [amended 2 June 2020]
- 1.7 For patients who still need to attend face-to-face appointments, ensure that existing arrangements to prevent cross-infection include COVID-19. See the [NICE clinical guideline on cystic fibrosis](#).
- 1.8 If patients need to attend face-to-face appointments, ask them to go alone if they can, or with no more than 1 family member or carer, to reduce the risk of contracting or spreading infection with COVID-19. They should avoid using public transport if possible.

Patients known or suspected to have COVID-19

- 1.9 If a patient has symptoms of COVID-19 on presentation or admission, follow [UK government guidance on investigation and initial clinical management of possible cases](#). This includes information on testing and isolating patients.
- 1.10 All healthcare workers involved in receiving, assessing and caring for patients who have known or suspected COVID-19 should follow [UK government guidance on infection prevention and control](#). This contains information on

using personal protective equipment (PPE), including visual and quick guides for putting on and taking off PPE.

- 1.11 Cystic fibrosis teams should report known or suspected cases of COVID-19 to the UK cystic fibrosis registry reporting hotline on the same day by emailing registry@cysticfibrosis.org.uk to request a call back.
- 1.12 Ensure that relevant members of the cystic fibrosis team are involved in decisions about the care of patients with suspected COVID-19, including escalation of treatment.
- 1.13 Patients with symptoms of COVID-19 should carry out airway clearance in a well-ventilated room, separate from other household members if possible, unless the patient needs assistance. This is because sputum generation is a potentially infectious aerosol generating procedure for COVID-19. Advise other family members not to enter the room until enough time has passed for aerosols to clear: follow [UK government guidance on infection prevention and control](#).
- 1.14 Explain to patients, their families and carers that when a nebuliser is used to administer an antibiotic, the aerosol comes from the fluid in the nebuliser chamber and will not carry virus particles from the patient. Tell families and carers to use appropriate hand hygiene when helping patients with their nebuliser mask. This is to prevent spread from a contaminated droplet that could be formed if the aerosol coalesces with a contaminated mucous membrane.

2 Treatment and care planning

- 2.1 Tell patients, their families and carers to continue with all their usual self-care arrangements including, for example:
- airway clearance techniques
 - prophylactic medication, including oral and inhaled antibiotics, and mucoactive agents
 - cystic fibrosis transmembrane conductance regulator (CFTR) therapies
 - diet, vitamins and pancreatic enzyme replacement therapy
 - home exercise (following [UK government guidance on shielding and protecting people defined on medical grounds as extremely vulnerable from COVID-19](#)).
- 2.2 Tell patients, their families and carers to wash their hands and clean equipment such as face masks and mouth pieces used for nebulised therapies, or face masks used for non-invasive ventilation, by regularly using washing-up liquid or following the manufacturer's cleaning instructions.
- 2.3 Prescribe usual quantities of medicines to meet the patient's clinical needs, normally 30 days' supply. Prescribing larger quantities of medicines puts the supply chain at risk.
- 2.4 Tell patients, their families and carers to follow the advice they have previously been given about what to do if they have an exacerbation, including taking rescue medication and contacting their cystic fibrosis team.
- 2.5 Monitor patients with cystic fibrosis closely, because they may be at greater risk of rapid deterioration if they contract COVID-19.

3 Modifications to usual care and service delivery

- 3.1 Think about how to modify usual care to reduce patient exposure to COVID-19 and make best use of resources (for example, workforce, facilities and equipment).
- 3.2 Specialist cystic fibrosis centres should maintain contact with network and outreach cystic fibrosis clinics using telephone or video consultations and meetings.
- 3.3 Specialist cystic fibrosis centres should maintain sufficient inpatient beds and facilities for those patients for whom a hospital admission is essential.
- 3.4 Day-case facilities may be necessary for procedures such as administering first doses of intravenous antibiotics for courses to be delivered at home and flushing totally implantable intravenous devices. Flushing frequency may be reduced from usual practice. If no alternative is possible, think about carrying out a home visit.
- 3.5 Ensure that sufficient clinical expertise and capacity remains within the cystic fibrosis team so that patients have the support they need, and their care can be safely managed out of hospital as much as possible.
- 3.6 When modifying individual patients' treatment plans:
 - take their clinical circumstances and preferences into account
 - involve all relevant members of the multidisciplinary team in the decision
 - record the reasoning behind each decision.
- 3.7 Discuss the risks and benefits of changing treatment plans with patients, their families and carers.
- 3.8 Only carry out lung function tests in hospital if the results will have a direct impact on patient care. Use home spirometry if possible.

- 3.9 Be aware that patients with cystic fibrosis can still access cystic fibrosis transmembrane conductance regulator (CFTR) therapies under the [NHS England policy statement for these drugs](#) (refer to devolved national policies if appropriate). Note that the assessments and data collection frequencies recommended in the [data collection agreement](#) are the ideal, and the minimum requirement is annual assessment and reporting. Conduct liver function testing and eye monitoring only when clinically essential.
- 3.10 Reduce the burden on frontline staff by following advice from NHS England and NHS Improvement about which data are not required (for example, the cystic fibrosis quality dashboard quarterly returns), and think about de-prioritising updating the cystic fibrosis registry (although note that centres will still need to meet the deadline of 31 January 2021).
- 3.11 Be aware that NHS England and NHS Improvement and NHS Blood and Transplant are working closely to enable limited access to transplants where this is feasible. However, given the severe capacity constraints because of managing the COVID-19 pandemic, the selection of patients from the waiting list for transplant will be considered on a case-by-case basis only. Referrals will only be seen in exceptional circumstances.
- 3.12 Defer the transition of young people to adult services until the pressures associated with the COVID-19 pandemic have passed.

4 Healthcare workers

- 4.1 Healthcare workers with known or suspected COVID-19, or who live in a household in which another person is known or suspected to have COVID-19, should self-isolate and only return to work in accordance with [UK government guidance on self-isolation for households with possible infection](#).
- 4.2 If a healthcare professional needs to self-isolate, ensure that they can continue to help if they are well enough to do so by:
- enabling telephone or video consultations and multidisciplinary team meetings
 - identifying patients who are suitable for remote monitoring and follow up, and those who are vulnerable and need support
 - carrying out tasks that can be done remotely, for example, updating the cystic fibrosis registry.
- 4.3 Cystic fibrosis teams should keep in touch with staff who are self-isolating, to support their mental wellbeing.
- 4.4 Provide all staff with visible leadership and supportive messaging, to maintain morale.
- 4.5 Take account of the information on the [NHS Employers website](#) about good partnership working and issues to consider when developing local plans to combat COVID-19.

Update information

Minor changes since publication

2 June 2020: We aligned recommendation 1.6 with current government advice on social distancing.

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