Lost in the system

IPF: the patient experience in England
About the British Lung Foundation

One person in five in the UK is affected by lung disease.

We offer hope and support at every step, so that no one has to face it alone. We campaign for positive change in the nation’s lung health. We fund vital research into new treatments and cures.

We are the UK’s lung charity.

Leading the fight against lung disease.

Help and support

Call us: 03000 030 555
Lines are open Monday to Friday from 9am to 5pm.
Email us: helpline@blf.org.uk

Thank yous

Boehringer Ingelheim is the founding partner of the British Lung Foundation IPF programme, and Intermune (which has now merged into Roche Products Ltd) is a supporting partner of the programme. Both companies contributed financially to the cost of the research and the production of this report.

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Executive summary

Idiopathic pulmonary fibrosis (IPF) is an incurable lung condition. Around 5,000 people are diagnosed in the UK each year, with the average life expectancy following diagnosis being just three years.

The British Lung Foundation launched its IPF Patient Charter in 2013, setting out the levels of care and support all IPF patients and their families have a right to.

This report looks at patients’ experiences. Patients told us how they can struggle to get a diagnosis and find themselves left alone to navigate their GP, local hospital and specialist centre for their care.

Patients with IPF can’t afford to waste time and energy going from service to service for the help they need. The National Institute for Health and Care Excellence (NICE) sets out a clear pathway for IPF care. By adhering to the pathway, offering accurate and reliable information, and, most importantly, ensuring every patient has the support of a specialist nurse, people with IPF can be supported to find their way through the system.
What is IPF?
Idiopathic pulmonary fibrosis (IPF) is a type of interstitial lung disease (a group of diseases that affect the ‘interstitium’ – a lace-like network of tissue that supports the air sacs in the lungs). In people with IPF scar tissue builds up in the lungs, making them thick and hard; this makes the lungs less able to take in oxygen. While some patients can live many years after diagnosis, the average life expectancy is just three years. There are around 15,000 people currently living with IPF in the UK, of whom around 5,000 are expected to die within the next 12 months. The number of people developing and dying of IPF in the UK is rising each year.

Who is responsible for IPF services?
The Health and Social Care Act 2012 saw services for people with IPF designated as specialised. This means that services are commissioned directly by NHS England and the standards these services have to meet are set out in a national service specification.

Our research
This report is based on the results of an IPF patient survey of 122 patient and carer respondents, and a freedom of information audit of 16 trusts in England providing tertiary IPF services. Not every trust was able to respond to every question posed. The results were supported by secondary research. While we have shared patients’ own stories throughout this report, for patient confidentiality purposes, their names have been changed.

Introduction

Average
LIFE EXPECTANCY
after diagnosis

15,000
People currently living
with IPF in the UK

1 in 3
EXPECTED TO DIE
within the next 12 months

3 YEARS
Number of people
developing and dying
of IPF in the UK is
RISING EACH YEAR
Getting a diagnosis

It is important that patients get an early diagnosis so they can access the support that can help them better manage their condition.

NICE requires that diagnosis be carried out by a specialist multidisciplinary team (MDT). However, to access an MDT, patients need to be referred by their GP.

Most patients responding to our survey were prompted to see their GP by a cough that wouldn’t go away (52 per cent) and/or regular breathlessness when doing any physical activity (50 per cent). However, following that initial visit to the GP, people can wait many months before being referred to a specialist.

Even after reaching a specialist, 39 per cent of patients reported a wait of three months or longer to be told they have IPF. This is all in the context of a condition with an average life expectancy of just three years from the point of diagnosis.

Patients also told us about problems with misdiagnosis. Some of the most common symptoms of IPF, including a persistent cough or breathlessness, are shared by other, more common, respiratory conditions, such as chronic obstructive pulmonary disease (COPD) or asthma. As a result, 26 per cent of patients responding to our survey reported being initially misdiagnosed.

As the first point of call for patients who are worried that they might have a respiratory condition, it is vital that doctors follow the NICE pathway for IPF, and refer people on to a specialist if IPF is a possibility.

Dave

“I gave up smoking a few years back, but had this cough I couldn’t shake. First I thought it was just a smoker’s cough. When I went to my GP he said it was just my lungs getting rid of all the ‘nastiness’ from years of smoking. It wouldn’t shift though, so I went back, but again was told not to worry. After a year it was still no better. This time I was sent for an X-ray, but again was told there was nothing wrong.

Then we moved and I saw a new GP. I was sent for a CT scan almost immediately, which showed that there was damage in my airways. I was told I had pulmonary fibrosis and referred to hospital. After a lung biopsy and further tests I was finally diagnosed, three years after my first visit to the GP.”

30 per cent of those responding to our survey said they had to wait six months or more for a referral to a specialist.

12 per cent reported waiting more than two years before being referred to a specialist.

Referral waiting times to specialist

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<th>0%</th>
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<td>More than 2 years</td>
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12 per cent reported waiting more than two years before being referred to a specialist.
All patients need information about their condition to help them take charge of their care and plan for the future. People with IPF are no different, and NICE is clear that both verbal and written information should be provided following diagnosis.

However, only 51 per cent of respondents to our patient survey said that, when they were first told they had IPF, the condition was explained by health care professionals in a way they could easily understand. A further 22 per cent found the verbal information about IPF difficult to understand, and 25 per cent reported that they did not have their condition explained at all (the remainder didn’t know).

There is a similar picture when it comes to written information; 49 per cent of people who responded to our survey were not given any written information about their condition. Of those trusts responding to our freedom of information audit, nine said that they provide patients with written information about IPF beyond clinical letters.

The British Lung Foundation produces free patient information on IPF that meets the NHS England Information Standard. Currently, six of the trusts responding to our audit offer patients the British Lung Foundation’s pack on IPF.

Every patient needs to be given accurate and reliable information on their condition, and all materials provided to patients should comply with the Information Standard.

**Finding out more**

I’m no stranger to lung disease. All my siblings and both my parents died of a lung condition. I’d heard of IPF before, but I still did not understand my diagnosis. The specialist simply said ‘you have what your brother was diagnosed with as well’. That was it. I felt like I was given the diagnosis and told to go home and manage it myself. As there is no treatment for IPF, I felt like I was being sent home to die.

I was told my life expectancy was two to five years and then I was scheduled for a check-up for six months later. I was referred to a physiotherapist to learn breathing exercises. That was it – no other information or support was given.”
Having someone to help

Interstitial lung disease (ILD) specialist nurses can help patients understand their IPF and coordinate their care. NICE says that all patients should have access to a specialist nurse. However, of patients responding to our survey, only 39 per cent reported that they have frequent contact with an ILD specialist nurse, and 36 per cent reported that they have no access to an ILD specialist nurse at all.

Of the trusts responding to our freedom of information audit, eight reported routinely allocating patients a named ILD specialist nurse within six months of diagnosis.

**Contact with an ILD specialist nurse**

- No access 36%
- Frequent contact 39%
- 19% Not as much as would have liked
- 6% Don’t know

John

“Until I met my specialist nurse, I felt alone and scared. My IPF nurse cannot do enough for me. She is always a phone call away to ask questions or talk to if I am worried. Admittedly she can’t always be there immediately but I can always leave a message and she will get back to me.”
Taking breath

Many people with IPF need access to oxygen therapy to help them breathe. Among patients responding to our survey, of those who do receive oxygen therapy, 25 per cent said it had improved their quality of life; 23 per cent said it helped them move about more; and 19 per cent said they were better able to do everyday tasks.

NICE says that patients should be initially assessed for oxygen when they are diagnosed and at any further follow-up appointments.

Eight trusts responding to our freedom of information audit reported that they assess people with IPF for oxygen therapy within six months of diagnosis, and 43 per cent of patients reported discussing oxygen therapy with a health care professional following their diagnosis. However, when we asked patients whether they had been reassessed for oxygen in the past year, only 39 per cent reported that they had.

It is important that NICE guidance is followed to ensure that all patients are able to benefit from the improved quality of life oxygen therapy can offer.

Sue

“When I was told I needed oxygen, I had reached the point where I couldn’t walk the 250 yards to the local shop. From the day I had the oxygen, I just tanked up and once I get that on, I can now do all the things I was doing before, in fact, more than I was doing before.

“I’ll go for a walk and I actually get some good mileage! I go to an over 50s’ keep fit class and I keep up with them. I have to have my oxygen on but I do everything that the other ladies and gentlemen do. I could not have done any of this before I had the oxygen, and to me it has been a lifesaver. It has been an opportunity to improve my fitness, in my interest, for the long run.”

Only 39% of patients surveyed had reported being reassessed for oxygen therapy in the past year
Getting fit

Pulmonary rehabilitation (PR) is an exercise and education programme that can help IPF patients cope with breathlessness, undertake more exercise and improve their overall quality of life. It is therefore an important part of treatment.

NICE says that people with IPF should be assessed for PR at the time of diagnosis and at all subsequent follow-up appointments. It also states that patients should be offered PR programmes designed specifically for IPF. This is because, while PR can help people with a wide range of conditions, it is important that it is tailored to each group’s particular needs. PR aimed at people with chronic obstructive pulmonary disease (COPD) is not ideal for patients with IPF.

Of the patients responding to our survey, 66 per cent had been assessed for PR following their diagnosis. However, when asked about the quality of their PR, 28 per cent rated it average or worse.

NICE guidance needs to be followed so that all patients are able to access PR and the courses they attend are tailored to their needs.

Lesley

“I heard about pulmonary rehabilitation on the grapevine but it was a struggle to actually find a course and get started. I had to do everything myself. Then when I next went to the consultant they said ‘yes, you should do that’ but they never mentioned it before!

“I was amazed at how useful it was; it told me things I feel my GP should have; things I needed to know. It gave me the confidence to go to the gym, even with my oxygen. I met a friend on the course and we went together and both exercised with our oxygen.

“Before, I was very lonely and desperate because of a lack of information. At the course I found out breathing techniques that I still use today, years later. It helped me to cope and accept my condition – even though I still have IPF, I know how to cope and keep going.”

“What would have made it even better was if the advice I had been given was IPF-specific; everything was geared to COPD. At the start I felt like I was just sitting in and not part of the group, and I had to interrupt to find out about how things worked for my condition.”
Taking control of your care

Currently, the single drug approved by NICE for the treatment of IPF is pirfenidone, which was approved for NHS use in 2013. A second drug, nintedanib, received European Commission approval this year. Although not everyone with IPF can be treated with pirfenidone, for those who can, it offers the potential to slow IPF’s progression and, in accordance with NICE guidance, should be offered to all eligible patients.

Of those patients responding to our survey, 66 per cent confirmed they were receiving some form of medication for their IPF, and 30 per cent of these had been prescribed pirfenidone.

Clinical trials are essential in the development of new drugs to treat IPF, and, promisingly, 42 per cent of all respondents had been given information on clinical trials. However, to really drive the progress needed in IPF treatment, all patients should be given information about clinical trials.

Palliative care means supporting both the patient and their family to manage and, where possible, provide relief from their pain and symptoms. It can include counselling, financial advice and support with pain management. It is distinct from end-of-life care, which refers more specifically to the care a person receives during the last phase of their life. Palliative care is provided locally. Patients can be referred by their GP, local hospital or specialist centre.

NICE says that people with IPF should have access to palliative care at any stage, not just as part of end-of-life care. Of those trusts responding to our freedom of information audit, some said 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. For patients this leaves no clear route into palliative care and the support they need: many have reported a lack of assessment for palliative care, despite the relatively short life expectancy common for IPF.

It is important that patients are also able to plan the transition from palliative to end-of-life care as their condition progresses. However, of the patients responding to our survey, 32 per cent reported that a lung disease specialist had never talked to them about the course their IPF could be expected to take.

NICE guidance is followed and that all patients are referred to palliative care early and then end-of-life care as their condition progresses.

Veronica

“My mother had IPF and she died in 2011. What we found really difficult was not getting a diagnosis, but the lack of care pathways and information for patients in the palliative care stage.

“There was a general lack of understanding of the condition’s progression by GPs and others, and no services available to support patients and their families at the end of life.

“In my view, the health system let my mother, and us as a family, down badly at the end of her life, due to disjointed services, breakdowns in communication, and the fact that there was no one person to coordinate health and care services. It was truly a traumatic experience.”
The priority for people with IPF is managing their condition as best they can and enjoying the time they have left with friends and family. Instead, patients find themselves struggling to navigate a system where their care may be provided through any one of three different NHS providers.

This is made even more frustrating by the fact that there are already clear guidelines and standards setting out what the best care looks like.

Three simple steps would transform care for patients with IPF:

1. All those providing IPF services to follow NICE guidance (including the quality standard and guidelines) and pathway.
2. All patients to be given Information Standard-approved information on IPF at the point of diagnosis.
3. All patients to have access to a specialist ILD nurse to help them navigate their care.

Our research suggests that, for patients with IPF and their loved ones, these three changes would make the world of difference. It would mean the difference between feeling lost in the system and being supported by it as they deserve to be.
References

1. National Institute for Health and Care Excellence, Idiopathic pulmonary fibrosis: The diagnosis and management of suspected idiopathic pulmonary fibrosis, June 2013


One person in five in the UK is affected by lung disease. Millions more are at risk.

We are the UK’s lung charity and we are here for every one of them, whatever their condition.

Lung disease can be frightening and debilitating. We offer hope and support at every step so that no one has to face it alone.

We promote greater understanding of lung disease and we campaign for positive change in the nation’s lung health.

We fund vital research, so that new treatments and cures can help save lives.

We are the British Lung Foundation.

**Leading the fight against lung disease.**

Get in touch with the British Lung Foundation to find support near you.

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