



Action for  
**Pulmonary  
Fibrosis**

# Our Vision

*A world in which everyone living  
with pulmonary fibrosis  
has a better future.*

**Supporting**

**Educating**

**Researching**

**Campaigning**



# What is Idiopathic Pulmonary Fibrosis?

Idiopathic pulmonary fibrosis (IPF) is a progressive and irreversible condition that results in scarring of the lungs. Over time, scarring makes the lungs stiff and leads to reduced lung capacity. Thickening of the gas exchanging parts of the lung (alveoli) means that transfer of oxygen from the air to the blood becomes impaired, which can lead to low blood oxygen levels. Common symptoms include breathlessness, cough, fatigue, and reduced exercise capacity. Eventually, everyday activities such as walking and climbing stairs become increasingly difficult.

IPF affects approximately 30,000 people in the UK, with 5000 new cases diagnosed each year. The chance of developing IPF increases with age and therefore it is mainly found in people over the age of 50. In rare circumstances, it can be found in younger individuals, especially if the disease runs in their family. Men are more commonly affected than women and over the last century, the number of people suffering from this disease has been steadily increasing.

The reason why IPF occurs is unknown, yet certain risk factors for its development have been identified. These include exposure to cigarette smoke and exposure to certain types of dust such as metal or wood.

IPF gets worse over time, although the speed at which this happens is highly variable. Some people may remain relatively free of symptoms for many years, while others may get rapidly worse.

For some, the outcome of this disease is worse than many cancers with an average survival of 3-4 years.

There are currently no treatments that cure or reverse IPF, but therapy with an approved drug (nintedanib or pirfenidone) can slow down its progress.

**Other treatment options may include an exercise and education programme (pulmonary rehabilitation) and oxygen therapy. In special circumstances lung transplantation may be appropriate.**

## The ideal IPF patient journey

Several guidelines exist which outline the care which patients with IPF in England should receive:

- The NICE quality standard on IPF<sup>12</sup>
- NICE Technical Appraisals for anti-fibrotic treatments<sup>13,14</sup>
- NICE IPF diagnosis and management clinical guideline<sup>9</sup>
- The interstitial lung disease (ILD) service specification<sup>10</sup>

Many IPF patients receive excellent care from dedicated staff in specialist and non-specialist hospitals in line with these guidelines. Action for Pulmonary Fibrosis' 2015 patient survey captured many of these positive experiences of the care and support<sup>15</sup>. All qualitative quotations in this report are taken verbatim from the results of this survey<sup>16</sup>.



Roche Products Ltd has funded an agency to help with the writing of this report and has also checked it for factual accuracy and ABR code compliance. Final editorial control rests with Action for Pulmonary Fibrosis (APF).

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## Dr Helen Parfrey

**APF founding Trustee**  
**Consultant Respiratory**  
**Physician, Papworth Hospital**

Action for Pulmonary Fibrosis was founded in 2013 by a group of patients, family members and medical professionals all with a personal or professional connection to the disease.

We bring a unique perspective to the job of running the charity, which gives us a real and genuine bond with patients and families.

APF was inspired by our late chair, pulmonary fibrosis patient Mike Bray, who felt there was a desperate need for a charity dedicated solely to this terrible disease; a charity that would put patients and families at the heart of everything it did.

Since 2013, we have worked hard to support patients, raise funds for research, educate health care professionals, raise awareness of the disease and campaign on behalf of those who don't have a voice.

In a very short time we have become recognised as the Voice of the Patient and are an authentic voice for those affected by IPF.



We listen to what patients and families are experiencing and learn from them.

This approach is central to how we operate and to how we deliver the support, services and advocacy that patients and families need.

We have achieved a great deal since we started the charity but we have bold ambitions and will continue to lead the search for better outcomes in the future and champion the best care available today.

To learn more about our Trustee Board visit our website at  
[www.actionpulmonaryfibrosis.org/trustees](http://www.actionpulmonaryfibrosis.org/trustees)

# We listen to and learn from the IPF community



## The APF Voice of the Patient Council

We are proud to be a charity that listens to patients. It is at the heart of everything we do. We do this through trustees and staff who regularly attend support group meetings across the UK to find out what patients are really experiencing in their treatment and care.

We regularly ask patients directly for their views through surveys and questionnaires and we encourage them to be part of our Voice of the Patient Council.

We contact Council members from time to time by email to ask their views on a variety of issues – it could be about pulmonary rehabilitation or oxygen therapy or about how we should support patients.

We hold a Council meeting once a year, usually around IPF World Week in the autumn.

But we also listen to and learn from others in the IPF community. Pulmonary fibrosis is an Interstitial Lung Disease, an umbrella term for diseases that affect the gas exchanging part of the lung.

We have a unique relationship with the ILD Interdisciplinary Network (ILD-INN), an organisation set up by a group of ILD specialist nurses dedicated to promoting ILD speciality practice through education and professional development and to influence the policy of ILD care.

We are working in partnership with them to deliver vital training to community and practice nurses and other health care professionals across the UK.



## Howard Almond

### IPF Patient & Trustee Lead on the Voice of the Patient Council

*"I was diagnosed with IPF in 2014 and I took the opportunity to get to as many informative conferences and other events as possible, and it is through this that I became a member of the Voice of the Patient Council and subsequently a Trustee.*

*I had never heard about IPF before I was diagnosed, and it became clear from an early stage, that a lot of medical professionals were not well-informed about it either. APF is working to improve key things in the day-to-day treatment and care of patients and to improve longer-term outcomes.*

*I now have Trustee responsibility for the APF Voice of the Patient Council and I am delighted to say that it plays a key role in helping guide the direction of the charity's efforts, highlighting difficulties and problems, and making suggestions about where we can use our resources with most effect."*

**If you are an IPF patient in the UK and would like to have an influence on the work of APF please sign up to our Voice of the Patient Council by visiting our website.**

## Support Groups

Extending the network of support groups for patients and families across the UK is at the heart of our charity. Having a disease like pulmonary fibrosis – or helping to care for a loved one who has it – can be very frightening and isolating. We know how valuable the support of others can be.

Since our charity began in 2013 we have been instrumental in growing the number of support groups from around six to over 40...and growing. Our website has a list of all the support groups in the UK, with contact details and an interactive map to help you find your nearest group. With a dedicated Support Group Co-ordinator and funding from Boehringer Ingelheim we will continue to extend the group network until everyone with pulmonary fibrosis has the support they need.



### Lorna McLauchlan

#### APF Support Group Co-ordinator

*"I've worked in respiratory care for 32 years and for most of those I was a respiratory ILD nurse specialist. I quickly learned how important patient support is and the benefits it has for those diagnosed with life changing respiratory conditions.*

*In 1995, I set up a respiratory support group and watched as it grew and touched the lives of so many people. Patients told me how the group decreased their feelings of isolation following diagnosis and helped them feel more in control of their lives.*

*It can be a lonely and confusing time following diagnosis and whilst a support group can never replace expert clinical advice it can provide a safe environment to chat to others in a similar position. The sharing of experiences can help people cope with day-to-day issues as well as making new friends, whilst education from expert speakers can help improve feelings of control and a sense of well-being.*

*Working with Action for Pulmonary Fibrosis allows me to carry on the great work of supporting the setting up of new groups across the country. I am again seeing the benefits for patients, their families, carers and friends. It gives me great job satisfaction to see this and I look forward to helping to establish many more groups across the UK."*

# ...it makes me feel I'm not alone

*"I really look forward to coming to the group. We learn a lot about the disease, not only from the experts but from each other."*

*"I look after my husband, who has IPF and it can be a real strain. The support I get at my group is fantastic. It makes me feel that I'm not alone and that it will be ok."*

*"I lost my dad to IPF a long time ago but I still go to my local group. The strength of the people who attend is inspiring and their humour and positive attitude, even when suffering from such a dreadful illness is amazing."*



## Steve Jones

### Lead Trustee on Support Group Development

*"I was diagnosed with IPF in 2008 and for the first five years, the disease progressed slowly. I continued to work and led a reasonably full life but in 2013 my condition deteriorated. I was fortunate to receive a single lung transplant in March 2016.*

*I am Secretary and Vice-Chair of the Papworth Hospital Pulmonary Fibrosis Support Group and I was delighted to have been asked to join Action for Pulmonary Fibrosis as a Trustee.*

*I really believe in the value of support groups and I am working hard with Lorna and other trustees to strengthen the network across the country and to reach out to people with IPF living in remote areas or who are otherwise isolated."*

# We educate health care professionals



One message which patients send us loud and clear is that there is a widespread lack of knowledge about pulmonary fibrosis within the health care community.

Many patients feel this has led to misdiagnosis and late diagnosis of their condition.

We listened and we set up a national training programme for community and practice nurses and other health care professionals.

The specialist respiratory nurses who are part of the ILD Interdisciplinary Network are our partners in this education project and deliver the training. This is an ongoing programme funded by Roche which will continue for as long as health care professionals need it.

## Increased knowledge of IPF will help early and accurate diagnosis

### What health care professionals say about our training programme

*"I now have a more in-depth knowledge and will be able to advise ILD patients where to go for advice and information. A very, very good presentation."*

*"I will keep ILD in the back of my mind when seeing patients whose symptoms are not responding to usual management."*

*"Very informative. I feel I can support my patients more affectively."*

*"I will be more inquisitive, more aware and ask more questions."*

**APF trustees also travel the country giving talks and presentations and runs regular media campaigns to help the wider public understand the disease and its devastating impact on lives.**



## The Mike Bray IPF Research Fellowship

The wonderful efforts of individual fundraisers and the generosity of people who have made memorial donations, one-off donations and who have signed up to make regular donations supports all of APF's work. After listening to patients, family members and fundraisers the charity has allocated a large proportion of money raised to research.

We have established the **The Mike Bray IPF Research Fellowship** in memory of the charity's inspirational founding trustee. This will be a rolling three-year fellowship to be awarded to an outstanding research project.

Mike, together with his wife, Elizabeth, was the driving force behind the establishment of Action for Pulmonary Fibrosis. He had been diagnosed with IPF in 2005 and received a single lung transplant at the age of 68 in 2010. He was passionate about fighting the patients' corner and was instrumental in the charity becoming recognised as the authentic voice of the patient. He helped shape the vision and values that underpin everything the charity does. Mike passed away after a short illness in January 2017 and he is missed by all those who were fortunate enough to know him.

## We award travel grants to researchers and health care professionals – Researcher Amanda Tatler attended the American Thoracic Society conference in Denver

*"During the conference I presented a scientific poster relating to my research investigating the potential anti-fibrotic effects of caffeine in the lung. In addition I used my time to learn more about emerging theories and clinical updates of IPF to enable me to continue to enhance my understanding of the disease. I learned lots and met many world-renowned researchers in the field of IPF. I am extremely grateful to Action for Pulmonary Fibrosis and all who donate and support for the opportunity to attend the meeting."*

**Read more about the Fellowship and APF's travel grants and bursaries and find out how to support them at [www.actionpulmonaryfibrosis.org](http://www.actionpulmonaryfibrosis.org)**

# Our Fundraisers!

## We couldn't do any of this without our amazing fundraisers

When we started Action for Pulmonary Fibrosis in 2013 we could never have imagined how much support we would get from our fundraisers and donors. It has been truly overwhelming and it is because of the people who have worked so hard and given so generously that we are able to fund major projects around the UK and make significant contributions to vital research.

We are grateful to every single person who has chosen to give to our charity – often at a very difficult time in their own lives – as we could not make a difference to the lives of those living with IPF and their families without you.

Thank you.



Find out more about fundraising for Action for Pulmonary Fibrosis by visiting our website

[www.actionpulmonaryfibrosis.org/fundraising](http://www.actionpulmonaryfibrosis.org/fundraising)

For a fundraising pack, advice on making one-off or regular donations or help with a fundraising event please contact our Fundraising & Marketing Co-ordinator Alex Croft at

[alex@actionpulmonaryfibrosis.org](mailto:alex@actionpulmonaryfibrosis.org)

...are amazing



# We are taking the fight to the seats of power

## Campaigning on behalf of patients is an important part of our work.

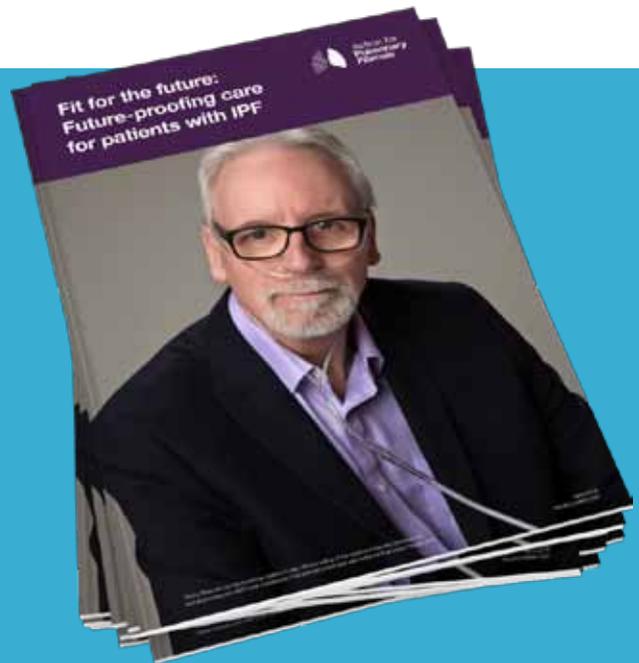
**We believe that taking the fight for better care and outcomes for IPF patients to the seats of power is our duty as a charity.**

One of our high profile campaigns - **Fit for the future: Future proofing care for patients with IPF** - was presented to MPs at a Parliamentary reception hosted for us by Stephen McPartland MP, Chair of the All-Party Parliamentary Respiratory Committee.

Mr McPartland has a personal interest in respiratory care and has met privately with trustees. He also helped launch a new patient support group in his own constituency of Stevenage.

*“The fact that as the NHS and government we do not know how many people are affected by this disease is totally unacceptable and I am not prepared to accept that. So I am going to work with Action for Pulmonary Fibrosis to push forward so that we have some kind of way of forcing NHS England to record this data so that we know where patients are with this disease and what we can do to support them throughout their condition.”*

*Stephen McPartland MP*



***APF Trustee Tony Gowland featured on the report cover and spoke about his personal experience of the disease to MPs***

# Raising awareness in Europe



The charity is a founding member of the EU IPF Federation, which brings together representatives of patient groups across Europe.

The Federation is committed to defending their vision of equal access to treatment and care for all IPF patients, regardless of geography, socio-economic status or age. It is estimated that around 110,000 people suffer from the disease across Europe.

One of the key achievements of the Federation was the development and launch of the European IPF Charter, which was launched at the European Parliament in 2014. The Charter lays down the rights of IPF patients and policy recommendations to ensure improvements in patients' quality of life, whilst supporting efforts to find a cure.

Learn more about the federation at [www.eu-ipff.org](http://www.eu-ipff.org)

## Call to Action

In order to plan the optimum services for patients with IPF, we need to have more accurate information about the incidence and prevalence of this terminal lung disease. It is vital that the NHS knows the number of patients suffering from IPF.

We ask NHS England to introduce the mandatory collection of data relating to the number of people diagnosed with IPF by a multi-disciplinary team (MDT). Through a better understanding of the true scale of IPF, the NHS can ensure that it makes best use of its resources to provide high-quality care both now and in the future.

Action for Pulmonary Fibrosis also contributes to the National Institute for Health and Care Excellence consultations relevant to IPF and patient care and took part in consultations on the **NICE IPF Quality Standard** which APF supported, as well as the review of the **NICE Clinical Guidance Note 163 - IPF in adults: diagnosis and management**.

Trustees have also taken part in consultations and have provided a patient representative at drug appraisals for the anti-fibrotic drugs Pirfenidone and Nintedanib.

**APF is keen to represent patients and to ensure that the disease is understood from the patient's perspective in any situations that influence the treatment and management of care.**

## We will always put patients and their families at the heart of everything we do

We speak to and hear from many patients and family members. They tell us their own personal stories and are always happy to help us spread the word about IPF. We never fail to be moved, humbled – sometimes shocked – but always inspired by what we hear.



### Anne Doran

*“Receiving the diagnosis of IPF left me feeling devastated and alone, then the fear kicked-in with immediate flashbacks to my father’s death from the same disease thirty years ago. I was feeling unwell with a heavy cold and feeling breathless.*

*I just assumed that I had ‘low grade pneumonia’ but it gradually got worse - I was finding it hard to speak and catching my breath. I felt exhausted and was gasping for breath after walking a short distance.*

*I had a scan and was told I had extensive lung changes that would require a lung biopsy but it was almost certain I had IPF. It never entered my mind I would have anything serious – I thought it was a chest infection and I’d be prescribed with a course of antibiotics. It really came out of the blue and was a massive bombshell.*

*My husband Tom collected me as usual from the hospital and I held the news in until we returned home and then told my family. It was the hardest thing I’ve ever had to do.*

*This disease is terribly frustrating. There’s so much I want to do. Not long ago I would have run around. Now a half hour trip needs several pit-stops. It’s do-able but I have to plan. I’m determined and always searching for a way round! I do have black days but I also have a good outlook on life and an incredibly supportive family.*

*You can spend your time focusing on all the negative aspects of this disease of which there are loads...unfortunately there is no ‘good news’, so you have to focus on the positive aspects of your life. What can you do? And how will you do it? Don’t cut yourself off from friends, keep active, keep going, keep fighting but most of all as they say, ‘keep calm and carry on.’*



# Listening to patients and families



## Elaine Grove

*"I am a Business Development Manager at Open College Network West Midlands. Each year we support a nominated charity and I am so happy that we have supported Action for Pulmonary Fibrosis 2016-17.*

*My mum, Sheila, was just 58 when she died from IPF in 2001, just 8 months after being diagnosed. She was fun to be with, and always had the ability to make me feel that any problem was halved, just by chatting to her.*

*Gosh, how I miss her. Back in those days very few people had heard of the disease and there was no support available for the patient or their families. Fast forward to the present day, and my goodness, how things have moved on. Social media, support groups, and of course Action for Pulmonary Fibrosis. Thank goodness!"*



## Peter Taylor

*"My dad was a professional footballer and manager at Nottingham Forest & Derby County, with his friend and partner, Brian Clough.*

*It was a crushing blow when he was diagnosed with IPF at the age of 59. After an active life he struggled to walk across the room without becoming breathless.*

*He died three years after diagnosis, in 1990. It was devastating for our family and in*

*those days there was zero knowledge about the disease and no support. I'm glad to say that treatments, outcomes and support for patients is so much better now.*

*I think the future is much brighter for IPF patients now than it was for my beloved dad."*

*Wendy Dickinson  
Peter's daughter and APF Trustee*



# Action for Pulmonary Fibrosis

**Action for Pulmonary Fibrosis is a charity set up by a group of pulmonary fibrosis patients, family members and medical specialists.**



Our aim is to raise the profile of this little-known disease, to campaign on behalf of patients, to support patients and families and to raise money for research. APF is working hard to improve the experience of patients and to ensure that everyone, wherever they live, receives the best possible treatment and care available.

Action for Pulmonary Fibrosis is leading the search for better outcomes and championing the best care today.

 **01543 442152**

 **[www.actionpulmonaryfibrosis.org](http://www.actionpulmonaryfibrosis.org)**

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**Your nearest Support Group**