# Shining a light on IPF

## The patient experience in Wales

British Lung Foundation

www.blf.org.uk

# 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 you

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# Contents

# Summary

Idiopathic pulmonary fibrosis (IPF) is an incurable lung condition. 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 in Wales.

This report looks at patients' experiences. Patients told us that they struggled to get a diagnosis, felt that GPs and other health care professionals had little understanding of their condition, and felt that they did not get the support and information they needed.

The Welsh Government's Respiratory Health Delivery Plan gives a commitment that IPF patients are managed through a specialist multidisciplinary team and that the National Institute for Health and Care Excellence (NICE) guidelines for IPF are implemented.[1] The Delivery Plan offers a once in a generation opportunity to transform services for IPF.

But it needs funding.

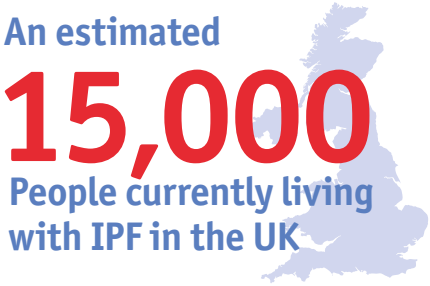
# Introduction

## What is IPF?

Idiopathic pulmonary fibrosis (IPF) is a type of interstitial lung disease. These diseases 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 absorb oxygen.

Research supports that there are an estimated 15,000 people currently living with IPF in the UK and that around 5,000 people die of it every year. Figures suggest the number of people developing IPF in the UK is rising each year. While some people can live many years after diagnosis, the average life expectancy is just three years.[2]





## Who is responsible for IPF services?

Services have developed organically in Wales in the absence of a single tertiary service.

In North Wales, patients can access the specialist centre in Liverpool and have much of their care managed from there with the support of local consultants here.

In Aneurin Bevan, Abertawe Bro Morgannwg, Cwm Taf and Hywel Dda Health Boards patients are usually treated and diagnosed locally, with selected patients being referred to the Cardiff and Vale multidisciplinary team (MDT) for further discussion.

## Our research

This report is based on the experiences of patients at focus group meetings in Caldicot, Cardiff and Llandudno. It includes feedback from patients in Swansea and references ongoing developments within the Respiratory Health Delivery Plan.

# 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.

The Respiratory Health Delivery Plan envisages patients being diagnosed by a multidisciplinary team and then supported by a specialist service. However to access a consultant and multidisciplinary team for diagnosis, patients need to be referred by a GP.

None of the patients we interviewed had heard of IPF prior to diagnosis. Every time they meet people they have to explain their condition. Most patients initially visited their GP with several different symptoms, the most common being breathlessness. Unfortunately many of these symptoms could be an indicator of another respiratory condition, so there was a range of initial misdiagnosis with asthma being the most common. A minority of people were initially diagnosed with anxiety.

Sue

**"My Dad had a diagnosis of emphysema a few years before, but then he kept getting chest infections over and over again. He was back and forth to the GP and was given antibiotics over and over again, but it didn't make a difference. By the end he could barely breathe and all he had was an inhaler. It took years to get a confirmed diagnosis [of IPF]."**

Steffan

**"When I left the meeting with the consultant [it was] like I knew nothing apart from the name of the condition and a prognosis. I was told to 'google it' and felt like I was on my own. I learnt everything I needed from the BLF and a booklet from Pulmonary Fibrosis Wales. I was terrified and felt like I should have had more support."**

The wait from initially visiting a GP to seeing a multidisciplinary team varied from three weeks to over a year. It depended on geography and the types of symptoms presented.

Most patients received a prompt diagnosis, but the amount of support they were given varied greatly. Some patients received a lot of support, had the condition explained, were referred to pulmonary rehabilitation immediately and had the opportunity to discuss treatment options.

A number of people told us that they received virtually no information or simply given the diagnosis and the prognosis. One person was told to 'Google it'. There was a geographic variation with patients diagnosed in Abertawe Bro Morgannwg UHB and Cardiff and Vale UHB feeling more informed that those diagnosed elsewhere in Wales.

Dafydd

**"At least three of the GPs in my practice were not even aware of IPF, while the others knew very little. They asked me what it was and I found that I was educating them. I learnt more at the conference in Treforest [BLF Meet the Experts event] than I learnt from my consultant. It was a great chance to meet doctors, nurses and other people with IPF."**

Bethan

**"My condition started in 2007. I'd had a minor, but persistent cough for the year before. My GP thought it was asthma and gave me an inhaler. It helped a bit, but then in 2013 I had a really bad infection that wouldn't go away and after seeing three different doctors, I finally saw a consultant and was diagnosed with IPF.**

**"Whenever I tell someone I have a lung condition, they assume it is COPD and tell me that I shouldn't have smoked. I have never smoked in my life, but no one has heard of IPF."**

# Knowing who can 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, yet many of the patients we spoke to had very little contact.

In South East Wales many patients were in contact with an oxygen nurse, but not an ILD specialist nurse. In North Wales patients living in Flintshire, Denbighshire and Wrexham spoke positively about having access to nurses as part of an ILD service in Aintree University Hospital (Liverpool), while in Anglesey, Conwy and Gwynedd it is was more difficult to access an ILD specialist nurse.

In Swansea and Llandough there are ILD specialist nurses who can support patients.

There are three pulmonary fibrosis patient support groups in South Wales, but none in North Wales. There are 13 Breathe Easy groups across Wales. These support patients living with all respiratory conditions, including IPF. Across Wales the frequency that people saw a consultant varied from every three months to annually depending one whether they were prescribed pirfenidone or not.

Sheila

**"If I need anything, I know I can speak to nurses at my hospital. They are very good and always look after us. You need to know the phone numbers. If they are not looking after you, you need to pester them."**

Derek

**"We have an excellent service from our ILD nurse. She is great. She is always there to answer any of my concerns and queries."**

Rhys

**"I feel I have no one to talk to. I saw a nurse during pulmonary rehabilitation, but I haven't heard anything since. The only number I have is my consultant's secretary, and he seems too busy to see me."**

# Taking breath

Many people with IPF need access to oxygen therapy to help them breathe. Doctors can prescribe oxygen therapy if the level of oxygen falls too low and the benefits can be life changing. The patients we spoke to described how they were unable to leave their homes and how oxygen can 'give them back their lives.' Oxygen has helped to improve confidence, helped with walking, reduced fatigue and had helped one patient to remain in employment.

The patients interviewed spoke highly of their oxygen nurses and the oxygen suppliers.

Martyn

**"Oxygen has helped me to continue living my life, but it has required a few adjustments. If we go on holiday, my wife and I need to ensure we can get oxygen wherever we are going, and if I am travelling in this country I need to take to take the equipment with me. It requires a lot of planning ahead, but everyone is so helpful."**

Pam

**"I can't fault the oxygen service, if ever I have a problem they send someone around to check my equipment. Oxygen has changed my life and I am far more mobile. However, you can't over do it. I work on the 3 Ps – pacing, planning and prioritising. You need to rest, you need to get your oxygen levels back up. You have to take it easy.'**

**"It's terrifying when you don't have enough oxygen. If I get too out of breath my oxygen levels drop, my hands shake and my knees go."**

# Keeping active

Pulmonary rehabilitation is an exercise and education programme that can help IPF patients cope with breathlessness, undertake more exercise and improve their overall quality of life.[3] It is therefore an important part of treatment.

NICE states that patients should be considered for pulmonary rehabilitation upon diagnosis and then at subsequent appointments.[4] While most patients were aware of pulmonary rehabilitation, some were not and had never been referred. Many of those who had been referred learnt about it later rather than at diagnosis.

Patients should be offered pulmonary rehabilitation that is designed specifically for IPF. Pulmonary rehabilitation is available, but the waiting lists can be long and often the information is not tailored to the needs of patients with IPF. Increasing capacity in pulmonary rehabilitation is a national priority in the Respiratory Health Delivery Plan.

Patients who had attended pulmonary rehabilitation spoke about the benefits of breathing techniques, increased mobility, better understanding of the condition, and meeting people with lung conditions. However they were often the only person with IPF in the room, and sometimes the professionals didn't understand their condition.

Gwilym

**"I am a member of three choirs and I found that singing really helps. When I am singing I forget about my condition and it really clears out your lungs. Particularly in a Welsh male voice choir, the 'amen' at the end of hymn is great way of pushing working your lungs."**

Gloria

**“The education was brilliant. When I went there was a physiotherapist, an occupational therapist and a nurse, and they tell you what exercise to do – nothing strenuous and you gradually build it up. They taught me to breathe through my nose and out through my mouth. This sounds simple, but was really hard at first. It has definitely made a difference and I feel far more mobile. It was great to meet other people with lung conditions, but I was the only person with IPF.”**

# Conclusion

Our patient experience work has demonstrated that services vary dramatically across Wales with some patients receiving an excellent service and some feeling that they are unsupported.

Services in Wales are not currently meeting the NICE guidelines, and few people are benefiting from a tertiary service as offered in many parts of England.

Patients need support, information and understanding from the NHS, while healthcare professionals need the resources to deliver a modern service.

In the summer, a group of consultants submitted a proposal to fund a tertiary service with additional ILD nurse time and multidisciplinary teams. We need this funding secured and guaranteed in future years.

Our discussions with patients have shown that they feel that their condition is poorly understood. GPs, non-specialist healthcare professionals and the public at large are in the dark as to what the condition is, how it can be treated and what support those living with IPF need.

The Welsh Government can make a difference through the Respiratory Health Delivery Plan and transform services for the better.

# We ask the Welsh Government to transform the care of patients with IPF

1. Funding for this year, and subsequent years, to develop a tertiary service for IPF and other ILDs

2. All patients to be given high-quality and accessible information on IPF at the point of diagnosis

3. All patients to have access to a named specialist nurse and consultant with a specialism in ILD

## References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