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 adults with interstitial lung disease, including idiopathic pulmonary fibrosis and pulmonary sarcoidosis, during the COVID-19 pandemic. It also aims to protect staff from infection and enable services to make the best use of NHS resources.

This guideline focuses on what you need to stop or start doing during the pandemic. Use it alongside your usual professional guidelines, standards and laws (including 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 adults with interstitial lung disease during the current COVID-19 pandemic.

We developed this guideline using the interim process and methods for developing rapid guidelines on COVID-19 in response to the rapidly evolving situation. We will review and update the recommendations as the knowledge base develops using the interim process and methods for guidelines developed in response to health and social care emergencies.
1 Communicating with patients and minimising risk

1.1 Communicate with patients, their families and carers, and support their mental wellbeing to help alleviate any anxiety they may have about COVID-19. Tell them they can get help and advice from charities (such as the British Lung Foundation, Action for Pulmonary Fibrosis, Pulmonary Fibrosis Trust and Sarcoidosis UK), support groups (including NHS Volunteer Responders) and UK government guidance on the mental health and wellbeing aspects of COVID-19.

1.2 Many patients with interstitial lung disease will have received a letter telling them they are at high risk of severe illness from COVID-19. Advise them or their carers:

- to follow the UK government guidance on shielding and protecting people defined on medical grounds as extremely vulnerable to COVID-19
- that their level of risk may change as a result of advice from their primary care team, specialists or changes in government guidance.

1.3 Discuss with patients who have been advised to shield whether the benefit of them attending services outweighs the risks.

1.4 Advise patients that if they think they have COVID-19 they should use the NHS 111 online coronavirus service (if in England), or call NHS 111. In an emergency they should call 999 if they are seriously ill. They should also talk to their hospital specialist team for advice about managing their symptoms and treatment.

1.5 Minimise face-to-face contact by:

- using telephone, video or email consultations whenever possible, following the NHS England clinical guide for the management of remote consultations and remote working in secondary care during the coronavirus pandemic
- cutting non-essential face-to-face appointments
• making multidisciplinary meetings virtual
• contacting patients via text message as well as telephone or email if patients are happy to do that
• using electronic prescriptions rather than paper
• using different methods to deliver prescriptions and medicines, for example, pharmacy deliveries, postal services, NHS Volunteer Responders or drive-through pick-up points for medicines.

1.6 Advise patients to keep a list of the medicines they take, and the conditions and any allergies they have, as well as a copy of a recent clinic letter, to give to healthcare staff if they need treatment for COVID-19.

1.7 Find out if people have advance care plans or advance decisions to refuse treatment, including do not attempt resuscitation decisions. Document this clearly and take account of this in planning care.

1.8 Consider asking local specialist palliative care teams for advice if needed.
2 Assessing symptoms of interstitial lung disease and COVID-19

2.1 Be aware that patients taking drugs that affect the immune response may have atypical presentations of COVID-19. For example, patients taking corticosteroids may not develop a fever.

2.2 Be aware that some adverse effects caused by antifibrotic drugs and immunosuppressants may be mistaken for symptoms of COVID-19, for example diarrhoea, fatigue and loss of appetite.

2.3 Be aware that symptoms of COVID-19 can be similar to the symptoms of the patient’s underlying condition.

2.4 Be aware that both interstitial lung disease and COVID-19 are associated with an increased risk of coagulopathy. Refer to local trust policies if available or see the British Thoracic Society’s guidance on venous thromboembolic disease in patients with COVID-19.
3 New referrals to interstitial lung disease specialist services

3.1 Hold the first outpatient appointment by telephone or video consultation. If that's not possible or suitable, offer a face-to-face appointment if the benefits outweigh the risks. If a patient has a face-to-face appointment, follow recommendations 5.1 to 5.4.

3.2 If blood test results from the last 6 weeks are available from the referral team or GP, and the patient's clinical condition has not altered considerably, use them to guide treatment decisions. If blood test results are not available and they are needed to guide urgent patient care, send the patient for blood tests (see recommendation 4.3).

3.3 If pulmonary function test results from the last 6 months are available from the referral team, and the patient's clinical condition has not altered considerably, use them to guide diagnosis and treatment decisions. If pulmonary function test results are not available and they are needed to guide urgent patient care, refer the patient for testing (see recommendation 4.1).

3.4 If a CT scan from the last 12 months is available from the referral team and the patient's clinical condition has not altered considerably, use it to guide diagnosis and treatment decisions. If a CT scan is not available and the results are needed to guide urgent patient care, refer the patient for a scan.
4 Interstitial lung disease investigations

4.1 Only carry out bronchoscopy and pulmonary function tests if the patient urgently needs them and if the results will have a direct impact on their care, because these tests have the potential to spread COVID-19. Plan investigations following NICE's guideline on arranging planned care in hospitals and diagnostic services or UK government guidance on infection prevention and control for COVID-19 as appropriate. [amended 6 November 2020]

4.2 If supervised spirometry is not possible, consider using home spirometry to get measurements needed for treatment decisions if the equipment and support are already available.

4.3 Consider alternatives to patients attending hospital for blood monitoring while on antifibrotic drugs or immunosuppressants, for example, community blood monitoring services.
5  Management: patients not known to have COVID-19

5.1 If patients have to attend a face-to-face appointment, on the day of the appointment first screen them by telephone and then again on arrival at the outpatient setting to make sure they have not developed symptoms of COVID-19.

5.2 Ask patients to help reduce the risk of contracting or spreading COVID-19 by:

- following the relevant parts of government advice on social distancing (this differs across the UK)
- coming to the appointment alone
- having only 1 person accompany them if they cannot come alone
- avoiding using public transport if possible.

5.3 Check patients' temperature when they arrive, ideally before they enter reception.

5.4 Minimise time in the waiting area by:

- careful scheduling
- encouraging patients not to arrive early
- texting or phoning patients when you are ready to see them, so that they can wait outside, for example, in their car
- providing a 'clean route' through the hospital to the department
- delivering clinical assessment and treatment promptly
- dispensing prescriptions rapidly.
Starting and continuing treatment

5.5 When deciding whether to start or continue an immunosuppressant, discuss the risks and benefits with the patient. Take into account the following in the context of the current prevalence of COVID-19 infection and the services that are available at the time.

- Is the patient's clinical condition stable?
- If treatment is needed, is there an alternative with a better risk profile?
- Is the required monitoring and review feasible?
- Will it be safer to delay starting the drug?
- Does the patient need to continue the drug?
- Can monitoring be done remotely or at a frequency that minimises the risk to the patient's safety and wellbeing?
- Are there any changes to the dose or route of administration that could make hospital attendance less likely?
- If continuing treatment, can the patient tolerate a reduction in the dose?
- If the patient has rapidly progressive interstitial lung disease, is intravenous therapy an option?

Involve all relevant members of the hospital specialist team in the decision and record the reasoning behind the decision.

5.6 Advise patients on immunosuppressive therapy to continue to take their treatment as prescribed to minimise the risk of their condition worsening.

5.7 For patients who have been advised to shield, making blood monitoring difficult, assess whether it is safe to increase the time between blood tests for drug monitoring if their clinical condition is stable on treatment.

5.8 Discuss with the patient the risks and benefits of being on an immunosuppressant with blood monitoring requirements. For patients with a condition that is responsive to immunosuppressants who are unable to attend
for blood monitoring, think about offering prednisolone alone.

5.9 Offer the lowest dose of prednisolone possible. If patients have been on prednisolone before, use the last dose that controlled their symptoms.

5.10 Offer antifibrotic therapy as usual if:

- the multidisciplinary team agrees a diagnosis of idiopathic pulmonary fibrosis
- eligibility criteria specified in NICE technology appraisal guidance on nintedanib and pirfenidone are met and
- blood monitoring can be safely carried out.

5.11 Advise patients already taking antifibrotic drugs that they should continue their treatment because there is no evidence they increase the risk of getting COVID-19 or make more severe disease more likely.

**Oxygen assessment**

5.12 Decide whether to carry out or defer an assessment for ambulatory or long-term oxygen therapy based on clinical need. Carry out oxygen assessments in the patient’s home if possible.

5.13 For patients already on ambulatory or long-term oxygen therapy, decide whether to carry out or defer an updated oxygen assessment, taking into account if their symptoms are worsening. Carry out oxygen assessments in the patient’s home if possible.

**Pulmonary rehabilitation**

5.14 Continue to offer pulmonary rehabilitation services to patients if available, including local services offering remote individualised education and exercise advice.

5.15 If remote pulmonary rehabilitation services are not available locally, think about using online pulmonary rehabilitation resources, such as the British Thoracic Society pulmonary rehabilitation resource pack. This covers self-management, home exercise and educational materials.
5.16 When face-to-face pulmonary rehabilitation services become available, discuss the risks and benefits of attending with the patient, taking into account the current prevalence of COVID-19.

**Lung transplantation referral**

5.17 Continue to refer patients for lung transplant assessment following the usual protocols.
6 Management: patients known or suspected to have COVID-19

6.1 When patients with symptoms of COVID-19 have been identified, follow UK government guidance on infection prevention and control for COVID-19. This includes recommendations on using personal protective equipment (PPE), patient transfers, and options for outpatient settings.

6.2 If COVID-19 is later diagnosed in a patient not isolated from admission or presentation, follow UK government guidance on management of exposed healthcare workers and patients in hospital settings.

6.3 If a patient has any signs or symptoms of a possible serious illness (for example, sepsis), assess and treat the illness in line with usual care.

6.4 If other possible diagnoses have been discounted, and COVID-19 is suspected, follow UK government guidance on investigation and initial clinical management of possible cases. This includes information on testing and isolating patients.

6.5 If a patient is diagnosed with COVID-19 pneumonia and shows consistent radiological changes, follow the British Thoracic Society's guidance on respiratory follow up of patients with a clinico-radiological diagnosis of COVID-19 pneumonia.

Continuing and stopping treatment

6.6 Before deciding to stop or adjust any treatment, if possible, contact the patient’s hospital specialist team for advice. If a medicine is stopped, ask the hospital specialist team when and how to restart it.

6.7 Continue antifibrotics if blood monitoring parameters are in an acceptable range and there is no other reason to stop, such as significant adverse effects.

6.8 If a patient is diagnosed with COVID-19, think about temporarily stopping immunosuppressants unless the risk of aggravating their underlying lung condition outweighs the benefits of stopping. When deciding whether to stop
treatment, discuss the risks and benefits with the patient.

6.9 Be aware that the half-life of some medicines means that the immunosuppressive effect will continue for some time after stopping treatment.

6.10 Advise patients on maintenance oral prednisolone that if they are diagnosed with COVID-19, they should continue treatment because stopping it can be harmful.

6.11 Be aware that some patients on long-term oral prednisolone may be at risk of an adrenal crisis and may need a higher dose if diagnosed with COVID-19.

6.12 Stop or adjust doses of medicines for interstitial lung disease if the patient develops acute kidney injury or deranged liver function tests because of COVID-19, in line with the BNF and the summaries of product characteristics.
7 Supplying medicines

7.1 Plan how to manage any disruption to normal routes for supplying medicines, such as homecare medicine delivery services.

7.2 Prescribe enough medicines to meet the patient's clinical needs. Prescribing larger quantities of medicines puts the supply chain at risk.
Update information

Minor changes since publication

6 November 2020: In recommendation 4.1, we linked to the NICE guideline on arranging planned care in hospitals and diagnostic services, and the UK government guidance on infection prevention and control for COVID-19 because NHS England's guide for managing respiratory patients during the pandemic was withdrawn.

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