Interstitial lung disease (ILD) is a debilitating condition affecting over 150,000 people in the UK (British Lung Foundation, 2019); it is classed as a rare disease by NHS England (Bit.ly/NHSEILDSpecification). The most common diagnosis for patients with ILD is idiopathic pulmonary fibrosis (IPF); in 2012, about 32,500 people had a diagnosis of IPF in the UK (BLF, 2019).

There is no cure for IPF; it is a life-limiting, progressive disease characterised by lung function decline and reduced quality of life (Oldham and Noth, 2014). The average life expectancy for patients with IPF is three years from diagnosis, although 20% survive for more than five years (National Institute for Health and Care Excellence, 2013a). NICE (2016, 2013b) recommends the antifibrotic treatments pirfenidone and nintedanib to slow down its progression.

The progressive nature of IPF means patients require local palliative care services and timely access to symptom management (NICE, 2015). Services are generally delivered at regional specialist centres, which are often a considerable distance from patients’ homes; this can involve round trips of up to 200 miles. Manchester University NHS Foundation Trust’s ILD healthcare teams support around 450 patients with IPF on antifibrotic treatment from around the north-west of England.

Barriers to treatment
Antifibrotic treatment aims to preserve lung function and, thereby, prolong life but it often results in side-effects and can cause liver toxicity. As such, patients require 1-3-monthly monitoring by specialist nurses to identify side-effects and provide support – this involves attending specialist outpatient clinics at the trust.

The average age of patients with IPF is 74 years, and many have significant comorbidities; they often require oxygen therapy and their lifestyle can be significantly restricted due to breathlessness (Navaratnam et al, 2011). All these issues make long journeys extremely challenging.

The trust’s ILD nursing team identified that several patients had great difficulty attending appointments and some of those eligible for disease-modifying treatment declined referral to the specialist clinic due...
to its distance from home and the monitoring schedule involved. To reduce the need for patients to travel to specialist clinics, some tertiary centres relied on GPs performing blood tests for patients with IPF. However, these looked only for drug toxicity, so patients received no side-effect management or face-to-face contact with a respiratory specialist service. Treatment adherence was also often hindered due to the complexities of managing side-effects caused by the treatment. Both factors were negatively affecting patients' survival.

What changes have we made?
Prompted by these concerns, discussions were held with the newly established North West ILD Network to identify essential referring areas. We contacted the specialist nurses and allied health professionals (AHPs) associated with a consultant with a special interest in ILD to find out if they were interested in collaborating with us to set up a local monitoring clinic. The response was hugely positive and, as more local respiratory services accessed the network, we identified more network sites.

We invited those interested in collaborating to Wythenshawe Hospital to observe how our nurse-led ILD clinic monitors patients on antifibrotic treatment, managing both medication side-effects and ongoing symptoms. We then organised training for health professionals from across the region on managing the side-effects of treatment for IPF, which include gastrointestinal disturbance, photosensitivity and fatigue (NICE, 2016; 2013b).

Working with the identified specialist nurses and AHPs from around the north-west of England, the ILD nurses developed a referral pathway to take IPF patients established on antifibrotic treatments at Wythenshawe Hospital back to their local respiratory nursing and AHP teams. In 2014, we referred patients back to local services by letter and email – we found, because there was no management protocol, we were being contacted regularly with the same queries and concerns. In 2015/16 we decided to develop a medication-specific structured protocol to provide a more equitable service for patients and support for the network teams.

A literature review was undertaken of NICE guidance, pharmaceutical evidence, clinical trial data and real-world experience papers to support development of the protocol document. This document includes:
- Patient demographics;
- First blood results on treatment;
- Pulmonary function tests;
- Oxygen and pulmonary rehabilitation (PR) status;
- A monitoring schedule with an aide memoire to help manage the most common side-effects.

The protocol was shared with the network teams and feedback about the support it provided was positive. We introduced an ILD nursing team secure email account to access help and support from us five days per week in addition to having telephone contact. In one area there was an interested consultant with no nurse support. We were unable to offer outreach nursing support from the tertiary centre because of the distance involved so these patients were supported via our nurse-led telephone clinic.

Nurse-led telephone clinics have been pivotal for our service to operate across the region. They give patients:
- Access to ILD specialist nursing care without the need to travel, thereby providing a lifeline;
- Support with antifibrotic treatment;
- Timely referral to resources such as PR and palliative care.

Patients are scheduled into the nurse-led telephone clinic and have 1-3-monthly appointments. The clinics do have shortcomings, particularly with older patients who may have difficulty hearing. Fortunately, an ILD specialist nurse and member of our network is now in post, monitoring patients on treatment closer to home.

What have we achieved?
As a result of our focus on improving access for patients with IPF we now have six successful monitoring sites across the north-west of England, with 120 patients being monitored locally. In 2016, we did a snapshot survey over one month, asking patients whether they preferred to be monitored locally in an IPF clinic: all said yes.

Our network team supports patients on antifibrotic treatment, thereby improving adherence to it. Support to local teams to manage patients with complex needs is provided via secure email and a telephone helpline. With these initiatives, we have achieved effective local care for patients who have IPF via easier access to oxygen, PR, a community matron and palliative care services – meeting NICE's (2015) IPF quality standards and enhancing patient satisfaction. As a tertiary service, nursing satisfaction has improved as a result of:
- Easier referral pathways;
- Improved knowledge of the availability of local patient services.

There are also ongoing development opportunities for local respiratory nurses and AHPs interested in ILD, including access to meetings of the North West and Northern ILD Networks, and interdisciplinary ILD network educational resources.

What are the next steps?
Looking ahead, we would like to expand the model for improving access to ILD services across the north-west of England, which involves developing the network and satellite clinic provision further.

We have also developed a structured IPF care pathway for use with patients throughout the disease trajectory. It aims to provide health professionals with evidence-based steps to follow, ensuring all patients receive equitable, high-quality care at each stage of the disease. NT

- If you would like to contact Helen Morris about this project, please email: helen.morris2@nhs.net

Advice for setting up similar projects
- Be realistic about your aims and goals
- Start small, with interested teams
- Produce a comprehensive pathway
- Establish clear lines of communication
- Be available for queries from healthcare teams
- Use all opportunities to network

References
National Institute for Health and Care Excellence (2015) Idiopathic Pulmonary Fibrosis in Adults. nice.org.uk/csp79
National Institute for Health and Care Excellence (2013a) Diagnosis and Management of Suspected Idiopathic Pulmonary Fibrosis. BJSys/ NICEIPF/GUIDeline