Showing how we care about idiopathic pulmonary fibrosis: Increasing support and awareness

Year 2: IPF project report

November 2014 - October 2015
Thank you to our founding partner Boehringer Ingelheim, and supporting partner Roche Products Ltd.
The companies have no influence over the activities or the content of this report.
We care about IPF

Idiopathic pulmonary fibrosis (IPF) is a long-term progressive disease where the lungs become irreversibly scarred over time. If it were a cancer, it would be the eighth most common in the world. Yet hardly anyone has heard of it. We want to change this.

IPF is a condition long neglected by politicians and health service managers. IPF is one of the most difficult respiratory diseases to manage and more effective treatments are urgently needed. We want to change this too.

In the second year of our IPF programme, we’ve continued to campaign for greater recognition for IPF and better support for those affected. We’ve worked with people with IPF, their families and health care professionals. We’ve provided comprehensive, reliable information for patients and their families, and training for health care professionals. We’ve raised awareness with policy makers, politicians and the public.

We’ve also funded research to improve our understanding of IPF and to develop better treatments and care. Our projects are scientifically robust and will make a real difference to people living with IPF.

Over the last two years we’ve built IPF into our day-to-day structure. An ILD specialist clinician now sits on our board of trustees. And we’ve set up a patient and carer Think Tank with IPF representatives and an ILD research advisory board with patients and health care professionals.

We’ve achieved a lot but there’s more to do. Together, we’ll show how we much care about IPF and bring about far-reaching change.

Dr Penny Woods
Chief Executive
Executive summary

One of our priorities at the British Lung Foundation (BLF) is interstitial lung disease (ILD), initially focusing on idiopathic pulmonary fibrosis (IPF). This report examines our second year of activity and action in our IPF project.

IPF is a chronic, progressive and incurable lung disease. Almost 50% of people will die within three years of diagnosis. However, individual survival is difficult to predict and some people will live a lot longer than this. Living with IPF can have a huge impact on the quality of life of both patients, their carers and family.

This year with the help of all our supporters, we continued to show how we care through providing information and support, raising awareness and funding research.

We built on what we started to provide the highest quality information and support. Our online IPF hub was Highly Commended at the British Medical Association (BMA) Patient Information Awards 2015. The hub was visited over 250,000 times between November 2014 and October 2015. This makes it the most comprehensive UK online resource for people affected by IPF.

All our health information, both online and in our booklets, is accredited by the NHS England Information Standard. This means that it is reviewed by both health care professionals and patients to make sure we produce easy to understand information that the public can trust. In one year we sent out over 13,000 IPF information packs, 22,000 IPF leaflets and 14,000 IPF booklets to those who needed them. We also supported people through our helpline, support groups and Meet the Expert events.

We continue to campaign for better care for people living with IPF and their families by influencing health care services and raising awareness. This year we told over half of MPs about IPF and launched our patient experience report to over 50 MPs and policy makers at Westminster. This was followed by an event at the Welsh Assembly to highlight the patient experience in Wales.

Throughout the year we generated over 140 pieces in the regional and national media with a potential media reach of 114 million. Many of these were personal stories showing how important it is to care about IPF. During IPF week social media posts with infographics, people’s stories and a call for more research were published every day on the theme of ‘#ICareAboutIPF’. Over 25,000 people saw our Facebook posts at the highest point during this week.

We drive research in areas where we feel we can make a real difference to patient outcomes. In our financial year 2014-15 we awarded £270,000 towards ILD research, thanks to the fundraising effort of all our supporters. This funding was spent on priorities decided through collaborative working with patients, carers and researchers. We ensure that the research projects we fund are scientifically robust and make the maximum difference to people living with IPF.

There has been an incredible amount achieved but there is still so much more to do. We will continue to work together with patients and their families, health care professionals, and policy makers to show how we all care about IPF and help improve lives.

Read about Ron’s fundraising challenge on page 18.

“The support I received from the team at the British Lung Foundation and those around me has been amazing. It gave me the confidence I needed to take on such an incredible challenge. I’m so glad I can give something back.”
Getting started

Background

In 2013 we decided to focus on five priorities, while ensuring that we continued to support everyone with a lung disease. One of these priorities is interstitial lung disease (ILD), focusing first on idiopathic pulmonary fibrosis (IPF).

Every year in the UK around 5,000 people will be diagnosed with IPF, and every year around 5,000 people will die from IPF. This is comparable to far better known conditions such as leukaemia.

Almost 50% of people will die within three years of diagnosis. However, individual survival is difficult to predict and some people will live a lot longer than this. About 20% of people live for more than 5 years. Since these figures were published, two new drugs have been approved by the National Institute of Health and Clinical Excellence (NICE) that slow the progression of IPF.

Living with IPF can have a huge impact on the quality of life of patients, their carers and family. Specialist health services may be difficult to access and the condition itself may make it difficult to meet others in the same situation.

Unfortunately, there is no cure yet for IPF. For a very few people, having a lung transplant might be an option. There are currently two drugs (nintedanib and pirfenidone), licensed for use by the NHS, that slow down the development of scar tissue in the lungs of people with IPF.

First steps

As a first step an IPF Patient Charter was developed with ten calls to action. See page 21 to read the charter. A two year project was then started to provide support and raise awareness. The activity in this project was funded by founding partner Boehringer Ingelheim, and supporting partner Roche Products Ltd. The companies have no influence over the activities or the content of our information.

Thanks to all the fundraising efforts of our supporters, we also continue to fund research into pulmonary fibrosis, with new research being funded every year.

There has been an incredible amount achieved in the first two years, and now we are planning further action as there is still so much more to do.

What is interstitial lung disease (ILD)?

‘Interstitial’ means the disease affects the interstitium, the lace-like network of tissue that supports the air sacs in your lungs. There are more than 200 different ILDs.

What is idiopathic pulmonary fibrosis (IPF)?

Idiopathic pulmonary fibrosis (IPF) is a type of interstitial lung disease. It is a progressive condition where scar tissue builds up in the lungs, making them thick and hard. People with IPF can feel breathless from simple everyday activities. Coughing is another common symptom.

Idiopathic - cause is unknown
Pulmonary - affects the lungs
Fibrosis - scar formation

“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... 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.”

Patient living with IPF

Navaratnam et al., 2011, Thorax, 66, 462-467
NICE., 2013, IPF: the diagnosis and management of suspected IPF
**Project aims**

The aims, objectives and outcomes for the project were developed to reflect the ten key calls to action in the IPF Patient Charter. Project activity was categorised to reflect the two main objectives of the project.

**Aim**

To enable the BLF to take a proactive approach towards developing a campaign, communications and support programme for IPF.

**Objectives**

- To ensure that people with IPF, their family and carers have the information and support they need to understand their illness, make decisions and make the most of their lives.
- To influence and shape NHS and other services to ensure they are accessible, integrated, appropriate and evidence based.

**Outcomes**

- Improved information and support for IPF patients, their families and carers
- Increased awareness of IPF and its symptoms
- Better patient experience for people with IPF along the patient pathway
- Improved timely referral at primary care level for diagnosis
- Progress towards the full implementation of the NICE guidelines for IPF

**Year 2 plans**

**Providing information and support**

- Evaluate the IPF information pack
- Pilot our IPF personal organiser
- Hold IPF Meet the Expert events around the country
- Help set up and support more pulmonary fibrosis support groups
- Develop our online resources for people with IPF and their families

**Influencing health care services and raising awareness**

- Use World IPF Week to raise even more awareness
- Launch the first ever IPF campaign report in parliament
- Continue to influence key policy makers
- Continue to promote IPF awareness through the media
- Encourage research into patient experiences
- Provide support and information for health care professionals
Key achievements

Summary of key achievements in 2014-2015

Providing information and support

- We had over 250,000 visits to our online IPF hub.
- There were over 13,000 IPF information packs ordered in the first year since launch, from Shetland to the Channel Islands, with excellent feedback from patients.
- This year over 450 people living with IPF and their families attended ‘IPF Meet the Expert’ events, with 97% telling us they had learnt more and 83% telling us they now feel more confident living with IPF.
- So far we have helped launch thirteen new pulmonary fibrosis support groups around the UK by providing a bursary and expertise, including the first ever pulmonary fibrosis support group in Northern Ireland.
- Our blog on ‘IPF the final stages’ by David was viewed over 17,000 times in the year.
- Our Helpline continued to support people living with IPF and their families, with one in ten calls being IPF-related.

Influencing health care services and raising awareness

- We reached over 50% of MPs leading up to the launch of our patient experience report, with over 50 parliamentarians and policy makers attending our event about IPF in Westminster.
- The success continued with the launch of our Welsh patient experience report at the Welsh National Assembly for Wales, with the Deputy Minister for Health giving a speech on the importance of pulmonary fibrosis services.
- During IPF week over 25,000 people saw our Facebook posts at the highest point during the week, compared to 5,000 on a normal week.
- We generated over 140 pieces in regional and national media with a potential media reach of over 114 million.
- Our study days for health care professionals covered ILDs and reached over 500 people.

Research

- We encouraged patient-centred research by collaborating with leading researchers on a journal article and supporting the recruitment of participants into a study for patient reported outcome measures.
- In our financial year 2014-15 we awarded £270,000 towards ILD research. We’ve awarded over £2 million to ILD research so far.
Evaluating our activity

Providing information and support

Information packs

The IPF information pack is designed to provide a high quality, comprehensive resource that is easy to hand out at diagnosis, and has space for local information to be added. In the first year since launch (June 2014 to June 2015) over 13,000 packs were ordered. Our tracking system shows that in the first year 6,712 of the packs were sent directly to patients while 6,580 were sent to health care professionals to distribute to patients. Those ordering 40 IPF packs or more were spread throughout the UK, showing that there has been a wide distribution of the IPF information packs. Smaller orders (under 40) were also widely scattered with requests from as far north as Shetland and as far south as the Channel Islands.

Most patients commenting on the IPF information packs thought the amount of information was about right. Comments about the balance of information included:

- “An excellent document with the right balance for its size and depth of coverage.”
- “I found the information very useful... it was very easy to read.”
- “No punches pulled advice and information.”
- “The presentation of the pack is middle of the road, and welcoming.”

We’ve also continued to distribute our IPF booklet ‘IPF and other interstitial lung diseases’ and our IPF leaflet separately. In the year from November 2014 to October 2015 over 22,000 IPF leaflets were ordered and 14,000 IPF booklets.

IPF personal organisers

We’ve developed an IPF personal organiser to help patients get organised, take notes and know what questions to ask their doctor, nurse or anyone else supporting them. We hope that this will help people living with IPF feel more in control of their condition. This organiser is being piloted in Aberdeen and Newcastle. We will have the results of this pilot later this year.

Our health information is accredited by the Information Standard

The Information Standard (IS) is a certification scheme, run by NHS England, that assesses the processes an organisation follows to make sure ensure it produces high quality health and social care information for the public.

All our health information is reviewed regularly by both health care professionals and patients to make sure we have easy to understand information that the public can trust.
Meet the Expert patient events

We held a series of seven Meet the Expert events across the UK, together with Action for Pulmonary Fibrosis (APF). These events provided an opportunity for people with IPF and their families to come together and understand the current environment of IPF care in the UK through talking to each other, listening to the speakers and taking part in question and answer sessions. Just over 450 people attended, 55% were those living with IPF and 39% were a carer, friend or family member.

Overall, 83% of participants rated the events as excellent and 17% rated them as good. When asked what they thought about the information and organisation given before the event 91% thought it was either excellent or good.

- 97% agreed or strongly agreed that they had learnt more as a result of the event
- 81% felt that the event had helped them feel more confident about living with IPF
- 93% thought the event enabled them find out more about where to get further support

“Registration at the North West IPF Meet the Expert event”

“I would like to thank you for putting together the event which I thought was excellent. I have reflected on this since returning home and feel a lot more comfortable with myself and my condition and slept soundly for the first time in ages. Some of my fears have now been articulated and explained.... I really did appreciate the drive and enthusiasm shown by the ‘experts’ and am really pleased I had the opportunity to attend.”

Patient living with IPF

“I learnt more today about IPF than previously and I try to read as much as possible in books/internet.”

Patient living with IPF

“The best seminar I have ever attended in my 80+ years.”

Patient living with IPF

“Great to see we are not on our own and to meet other partners of people with the disease.”

Carer for someone living with IPF
Support groups

There are now around thirty pulmonary fibrosis support groups in the UK and the number continues to grow. We have so far supported thirteen pulmonary fibrosis support groups to set-up with start-up bursaries and expert advice. We have also helped other groups by providing health information and giving talks about the support that is available.

One of the groups to hold their first meeting in 2015 was the North-West Northern Ireland IPF support group. This is the first ever support group in Northern Ireland. At their first meeting they had 59 patients and carers attending.

The groups provide support to their members and are involved in a range of activities such as handing out flyers to GP practices and inviting their local MP to speak. One group (the Wessex Interstitial Lung Disease support group) was shortlisted for a Nursing Times award. They are also conducting research on the impact of the support group on patients, which they plan to publish.

Online resources

Our IPF information hub on our website was Highly Commended at the British Medical Association (BMA) Patient Information Awards 2015. This hub was visited over 252,519 times between November 2014 and October 2015. This makes it the most comprehensive UK online resource for people affected by IPF.

Altogether 13 blog posts talking about IPF were published during this period. The most popular blog was ‘9 things people with lung disease want you to know’ written by Ann Bennett who lives with IPF. This blog struck a chord with so many people living with a lung condition. It was widely shared on social media, had 47 online responses and was viewed 12,712 times since being launched in May 2015.

One of our continuing most popular blogs is ‘IPF the final stages’ by David Forder. It was viewed by 17,013 people during the year. This shows the importance of highlighting end of life experiences in a personal and compassionate way.

Helpline support

Our helpline is there for anyone who wants to get in touch and talk about any problems they may be having whether it is medical, emotional or financial. We have fully qualified nurses and expert advisors who are able to help. Around one in ten calls to our helpline are IPF-related.

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“I lost my husband a few weeks ago to IPF not that old, 54. The thing we both hated was the fact he looked fairly healthy normally, especially if he didn’t have oxygen on people wouldn’t believe he was ill or had a terminal illness. I would never judge anyone like that, it was so wrong. Your post [Ann’s blog] is spot on.”

Blog comment
Influencing health care services and raising awareness

World IPF Week 2015

IPF week continues to grow. This year we focused on ‘#ICareAboutIPF’ to encourage people to interact with us and share stories of how they care. The theme was really well received by patients, their families and health care professionals. Through a radio day we shared the stories of two patients and asked our trustee Dr Toby Maher (a leading specialist ILD consultant) to share his experiences on IPF. This resulted in over 50 radio pieces across the UK, including BBC Radio 5 Live. We also collected 20 case studies from around the UK and sent their stories out to local newspapers which resulted in over 45 articles. Our article on ‘Lack of funding for rare lung diseases’ was published on the front page of the health section on the BBC News website.

With our social media campaign, around 25,000 people saw our Facebook posts at the highest point during IPF week, compared to around 5,000 on a normal week. IPF-related posts with infographics, people’s stories and a call for more research were published every day throughout IPF week and in the build-up. The IPF Week Facebook banner with patients showing signs saying ‘ICareAboutIPF’ had the most shares of any Facebook post we’ve done.

We published two short films of people’s experiences living with IPF on our website and on our YouTube channel. They were viewed over 1,800 times each within the first two months. We also sent out two supporter emails to over 20,000 contacts with an open rate of 20%.

We reached out to health care professionals with the Twitter chat during the week reaching a potential audience of over one million with the Association of Respiratory Nurses (ARNS), Primary Care Respiratory Society (PCRS-UK), the Association of Chartered Physiotherapists in Respiratory Care (ACPRC) and others taking part. We also sent out over 400 IPF week packs to pharmacists across Northern Ireland.

Pulmonary fibrosis and Breathe Easy groups raised awareness by running stalls at their local hospitals and fundraising events. We launched two new support groups just after the week and sent out press releases.

Celebrities also got involved and we featured in West Ham United’s match day programme. Actor David Oakes shared his #ICareAboutIPF photo with his social media followers. While singer Andrea Corr shared her blog on IPF as well. Our Chief Operating Officer took part in a web TV interview alongside Boehringer Ingelheim.

Both the press and social media activity meant that there was an 84% increase in the number of views of our IPF health information. Showing that as well as raising awareness of IPF to the general population, we also highlighted the information available to support people with IPF and their families. The number of people viewing our IPF health information increased compared to last year, with IPF week going from strength to strength.
Parliamentary awareness

Lost in the system: IPF the patient experience in England

We launched the first ever patient experience report ‘Lost in the system: IPF the patient experience’ at Westminster in February 2015. To research this report we conducted a comprehensive patient survey, sent out freedom of information requests to de facto specialist centres and completed a small survey of nurses with an interest in ILD.

Over 50% of MPs were reached by our e-action encouraging people to write to their MPs about IPF. At the report launch 47 parliamentarians attended (44 MPs and 3 peers), two policymakers from NHS England and two from NICE. We invited patients and carers from around the UK to tell their story and meet their MP. We also asked health care professional representatives to attend. An Early Day Motion on IPF (which allows MPs to draw attention to a cause) was started online and was signed by 47 MPs.

Shining a light on IPF the patient experience in Wales

Later in the year during IPF week, we launched our Welsh patient experience report ‘Shining a light on IPF the patient experience in Wales’ at the National Assembly for Wales. We held three focus groups before the launch to get patients’ views. The Deputy Minister for Health gave a speech on the importance of pulmonary fibrosis services. Nine Assembly Members came along to support us alongside people living with IPF and their families.

“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.”

Carer for someone with IPF
Commissioning and guidelines

We continue to influence key policy makers for people with IPF and their families. We published an article on understanding IPF in The Commissioning Journal, which is sent to every Clinical Commissioning Group (CCG) and other commissioners.

In England we sit on the Specialised Respiratory Clinical Reference Group. This group is looking at respiratory specialist services in England such as those for ILDs. In April 2015 the BLF responded to NHS England’s consultation on the prioritisation framework for specialised commissioning. This consultation focused on the proposed principles to be used when deciding how the limited specialised commissioning budget is spent.

We also spoke to the NICE Implementation Team. This team is dedicated to increasing the dissemination and uptake of NICE guidance. Potential areas for future joint work were identified, and the BLF were asked to share examples of best practice service delivery for IPF.

The policy team continues to monitor developments in specialised commissioning policy and to engage with the Specialist Healthcare Alliance as part of this. Locally we have attended specialist ILD commissioning events and meetings with ILD leads to help provide patient and BLF perspective for establishing regional networks and clinical pathways.

In Scotland we continue to support the Scottish Interstitial Lung Disease Group, which provides a forum for discussion and sharing expertise for healthcare practitioners. In Northern Ireland we continue to promote and support the introduction of support groups for people and their families living with pulmonary fibrosis, which we are using to help drive service improvement. In Wales we have played a key part in the formation of the Welsh Respiratory Health Delivery Plan and pushing for the funding of a tertiary ILD service.

Media activity

This year we were able to significantly expand the profile of IPF in the media. We generated over 140 pieces in regional and national media, compared with 70 the previous year. This resulted in a potential total media reach of over 114 million.

We have over 12,000 followers on the BLF Facebook page and over 11,000 followers on Twitter (@lunguk). We post regular items about IPF on both these accounts on new research, personal stories, campaigns and events. This included holding a Twitter chat reaching a potential audience of over one million. We also communicate with other pulmonary fibrosis patient group pages on Facebook. Our online web community (HealthUnlocked) has also seen an increase in activity from pulmonary fibrosis patients and carers. This community provides valuable support for those affected by IPF.

Ashley being interviewed by BBC Radio Solent
Supporting health care professionals

Our series of health care professional events which include a focus on ILDs and palliative care continues. We held nine study days around the UK with over 500 health care professionals signing up to attend. All these study days had at least one talk on ILDs as well as other lung conditions. This enabled us to reach health care professionals who may not have considered learning about ILDs. Two events - in London and Exeter - focused only on IPF, with 95 health care professionals signing up to attend.

We led a session on ILDs at the Primary Care Respiratory Society (PCRS-UK) conference, and asked a patient representative to talk about his experiences of living with IPF to the primary care, health care professionals attending.

We also launched a new pulmonary fibrosis health care professional area on our website, and are building on this to become a comprehensive resource for health care professionals to find out more about IPF and ILDs.

“Learning more about IPF, management and treatment, and how my professional role is involved in this condition and what I can do to help my patients manage their condition. Very useful having a patient speaker”

Health care professional

“Ron’s experience and perspective on living with IPF was inspirational and thought provoking - thank you. Great to meet other professionals working in very different areas of respiratory care to me. Learnt a lot! Mostly see COPD patients so the “basic” information about IPF - aetiology and treatment - was terrific and appreciated”

Health care professional
My experience of a health care professional study day

Sarah, health care professional and BLF Professional

Sarah shares her experience of helping to organise our health care professional study days.

I’ve supported our local Breathe Easy group and put on awareness raising events with the British Lung Foundation for years, so I was really pleased to be approached about becoming a BLF Professional.

I specialise in treating people affected by interstitial lung disease (ILD), a group of conditions that cause inflammation or scar tissue to build up inside the lungs. Most health care professionals see fewer patients with ILD than more common conditions like chronic obstructive pulmonary disease (COPD), and that means there is much less training in this area. I felt that other respiratory health care professionals working in my region could really benefit from an educational event focussing on ILD.

I got in touch with the BLF and suggested there was an opportunity for a study day like this in the South West. So when Bethany, the manager of the BLF Professionals programme, contacted me about organising one, I leapt at the chance to get involved.

Getting involved

The BLF’s health care professional study days offer the opportunity to update your knowledge, learn new skills, network with regional colleagues and share best practice. They consist of a mixture of talks and workshops delivered by specialists, tailored to the needs of health care professionals in that region.

At the event I helped to organise, our aim was to give health care professionals the information and confidence to improve the holistic care of their ILD patients. We covered topics like current and emerging therapies, facing difficult conversations, multi-disciplinary team diagnosis, and the roles of oxygen and pulmonary rehabilitation. We also had the privilege of a hearing a patient living with ILD share her experiences with us, which gave us real insight into the reality of living with this condition.

Inspiring success

The day proved to be a great success. We had loads of positive feedback, and after the event people told me about the changes and improvements they would be making to their practice. One comment that really stayed with me was “the day has inspired me to fight even harder for my patients’ rights”.

I found organising the day with the BLF hugely satisfying, and it was really useful to be able to tailor the education to local needs. And of course, helping to run the event was also a great way of supporting the charity and the work they do to improve treatments for lung disease.

If you want to take part in a BLF study day, I would really encourage you to join the BLF Professionals programme. As well as getting to attend the health care professional study days for free, as a BLF Professional you can have a say on what training you feel you need, which helps them to decide on the education they offer.

As a BLF Professional you can also apply to shadow a respiratory service in another part of the country and get a reduced membership to organisations like PCRS-UK and ACPRC.

When I first started working with the BLF, I didn’t realise what a difference it would make. They really listen to you, and if they can help you to help your patients, they will.
Research into pulmonary fibrosis

Research into patient experiences

This year we collaborated with leading ILD researchers to produce a journal article on ‘The need for patient-centred clinical research in idiopathic pulmonary fibrosis’ published in a special edition of BioMed Central Medicine. We also presented three posters on quality of life experiences at the Pulmonary Fibrosis Foundation Summit in Washington DC and supported an ILD Research Nurse to successfully recruit participants for a study into patient-reported outcome measures (PROMs).

Research studies awarded in 2014-15

Thanks to all our supporters who fundraise for the BLF we continue to fund research into ILDs including IPF. We’ve awarded over £2 million to research so far. In our financial year 2014-15 we awarded £270,000 towards ILD research. We aim to drive research in areas where we feel we can make a real difference to patient outcomes.

We have set up an ILD research advisory board with health care professional, patient and carer representatives. This group has been set up to identify research priorities in ILD. The research advisory board has identified a set of priorities that will ensure we fund research projects that will not only be scientifically robust but that will make the maximum difference to people living with a lung disease.

Can we reduce lung scarring in pulmonary fibrosis? (lab. study)

*Lead researcher:* Prof. Simon Johnson
*University of Nottingham*

*Amount:* £49,781
*Duration:* 12 months, starting Mar 2015

This pilot study will examine the extra-cellular matrix (ECM) protein in the lungs. In fibrosis, cells known as fibroblasts produce too much ECM protein making the lungs smaller, stiffer and reducing oxygen uptake leading to breathlessness, cough and respiratory failure. The ECM in IPF may be physically different which prevents its removal by normal mechanisms. This difference may be the action of proteins which link ECM proteins together stabilising the structure. The study will investigate if blocking the proteins that cross-link the ECM reduces fibroblast growth, fibrotic signals and allows ECM to be broken down normally both in isolated cell preparations and in a mouse model of fibrosis.

Better prediction and treatment of IPF (lab. study)

*Lead researcher:* Prof. Donna Davies
*University of Southampton*

*Amount:* £119,975
*Duration:* 24 months, starting Apr 2015

One obstacle both to development of new treatments to IPF is that the disease course can vary considerably in different patients. Doctors currently have no way of predicting the disease course. This study will evaluate lung cells and lung washings from affected and unaffected regions of patients’ lungs to identify biomarkers that might give insight into disease mechanisms and also try to identify patients who may have disease that is present in what are currently considered ‘normal’ regions of their lungs. This information will help with clinical trial design in the long term provide effective targeted therapies and biomarkers for IPF patients.

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3 Our research funding year aligns with our financial year from July 2014 to June 2015
## Improving clinical trials in ILD through the application of bioinformatics and systems biology

**Lead researcher:** Dr Toby Maher  
Imperial College London  

**Amount:** £100,000  
**Duration:** 36 months, starting Oct 2015

With funding from the Richard Mintz Fellowship

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Improved phenotyping (understanding individual characteristics) of fibrotic interstitial lung disease, including IPF, should provide novel disease insights and improve trial design and delivery. It should also help simplify diagnosis and treatment. This study will use clinical bioinformatic methodologies to re-evaluate current diagnostic algorithms and disease categorisation in ILD. It will also conduct comprehensive disease phenotyping including; transcriptomics, genomics, proteomics and assessment of bronchoalveolar lavage fluid, in individuals with fibrotic ILD.

## Initial research studies awarded in 2015-16

We are part of the way through our funding year for 2015-16 and have funded several more ILD research studies.

### Molecular profiling of the pulmonary epithelium in IPF (lab. study)

**Lead researcher:** Prof. Rachel Chambers  
University College London

**Amount:** £24,726  
**Duration:** 6 months

Understanding disease mechanisms and identifying new drug targets while developing new techniques to study free circulating DNA in alveolar epithelial cells.

### UK Collaboration to Study Inflammatory and Fibrosing Lung Disease in people with Immune Deficiency

**Lead:** Dr John Hurst  
University College London

**Amount:** £24,672  
**Duration:** 24 months

Creating a UK network of clinicians and patients to develop good practice guidelines on granulomatous-lymphocytic interstitial lung disease (GLILD). As well as creating a biobank, developing patient information and encouraging further research.

### A human model of lung fibrosis (lab. study)

**Lead:** Dr Katy Roach  
University of Leicester

**Amount:** £24,072  
**Duration:** 12 months

Ensuring that a model of a human lung with IPF is suitable and can be used to study and develop effective treatments for IPF.

### Immune cell regulation of fibrotic lung disease (lab. study)

**Lead:** Dr Robin McAnulty  
University College London

**Amount:** £25,000  
**Duration:** 18 months

Inhibiting a cell signalling molecule (STAT3) to see if this blocks a type of white blood cell important in immune protection (B-cells) and examining the effect on lung fibrosis.
My challenge taking on RideLondon

Ron is living with idiopathic pulmonary fibrosis, but it hasn’t stopped him taking on the ultimate triathlon challenge.

When I was diagnosed with idiopathic pulmonary fibrosis (IPF) in April 2015, I didn’t think that a year later I’d be writing about completing a ‘Triath-Ron’. Little things like walking upstairs (just normal walking, not running) leave me completely out of breath.

Positive action

I was determined to raise the profile of IPF. I wanted to do more. I wanted to take on a challenge, to show what can be achieved in the face of a seemingly devastating illness. I wanted to take on a challenge not just for myself, but also to help find a cure.

Claire, my yoga teacher, decided she would join me and together we chose to do a triathlon. We started by walking the distance of a half-marathon in May. I can’t run any more, but it’s the distance that counts. Friends from my pulmonary fibrosis support group joined us throughout the day. It was tough.

Next up in June was an epic swim. Just over 2km at 4:30 in the morning! I enjoyed the challenge of swimming against the flow. It was dark, cold and very lonely. For much of the way I was swimming on my own, as most people quickly passed me. I was so proud of myself – I’m not going to surrender to IPF!

Taking on RideLondon

Finally, August was the big one - a huge 193km (120 miles) bike ride from London to Surrey and back again.

No matter how much training I got under my belt, hills would always be a problem and cause my oxygen levels to plummet. But everything that goes up must come down, and riding downhill was always a relief.

The big day arrived. As I waited to cross the starting line, I knew that whatever happened, I would be an emotional wreck. As I expected, the hills and inclines were my biggest challenge. My heart rate increased considerably and my oxygen levels dipped to extremely low levels.

Within the first 50 miles I had severe cramp 4 times in both legs and regrettably had to concede that enough was enough. My consultant has since explained that oxygen gets rid of the lactic acid which causes cramps, but I simply couldn’t take enough in to beat it.

I was devastated at having to finish early. But despite being upset, I was also very proud. I might not have made the full course, but cycling 50 miles with IPF is a huge achievement.

More determined than ever

The support I received from the team at the British Lung Foundation and those around me has been amazing. It gave me the confidence I needed to take on such an incredible challenge. I’m so glad I can give something back.

So far my triathlon has raised almost £3,000. I know this will make a big difference to the support people like me receive, and will also help fund much-needed research towards new treatments and cures.

My challenge has shown me just how many people do care, as the number of friends, family and even strangers who have supported me has been unbelievable. Being able to do this triathlon has made me even more determined to do the very best I can on behalf of everyone who can’t.
**Recapping highlights from Year 1**

Further information about the first year of the project can be found in ‘Providing better support and awareness for people with idiopathic pulmonary fibrosis (IPF) Year 1: IPF project report’.

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<tr>
<th>Getting started</th>
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<tr>
<td>• IPF Patient Charter with ten key calls to action after a wide ranging consultation.</td>
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<th>Providing information and support</th>
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<tr>
<td>• The first ever IPF information pack produced, with over 6,000 packs ordered in the first 6 months.</td>
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<td>• ‘Meet the Expert’ events for people with IPF and their families, with over 77% of those attending telling us that their understanding of IPF had improved.</td>
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<td>• Seven new pulmonary fibrosis support groups launched around the UK so far, providing a bursary and expertise.</td>
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<td>• Over 120,000 visits to the IPF information on our website in one year, with a new IPF hub launched half way through the year.</td>
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<td>• Over 16,000 visits to our IPF blogs.</td>
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<td>• National media reach of over 14 million people during IPF week, and getting our supporters, celebrities and health care professionals involved.</td>
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<th>Influencing health care services and raising awareness</th>
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<td>• Over 70 pieces on IPF into national and regional media, with a total media reach of over 40 million people.</td>
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<td>• Mapping the locations of ILD services and pulmonary fibrosis support groups around the UK and making this information available online.</td>
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<td>• Strengthening relationships for the continued development of ILD services and supporting policy development around the country in Scotland, Wales, Northern Ireland and England.</td>
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<td>• Over 300 of our supporters, including over 150 people with IPF and their carers giving us their views so we could feedback on NICE quality standards.</td>
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<td>• Holding our first Westminster parliamentary event on IPF giving MPs a chance to speak to people living with IPF and their carers; as a result over ten parliamentary questions were asked.</td>
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<td>• Providing training for nurses and affiliated health care professionals on IPF and ILD, with delegates telling us that as a result of the day they are now more knowledgeable about the diagnosis, treatment and prevalence of IPF/ILD.</td>
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Looking forward

We are continuing to build on our work on pulmonary fibrosis and IPF and we are committed to supporting people living with IPF, their families and carers. We will continue to call for action to policy makers, politicians and health care professionals. Three simple steps would transform care for patients with IPF:

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

We will strive to improve our support for everyone affected by pulmonary fibrosis and IPF and raise funds to support this important work. We could not do all that we do without continuing input and fundraising from all our supporters, thank you.

Resources

The BLF has lots of information about IPF and living with a lung condition.

You might find these useful:

- IPF information pack
- Idiopathic pulmonary fibrosis leaflet
- Idiopathic pulmonary fibrosis and other types of interstitial lung disease booklet
- Looking after someone with a lung condition booklet
- Coping with the final stages of chronic lung disease booklet
- Breathlessness booklet
- Oxygen treatment booklet
- Living with a lung condition booklet
- Going on holiday with a lung condition booklet

If you would like any of these health information resources you can order online or telephone.

- To order online go to shop.blf.org.uk/collections/lung-health-information
- To order by telephone call our helpline on 03000 030 555

We also have lots of information about IPF available on our website at www.blf.org.uk/IPF

Whether you have a lung condition or care for someone who does, we’re here to help. You can get in touch with our helpline at 03000 030 555.

Acknowledgements

Boehringer Ingelheim is the founding partner of the British Lung Foundation IPF programme, and Roche Products Ltd. is a supporting partner of the programme. Both companies contributed financially to the IPF project work. Neither company had any influence over the content.

Thank you to all our supporters who have helped raise funds for our research and work on IPF. We couldn’t have done all our activity and funded all our research without you.

We really appreciate the help and views from our supporters living with IPF, their family and carers who have taken part in our parliamentary events and focus groups. A special thanks to members of our IPF advisory group who have given us invaluable advice in the first two years of the project.
Our IPF Patient Charter

Idiopathic pulmonary fibrosis (IPF) is a chronic, progressive and incurable lung disease.

Killing over 5,000 people a year in the UK - it is on a par with far better-known conditions such as leukaemia. The condition has been on the rise for decades, but remains poorly understood and with limited treatments and no cure.

The disease often acts with devastating speed, with barely half of patients surviving the three years after diagnosis.

With rates of IPF continuing to rise, the British Lung Foundation (BLF) is calling for more to be done to help fight IPF and the devastating impact it has on the lives of patients and their families.

The BLF believes that people with IPF and their families have the right to:

1. Timely and accurate diagnosis and care, involving an appropriately skilled, specialist multidisciplinary team
2. Access to seamless, well-integrated health and social care services, including ambulatory and domiciliary oxygen services personalised to the needs of IPF patients
3. High quality information about the condition, including full details of all treatment, clinical trials, transplant, support and service provision options available to them
4. Specialist palliative care and end-of-life care; supported by the Gold Standards Framework where appropriate
5. Access to dedicated peer support networks, both for patients and their carers, in person or digitally

The BLF calls upon UK governments and NHS leadership to:

6. Ensure sufficient funding is made available for the FULL implementation of the NICE guidelines for IPF
7. Build and resource clinical networks to ensure seamless care between providers at all stages of the patient pathway, and a co-ordinated approach to IPF management including access to clinical trials
8. Increase funding for IPF research to a level that will tackle the considerable, and growing, impact of the disease in the UK
9. Recognise the urgency of need in patients with rapid disease progression, via swift access to specialist care, appropriate prioritisation on transplant waiting lists, and prompt social care assessment and response
10. Conduct awareness campaigns to improve public and primary care recognition of the condition and its symptoms, and encourage employers and providers of insurance, travel and financial services to better meet the needs of people with IPF
Supporting us

Ruth Sabella, IPF Project Manager

“Thank you to everyone who has supported us. I’ve heard many inspiring stories, but also numerous cases when care should have been better. A single bad experience is one too many. We’ll continue campaigning for everyone to have the support they need.”

Corporate partnerships

If you or your company would like to find out about becoming a partner or supporter of the IPF project, please contact our corporate partnerships team.
Email: corporate@blf.org.uk

Patients or carers

If you have IPF or know someone who does and would like to help us continue to raise awareness of the condition, there are lots of different ways you can get involved. You can tell us your story, join a focus group or fundraise for IPF research and support.
Email: patientvoice@blf.org.uk

Health care professionals

If you are a health care professional and would like more information on the IPF project, please contact our stakeholder engagement team. You can also become a BLF Professional and support our work. Or you can fundraise for us.
Email: HCP@blf.org.uk

“Our dad, Rod, died suddenly from IPF. It was a massive shock. We shared an unshakeable bond as a family. Ever since dad’s death we have worked to raise funds for the British Lung Foundation for research into IPF. Something good must come from our loss – that’s our promise to him.”

Kate and Tracey, pictured with their mum Pam

Donate today at: www.blf.org.uk/ipf-appeal or text IPF to 70500 to donate £5