



# **A map for better care:** making effective care pathways for people with interstitial lung disease

September 2017

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# About the British Lung Foundation

We're the only UK charity looking after the nation's lungs. We offer hope, help and a voice. Our research finds new treatments and cures. We help people who struggle to breathe to take control of their lives. And together, we're campaigning for better lung health.

With your support, we'll make sure that one day everyone breathes clean air with healthy lungs.

## Special thanks to everyone who attended our roundtable and fed into this report:

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# Foreword - Angela Gray

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My husband, Bob, had one of the most common types of Interstitial lung disease (ILD), idiopathic pulmonary fibrosis (IPF). Most people have never heard of it. And neither had we, until Bob went to the hospital for what we thought was a harmless cough and left with a diagnosis of this deadly disease.

IPF completely changed our lives. In a year and a half, we went from walks with our dog and ballroom dancing to arranging end of life care. IPF is not only fatal, it is also exhausting. We tried to continue as normal with our lives – we planned holidays and trips together. We tried to be hopeful. But after a while, the fatigue and breathlessness that comes with IPF restricted what Bob was able to do.

Since Bob died, I've been left with so many questions about his care. Could more have been done to improve his quality of life? Why was he never offered pulmonary rehabilitation? Why did it take so long for people to explain how deadly IPF was? Why did it feel like we were being passed from pillar to post around the health service? That's why I'm so pleased to have worked with other carers, patients, professionals and the British Lung Foundation to write this report.

Bob died three years ago. Since then changes have been made to the way services are delivered, and new treatments are available. This report outlines this progress and identifies where challenges still exist. It uses insights from professionals, patients and their families to recommend where and how services could be improved.

Bob was the kind of person who never complained (except about my driving in his car!). When he needed the most support he was too exhausted and breathless to demand it. We need to put in place this report's recommendations so that people with ILD don't have to spend the short time they have left finding their way around the health care system. It might be too late for Bob, but ILD isn't going anywhere. This year, more than 6,000 people in the UK are likely to be diagnosed with IPF.

Together, we can make the health care system work for everyone living with ILD.

**Angela Gray** *Former Chair of Northern Region IPF Support Group*

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## Note from the British Thoracic Society

"The British Thoracic Society was pleased to be invited to take part in the roundtable exercise in November 2016. BLF has raised important issues aimed at improving the care of patients with ILD and the development of effective

services across the UK. BTS hopes to play its part in this process."

**Dr Michael Gibbons** *Chair, BTS Interstitial and Rare Lung Disease Advisory Group*

# Executive summary

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Idiopathic pulmonary fibrosis (IPF) is currently an incurable lung condition with no known cause or cure. The average life expectancy in the UK following diagnosis is three years; a poorer prognosis than cancer of the colon, breast or ovary.

During the last decade IPF was responsible for approximately 5,000 deaths a year in the UK – or, put another way, 1 in every 100 deaths each year in the UK is due to IPF.

Access to services, treatment and support for patients with IPF is fragmented. This urgently needs to change. We need to prioritise respiratory services and make health systems work for everyone living with IPF. Patients can't afford to waste the precious time they have left going from service to service for help. They need speedy, seamless access to the care they need, when they need it.

Many patients tell us that even medical professionals don't understand their condition; that they didn't receive any clear information about the disease; that their diagnosis was delayed; and that treatments are out of reach. Following an initial visit to their GP, people may have to wait many months before being referred to a specialist.

We can and must improve services for patients. We recommend the following actions:

- 1. Establish taskforces for lung health** in England and Scotland to write five-year strategies to improve respiratory outcomes. In Wales and Northern Ireland, integrate a national plan for ILD services into existing and future respiratory plans.
- 2. Create local ILD networks** across the UK. These networks should bring together health care professionals, policy makers, commissioners, charities and patients to improve local ILD plans and services.
- 3. Develop ILD pathways.** ILD networks should develop and publish these pathways. They should be designed around the key principles in this report:
  - Tailored interventions
  - Collaboration and integration
  - Equity of access
  - Patient-centred care and communication
  - Transparent data and information.

**4. Improve access to personalised treatments, diagnosis and support**

including anti-fibrotic drugs, ILD specialist nurses, multi-disciplinary diagnosis, oxygen therapy, pulmonary rehabilitation, peer support groups and palliative care.

**5. Evaluate and improve existing ILD services**

by improving the BTS ILD Registry, data recording and sharing, publishing a list of ILD services, amending the tariff system to incentivise best practice, evaluating existing services and publishing findings.

These recommendations have been drawn together from a roundtable discussion held by the British Lung Foundation in November 2016. The report examines care for all ILDs. IPF is the most common type of ILD, so is used as an illustrative example throughout the report.



# Abbreviations

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<b>BTS</b>	British Thoracic Society
<b>CCG</b>	Clinical commissioning group
<b>CF</b>	Cystic fibrosis
<b>COPD</b>	Chronic obstructive pulmonary disease
<b>CQUIN</b>	Commissioning for quality and innovation framework
<b>CTD-ILD</b>	Connective tissue disease-associated interstitial lung disease
<b>FVC</b>	Forced vital capacity - measure of lung function
<b>ILD</b>	Interstitial lung disease
<b>IPF</b>	Idiopathic pulmonary fibrosis
<b>MDT</b>	Multi-disciplinary team
<b>NHS</b>	National health service
<b>NICE</b>	National institute for health and care excellence
<b>QALY</b>	Quality-adjusted life year
<b>SMC</b>	Scottish Medicines Consortium
<b>STP</b>	Sustainability and transformation plan



# Introduction

## What is ILD?

Interstitial lung disease (ILD) is an umbrella term for more than 200 conditions that affect the interstitium, a lace-like network of tissue that supports the alveoli in lungs.<sup>1</sup> ILDs are largely classified into those where the cause is known and those where the cause is not. IPF is the most common type of ILD,<sup>2</sup> and does not have a known cause.<sup>3</sup>

### Classification of interstitial lung disease (ILD)

#### Major ILDs of known cause (around 35% of all patients with ILD)

- Pneumoconiosis (e.g. asbestosis, silicosis)
- Extrinsic allergic alveolitis (hypersensitivity pneumonitis)
- Latrogenic ILD caused by drugs and/or radiation
- Post-infectious ILD

#### Major ILDs of unknown cause (around 65% of all patients with ILD)

- Sarcoidosis
- Idiopathic interstitial pneumonias (IIPs), of which the most common are:
  - Idiopathic pulmonary fibrosis (around 55% of IIPs)
  - Nonspecific interstitial pneumonia (around 25% of IIPs)
  - Respiratory bronchiolitis ILD, occurring in smokers (around 10% of IIPs)
  - Desquamative interstitial pneumonia (around 5% of IIPs)
  - Cryptogenic organising pneumonia (around 3% of IIPs)
  - Lymphoid interstitial pneumonia (around 1% of IIPs)
  - Acute interstitial pneumonia (around 1% of IIPs)
- ILD in connective tissue diseases (CTDs) and in collagen-vascular diseases, of which the most important are:
  - ILD in rheumatoid arthritis
  - ILD in progressive systemic sclerosis

Source: European Respiratory Society White Book

“I had a cough for about 6 years before I was diagnosed with IPF. When I had the first X-ray I was told I had lung damage. Eventually I had another X-ray and a specialist told me I had IPF. I used to be a weight-lifter and didn’t smoke. I couldn’t believe I now had a deadly lung condition.”

### Roy from Penyffordd living with IPF

Typically patients with ILD present with breathlessness, a cough and reduced exercise capacity. People with IPF develop scarring (fibrosis) in their lungs, making it harder for them to absorb oxygen.<sup>4</sup> Some experience clubbing of the fingers, as well as crackles in their lungs when breathing.<sup>5</sup>

### Difficult to diagnose

Some ILDs are difficult diseases to diagnose due to their relative rareness and the need of a multi-disciplinary team (MDT) to reach a diagnosis.<sup>6</sup> The symptoms associated with ILD, particularly breathlessness, are often misdiagnosed as other lung conditions.

IPF is mostly diagnosed in people between 71 and 80, with 85% of diagnoses made in people aged over 70.<sup>7</sup> It is rare for people under 51 to be diagnosed.<sup>8</sup>

Before the availability of specific treatments, studies showed that almost half of people with IPF in the UK died within three years of their diagnosis.<sup>9</sup> However, about one in five people lived for more than five years after they were diagnosed. Many clinicians believe the treatments now available will mean that people diagnosed today will survive longer.<sup>10</sup>

### No cure but treatment can slow progression

Lung scarring from some ILDs, such as IPF, is often irreversible and treatments are not always effective in preventing progression. For IPF, two new drugs have recently become available to slow lung scarring and subsequent decline in lung function.<sup>11</sup> Interventions such as pulmonary rehabilitation can help improve physical function and lessen the impact of the disease.<sup>12</sup> Some people with IPF and other ILDs may

be eligible for lung transplantation, although the long waiting list for transplants and poor post-operative prognosis means that this is only an option for a minority of patients.<sup>13</sup>

People living with IPF can often experience long periods of stability which are punctuated by episodes of dramatic and acute deterioration. These episodes often prove life-threatening.

### IPF is more common than we thought

The British Lung Foundation’s 2016 report – *The Battle for Breath* – identified that around 32,500 people in the UK live with IPF, which is more than double NICE’s previous estimates.<sup>14</sup> The report found that there are 6,000 new diagnoses of IPF each year and 5,300 deaths. IPF is 50% more common in men than women; the reasons for this are not known.

Prevalence is highest in Northern Ireland, north-west England, Scotland and Wales, but least common in London; again, the reasons are unknown. The report also identified that IPF incidence is not influenced by socio-economic status, which may be considered surprising given that there had been previous indications that IPF is linked to industry and manufacturing.<sup>15</sup> Some evidence suggests a correlation with tobacco smoking.<sup>16</sup> There are 9,000 yearly hospital admissions for IPF, accounting for 86,000 hospital bed days.

Although this dataset is the most authoritative on IPF in the UK, we must exercise some caution due to the broad range of possible codings that could be used by health care professionals to record IPF, as well as changes that have been made to the way IPF is coded.

“Unlike most people, I did know what IPF was when I was diagnosed. I helped my mother, brother and sister battle with this devastating condition. There is no known genetic link, but IPF has certainly ripped my family apart. I’m going to fight as long as I can to improve services for people who can’t help themselves.”

**Una from Belfast living with IPF**



## Progress in ILD service provision across the UK

The availability of new treatments has substantially changed the services available to ILD patients in the last few years.

### Tailored interventions

Improvements in the access to, and quality of, interventions such as pulmonary rehabilitation, lung transplantation, oxygen therapy and palliative care were targeted by NICE in IPF guidance published in June 2013 and January 2015.<sup>17 18</sup> This guidance is applicable in England, Wales and Northern Ireland. It is also used to guide care in Scotland, but is not formally applicable. The NICE guidelines led to the development of NHS England's service specification on ILD, published in 2013, which also sets out that services must offer these treatments across the care pathway.<sup>19</sup>

The last few years have also seen the approval by health care regulators of new drugs to slow the progression of IPF – pirfenidone in July 2013 and nintedanib in January 2016. These drugs have been shown to reduce annual decline in lung function (forced vital capacity – FVC) by 50%, but are only available for IPF patients with a predicted FVC range of between 50% and 80% in England, Wales and Northern Ireland and lower than 80% in Scotland. This is despite evidence showing these drugs can be effective for patients with over 80% FVC.

### Collaboration and integration

The NICE IPF guidelines and ILD service specification in England aim to support collaboration in ILD diagnosis and service delivery. ILD specialist centres have become leading experts in

care and are supporting more patients than ever before.

The NICE IPF guidelines and NHS England's service specification guided the establishment of these specialist centres. They have also guided progress in Northern Ireland, where the Public Health Agency is currently revising the ILD pathway to consider new models of care. Health services in Wales are expected to collaborate to deliver services for people with respiratory disease, as outlined in the Welsh Government's respiratory health delivery plan.<sup>20</sup> In Scotland, however, progress has largely been driven by local experts. Specialist centres have emerged without support from a national strategy or guidelines. A policy roundtable in Holyrood, organised by the BLF and Boehringer Ingelheim in 2014, attributed this to a lack of national prioritisation in lung disease.<sup>21</sup> This differing pace of change shows that ILD services require support and investment from national governments to achieve better care for patients.

### Equity of access

NHS England, as well as the Welsh and Northern Irish governments, has set out national frameworks to improve equity of access to ILD care – most notably diagnosis through a MDT. NHS England's 2013 ILD service specification specifically sets out an objective to improve equity of access to specialised therapies for all patients with ILD in England.<sup>22</sup> It also sets out that centres must provide access to specialist MDTs to increase access to specialised therapies.<sup>23</sup> A framework to ensure equity of

access to ILD care in Scotland will need to be developed in the future.

The Welsh Government's Respiratory Health Delivery Plan, published in 2013, also sets out that local health boards should ensure ILD patients are managed through a MDT, to improve diagnosis times and reductions in mortality rates. This strategy prompted the establishment of the South Wales tertiary ILD service in April 2016. This in turn led to a marked reduction in the mean time from referral to MDT diagnosis from 19 weeks between June and December 2015 to 2.2 weeks from April 2016 onwards.<sup>24</sup> A tertiary ILD service in North Wales also became functional from April 2017.

In Northern Ireland, the publication of a service framework for respiratory health and wellbeing for 2015-2018 set out national targets for all health and social care trusts to meet by set dates.<sup>25</sup> These included access to radiologists with thoracic imaging, access to pathology services, a named lead consultant with an interest in ILD, a named specialist nurse with an interest in ILD and a percentage of people with suspected ILD/IPF who have had a case discussion with a local MDT.

### Patient-centred communication and care

There have been limited national directives to improve patient-centred communication and care. NICE

guidance published in 2013 and 2015 on IPF specifies that written and oral information and support must be provided for people with IPF (and their families and carers) on diagnosis, as well as on an ongoing basis. This guidance is applicable in England, Wales and Northern Ireland. Although the number of ILD support groups across the UK has increased, this increase has been locally driven, with support from the British Lung Foundation and Action for Pulmonary Fibrosis. Similarly, initiatives to produce high quality, patient targeted information on conditions have been driven by the third sector, with Information Standard accredited ILD specific information published by the BLF.

### Transparent data and information

The British Thoracic Society's ILD Registry was launched in England in February 2013. It signalled a major improvement in the collection, sharing and usage of patient data.<sup>26</sup> This is a registry of patients in secondary and tertiary care with a diagnosis of IPF, suspected IPF, and sarcoidosis, which contains patient information, clinical information on lung function, and follow-up information. It has helped assist clinical practice and improve standards of care, as well as identify areas requiring improvement and benchmarking across service providers.<sup>27</sup>

## What does the ILD care pathway look like?

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The NICE guidelines give us a good understanding of the way IPF care pathways should be managed, but care should also go above and beyond this framework. Care pathways will be different for every person with ILD; they require overarching principles to guide them and local networks to embed them for consistency and integration.

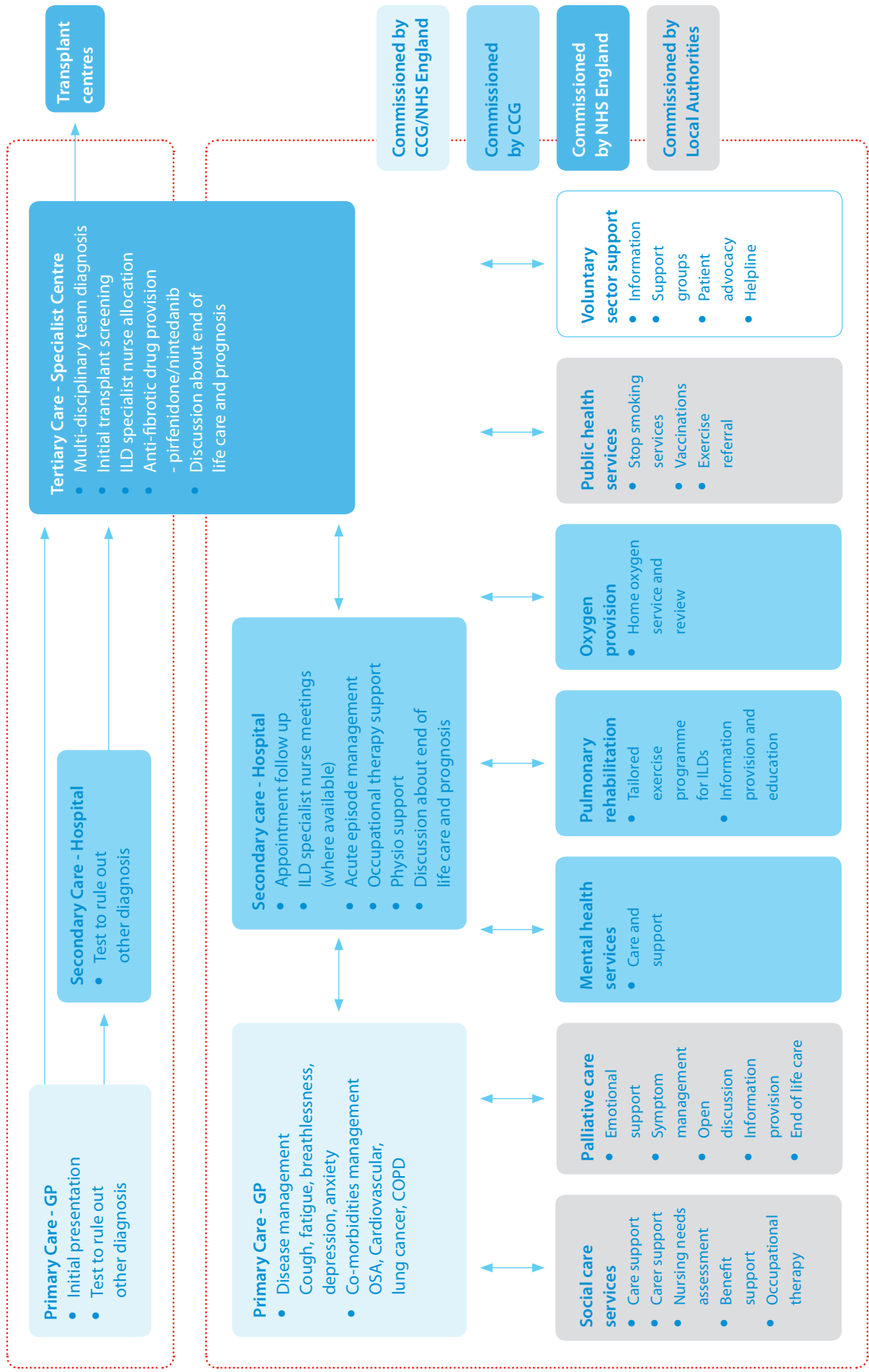
Across the health sector, 'care pathway' is often used interchangeably with other terms such as clinical pathway, critical pathway, integrated care pathway, care journey or care map. These terms all refer to a process of systematically planning and evaluating a focused patient or client care programme.<sup>28</sup>

Care pathways aim to enhance the quality of care for different groups of patients. They tend to focus on improving patient outcomes, promotion of patient safety,

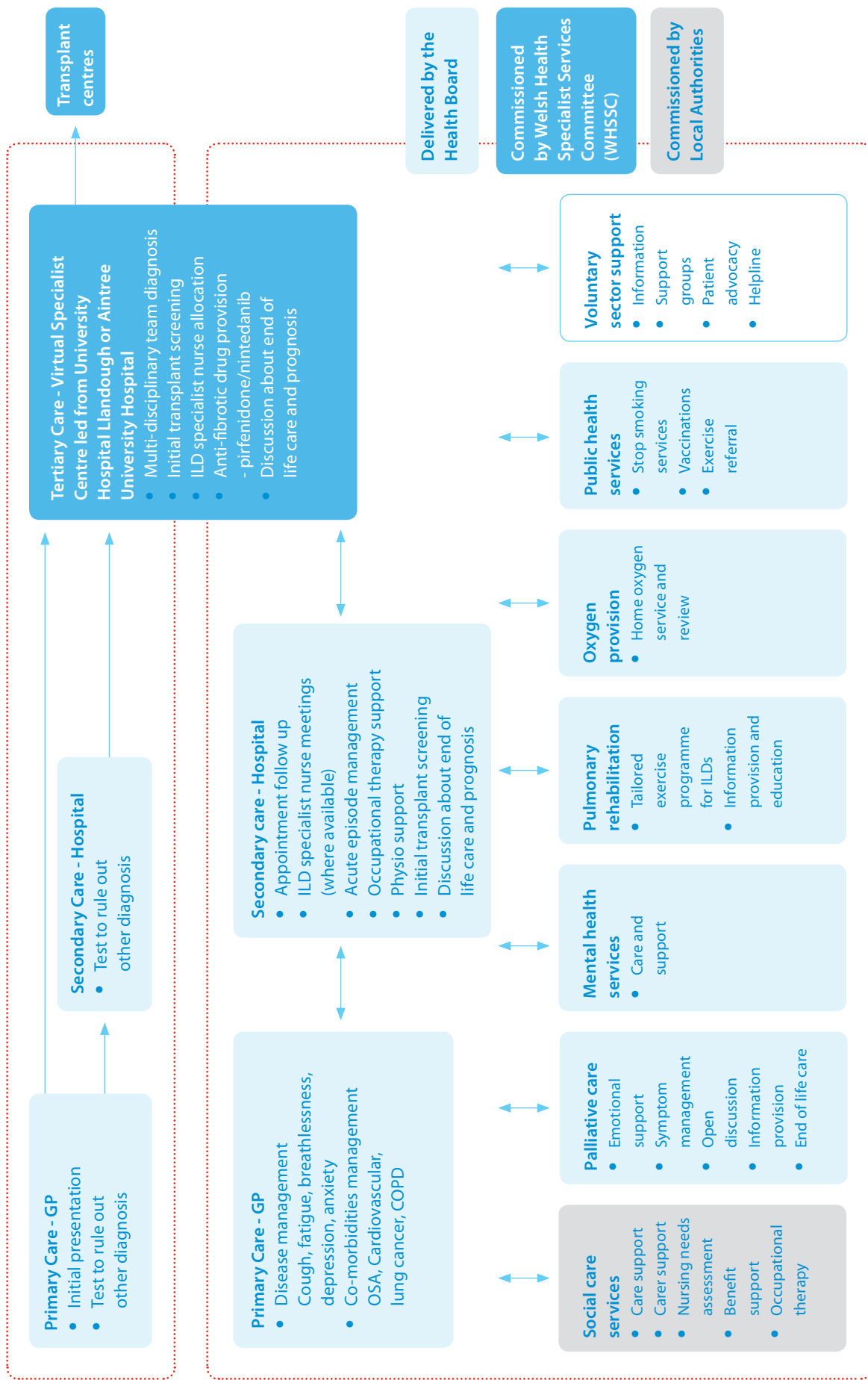
increased patient satisfaction and optimisation of resources. This report is rooted in the European Pathway Association's definition of a care pathway, but does not seek to prescribe what these pathways should look like in different areas.

The following diagrams have been produced for this report to illustrate the current pathways for people living with IPF. The diagrams demonstrate the range of services that a person with IPF may encounter and rely upon in each nation. The service providers vary across nations, but the core components of care remain the same. These diagrams will vary for every person living with IPF. This report has used the NICE guidelines to demonstrate the services at each stage of the pathway, but in reality not every patient will receive these services. We will elaborate on this inconsistency and variation further in the report.

# Current IPF care pathways in England - a patient perspective

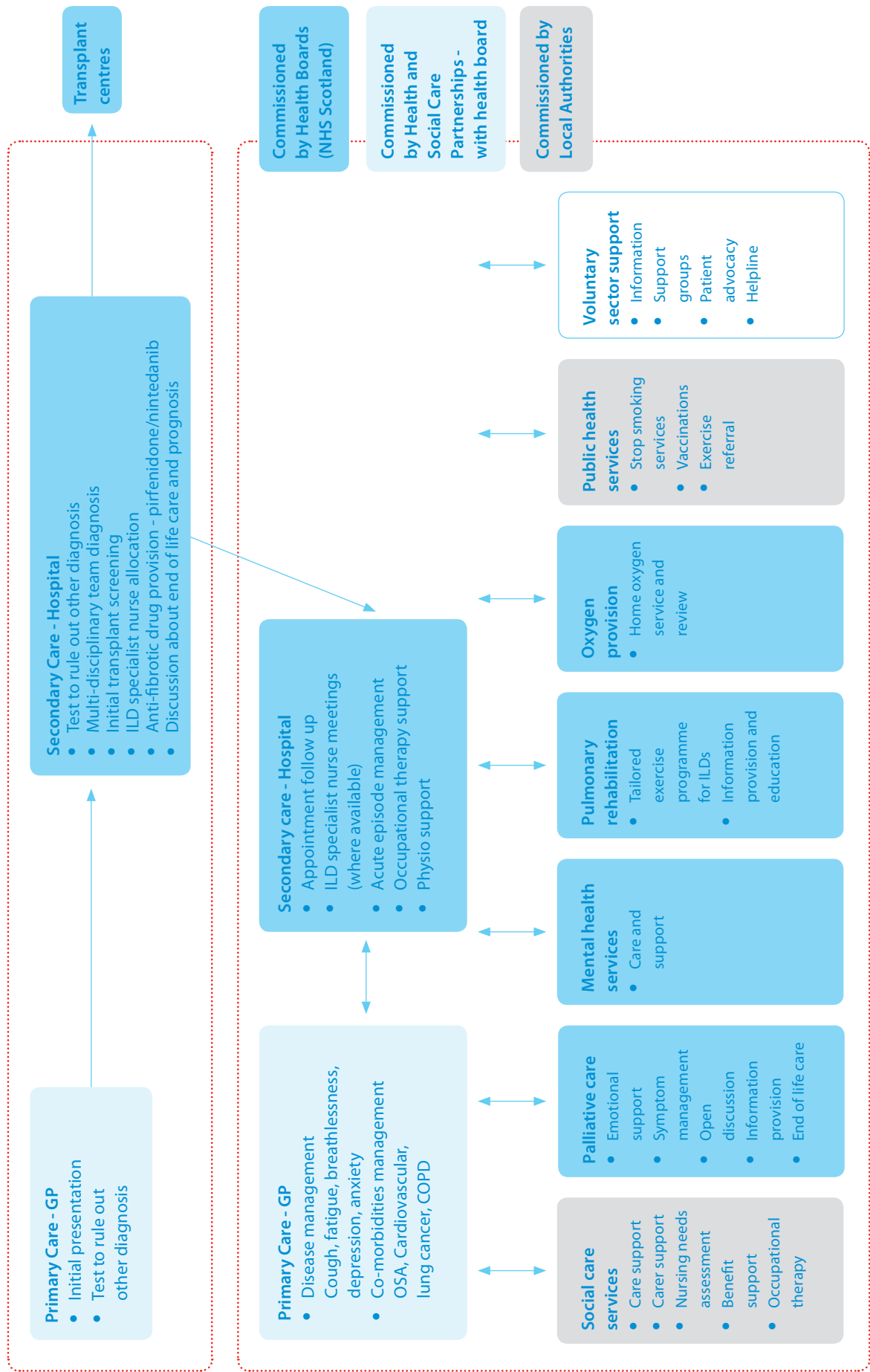


# Current IPF care pathways in Wales - a patient perspective

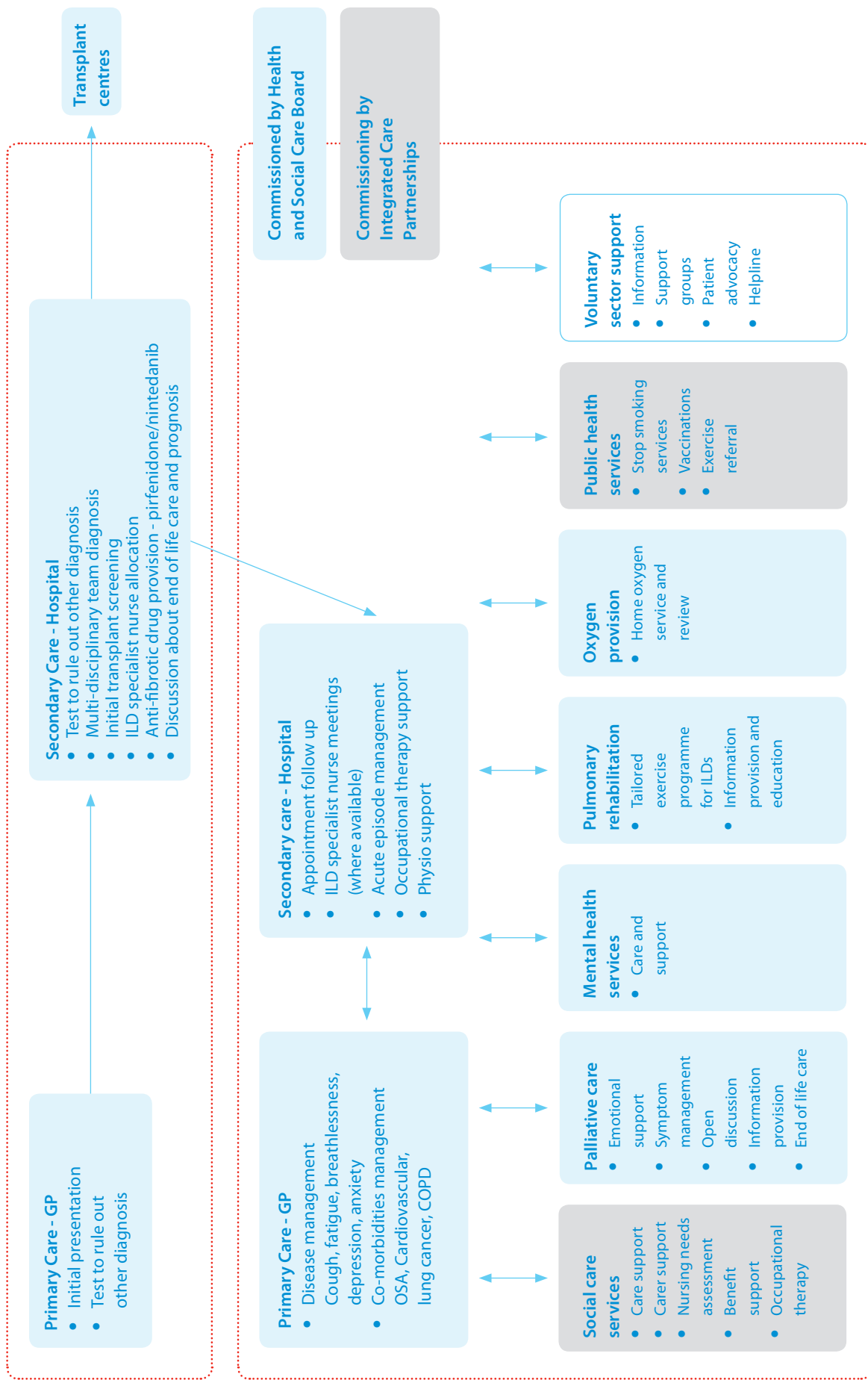




# Current IPF care pathways in Scotland - a patient perspective



# Current IPF care pathways in Northern Ireland - a patient perspective



# Key principles of care

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The delivery of ILD care is complex and touches on many parts of health and social care services. This requires local health teams, policy makers, commissioners, specialist centres and providers to work together to organise, plan and deliver a full range of integrated services. This section of the report identifies areas for improvement and uses insights from our roundtable to suggest solutions and practical measures to counter these challenges. It also showcases examples of best practice in areas of the UK.

## 1) Tailored interventions

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Early, timely and accurate diagnosis is essential for all ILD patients. Once this diagnosis is received, all patients should receive a holistic needs assessment for the services that they require, before being offered a range of tailored services that have been commissioned in line with national guidance.

### i) Access to drugs

Two drugs for IPF have been approved by NICE (and endorsed for use in Wales and Northern Ireland) and the Scottish Medicines Consortium (SMC) for IPF treatment on the NHS – both are recent innovations and the first major treatments for IPF. These are:

- Pirfenidone (approved in 2013 by both NICE and the SMC)
- Nintedanib (approved in 2015 by the SMC and 2016 by NICE).

**We need all patients who could benefit from anti-fibrotic drugs to be offered them**

The BLF's 2015 report, *Lost in the system*, found that only 66% of all patients responding to a survey conducted as part of the report were receiving some form of medication for their IPF, and only 30% of these had been prescribed pirfenidone.<sup>29</sup>

In addition, not all IPF patients are eligible for these anti-fibrotic drugs. NICE recommends prescribing pirfenidone and nintedanib to treat IPF only if the patient has a forced vital capacity (FVC) of between 50% and 80%. The SMC recommends prescribing pirfenidone and nintedanib for people with IPF if FVC is 80% or less.<sup>31</sup> Unfortunately, these limits mean that patients in the early stages of diagnosis with lung function above 80% are not able to benefit from treatments.

“Participating in IPF research trials and keeping up with new developments gives me hope for the future. Attending research conferences helps me remain positive. They give me a chance to make my voice heard. I was fortunate to have a lung transplant in 2015. I know many other people won’t get this chance. That’s why I’m determined to help IPF research in any way I can.”

**John from Staffordshire living with IPF**

Studies have shown that patients with lung function over 90% have received the same benefit from these drugs as patients with more impaired lung function,<sup>32,33</sup> yet they are unable to receive treatment until their FVC declines. One patient described the current situation as like telling a cancer patient they cannot have treatment until their tumour gets larger. Patients should not have to wait until their disease gets worse before being prescribed these life-extending drugs.

**We want all patients to be given information about and access to clinical trials for promising new drugs**

Clinical trials of promising new drugs are not guaranteed to provide results for patients. But access to novel treatments can provide them with hope and reassurance that every possible option that could have been taken, has been taken. However, *Lost in the system* found only 42% of patients had been given information on such clinical trials.<sup>34</sup> This suggests scope for more widespread provision of information and access to clinical trials.

**National health care regulatory bodies should:**

- Amend the criteria for prescribing pirfenidone (Esbriet) and nintedanib (Ofev) to ensure that anyone who can benefit from them has access to them.

**Health care professionals should:**

- Ensure that patients who can benefit from drugs have access to them.
- Discuss and agree the most appropriate drugs to use throughout a patient’s care journey.
- Give patients information on clinical trials.

## ii) Pulmonary rehabilitation

Pulmonary rehabilitation is a six-to eight-week multi-disciplinary programme of supervised exercise classes and education designed to improve physical function, improve confidence to exercise and create sustained behaviour change around physical activity and self-management.

We need quicker and more widespread access to pulmonary rehabilitation, as well as better tailoring to ILD, particularly IPF.

We learned from the roundtable that pulmonary rehabilitation tailored specifically for IPF is not routinely offered. This is despite NICE guidance recommending access and tailoring, outlining that IPF focused pulmonary rehabilitation programmes can contribute to improved health-related quality of life and exercise capacity.<sup>35</sup> Data from the BTS ILD Registry found that 53% of patients at participating centres were referred to pulmonary rehabilitation.<sup>36</sup>

Evidence from patients and health care professionals indicate that pulmonary rehabilitation services are sometimes structured to focus on other lung conditions, largely COPD.<sup>37</sup> This can lead to patients feeling that they are being offered treatment for COPD, rather than for IPF.

The commissioning of all pulmonary rehabilitation services for ILD patients must be improved, with better referral and improved communication of its benefits.



“I had been managing my condition quite well with medication and regular exercise, but it was after I was hospitalised for a chest infection that I needed some extra help, so I was immediately referred to pulmonary rehabilitation. I thoroughly enjoy each class and have made some great friends. With both oxygen and exercise I feel like I’ve been able to take control of my condition. I live life to the full.”

**Jim from Glasgow living with ILD**

“I’ve lost count of the number of times I’ve been told: ‘you look really well, I can’t believe you’re ill’. It’s hard to convince people that I have a serious lung condition – even family. Being really breathless is the most frightening thing about having a lung condition. My energy levels might dip, but it will never stop me being me. I might be ill, but my illness doesn’t define me. I’ve learnt that being breathless is good for me. Exercise helps me get more oxygen. It helps me get stronger. I just need to do things at my own pace.”

**Ann from Falmouth living with IPF**

#### We need to invest in research to show which patients with IPF are most likely to benefit from tailored pulmonary rehabilitation

Existing academic evidence suggests that pulmonary rehabilitation is an effective intervention for people with IPF. However, there are several gaps in research, notably:

- Evidence from UK-based studies
- Evidence showing which ILD patients are suitable
- Effectiveness at different stages of an ILD patient’s treatment journey – a major factor in the effectiveness of interventions
- The longer term benefits for ILD patients.

So far, we only know that pulmonary rehabilitation is more effective when delivered early. It can improve six minute walking test (6MWT) results and quality of life for some, but not all, people with IPF.<sup>38 39 40</sup>

It was suggested at the roundtable that gaps in the evidence make it difficult to commission ILD focused pulmonary rehabilitation. We need to establish a robust evidence base similar to that for COPD. For COPD, we know that it is one of the most cost-effective interventions at £2,000 - £8,000 per QALY,<sup>41</sup> and that it can reduce hospital admissions,<sup>42</sup> reduce anxiety and depression,<sup>43</sup> and improve physical function.<sup>44</sup>

#### National policy makers should:

- Support monitored access to pulmonary rehabilitation for people with ILD. This will enable research to assess:
- Which patients with IPF are most likely to benefit from pulmonary rehabilitation, to what extent they can benefit, and which metrics are most likely to see improvements

- What the long term benefits of pulmonary rehabilitation are for people with ILD
- How pulmonary rehabilitation can be tailored to improve outcomes for people with ILD.

#### Commissioners of pulmonary rehabilitation services should:

- Tailor services for ILD with sufficient capacity, resources and expertise, in line with NICE guidance.
- Ensure that enough services and places exist on tailored IPF programmes.

#### Health care professionals should:

- In line with NICE guidance, ensure that patients are aware of the benefits of pulmonary rehabilitation, both in terms of education and improvements to physical function, and refer them for an assessment.
- Ensure that ILD patients are referred for an assessment for pulmonary rehabilitation.

### iii) Lung transplantation

Lung transplantations are an option for some patients with IPF, but do not represent a permanent cure and come with their own risks. Often people with IPF who could receive a transplant, it is likely that two will die shortly afterwards, with a further three in five years, and a similar number within ten years.<sup>45</sup>

Transplantations must be conducted at a time when the patient is most likely to survive the procedure, and also before a patient deteriorates to the point where a transplant is a critical need.<sup>46</sup> Some clinicians believe that if a patient’s lungs are likely to stop working within two years then it is time to consider transplantation.<sup>47</sup>

IPF lung transplantations are most frequently carried out in those aged 55-59 years old.<sup>48</sup>

Patients may be considered ineligible because of a range of factors, including if they are a current smoker (a minimum of six months abstinence is required) or have other co-morbidities.<sup>49</sup>

**We need patients to be promptly assessed and referred for lung transplantation**

Only 188 lung transplants for all lung conditions were carried out in 2015/16 and on 31 March 2016 there were 330 people on the active lung transplant list.<sup>50</sup> These low levels of transplantation have been attributed to donor shortage,<sup>51</sup> to under-use of viable lungs, and to systemic challenges in gaining patient and family consent.<sup>52</sup> Data from the Freeman Hospital in Newcastle shows that just 29% of 486 transplantation referrals for IPF patients between 1987 and 2012 resulted in transplantations.<sup>53</sup>

Some patients at the roundtable told us they feel like they weren't considered for transplant because of their age. Given that one of the core determinants for post-transplantation survival is the quality of a patient's health,<sup>54</sup> this should be the main criteria, rather than age.

**We need more health care professionals to discuss lung transplantation with patients shortly after diagnosis**

NICE guidance outlines that specialist health care professionals should discuss lung transplantation with patients within six months of diagnosis, unless it is not suitable for them.<sup>55</sup> However, not all patients are made aware of the option of a transplant and many are not

provided with a satisfactory and tactful explanation of why they are not considered suitable. This can result in a poorer experience for the patient and their family/carers, as they may feel they have missed a vital treatment opportunity.

**We need national governments to increase the number of lungs being used in transplants and ensure that all eligible IPF patients are prioritised for treatment**

Only 25% of lungs from donors are used in transplantation. The remaining 75% are often clinically viable and suitable for use, but get discarded. A report from the Cystic Fibrosis Trust found that many clinicians are developing new techniques which mean more lungs can be transplanted that wouldn't have been used before.<sup>56</sup> National governments should ensure clinicians have access to the best scientific evidence and global examples on lung transplantation. This will help surgeons develop new techniques to increase utilisation rates and carry out innovative procedures.<sup>57</sup>

Given the poor prognosis many IPF patients have, they should be prioritised for lung transplantation where they are eligible and it is deemed effective. Currently, lungs retrieved from donors are allocated to the closest transplant centre. We support the creation of a national list which would allocate lungs to patients who need them most.

**National governments should:**

- Establish a national allocation system for lung transplantation that prioritises patients who are most suitable for transplants and also require them the most.
- Ensure clinicians have access to the best scientific evidence on lung transplantation

to enhance practice and improve utilisation rates.

#### Health care professionals should:

- Discuss lung transplantations with patients within six months of diagnosis, in line with NICE guidance.

#### iv) Oxygen therapy

The majority of people with IPF will experience breathlessness.<sup>58</sup> This can impact on their ability to perform day to day activity and can have a significant impact on their quality of life. Chronic hypoxemia (low concentration of oxygen in the blood) can also occur in people with severe ILD.<sup>59</sup> This can result in poor tissue oxygenation and the development of complications such as pulmonary hypertension.<sup>60</sup>

Oxygen therapy can be used to address hypoxemia, and to support people to be active and take part in pulmonary rehabilitation.<sup>61</sup> Oxygen can be delivered either at home from a static position within a residence (home oxygen therapy) or outside and on the move (ambulatory therapy). NICE recommends that patients require an assessment of home and ambulatory oxygen therapy at each follow-up appointment and before they leave hospital.<sup>62</sup> This is intended to identify changes in their oxygen needs and could prevent further exacerbations.

#### We need patients to receive a full, tailored assessment of their oxygen needs in their own home

Evidence gathered from patients suggests that assessments for home oxygen do not always adequately take into account a person's individual living circumstances. This

can include the patient's choice of equipment used to deliver oxygen, and the setup of oxygen provision within their own home.

Health care professionals should also ensure that a patient's smoking status is recorded, and that if they smoke, they should be referred to stop smoking services. This is because smoking can accelerate lung function decline,<sup>63</sup> and negate the impact of oxygen therapy.<sup>64</sup>

#### Health care professionals should:

- Ensure that all patients are referred for home oxygen assessments and that oxygen provision is tailored to IPF.
- Ask people with ILD their smoking status and, if they do smoke, refer them to a stop smoking service.

#### v) Palliative care

Palliative care refers to a holistic set of services that are designed for people with complex and terminal conditions. These services offer patients and their family support in managing pain and symptoms, as well as emotional and psychological support. Patients can be referred by their GP, local hospital or specialist centre. This is distinct from end of life care, which refers more specifically to the medical care a person receives during the last phase of their life.

#### We want patients to be referred for individualised palliative care

Although NICE outlines that people should have access to palliative care at any stage, and not just as part of end of life care, referrals are not always made. In 2015, we asked all NHS trusts about their approach to palliative care.<sup>65</sup> Of those that answered, some said



“IPF has made a big difference to my life. I’d always been active and enjoyed exercise. Now I get breathless quickly and very tired. I get so frustrated when I can’t do the things I used to be able to. It’s very hard for my wife and my family. We just take each day as it comes. Without them, I don’t know how I would have coped.”

**Henry from Bridgend living with IPF**

that GPs would make the referral to palliative care; some had palliative care as part of their patient pathway; and some addressed palliative care on a case by case basis.

Many patients have reported to us that they have not even been assessed for palliative care. *Lost in the system* noted that 32% of patients reported that a lung disease specialist had never talked to them about the course their IPF could be expected to take.<sup>66</sup> Some patients interviewed for the report identified a “general lack of understanding of the condition’s progression by GPs and others, and a lack of available services to support patients and their families at the end of life”. It is clear that national bodies need to ensure there is a standardised and clear route to palliative care for all patients. This will ensure all patients can live and die well.

**NICE should:**

- Ensure that the NICE IPF guidelines are updated when the new NICE palliative care guidelines are published in 2018.

**Commissioners should:**

- Ensure that there is a clear, timely and standardised pathway to palliative care for all patients, which includes a holistic needs assessment.

**Providers should:**

- Provide health care professionals with the training, expertise and confidence to offer a holistic needs assessment and discuss palliative and end of life care with all ILD patients with a terminal and progressive condition, as well as their family and carers.

**Health care professionals should:**

- Provide access to palliative care for people with IPF, in line with NICE guidance on IPF, end of life care and palliative care guidelines - due to be published in 2018.
- Offer frequent holistic needs assessments, tailored to individual needs, with referrals to palliative care specialists when necessary. This should happen as early as possible after diagnosis.

## 2) Collaboration and integration

Delivering integrated care for people with ILD requires local and national leadership. Local networks of care should facilitate collaboration. They should ensure all relevant members of the MDT and health systems are included in developing and delivering care pathways. There are four specific points in patients’ pathways where embedding collaborative practices would reap rewards: at the commissioning stage; at diagnosis; in training delivery; and across local networks.

### i) Collaborative commissioning across respiratory services

NHS England identified collaboration in commissioning as a key solution in the *Five Year Forward View* and is trialling this approach in sites across England.<sup>67</sup> However, The King’s Fund research shows integration in commissioning remains rare and tends to be restricted to a small number of service areas.<sup>68</sup> They found that the intensity of

work, negotiation and innovation required to join up commissioning under financial challenges and rising demand are the biggest barriers to achieving collaboration.

#### We need to support collaboration across service provision

Local and national commissioners must be given the time, knowledge, expertise and skills to work together to commission services across people's care journey. This means ensuring that ILD care delivered through specialist centres is integrated with broader respiratory services. Specialist centres only make up part of someone's ILD pathway. Increasing access to services such as long term care, monitoring and support requires a collaborative

approach with local commissioners, NHS area teams and national bodies. Clear lines of accountability need to be established across local pathways.

#### ii) Improving diagnosis

In the BLF's 2015 patient survey, 26% of people reported being initially misdiagnosed.<sup>69</sup> This wastes valuable time that IPF patients do not have. Many people are put on the pathway for COPD for numerous months until a health care professional identifies there has been little improvement. Twenty-three per cent of patients faced 12-24 months of chest symptoms before they reached a specialist team for diagnosis.

#### Case study: Proactive case-finding for IPF – Lambeth and Southwark CCGs

This collaborative approach has been successfully used by Lambeth and Southwark clinical commissioning groups (CCGs), who have used existing COPD registers to case-find people with IPF. Proactive case-finding has been used for many years to identify people at risk of COPD. Case-finding in COPD has been shown to be an efficient way to reach target groups and produce high yields.<sup>71</sup>

In Lambeth, health care professionals are using virtual clinics for primary care to review asthma and COPD registers to discuss patients who have persistent breathlessness and chronic cough. They are developing an algorithm for GPs that will flag up potential ILD for people who present with breathlessness and persistent cough. This case-finding approach uses existing registers and requires relatively low resource.

#### Case study: Proactive case-finding for IPF – breathlessness clinic at Imperial College NHS Trust

A breathlessness clinic has been established by the Imperial College NHS Trust on the same site as the specialist centre. Anyone who presents with breathlessness to their GP or secondary care clinician in the North West London area is referred to this integrated community cardio-respiratory service at Imperial for further tests.

The staff at the clinic have a good awareness of IPF and have been able to actively case-find for IPF among people who have been referred for other conditions. The breathlessness clinic has helped streamline the care pathway by:

- Upskilling staff in both services to be able to identify and diagnose IPF
- Building good relationships between services
- Facilitating informal and formal advice sharing between services
- Enabling priority cases to be identified so they can be allocated more quickly to specialist care.

### Case study: ILD Interdisciplinary Network

The ILD Network started out as an informal group of ILD specialist nurses to provide peer support, ILD educational study days, and shadowing opportunities for newly appointed nurses working in ILD care. Since publication of the NICE guidelines and quality standards, the demands on ILD clinical nurse specialists, specialist pharmacists and allied healthcare professionals with an interest in ILD has increased. In response the ILD Interdisciplinary Network was launched in autumn 2015. The purpose of the group is to provide a supportive network, to promote ILD specialty practice through education and professional development, and to influence the policy of ILD care. They support best clinical practice and the academic advancement of ILD professionals.

The commitment to delivering high quality study days and training events continues. The annual conference in October brings together over 100 ILD clinical and academic nurses and AHPs. This provides an opportunity to support the specialism, increasing knowledge and strengthening the ILD voice. The network now has over 200 members from across the UK and Europe. Find out more: [ild-inn.org.uk](http://ild-inn.org.uk)

### Case study: Wessex ILD Symposium

Across the Wessex region a group of general respiratory and specialist ILD professionals has been established with the aim of improving collaborative working and sharing expertise. The group is comprised of health care professionals from both general hospitals and specialist centres, specialist and community nurses, and radiologists. It meets twice a year for an evening. The group aims to provide a space to socialise and meet peers, as well as share examples of challenging practice. External speakers update the group on wider developments and internal presentations are given to improve knowledge and skills.

As a group, the symposium has successfully rewritten local ILD drug guidelines for the region, and has streamlined the referral pathway and opened access to the MDT; this supports the care of patients with ILD closer to their home by adopting a collaborative region-wide approach.

#### We want all patients to receive early and accurate diagnosis

A collaborative and co-ordinated diagnosis approach from presentation helps ensure other services are quickly brought into a patient's care pathway.

A delayed diagnosis for people with ILD results in lower survival rates, regardless of the severity of the disease or other prognostic factors. Every misdiagnosis also costs our health services money.<sup>70</sup>

#### iii) Training and resources for health care professionals

Training and expertise on ILD often rests with experts who are based in specialist centres. This reflects the rarity of the condition and current service framework.<sup>73</sup> Specialist

centres and teams are largely the local experts in care networks.

#### We want all health care professionals in the MDT and patient's pathway to be informed on ILD care

It is not feasible for specialist centres alone to service large and diverse geographic regions: expertise should be shared across areas. There should be a focus on improving information and training outside of specialist centres - particularly for GPs, commissioners and community care practitioners.

#### iv) Local networks of care

Networks of care should be established to complement existing local plans such as sustainability

and transformation plans (STPs). Only when specialist ILD teams are integrated with other parts of the health system will they be able to fulfil their potential and ensure that patients are receiving high quality care across the system.

In NHS England's *Five Year Forward View* integration is a key way of improving service delivery and design. Integrated "new models of care" are being trialled through 50 partners in "vanguard sites" across the UK.<sup>73</sup> Learnings from these models should be communicated clearly and widely across the UK. The 2017 update on the *Five Year Forward View* acknowledges the need for collaboration even further and emphasises regional co-ordination in service provision. It states that some leading STPs will become Accountable Care Systems. In these areas, more control for specialised commissioning may be given to local decision-makers. Potentially, this could help integrate commissioning for ILD services.

**We want local networks of care to be established to ensure a collaborative and integrated approach is taken to ILD care planning**

Governments should facilitate integrated models of care throughout the UK. Our overall vision is for everyone with ILD to receive the highest quality care in the best settings. For this to happen, local models should be developed in line with the guidelines in this report and local expertise. There should be a clear focus on establishing a whole-system approach, where services are integrated and streamlined. From a patient's perspective, the ILD pathway should provide seamless care and enable and support them to self-manage their condition. By creating these holistic and tailored local networks, areas can ensure

that all members of the MDT are involved with people's care. This should include pharmacists who are often not included or budgeted for in service level agreements.

### Case study: Northern Ireland ILD steering group

In Northern Ireland, trusts have development goals set out on the ILD pathway, and the Public Health Agency provides support to achieve these. These goals are clearly set out in a national respiratory strategy which is helping drive political change.

Additionally, the Public Health Agency in Northern Ireland has established a cross-sector ILD steering group to develop care pathways that work for patients. This steering group comprises patient groups, the government, and health care professionals from Northern Ireland, Scotland, and England. By taking this collaborative approach from the planning stage, the steering group has been able to develop pathways that work for professionals across the sector. It has also meant that planning has been integrated with work that it is happening outside of Northern Ireland.

### Case study: Papworth Hospital and pathways for ILD-CTD

Papworth Hospital has been trialling its own patient pathways which have helped improve engagement across their local network. It has also established cross-working ties with the specialist rheumatology network to improve care and treatment access for patients with connective tissue disease associated with ILD. Despite similarities in clinical and pathologic presentation, the prognosis and treatment of CTD associated ILD (CTD-ILD) can differ greatly from that of other forms of ILD, such as IPF.<sup>74</sup> These nuances in patients' conditions across different ILDs must be reflected in different care pathways.

#### UK governments should:

- Provide resources for specialist centres and local commissioners to collaborate and form local networks of care to deliver high quality care, close to home.
- Establish clear lines of accountability for ILD across the pathway, including outcomes for specialist centres and local commissioners. This will ensure no patients fall through the gaps between commissioning organisations.
- In England, establish a national plan for specialised commissioning that scrutinises existing ILD services and evaluates their effectiveness.

#### Commissioners, NHS area teams and health boards should:

- Lead the establishment of local networks' health services, ensuring that all members of the MDT are involved as well as patients and charities. These networks should develop holistic

care pathways that seek to integrate services and improve access to services and diagnosis.

- Provide training and resources across ILD networks, to upskill professionals outside of specialist centres and facilitate the sharing of expertise across the pathway.

#### Providers should:

- Work with health care professionals, commissioners and local policy makers to establish local networks of care.
- Work collaboratively to develop local care pathways that aim to deliver seamless and integrated care.

#### Health care professionals should:

- Establish local networks of care that seek to share expertise, resources, and training.
- Carry out proactive case finding for ILD through breathlessness clinics.

## 3) Equity of access

Achieving equity in health service provision should be a core consideration when designing ILD care pathways. Local ILD networks should look at designing holistic services that work for local populations and circumstances. Pathways should aim to improve access to MDT diagnosis, treatments and patient-centred support for all patients. 'Equity' refers to a focus on reducing geographical variations in health outcomes and provisions, as well as inequalities of access for different groups within society.

On a national level, reducing variation and achieving standardisation in service provision were key drivers

to the centralisation of specialised services.<sup>75</sup> Yet it is difficult to measure improvements in any of these areas as services have not been centrally audited or recorded. All ILD services should be scrutinised and evaluated to identify where progress has been made for patients and to improve provision across the UK. National governments should establish frameworks for ILD care that support and strengthen local networks to enable more people to access diagnosis, treatment and support outside of specialist centres.

### i) Regional variation

Fragmentation of services and lack of equity in provision are well-

“The primary purpose of the NHS is to improve the outcomes of health care for all: to deliver care that is safer, more effective, and that provides a better experience for patients.”

**NHS England: *Equity and Excellence: Liberating the NHS***

documented for specialist services. In February 2016 in England, only 83% of all specialist services were compliant with service specifications, varying from 74% in the North West and to 95% in the East Midlands.<sup>76</sup> This variation reflects a systemic equity challenge across the health system. It shows that service standards and specifications alone do not always lead to an improvement in outcomes.

A comprehensive list of ILD services across the UK is not publically available. This makes evaluation of equity especially challenging. We have included a list of key ILD centres in this section to demonstrate the spread of services the BLF is aware of, but acknowledge this is by no means a full list.

For conditions where good data is available, service variation is well documented. For instance, there is considerable variation in the number of people receiving chemotherapy for cancer across CCGs.<sup>77</sup> Given that specialised cancer care receives the highest amount of funding from NHS England, and specialised respiratory care is not even in the top ten funded areas, it is likely that these trends are similar or worse for ILD patients.

**We want all patients to receive specialist treatment and care in accessible locations**

When commissioning an ILD specialist centre, local needs, travel times and geography should be taken into consideration to help improve equity. There is often an expectation that patients will travel further for specialised services to access expert care. Travelling

long distances with a condition as debilitating as IPF can become very tiring and overwhelming for patients and their carers.

Research on other conditions has found that centralising services can increase the practical and financial hardship of travel for patients and can be associated with delayed interventions. This is often more acute in less affluent regions.<sup>78</sup> To reduce the impact centralised care may be having on health inequalities, local health leaders should build strong local networks around specialist centres.

Current ILD service frameworks rely on a small number of centres to cater for a diverse range of patients. These centres are essential for ensuring that expert and specialised care is delivered for ILDs. But we also need to look at strengthening services around specialist centres so that other aspects of care can be managed in a way that benefits different patients. Where possible, we should look at shared care between local services and specialist centres.

Table 1: A list of ILD key centres across the UK<sup>79</sup>

England	
Area*	Location of centre
Cheshire and Merseyside	Aintree University Hospital NHS Foundation Trust
East Midlands	University Hospitals Leicester NHS Trust
East Midlands	Nottingham University Hospitals NHS Trust
East of England	Papworth Hospital NHS Foundation Trust
South West	Royal Devon & Exeter Foundation Trust
South West	North Bristol NHS Foundation Trust
London	University College London Hospitals NHS Foundation Trust
London	Royal Brompton & Harefield NHS Foundation Trust
London	London Guy's & St Thomas' NHS Foundation Trust
London	Imperial College Healthcare NHS Trust
Greater Manchester, Lancashire and South Cumbria	University Hospital of South Manchester NHS Foundation Trust
Thames Valley	Oxford University Hospitals NHS Foundation Trust
Wessex	University Hospital Southampton NHS Foundation Trust
Wessex	Portsmouth Hospitals NHS Trust
Yorkshire and Humber	Leeds Teaching Hospitals NHS Trust
Yorkshire and Humber	Sheffield Teaching Hospitals NHS Foundation Trust
Yorkshire and Humber	Hull and East Yorkshire Hospitals NHS Trust
Northern England	Newcastle upon Tyne Hospitals NHS Foundation Trust
West Midlands	University Hospitals Birmingham NHS Foundation Trust
North Midlands	University Hospital of North Midlands NHS Trust
Scotland	
Area*	Location of centre
Scotland	Greater Glasgow & Clyde
Scotland	Grampian
Scotland	Lothian
Wales	
Area*	Location of centre
Wales	Cardiff and Vale University Health Board
Wales	Aneurin Bevan University Health Board
Wales	Abertawe Bro Morgannwg University Health Board
Wales	Betsi Cadwaladr University Health Board
Northern Ireland	
Area*	Location of centre
Northern Ireland	Western Trust
Northern Ireland	Northern Trust

\*Areas are indicative only. Many centres may cover additional surrounding areas depending on local agreements and partnership working.

## ii) Models of service delivery

In other conditions, challenges with service provision and equity have been improved by looking at models of service delivery. In particular, 'hub and spoke' models can offer services and facilities to the wider community as well as those in a specialised centre. This means support can continue to be delivered from a central point over a defined area, but that it can also link up with smaller services to create a larger network that can be both more sustainable and cost-effective.

These models are used for other health conditions including cancer services, stroke services and dental services. Where evidence exists it suggests that these models can:

- Increase numbers of people accessing and engaging in treatment
- Get patients into treatment faster
- Be more cost-effective.

### We need research into the efficacy of different service models for ILD

Most existing models are based around a conventional understanding of a hub and spoke model, where there is one centralised hub and spoke centres supporting it. However, this should be designed and adapted to suit regional needs.

### Case study: Exploring a hub and spoke model for ILD services in the South West

A local network in the South West – including local commissioners, physicians, trusts, ILD specialists, and charities including the BLF – is looking at how to revise their services to meet the demographics of the region. Large numbers of the population live in rural areas and there are high rates of respiratory disease in many of the CCGs. There are currently 11 CCGs with a population of 4.7 million people, spanning over 23,800km<sup>2</sup> and accommodating for over 51,000 respiratory outpatients on a yearly basis. These demographics make it challenging to commission one specialist centre to meet the entire region's needs.

The local network is exploring the use of a hub and spoke model to enable links to be formed and strengthened across the region. This will ensure that hospital and primary care that is currently outside of the specialist network remains supported. It will enable a tailored solution at a local level that could better address the needs of the local population.

They are therefore proposing that a shared care model be used, which will allow management of ILD patients closer to home. Patients will be assessed with standardised protocol, discussed in regional MDT and seen at a specialist centre if/when appropriate. Subsequently individual management plans can be developed for patients and shared care arrangements made with their nearest affiliate centre (or spoke) for follow up.



### iii) ILD specialist nurses

The NICE IPF quality standards state that all “people with IPF should have an ILD specialist nurse available to them” and the ILD service specification supports this by stating ILD specialist nurses should be part of each MDT. ILD nurses play an essential role in helping people co-ordinate their care and understand their condition, yet in our 2015 survey only 39% of people reported they had frequent contact with an ILD nurse and 36% of people said they had no access.

#### We want all ILD patients to have access to a specialist nurse

Research by the Royal College of Nursing (RCN) and National Voices found that patients consistently rated specialist nurses higher than any other health care professionals in understanding patient needs, designing and implementing care pathways, obtaining patient feedback, and being transparent and honest.<sup>80</sup> Further research by the RCN found that specialist nurses reduced waiting times, led to reductions in avoidable hospital admissions, reduced patient drop-out rates, and more.<sup>81</sup> A short study of patient views of ILD specialist nurses found that all of those surveyed (50 ILD patients – 42 with IPF, eight with hypersensitivity pneumonitis and one with sarcoidosis) thought that the specialist nursing service was very important to them, and 90% wanted to be seen at least every three months.<sup>82</sup>

#### We want the role and remit of ILD specialist nurses to be standardised

Across the UK, specialist nurses are often one of the professional groups hardest hit by national and local budget cuts.

There is no standardised definition of an ‘ILD specialist nurse’, which often leads to considerable variation for both practitioners and patients across the UK. National standards would help providers and networks provide effective training and resources. Additionally, it would help standardise the role across regions and workplaces. Standards should be written that embed best practice and ensure the roles are designed to work in the best interest of patients. It would also help create a clear career pathway for practitioners.

#### We need more specialist nurses with ILD training and expertise

ILD nursing is a challenging field that requires a wide range of knowledge and communication skills. ILD patients often have complex needs and co-morbidities, meaning nurses require tailored and extensive training to enable them to deliver optimal care.

Our 2015 survey showed that there are not enough ILD nurses to meet demand. For instance, in Northern Ireland there is only one nurse who is classed as an ILD specialist. We need more specialist nurses with specific training and expertise in ILD care. This will improve the provision of information and support available to families and carers, particularly in managing acute episodes and palliative care. Attendees at our roundtable also felt that more needs to be done to make the ILD specialism an attractive career path. This could be done through tailored training programmes and incentives in regions that need more specialists.

#### UK governments should:

- Support existing ILD nurse networks across the UK with a focus on more training for nurses.

- Write national standards and a career pathway for the role of an ILD specialist nurse.
- Carry out research to establish an optimum ratio of patients to ILD specialist nurses.
- Publish a list of ILD specialist centres, to increase transparency and give patients more choice about where they are treated.
- Commission research into hub and spoke models for ILD service provision.

#### Local networks should:

- Publish local pathways and specialist centres.

#### Commissioners and providers should:

- Deliver training programmes for ILD specialist nurses and establish regional standards for career development.

### iv) Multi-disciplinary team

The MDT approach for accurately diagnosing and managing individuals with ILDs is considered the 'gold standard'.<sup>83</sup> It is the first call in the BLF IPF Patient Charter.<sup>84</sup> NICE provides guidance on the minimum composition of a MDT involved in diagnosing IPF.<sup>85</sup> This varies depending on diagnosis method, but typically will involve a consultant

respiratory physician, a consultant radiologist, an ILD specialist nurse, and a MDT team co-ordinator.

A single-centre retrospective five year review in the UK found that a MDT approach can establish a diagnosis in 76% of cases when prior diagnosis is uncertain.<sup>86</sup> This review also found that a prior diagnosis of IPF is deemed inaccurate in over 50% of cases after MDT discussion, and showed that a MDT approach can establish a diagnosis without lung biopsy. MDTs play an important role in management decisions relating to a patient's care, as they can determine which interventions are most appropriate.

A UK study in 2016 surveyed all 20 NHS England commissioned ILD centres, plus nine specialist centres in Scotland, Wales and Northern Ireland. They found considerable differences in workforce composition and frequency of MDT meetings. In 57% of centres, MDTs were co-ordinated by the ILD lead consultant, and in 26% by a medical secretary. Only 17% were directed by a MDT co-ordinator. In 78% of centres peripheral hospitals participated in MDTs, in person, via video-link, or through paper referrals. However, the majority of MDT cases were discussed and reviewed at the specialist centre.<sup>87</sup>

### Case study: Service model and MDT approach in South Wales

The service operates a network approach to the care of ILD patients. Each health board in South Wales has an ILD lead and most units also have a local ILD MDT with a local radiologist. There is a weekly ILD MDT based in Cardiff which is video-linked to local ILD leads. In addition, there is a monthly tertiary ILD clinic for patients who would benefit from a central face to face evaluation.

This model has helped facilitate timely access to specialist ILD MDT discussion. The average time from local referral to central MDT discussion is less than three weeks. Video links with local MDT leads enables them to be more involved in the diagnostic process. This model has also helped facilitate education and consistency across the region. Prescription of high-cost therapy remains with the local ILD teams, allowing the development of local expertise in managing anti-fibrotic therapy. In turn, this has developed a good network of regional ILD leads who are able to share best practice and expertise across the pathway.

### All patients should have timely and accurate access to MDT diagnosis

The BTS ILD registry finds that the average time for IPF patients, from referral to first clinic appointment, is 7-10 weeks.<sup>88</sup> IPF has a prognosis worse than many cancers, so access to MDT diagnosis and services should be fast-tracked in line with standards that have been set for cancer patients.

### National governments should support and invest in digital solutions for IPF care, particularly for use in virtual MDTs

Some centres in the UK have begun to trial virtual MDTs. These are not well defined as a

concept, but generally involve the use of technology to bring together clinicians from various different locations.<sup>89</sup> This helps address concerns regarding the time and cost involved in bringing together an MDT.

### Virtual appointments should be explored as a way to improve access to services

Virtual appointments could be used where possible to improve people's ability to access services. Many IPF patients struggle to travel because of breathlessness and/or oxygen equipment, therefore in places where this approach is being trialled it has tended to be a popular solution.

## Case study: Imperial College Healthcare NHS Trust – digital solutions and telemedicine

Imperial College Healthcare NHS Trust has around 600 ILD patients on their caseload across the North West London region. The centre is trialling the use of telemedicine with their patients through the use of an online patient hub. The hub has been designed to meet all NHS data requirements, the bespoke needs of the centre and patients. It acts as a portal where patients are able to access all their medical records, health advice, a shared calendar and self-management advice. This is aligned with patients' self-management plans and can be linked up with other smart devices, such as fitbits. Through the portal, patients are

able to directly contact a specialist at the centre; and vice-versa, clinicians are able to communicate with patients. This communication can be done through a secure video link or through messages.

The portal provides patients with a highly personalised experience. It ensures that all health care professionals are able to access a patient's clinical background. It has made it easier for district general hospitals to access specialist information and enable the safe sharing of information ahead of MDT discussions. The data collected on the portal will also help supplement academic research in this area. The centre plans to use the portal to carry out video conferencing and safe sharing of patient records for virtual MDT discussion.

### Trusts should receive enough reimbursement to cover the cost of MDTs and collaborative working

Payment mechanisms for ILD care should be amended to reward good outcomes and to increase incentives for effective MDT provision. NHS England identified improvements to the payment system as a critical area of focus for successful delivery of the *Five Year Forward View*.<sup>90</sup>

In a 2016 survey of ILD specialist centres, all respondents agreed that the available MDT time was insufficient. The most common reasons were cited as: lack of dedicated MDT funding (83%); lack of sufficient respiratory radiologist consultant time (78%); and lack of dedicated administrative support (61%). In 96% of cases there is no local tariff in place to fund MDT

discussion and all respondents agreed that a dedicated tariff would improve MDT provision.<sup>91</sup>

These findings are echoed in a 2014 study which found that the current NHS tariff for IPF services is not sufficient to meet the standards in the service specification. The estimated average cost per patient for first episodes of diagnosis, management and monitoring was £1,384, which is approximately £408 (42%) more than is reimbursed by the tariff. The study found that the cost of the NICE and service specification pathway was approximately £477 (41%) more than is reimbursed by the tariff. On average, the costs of one diagnosis, management and monitoring episode in line with the requirements in the NICE guidelines and ILD service specification are £245 more than the tariff received by specialist centres. This is largely because the tariff does not account for the staff time that a MDT requires.<sup>92</sup>

Trusts are operating within increasingly tight budgets - in 2015-16, 88% of NHS trusts opted for a tariff arrangement that included a 3.5% efficiency saving. The National Audit Office found that the current funding arrangements for specialised services are an obstacle for transparent reporting and lead to variations in prices across the country. In particular, the national tariff in its current form does not provide payment relating to costs of co-ordination or incentives for innovation in care.<sup>93</sup>

**We want payment mechanisms to be amended to improve ILD patient outcomes, including the national tariff and CQUINs**

In some cases, patients are being forced to travel to centres in order for the centre to receive payment for the MDT. Trusts should be reimbursed for each patient MDT meeting, irrespective of where it is held. Tariffs should be designed to achieve the best results for patients. These amends would enable more professionals to explore the use of virtual MDTs. In comparison, in cystic fibrosis (CF) care, a separate currency has been designed to support specialist CF MDTs. It aims to provide care in a seamless and patient-centred way.<sup>94</sup>

Additionally, the existing Commissioning for Quality and Innovation (CQUINs) payments framework does little to incentivise providers to work across ILD patient pathways. For other conditions these targets have been established. They largely aim to improve patient care, incentivise best practice models, enable collaborative working, and improve transparency and regional variation. For instance, for auto-immune conditions, a target exists to support the development of co-ordinated MDTs and better data collection. Such incentives should be explored for ILD care. A CQUIN target around increased access for IPF patients to the MDT and better data collection could help tackle systemic challenges in the current framework.

#### UK governments should:

- Amend the tariff and specialised top-up to cover the full cost of MDTs.
- Amend the tariff to incentivise patient-centred approaches, such as virtual MDTs.
- Establish a CQUIN target for ILD care to improve MDT access, and incentivise best practice and collaboration.
- Optimise the use of technology to help improve timely communication, diagnosis, and outcomes.
- Work with providers to improve ease of patient access when deciding where to locate specialist centres and consider establishing 'sub-hubs' where centres cover particularly large geographical areas or areas with poor transport.

#### Commissioners should:

- Comply with NICE guidelines and establish sufficient workforce capacity to ensure that patients with ILD have access to a MDT.



“I’m going to be 50 next year. I was hoping to go away with my son to Florida but I can’t now because it’s so hard to get oxygen on a plane. My son is 15, I don’t think he’s really absorbed what’s happening to me. I work part-time and try to carry on as normal. It’s difficult. My diagnosis has been very stressful. My nails and my hair were my pride and joy – now my nails have started to club and I’m losing my hair from the stress. I’m lucky I qualify for support. Having things like a blue badge has helped me get my life back.”

**Fiona from Falkirk living with IPF**

## 4) Patient-centred communication and care

Patient-centred communication and care along a patient’s care pathway are absolutely vital to ensure that patients and their family and carers have a positive experience of care.

Although there is no single definition for patient-centred care, a set of key principles identified by the Health Foundation should help local networks shape and deliver ILD care. These are:

- Affording people dignity, compassion and respect
- Offering coordinated care, support or treatment
- Offering personalised care, support or treatment
- Supporting people to recognise and develop their own strengths and abilities to enable them to live an independent and fulfilling life.<sup>95</sup>

Structured communication between health care professionals and patients is one aspect of patient-centred care that is vital to ensure people receive the best possible support and treatment.

However, effective communication between patients and care professionals is not always carried out satisfactorily.<sup>96</sup> This is despite the provision of high quality Information Standard accredited information produced by the BLF. Poor communication can have a significant negative effect on patients with IPF, who face a “relentless and unpredictable” progression of their disease.<sup>97</sup> Studies on communication with IPF patients have shown as a consistent theme the need for more information and disclosure with

patients.<sup>98</sup> Many patients and carers reported difficulty in translating health information and knowledge of IPF to their own disease pathway.<sup>99</sup> They did not feel as informed about psychological and physical treatment paths. These studies all confirm the importance of a holistic approach to care that focuses on optimising patients’ quality of life.

### i) Provision of information and support

The provision of clear, concise, and easy to read information and support to a patient on their diagnosis, prognosis, and management is vital. This helps ensure that they are kept informed and in the best possible control of their condition, while also reducing feelings of anxiety and hopelessness.

Patient information can be provided in writing – through leaflets, information packs, webpages, and other means, or in person – delivered over the phone or face-to-face.

**Health care professionals should have open, honest and informed conversations with patients about their prognosis and condition**

*Lost in the system* identified that only 51% of patients had their condition explained in a way they could easily understand.<sup>100</sup> A further 22% surveyed as part of the report found the verbal information about IPF difficult to understand, and around 25% said that they did not have their condition explained at all.<sup>101</sup> This contributed to considerable anxiety and patients feeling that they were being left behind.

“We knew it was terminal. I’d seen articles online putting life expectancy at 3-5 years. Mum was told she had months to live – but when she came out of the hospital she started feeling much better. She wasn’t. Mum passed away just before Christmas. That was only 7 months after she was diagnosed. She was 56. There’s so much to process with IPF. When people say cancer you know what that means. They told us her ‘lungs were scarred’. We just didn’t know what to expect.”

**Charlotte, daughter of Pauline from Stovehaven**

**We want health care professionals to be able to have an open discussion on a patient’s care journey**

Open discussion needs to be a central theme in a patient’s care journey. From diagnosis onwards, careful discussion about patient and carer needs should take place, plus time and information should be provided to allow patients to digest this. The success of interventions will depend on patient factors such as expectations, experiences and motivations.<sup>102</sup> Family caregivers are the main support for the patient so should be included from the outset.<sup>103</sup> Decisions on a patient’s care must be made with the involvement, consent and understanding of the health care professional as well as the patient and their family and carers.

This requires health care professionals to be provided with training on how to communicate effectively and avoid further distress for the patient. This training should be similar to the SAGE and THYME model which is currently being deployed in cancer care.

**We want patients to be provided with Information Standard accredited written information about their condition**

*Lost in the system* identified that 49% of people who responded to our survey were not given any written information about their condition.

The poor provision of written information on IPF by health care professionals is increasingly unacceptable as the body of patient literature grows. Since 2013, the BLF has produced booklets and online information on ILD, providing the most comprehensive, Information Standard accredited, information on ILD in the UK. The Information

Standard is an NHS England led scheme to ensure high quality patient information. This means it has been peer reviewed by both health care professionals and patients. The BLF’s IPF information hub was Highly Commended at the British Medical Association (BMA) Patient Information Awards 2015.

Many more patients and their families are accessing high quality written information; however there is still more work to be done.

**Providers should:**

- Ensure that ILD specialist nurses are available to have face-to-face discussions with patients on their condition at any point throughout their care journey.
- Ensure that pathways are in place for non-respiratory specialists to refer patients with ILD to an ILD specialist nurse for a discussion on their condition.
- Ensure that all patients are offered appropriate Information Standard accredited information on ILD at diagnosis, and at times throughout their care journey.

**Health care professionals should:**

- Ensure that decisions on a patient’s care are made with the involvement, consent, and understanding of the patient and their family or carers.
- Provide patients with hard copies of Information Standard written information, or direct them to written information online.

## ii) Peer support

Access to dedicated peer support networks, both for patients and their carers, in person or digitally, is one of the core elements of the BLF IPF Patient Charter. This was launched in 2014 and developed

by the BLF in co-ordination with the Pulmonary Fibrosis Trust and Action for Pulmonary Fibrosis.

Peer support is available in the UK through over 230 Breathe Easy support groups, which are attended by 6,000 people annually.<sup>104</sup> These groups cover a range of conditions – including ILD – and provide a chance for people living with a lung condition to socialise and discuss disease management. Some of these groups are integrated into a patient's care pathway and have health care professional involvement. Both types of group have been assessed as cost-effective and produce positive outcomes in terms of self-efficacy, health outcomes and cost savings.<sup>105</sup>

Other support groups are much more specialised, and only provide support for people with an ILD, or for people more specifically with IPF. There are over 40 of these groups across the UK, which are organised and run by health care professionals and patients. Like Breathe Easy groups, ILD and IPF support groups take place on a regular basis and can include health care professionals as speakers.

#### We want all patients to be aware of, and have access to, group peer support

Although the number of peer support groups has increased, some patients do not have local access to a peer support group or are not aware of their local group. An online poll by the BLF of its web community members identified that of 433 respondents, 18.7% did not have a group in their area and 24.7% had not heard of Breathe Easy groups.<sup>106</sup> Patients should receive clear information about, and signposting to, local groups.

### Case study: Wessex ILD patient support group (WILD)

The WILD support group was set up in September 2014 for patients and carers in Southampton and Portsmouth, in partnership with Southampton General Hospital and Queen Alexandra Hospital, Portsmouth. WILD aims to provide information, support, collaboration, and a voice to patients with ILD. The group was organised by doctors, physios, ILD nurses, Southampton bio-medical research unit, and the University of Southampton department of clinical psychology.

#### Patient-led agenda

The group is attended by 50-60 people – about half are ILD patients and half are carers. Meetings are held quarterly for 3 hours. In between each support group meeting, a steering group of patients, carers, and health care professionals meet to discuss the ongoing structure and agenda for the support group. This is based on feedback from attendees and discussion at the meetings.

#### Breakout sessions for carers and patients

Each group meeting features a 45-minute session where carers and patients go into separate breakout groups. These breakout sessions are facilitated by a psychologist who is not an ILD professional or WILD organiser. This aims to encourage open and honest discussion between patients. Patients are given the option not to partake but, so far, all patients attend and participate in these discussions.

#### Access to information and support through peer and professionals

Initial findings from the group suggest that patients find the support group particularly beneficial in accessing information and social support from WILD members and health care professionals (HCPs). This was particularly important for newly diagnosed ILD patients who were able to access information and opinions about treatment, self-care and symptoms. The combination of both peer support and professional guidance has been emphasised by many attendees.



**Commissioners should:**

- Integrate existing patient support groups into local care pathways.

**Health care professionals should:**

- Work with patients and charities to establish new peer support groups.
- Signpost patients to peer support groups following diagnosis.

**iii) Self-management**

Self-management refers to actions taken by patients to recognise, treat, and manage their own health. Patients must be supported by health care professionals through the development of self-management plans, goal setting, education, ad-hoc support, and general case management.

For people with IPF, self-management could include:

- Monitoring symptoms on a day-to-day basis, including breathlessness and cough
- Taking action to reduce the impact of these symptoms, through methods such as daily exercise, avoiding tobacco smoke, and planning activities in advance
- Ensuring awareness of what to do if these symptoms get worse
- Support on managing the side-effects of treatments.

**We want widespread self-management support for patients with ILD across the UK**

Anecdotal evidence suggests that self-management support is not being implemented in routine clinical practice for patients with IPF. A 2012 survey of respiratory health care professionals in London found that although support for self-management is strong, it was only discussed with around half

of patients who had asthma or COPD, with a similar proportion receiving a written action plan.<sup>107</sup>

The survey found that common barriers to the implementation of self-management support in routine clinical practice include time constraints, lack of training, lack of belief in patient's ability to self-manage, and lack of confidence in completing self-management plans.<sup>108</sup>

Tools such as the BLF's pulmonary fibrosis personal organiser, which helps patients record notes on their condition, track their care, and know what questions to ask their health care professional, are useful in helping patients feel more in control of their condition. This was recently piloted by the BLF with excellent feedback, and is now available online.<sup>109</sup>

**We want further research on the effectiveness of the self-management of ILDs**

There has been no formal research on self-management among patients with ILD. This includes the effectiveness of self-management, the perceptions and challenges of self-management among patients and health care professionals, and the provision of self-management plans. UK-based research covering this data could go some way to informing service improvement, through providing health care professionals with information on what is working with current approaches and what is not, and supporting commissioners to invest in ensuring that health care professionals are well-equipped to support self-management.

**Commissioners and providers should:**

- Ensure that health care professionals have the training and support to offer patients self-management plans and support their use.
- Ensure patient representatives are included in all planning and pathway development.
- Signpost patients to peer support groups following diagnosis.
- Support the development of self-management plans and guidance that are easy to read, promote behaviour change, and provide information on managing the condition and treatment side effects.

**Health care professionals should:**

- Provide advice, support, and counselling on living with a terminal condition.

## 5) Transparent data and information

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Effective and transparent sharing of data and information is fundamental in ensuring successful communication within local networks of care. This involves robust collection by health care professionals and data systems which allow sharing. This will help support a patient's transition along their care pathway.

Concerns around the collection and sharing of patient data and information are long-running and constitute a systemic challenge in our health systems. These concerns are not restricted to ILD – they negatively impact on most health services across the UK.

NHS England's *Five Year Forward View* called the "information revolution" one of "the three major economic transitions in human history", but noted that there has been slow progress in capitalising on this to improve patient care.<sup>110</sup> The *Five Year Forward View* stressed that data and information can act as the "electronic glue" that enables parts of the NHS to work together.

Improvements to the collection and sharing of data have the potential to lead to better outcomes for patients with ILD by ensuring a patient's seamless progression through their care pathway.

### i) Data collection and coding

Data already collected through the BTS ILD Registry – launched in February 2013 – is being used to facilitate the sharing of best practice, benchmarking against NICE quality standards, and clinical audit, allowing centres to monitor and improve standards of care.<sup>111</sup> It helps clinicians improve services, and was viewed by contributors at the roundtable to be a very useful recent innovation that has helped address the previous lack of data.

The registry is still developing as a resource, with a growing number of sites participating (approximately 40) as of February 2017, with almost 1,000 IPF patient records.<sup>112</sup> It does not yet cover Northern Ireland.

We want further resources and incentives to ensure data on the diagnosis of ILD and treatment offered can be recorded in a digital, standardised manner

There is a lack of data on the number of people diagnosed with IPF with a multi-disciplinary team, people's disease progression, management plans, and other areas. There are no mandatory requirements for ILD data to be collected. There are also no financial incentives to encourage better ILD patient data recording, meaning that resource-limited staff are more likely to prioritise other activities. This makes it harder to identify trends, build an evidence base for commissioners to commission effective services, and plan patient care. Inputting ILD data should be mandatory.

Issues around data recording are common across health services in the UK, particularly with respiratory conditions. The most comprehensive epidemiological report on lung disease in the UK – *The Battle for Breath* by the BLF – identified at least eight different codes in primary care for IPF.<sup>113</sup> Issues around data collection were also identified in the APPG on Respiratory Health's inquiry into respiratory deaths.<sup>114</sup>

Steps should be taken to better enable data entry. This should involve material support being given to providers for there to be a sufficient number of administrative staff in the workforce to enter data. Governments should consider financially incentivising providers to record ILD patient data.

**UK governments should:**

- Provide support to the BTS to develop the current BTS ILD Registry and ensure it

is mandatory (with opt-out exceptions), with material support for health care professionals to complete.

**Providers and commissioners should:**

- Ensure that trusts and health boards with existing data collection capacity participate in the BTS ILD Registry. Those that do not should take steps to ensure that they can in the future.
- Ensure that health care professionals are mandated and have the time to enter patient data onto a central registry.

**Health care professionals should:**

Ensure that diagnostic and treatment data on IPF and sarcoidosis is recorded on the BTS ILD Registry.

## ii) Data, information and knowledge sharing

Sharing patient data across local networks is often recognised as a way of speeding up treatment and care, while also reducing the duplication of activities.<sup>115</sup> The relative rarity of IPF means that care is often delivered by different providers, and this means that data sharing is even more pertinent.

We want an improved framework for sharing patient information between clinicians

We learnt from attendees at the roundtable that the sharing of patient data across NHS organisations is not always straightforward, and that this can lead to delays in treatment, as well as patients being unable to access specialist services. For example, patients who receive treatment from one NHS trust might not have their records accessed by clinicians in another NHS trust, and in turn may not be prescribed anti-fibrotics when they need them. There

were also issues highlighted around sharing data with general practice, with scope for improvement.

#### UK governments should:

- Explore mechanisms to support sharing ILD patient data and information, including using existing innovations such as the BTS ILD Registry.

#### Providers should:

- Require MDTs to produce a document for GPs summarising a patient's diagnosis, treatment, and care, with guidance for GPs on aspects of care that will require further monitoring.

### Case study: Heart of England NHS Foundation Trust

Wye Valley NHS Trust in Hereford employs an ILD specialist nurse and provides care for patients with IPF, but the MDT that plans treatments and determines individual patient care pathways is based in Heart of England NHS Foundation Trust. The distance between the two and the lack of framework for sharing patient data meant that many patients either did not receive the best quality care, or there were delays in treating them.

Both NHS trusts set up data sharing arrangements. This involved using Dendrite Clinical Systems as a pro-forma

for MDTs. Although this took a year to implement, it allowed Wye Valley to refer patients to Heart of England for the MDT to determine the care pathway, before the patients were redirected to Wye Valley. Consultants at Heart of England would see patients each year and provide anti-fibrotics if necessary. Heart of England report that most patients have a positive experience of care following these changes.

Several future changes were identified as necessary to help enhance the process further. This includes rolling out Dendrite Clinical Systems to other hospitals in the West Midlands, although data protection concerns are an obstacle. Better data sharing with GPs was also identified as a challenge.

### iii) Specialist centres

The relative rarity of some ILDs means that treatment is often delivered by dedicated, specialist centres which have the necessary levels of expertise and MDT workforce. These centres exist across the UK, split on a geographical level. Research by the BLF found that there are 29 specialist centres, the vast majority of which have a MDT/ILD specialist nurse and local pulmonary fibrosis support group nearby.<sup>116</sup>

#### We need more transparency on which ILD specialist centres exist in the UK

There is no comprehensive list of ILD specialist centres across the

UK. This has implications for both patients and clinicians – both of whom as a result could be less aware of which services are available in their local area. An accessible and informative list of ILD centres would improve evaluation and measurement of service provision.

#### UK governments should:

- Publish a full list of specialist centres, including whether there is a MDT located on site, whether this includes an ILD specialist nurse, and whether there is a local pulmonary fibrosis support group nearby.

# What needs to happen?

## National governments and government bodies

Establish service frameworks for ILD care that support, provide and standardise care and improve existing frameworks.

Establish taskforces for lung health in Scotland and England to write comprehensive five year respiratory strategies. Existing strategies in Wales and Northern Ireland should be reviewed and improved upon.

Evaluate ILD services to determine their effectiveness for patients and for governments. These results should be published and made publicly available.

In Northern Ireland, the current respiratory strategy should be built upon to develop and support ILD pathways.

In Scotland, a service framework – including care and service guidelines – urgently needs to be put in place.

Scottish Government should set clear and ambitious evaluation and outcome targets in a new service framework for ILD care.

In Wales and Northern Ireland, existing respiratory plans should include ambitious targets and evaluation for ILD care and services.

In England, the current ILD service specification should be extended to cover care outside of specialist centres. It should seek to strengthen local networks and incentivise the creation of care pathways.

In Wales, the next Respiratory Health Delivery Plan should support and strengthen ILD service pathways.

NHS England should establish a strategy for specialised commissioning that sets evaluation and transparency measures for ILD care.

## National governments and government bodies

Tailored interventions			
<i>Pulmonary rehabilitation</i>	<i>Access to drugs</i>	<i>Palliative care</i>	<i>Lung transplantation</i>
<ul style="list-style-type: none"> <li>• Provide researchers with access to monitor pulmonary rehabilitation for people with ILD, to enable research to assess:               <ul style="list-style-type: none"> <li>• Which patients with IPF are most likely to benefit from pulmonary rehabilitation, to what extent they can benefit, and which metrics are most likely to see improvements</li> <li>• What the long-term benefits of pulmonary rehabilitation are for people with ILD</li> <li>• How pulmonary rehabilitation can be tailored to improve outcomes for people with ILD</li> </ul> </li> </ul>	<ul style="list-style-type: none"> <li>• Amend the criteria for prescribing pirfenidone (Esbriet) and nintedanib (Ofev) to ensure that anyone who can benefit from them has access to them</li> </ul>	<ul style="list-style-type: none"> <li>• Ensure that the NICE IPF guidelines are updated when the new NICE palliative care guidelines are published in 2018</li> </ul>	<ul style="list-style-type: none"> <li>• Establish a national allocation system for lung transplantation that prioritises patients who are most suitable for transplants and also require them the most</li> <li>• Ensure clinicians have access to the best scientific evidence on lung transplantation to enhance practice and improve utilisation rates</li> </ul>
<b>Collaboration and integration</b>	<b>Patient-centred communication and care</b>	<b>Equity of access</b>	<b>Information, data and transparency</b>
<ul style="list-style-type: none"> <li>• Provide resources for specialist centres and local commissioners to collaborate and form local networks of care to deliver high quality care, close to home</li> <li>• Establish clear lines of accountability for ILD across the pathway, including outcomes for specialist centres and local commissioners. This will ensure no patients fall through the gaps between commissioning organisations</li> <li>• In England, establish a national plan for specialised commissioning that scrutinises existing ILD services and evaluates their effectiveness</li> </ul>	<ul style="list-style-type: none"> <li>• Integrate existing patient support groups into local care pathways</li> </ul>	<ul style="list-style-type: none"> <li>• Support existing ILD nurse networks across the UK with a focus on more training for nurses</li> <li>• Write national standards and a career pathway for the role of an ILD specialist nurse</li> <li>• Carry out research to establish an optimum ratio of patients to ILD specialist nurses</li> <li>• Publish a list of ILD specialist centres, to increase transparency and give patients more choice on where they are treated</li> <li>• Commission research into hub and spoke models for ILD service provision</li> <li>• Amend the tariff and specialised top-up to cover the full cost of MDTs</li> <li>• Amend the tariff to incentivise patient-centred approaches, such as virtual MDTs</li> <li>• Establish a CQUIN target for ILD care to improve MDT access, and incentivise best practice and collaboration</li> </ul>	<ul style="list-style-type: none"> <li>• Provide support to the BTS to develop the current BTS ILD Registry and help make it a mandatory registry.</li> <li>• Explore mechanisms to support sharing ILD patient data and information, including using existing innovations such as the BTS ILD Registry</li> <li>• Publish a full list of specialist centres, including whether there is a MDT located on site, whether this includes an ILD specialist nurse, and whether there is a local pulmonary fibrosis support group nearby</li> </ul>

## Local commissioners, local NHS England teams and health boards

These people should have primary responsibility for leading on the establishment of local networks and ensuring that ILD services are integrated with other respiratory, health, and social care services.

Tailored interventions			
<i>General</i>	<i>Pulmonary rehabilitation</i>	<i>Palliative care</i>	
<ul style="list-style-type: none"> <li>Tailor services for ILD with sufficient capacity, resources, and expertise, in line with NICE guidance. This should include pulmonary rehabilitation and the provision of oxygen</li> </ul>	<ul style="list-style-type: none"> <li>Ensure that health care professionals are resourced and able to refer patients with ILD for an assessment for pulmonary rehabilitation</li> </ul>	<ul style="list-style-type: none"> <li>Ensure that there is a clear, timely, and standardised pathway to palliative care for all patients, which includes a holistic needs assessment</li> </ul>	
Collaboration and integration	Patient-centred communication and care	Equity of access	Information, data and transparency
<ul style="list-style-type: none"> <li>Lead the establishment of local networks health services, ensuring that all members of the MDT are involved as well as patients and charities. These networks should develop holistic care pathways that seek to integrate services and improve access to services and diagnosis</li> <li>Ensure that enough services and places exist on tailored IPF programmes</li> </ul>	<ul style="list-style-type: none"> <li>Work with providers to ensure that health care professionals have the training and support to offer patients self-management plans and support their use</li> <li>Integrate existing patient support groups into local care pathways</li> <li>Work with commissioners to ensure patient representatives are included in all planning and pathway development</li> </ul>	<ul style="list-style-type: none"> <li>Work with providers to improve ease of patient access when deciding where to locate specialist centres and consider establishing 'sub-hubs' where centres cover particularly large geographical areas or areas with poor transport</li> <li>Work with providers to deliver training programmes for ILD specialist nurses and establish regional standards for career development</li> <li>Comply with NICE guidelines and ensure that there is sufficient workforce capacity to ensure that patients with ILD have access to a MDT</li> <li>Optimise the use of technology to help improve timely communication, diagnosis and outcomes</li> </ul>	<ul style="list-style-type: none"> <li>Work with providers to ensure that trusts and health boards with existing data collection capacity participate in the BTS ILD Registry. Those that do not should take steps to ensure that they can in the future</li> <li>Work with providers to ensure that health care professionals are mandated and have the time to enter patient data onto a central registry</li> </ul>

## Providers

Tailored interventions	Collaboration and integration	Patient-centred communication and care	Equity of access	Information, data and transparency
<i>Palliative care</i>				
<ul style="list-style-type: none"> <li>Provide health care professionals with the training, expertise, and confidence to provide a holistic needs assessment and discuss palliative and end of life care with all patients with a terminal and progressive condition, as well as their family and carers</li> </ul>	<ul style="list-style-type: none"> <li>Work with health care professionals, commissioners, and local policy makers to establish local networks of care</li> <li>Work collaboratively to develop local care pathways that aim to deliver seamless and integrated care</li> </ul>	<ul style="list-style-type: none"> <li>Ensure that ILD specialist nurses are available to have face-to-face discussions with patients on their condition at any point throughout their care journey</li> <li>Ensure that pathways are in place for non-respiratory specialists to refer patients with ILD to an ILD specialist nurse for a discussion on their condition</li> <li>Ensure that all patients are offered appropriate Information Standard accredited information on ILD at diagnosis, and at times throughout their care journey</li> <li>Work with commissioners to ensure that health care professionals have the training and support to offer patients self-management plans and support their use</li> <li>Work with commissioners to ensure patient representatives are included in all planning and pathway development</li> </ul>	<ul style="list-style-type: none"> <li>Work with commissioners to deliver training programmes for ILD specialist nurses and establish regional standards for career development</li> <li>Work with commissioners to improve ease of patient access when deciding where to locate specialist centres and consider establishing 'sub-hubs' where centres cover particularly large geographical areas or areas with poor transport</li> </ul>	<ul style="list-style-type: none"> <li>Work with commissioners to ensure that trusts and health boards with existing data collection capacity participate in the BTS ILD Registry. Those that do not should take steps to ensure that they can in the future</li> <li>Work with commissioners to ensure that health care professionals are mandated and have the time and resources to enter patient data onto a central registry</li> <li>Require MDTs to produce a document for GPs summarising a patient's diagnosis, treatment, and care, with guidance for GPs on aspects of care which will require further monitoring</li> </ul>



## Healthcare professionals

Tailored interventions				
<i>Pulmonary rehabilitation</i>	<i>Access to drugs</i>	<i>Oxygen therapy</i>	<i>Palliative care</i>	<i>Lung transplantation</i>
<ul style="list-style-type: none"> <li>In line with NICE guidance, ensure that patients are aware of the benefits of pulmonary rehabilitation, both in terms of education and improvements to physical function, and refer them for an assessment</li> <li>Ensure that ILD patients are referred for an assessment for pulmonary rehabilitation</li> </ul>	<ul style="list-style-type: none"> <li>Ensure that patients who can benefit from drugs have access to them</li> <li>Discuss and agree the most appropriate drugs to use throughout a patient's care journey</li> <li>Give patients information on clinical trials</li> </ul>	<ul style="list-style-type: none"> <li>Ensure that all patients are referred for home oxygen assessments and that oxygen provision is tailored to IPF</li> <li>Ask people with ILD their smoking status and, if they do smoke, refer them to a stop smoking service</li> </ul>	<ul style="list-style-type: none"> <li>Offer frequent holistic needs assessments, tailored to individual needs, with referrals to palliative care specialists when necessary. This should happen as early as possible after diagnosis</li> </ul>	<ul style="list-style-type: none"> <li>Discuss lung transplantations with patients within six months of diagnosis, in line with NICE guidance</li> </ul>
Collaboration and integration		Patient-centred communication and care		Information, data and transparency
<ul style="list-style-type: none"> <li>Establish local networks of care that seek to share expertise, resources, and training</li> <li>Carry out proactive case finding for ILD through breathlessness clinics</li> </ul>		<ul style="list-style-type: none"> <li>Ensure that decisions on a patient's care are made with the involvement, consent, and understanding of the patient and their family or carers</li> <li>Provide patients with hard copies of Information Standard written information, or direct them to equivalent information online</li> <li>Signpost patients to peer support groups following diagnosis</li> <li>Work with patients and charities to establish new peer support groups</li> <li>Provide advice, support, and counselling on living with a terminal condition</li> <li>Support the development of self-management plans and guidance that are easy to read, promote behaviour change, and provide information on managing the condition and treatment side effects</li> </ul>		<ul style="list-style-type: none"> <li>Ensure that diagnostic and treatment data on IPF and sarcoidosis is recorded on the BTS ILD Registry</li> </ul>

# Final thoughts

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Health systems across the UK are facing considerable financial pressures and the challenges of a rapidly ageing population. People are living longer, with more long-term conditions and complex care needs. Lung disease plays a major role in this and is only likely to increase. It is therefore vital that UK policy makers and health leaders prioritise respiratory care across the board and put in place robust policies to improve outcomes. We need a taskforce for lung health in both England and Scotland to develop a five year strategy to improve respiratory services and the nation's lung health.

The radical system changes we have seen in health care over the last five years should be used as an opportunity to embed patient-centred care and deliver excellence in service provision for ILD. A dedicated network of experts and professionals are carrying out exemplary care for people with ILD across the UK. This needs to be rolled out across all regions so that every person with ILD is able to access consistent, transparent, and effective services. Clear pathways for people with ILD must be established to create a seamless and integrated system of care.



# Appendix

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## How are ILD services commissioned across the UK?

Health is a devolved responsibility in the UK, leading to differing commissioning arrangements for ILD services in each of the four nations. An overview of these commissioning arrangements can be found below. This only looks at services which are formally designated ILD services – in practice these services are likely to form only part of a patient’s care pathway.

### England

NHS England is responsible for commissioning ILD services in England, with support from a clinical reference group (CRG) – a group of clinicians, commissioners, patients, and carers – that provide input into policy development for all specialised respiratory services. The BLF is a member of this CRG.

As ILD is less common than other conditions, it is commissioned as a ‘specialised service’. This means that specialist ILD services are not routinely provided by all acute settings, but only by relatively few hospitals – designated as ILD centres – spread across the country. These centres provide care through expert clinicians, typically a MDT which works in partnership with local hospitals and GPs to deliver care. These centres are intended to provide regular, easy to access support to people with ILD, while ensuring that ILD clinical specialists are clustered in a way that supports their work.

Specialised services can be expensive to provide, and account for around 14% of the annual NHS budget – £14.6 billion in 2015-2016.<sup>117</sup> This significant resource is directed towards improving care for a vast range of complex conditions and the establishment of ‘centres of excellence’ for these conditions. Some specialised services, such as those for cystic fibrosis, cover the majority of care patients with those conditions need. For ILD, specialised services only cover part of the pathway.

NHS England has established the standards and services it expects in centres treating patients with ILD, as part of the NHS standard contract.<sup>118</sup> NHS England has asked commissioners in each region to decide which centres should lead on ILD services. It is not clear how many centres there are in England or where they are situated.<sup>119</sup>

Recommendations on treatment options for IPF, as well as guidance for managing the disease, are determined by NICE. NICE publishes clinical guidelines, quality standards, and technological appraisals.

### Scotland

Local health boards are responsible for commissioning ILD services in Scotland. There is no formal overarching ILD service framework in Scotland; however three hospitals in Aberdeen, Edinburgh, and Glasgow have evolved into more specialised centres.

There is also a Scottish ILD Group, made up of clinicians across Scotland, that meets twice a year to share and develop best practice informally. The ILD group works with the National Advisory Group to develop best practice standards for ILD services as part of the proposed National Respiratory Action Plan for Scotland.

Scotland does not have a direct NICE equivalent and NICE guidelines do not apply in Scotland; they are however often used as a guideline. The Scottish Medicines Consortium (SMC) oversees the approval of medicines in Scotland. The Scottish Intercollegiate Guidelines Network (SIGN) produce guidelines for treatment, but there are currently none for IPF or ILD. Health Improvement Scotland (HIS) issues alerts to NHS Scotland on the publication of NICE guidance and advises on its applicability to Scotland.

### Wales

Health boards are required to commission services for people with ILD, as outlined in the Welsh Government's 2014 Respiratory Health Delivery Plan.<sup>120</sup> This plan sets out that people diagnosed with ILD must be managed through a MDT that works to national guidelines. This will involve local health boards ensuring that ILD patients are managed through a MDT framework and have access to specialist nursing support for appropriate conditions.

There is one specialised centre in Wales, delivered through Cardiff and Vale University Health Board.

The provision of ILD treatments in Wales is determined by NICE, which has a service level agreement with the Welsh Government, and the All Wales Medicines Strategy Group

(AWMSG). NICE works with the AWMSG to develop an appraisal work programme, implementation, support of guidance, patient access schemes, and value based pricing.

The AWMSG funds virtual MDT meetings to discuss cases of ILD. This means that while patients may be treated by a local consultant, they can be diagnosed by a team of experts from across Wales.

### Northern Ireland

Health and social care trusts are required to commission services for people with ILD, as specified in the Department of Health's 2015-2018 respiratory framework.<sup>121</sup> This strategy sets objectives for people with known or suspected ILD to be under the care of a respiratory MDT. It also sets objectives that ensure the MDT contains the appropriate expertise to deliver care.

The provision of ILD treatments in Northern Ireland is ultimately determined by the Department of Health, which has the option to endorse NICE guidance (either fully or with caveats) for use in Northern Ireland, or reject it. If approved, it works with NICE to support and facilitate its implementation. The Department of Health endorsed the NICE clinical guideline on IPF in August 2013.<sup>122</sup>

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