

British Thoracic Society Advice for Managing Interstitial Lung Disease Patients during COVID-19 pandemic

Purpose: This advice is designed to help clinicians looking after Interstitial Lung Disease (ILD) patients during the COVID-19 pandemic.

Principles and Scope: This is a fast evolving situation and this document will be updated regularly.

All advice is general. Clinicians, after discussion with their patients, may choose alternative actions in some situations due to specific individual circumstances. This is perfectly acceptable practice.

Advice and support can be sought from your local specialist ILD centres. Expected staffing shortages however may mean response times will be slowed.

UK outpatient services have now been severely affected by the pandemic. New ILD referrals to specialist centres ideally will be queued. Delays are expected post pandemic to clear referral backlogs.

Update: All patients with ILD and sarcoidosis are advised to ‘shield’ where possible

On March 21st 2020 [a letter was sent to all English medical and nursing directors](#) from Ruth May, Chief Nursing Officer, England and Professor Stephen Powis National Medical Director, NHS England and NHS Improvement.

The letter “Caring for people at highest risk during the Covid-19 incident” specified which patient groups the government advised should ‘shield’ to try and maximally protect themselves from infection. Several respiratory disease categories were included in this advice for example: lung cancer, lung transplant, severe asthma and COPD and cystic fibrosis.

After discussion between the Royal College of Physicians London and BTS it is now advised that ALL ILD and sarcoidosis patients should be offered the same advice to shield due to their inherent risk factors. These disease categories however are not fully covered by the letter that was released. Some ILD patients were covered by virtue of the fact that patients on immunosuppression were mentioned in the letter. That statement does not cover fully patients however with Idiopathic Pulmonary Fibrosis, for example, who take anti-fibrotic drugs rather than ‘traditional’ immunosuppressant drugs.

All ILD and Sarcoidosis patients are now accepted to be relevant for inclusion in Group 4 of the list of vulnerable people and should be sent an At Risk letter as a matter of urgency – further details are available on the BTS website.

It is up to patients clearly to decide whether or not to take the advice being offered. ILD Specialist centres may receive phone calls from patients questioning this advice. Clinicians may need to assess and advise on a case by case basis for some patients. For the majority of ILD and sarcoidosis patients however the advice will be to shield.

It is also important to note that this advice has been agreed for NHS England but the NHS in our devolved nations, Wales, Scotland and Northern Ireland may decide to give advice to this patient group in a slightly different way. We will provide updates regarding devolved nations intended actions as soon as we have them confirmed. For ILD and sarcoidosis patients covered by NHS England however the advice is for patients to shield and receive a letter confirming this. Latest advice on Social Shielding (added 23/3/2020) is available here:

Guidance on shielding and protecting people defined on medical grounds as extremely vulnerable from COVID-19: <https://www.gov.uk/government/publications/guidance-on-shielding-and-protecting-extremely-vulnerable-persons-from-covid-19/guidance-on-shielding-and-protecting-extremely-vulnerable-persons-from-covid-19>

1. ILD Outpatient clinics – general points

A variety of actions have already been taken around UK ILD OPD activity already. Most organisations have now moved to telephone only (or other non-face to face methods) consultations already. Some centres have, or may be about to, shut down OPD activity except for only the most urgent cases. Government advice is for non-essential movement to cease. Most trusts have had to shut down routine spirometry/PFTs/bronchoscopy/CT scanning – urgent cases only getting tests.

Advice:

- Where possible conduct routine ILD OPD by non-face to face means.
- If this is not possible re-book patients later in year, if safe to do so.
- It remains unclear when normality will be restored.
- Going forward all OPD activity is suspected to pretty much cease in many places at some point as number of admissions rise and respiratory teams will be needed for inpatient care.
- Some appointments will be deemed clinically urgent e.g. rapidly declining patients and if patient agrees to attend then teams may see some of these on a case-by-case basis. This would be defined as essential travel/movement.
- Screening questions should be asked to ensure no suspected COVID-19 cases come to OPD.
- Ask about new cough, shortness of breath, fever, muscle aches, loss of taste and smell and headaches.
- We advise patients attending OPD areas have their temperatures checked on arrival.
- Admission avoidance ILD OPD interventions would be wise at this time to preserve beds where possible.
- What is possible will depend on local facilities and any risks a patient may be exposed to in attending an OPD visit.
- Essential clinic attendances for patients remain acceptable. This includes blood monitoring visits for essential medication.

2. Idiopathic Pulmonary Fibrosis (IPF) patients already on anti-fibrotic drugs

The anti-fibrotic drugs used are pirfenidone and nintedanib.

This group is not thought to be at any greater specific COVID-19 risk due to their treatment. Their risks may be their age, underlying chest disease +/- other co morbidities. No need to alter drugs due to outbreak.

ILD specialist centres not already using homecare systems should drive towards this now to ensure drug delivery to homes avoiding hospital visits to collect medication. Family members/friends may be able to collect drugs for those at risk if hospital is only source of medications. Continue drugs where feasible e.g. no major side effects etc.

Routine OPD visits for relatively stable patients should, where possible, be conducted by phone. Blood monitoring should continue. Where it cannot continue a risk assessment is required as to whether it is safe to continue drugs or not.

Pulmonary function testing for non-urgent cases has now ceased. Completing annual NICE checks re 10% FVC fall on anti-fibrotics is now not possible. This is not deemed urgent. Post telephone consults advise patients spirometry will be rechecked as soon as feasible post outbreak. Continue drug in meantime if clinically appropriate to do so.

As clinical review may already have been done by phone and next OPD review may be in 4-6 months - patients in many places can remotely attend for a spirometry result without having to be seen that day in OPD after pandemic has subsided. This may allow teams to collect FVC data needed for Bluteq prescription system without extra work of OPD visit in some cases. Some patients however may need to be seen particularly if FVC has dropped beyond 10%. Case by case basis management is needed. Managing pressure on ILD OPD services post outbreak will be important so try and plan to reduce workload where possible to reduce catch up later. Teams have a 12 month period to enter annual FVCs so drug supply can continue without pressure while systems catch up.

3. IPF patients already on anti-fibrotic drugs and diagnosed with COVID-19

If an IPF patient on anti-fibrotic drugs is diagnosed with COVID-19 and admitted to hospital it is not considered harmful to pause the drugs for short periods of time (4-8 weeks). However, any reintroduction of drugs in the recovery phase must be supervised by the patients ILD Specialist team. Please contact local specialist team to help. Anti-fibrotic drugs can be associated with deranged liver function tests and are generally not advised if creatinine clearance falls below 30mls/min.

4. New referrals that have suspected IPF

At the moment movement/travel for 'essential events' is permitted. Many would deem a new IPF case that appears suitable by FVC criteria for treatment as an 'essential event'. Cancer clinics (similar prognosis to IPF) for example are continuing to run in many places.

It would be sensible to run these cases through a 'pre OPD MDT' where possible to ensure all information is available pre OPD consult (some places do this as routine others do not). If teams have PFTs already pre OPD visit, from their centre or from a referring team, that show FVC is in criteria (i.e. 50-80% predicted) from last 6 months it is acceptable to use that value to decide upon treatment and use that value for bluteq system. If can get test on day – do – but if cannot can use pre-existing values.

Where IPF is diagnosed but no previous FVC, or FVC from past is above criteria, can reasonably request PFT as urgent from labs (like for cancer patients) as long as patient is not showing signs of infection with COVID-19. Using pre OPD MDT and a telephone consultation remote start-up of drugs using homecare is possible. Recent blood test results within 6 weeks of consultation would also need to be seen however. These may be available from GP who may have done bloods recently for other reasons. Some hospitals have access to GP blood results others do not. Paperwork to get patients signature for homecare can be obtained by post.

There is no reason at the moment not to offer patients start-up of anti-fibrotic drugs **as long as safety monitoring bloods can be obtained**. This may become a challenge later if pandemic runs on. In reality few new start-ups are likely to occur as staff may be shifted from OPD services to cover in patients and the risk of delaying starting an anti-fibrotic needs to be balanced by the risk of starting a new drug with possible adverse effects in the current environment.

5. ILD patients already on immunosuppression

The general advice for ILD patients already established on immunosuppression (IS) is to keep taking them unless they are experiencing symptoms of infection, monitoring bloods dictate problems or side effects are an issue. The risk being if stopped or reduced their background disease control could deteriorate.

It is important that ILD teams where possible aim to keep IS dosing as low as possible to maintain disease stability.

Any IS patient is at increased risk of infection. It is thus particularly important that ILD patients on IS socially isolate now and may need to do for some time to come. This group may experience a higher rate of post viral pneumonia or chance of clinical decline. At the first signs of deterioration or symptoms of a lower respiratory tract infection this group should seek early medical advice and if appropriate be treated early with broad-spectrum antibiotics. They should seek early medical advice about when/if to pause IS drugs due to co-existing infection or follow

advice already given to them by their clinics. A back up pack of antibiotics for this group would not be unreasonable where they can be supplied.

If IS/ILD cases require hospitalisation usual guidance regarding pausing IS drugs should be followed. This is usually stop IS medication during infection and for usually 2 weeks after to permit recovery. Sometimes longer pauses are merited on a case by case basis.

If doing consults carefully consider whether any IS in a patient has altered their disease trajectory. Could IS be weaned or stopped, even temporarily, for a few months taking into account COVID-19 risks? Could a sarcoid patient tolerate a slight drop in their IS? Do patients really need prednisolone escalation now? If original indication for IS was deemed *weak*, or, where there was a historical commencement of IS that is now weakly justified, it may be a good time to consider discontinuation of therapy.

If the pandemic persists over a longer term safety blood monitoring may become more difficult to access regularly for patients. On a case by case basis consider if you can switch patients back to prednisolone (at last dose that controlled them) and stop 2nd line IS drugs such as MTX or MMF for next few months. Consider how patients will collect new prescriptions.

6. Specific group: sarcoidosis patients with long standing lymphopaenia (on or off immunosuppression)

Longstanding low lymphocyte counts in sarcoidosis patients (whether on or off IS) is fairly frequent and not considered by itself to be a risk to patients with Covid-19 as we currently understand it. If a patient is receiving IS lymphopaenia is not a reason to alter IS specifically. It is noted that this was recently raised on social media. A recent study did not find this to be a specific issue.

7. Immunosuppression – new start ups

General advice in a non-rapidly progressing ILD patient

Starting a new ILD patient on IS during a viral pandemic raises concerns for both patient and doctor. Where patients appear stable (although maybe impaired) and evidence for effectiveness of IS is weak e.g. chronic hypersensitivity pneumonitis - if it is clinically safe to do so consider delaying any start-ups if possible until pandemic is moving on.

If oral IS is required consider oral prednisolone at doses of only 20mg or preferably less where clinically safe to do so. Higher dose steroids should be avoided due to associated poorer outcomes if concurrent COVID-19 infection ensues. Aim to go 'low and slow' in relation to steroids where feasible. In situations where you may normally introduce a 2nd line drug for e.g. methotrexate (MTX), mycophenolate mofetil (MMF) it may be safer to wait until pandemic is resolving to do this and leave the patient on prednisolone with counselling around any additional side effects that may be induced. This will reduce any burden on health services re blood monitoring required for drugs such as MTX/MMF. If a national lockdown does ensue in

future this will reduce the need to have bloods checked for safety. Second line agents e.g. methotrexate have longer half-life and take some time to be removed by body and have a longer impact on immune system than steroids might once stopped.

General advice in a rapidly progressing ILD patient

In rapidly progressive ILD cases e.g. vasculitis, connective tissue disease ILD - consider intravenous therapy on a case by case basis. Discuss risks versus benefits with patient. Where steroid based regimes fail to control disease longer acting drugs such as IV cyclophosphamide may be required (with some risks). IV rituximab should be a last resort, due to its very long half-life, at least until pandemic is starting to resolve. Prophylactic antibiotics are recommended with IV regimes most units have protocols in place regarding this.

General advice re steroids

If patients are on long term steroids usual dose increases ('bump ups') should be considered if ill to reduce risk of adrenal crisis. 'Bump down' once recovery is established. Patients are usually issued guidance around 'bumps' when given treatment initially.

8. Clinical Trials

General guidance is now out from MHRA and R&D departments. Pause observational studies where possible. Suspend new recruits into CTIMP studies. This will minimise workloads. It is anticipated that research staff, where possible, will be diverted to deliver other aspects of clinical care as pandemic progresses. Cessation of routine spirometry means new recruits often cannot meet screening criteria anyway. Patients under follow up in CTIMP studies may continue visits, where possible, with the primary aim to maintain safety. Patients must be screened by phone before attending research units for symptoms of possible COVID-19 infection. Spirometry will not be available routinely for these visits. Visits should be remote where possible. Teams can contact the study sponsor for advice on how to handle individual visits and what can be delivered remotely. Where safety bloods cannot be obtained contact sponsors for advice re managing individual cases. If safety cannot be maintained IMPs may need to be discontinued. Drug delivery can often be achieved via courier supported by sponsors.

9. Post COVID-19 pulmonary fibrosis

Acute Respiratory Distress Syndrome (ARDS) is known to promote the development of fibrosis in susceptible people that may relate to pre-existing undiagnosed fibrosis or genetic susceptibility. Given the large number of people expected to develop ARDS as a result of COVID-19 it is likely that there will be a substantial proportion of patients developing post COVID-19 fibrosis. The ILD community needs to prepare for this likely rise in referrals to services post pandemic. A further discussion later is also merited nationally with specialist teams to decide how best to follow these patients up.

10. NHSE Speciality Dashboard submissions

NHSE are fully aware of the pressures services will be under in the coming months. Full discretion will be applied for services who find themselves unable to submit NHSE ILD dashboard items during the pandemic period.

11. Respiratory Support and Intensive Care

Management advice re the use of CPAP, NIV or Hiflo oxygen systems is not ILD specific and is outside the remit of this scope. Information about respiratory support and Intensive care during COVID- 9 pandemic is available elsewhere on the British Thoracic Society website:

<https://www.brit-thoracic.org.uk/about-us/covid-19-information-for-the-respiratory-community/>

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