Restoring Lung Function testing for management of ILD

Background

Patients with a diagnosis of Idiopathic Pulmonary Fibrosis (IPF), diagnosed by a specialist ILD centre, require an FVC of between 50-80% predicted to be eligible to start anti fibrotic drugs. Without the relevant FVC value being provided anti-fibrotic drugs cannot be started. Restoration of timely access to lung function tests in this group of patients is thus a priority for drug access to be maintained. With IPF having cancer like outcomes in terms of mortality, access to diagnostics for IPF patients should be on a par with the access already provided for cancer patients. We suggest that breathing tests for IPF patients should be completed within a 2-4 week period of receipt of request.

Patients with ILD have been shielding and are understandably anxious about attending for diagnostic tests. Consideration needs to be given as to how best to provide pulmonary function tests as part of a COVID minimised pathway at a local level.

Respiratory function tests are required in a variety of clinical situations as below.

1] New IPF referrals to specialist centres

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Most centres obtain a forced vital capacity (FVC) at the initial attendance at the specialist clinic. This is essential to decide about commencing anti-fibrotic drugs. With the current limited access to pulmonary function tests (PFTs), as a consequence of them being aerosol generating procedures (AGP), there is reduced capacity in physiology departments. Clear *prioritisation* for ILD patients is thus now required to ensure timely access to obtaining an FVC value that can be used to prescribe drugs if suitable. Having an FVC on the day of attendance at any face to face clinics is important as it limits the number of visits for this vulnerable group. Some PFTs will be needed remote to any face to face attendance, as telephone consultations will be used instead. Whilst full lung function tests (e.g. gas transfer, lung volumes) are the best for monitoring disease progression, drug prescribing is based upon the FVC, which is the minimum requirement.

Recognising the limited access to pulmonary function testing that the COVID outbreak was going to cause, it was agreed in March 2020 by the specialised services commissioner that it was acceptable to use an FVC that was up to 6 months 'out of date' from the start date on the patients anti-fibrotic drug prescription. This seems to have worked well so far and centres have been able to perform remote drug start-ups. However, if the FVC result is greater than six months old, a further FVC needs to be obtained. The prescribing system that generates the funding confirmation with NHSE will not accept a lung function result with a date outside the 6 month window.

Equally if a patient is referred for their assessment at specialist centre, with a prior FVC greater than 80%, this needs to be repeated to determine access to therapy.

2] New or suspected IPF cases diagnosed at District General Hospitals (DGHs)

If a patient is 'suspected' of having IPF in a DGH setting they merit the same semi urgent priority for testing i.e. a 2-4 week timeline ideally spirometry, lung volumes or V_A together with gas transfer to establish if they are in range to start drug treatment. If this is not possible then the spirometry should be obtained as a minimum.

If appropriate, a prompt referral to a specialist centre can then be made. The specialist centre will be able to use this FVC as long as they see or assess virtually the patient within 6 months of PFT being done.

3] Known IPF cases in DGHs or Specialist centres that are being monitored as FVC previously too high to access treatment

There are many IPF patients that have confirmed or highly suspected IPF who are eligible for anti-fibrotic treatment, but are excluded as their FVC is currently too high i.e. > 80% predicted. Specialist centres have a number of 'high FVC IPF patients' that they are monitoring for a fall in FVC, which will trigger starting anti-fibrotic treatments as per the relevant NICE Health Technology Appraisal. This cohort of patients would normally have their FVC monitored approximately every 6 months. If unstable or in those that have exacerbated, a 3 monthly monitoring test may be in place. These patients are also a priority for FVC measurement, either on day of clinic or within 2-4 weeks of a virtual appointment.

In secondary care these patients also need to be identified and the FVC performed to *ensure equity* of care and allow onward referral. Whilst the population is potentially large, it is important to prioritise a] those IPF patients with a previous FVC of ≤ 90%, but above 80% and b] patients with a high FVC who report a significant decline or exacerbation and in whom their FVC may have fallen significantly. Testing should not be performed until 4-6 weeks post exacerbation.

Some patients, with limited training, may be suitable to be issued with a basic portable spirometer to measure FVC on a weekly/fortnightly basis at home. Several devices enable the spirometer to Bluetooth to a phone App that is linked to the patients' electronic medical record. This would reduce hospital visits and give confidence to the patient, whilst enabling remote clinics to work

4] Other ILD conditions

New ILD referrals with diseases usually managed with immunosuppression who are symptomatic with breathlessness e.g. sarcoidosis, chronic hypersensitivity pneumonitis and CTD-ILD require full PFT's to help inform treatment decisions and assess response. Some of these cases will also deserve priority testing and respiratory physicians should discuss urgency on a case by case basis with their lung function lab.

BTS
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