Inspiration

INSIDE

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Inspiration

Idiopathic Pulmonary Fibrosis (IPF)
Report February 2016

Action for Pulmonary Fibrosis

www.actionpulmonaryfibrosis.org

British Lung Foundation

www.blf.org.uk

Boehringer Ingelheim

www.boehringer-ingelheim.co.uk

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Respiratory diseases affect one in five people in the UK and cost the NHS a staggering £4.7 billion per year[1]

Yet while many people will be familiar with asthma, COPD and lung cancer – there are a host of other lesser known respiratory conditions that are both under-recognised and under-funded, which represent a growing burden for the NHS.

One such condition is idiopathic pulmonary fibrosis (IPF) – a devastating and incurable disease where the lungs become irreversibly scarred over time. Progression can be rapid and average survival is just three years from diagnosis. If IPF were a cancer it would be the eighth most common cancer in the world, but most people have never heard of this condition.

So why should healthcare professionals, policymakers and the general public sit up and take notice of IPF and other rare lung diseases?

Firstly, IPF kills more people every year than leukaemia or ovarian cancer. Certainly the relief often felt by patients on receiving a diagnosis of IPF can be short-lived when they realise the prognosis may be as bad as, or even worse than, it is for lung cancer.

There are limited treatment options and no cure. Worse still, no-one knows the cause.

On top of this the numbers of people with IPF and other interstitial lung diseases are increasing every year. But I know as a GP there are huge gaps in awareness, and difficulties in diagnosis, so this is likely to be just the tip of the iceberg. Indeed, it is forecast that there will be 20,000 hospital admissions related to IPF in 2020, costing the NHS £20 million, and patient groups Action for Pulmonary Fibrosis and the British Lung Foundation believe much more could be done.

So I welcome this initiative by Boehringer Ingelheim, working with patients, patient groups and leading clinicians to help put IPF in the spotlight and call for greater awareness and understanding of this devastating condition.

Isn't it time we sat up and paid attention?

Dr Hilary Jones

GP, presenter and writer

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The hidden threat of IPF

IPF has a devastating impact on the lives of both patients and families. Patients are often unfairly marginalised because of the social stigma attached to what has been thought to be a smoker's disease.

It's the disease no-one has heard of, yet the number of people being diagnosed with idiopathic pulmonary fibrosis (IPF) in the UK is steadily increasing year on year.[1]

Indeed, if IPF were a cancer it would be the eighth most common tumour type globally with a similar mortality rate to leukaemia and ovarian cancer. The average survival rate for IPF is little better than inoperable lung cancer[2] - with half of those diagnosed dying within the 3 years of their diagnosis.[3]

But by following a simple and clear disease management pathway, it is possible to limit the brutal effects of this chronic, progressive and irreversible lung disease for both the patient and stretched healthcare resources.

At the forefront of the issue is poor awareness of the condition and the consequent high levels of misdiagnosis. And, although the National Institute for Health and Care Excellence (NICE) has issued guidelines on how best to diagnose and manage IPF,[3] there are still significant hurdles to overcome before patients receive the care they are entitled to – and the same level of care afforded to patients with other life-threatening diseases.

So what exactly is IPF and why haven't we heard of it?

The nature of the disease

IPF is an incurable lung disease with no known cause, although it is believed to have both genetic and environmental links including exposure to certain viruses, such as Epstein-Barr, and contact with industrial hazards.[4] Patients with the disease suffer irreversible scar tissue formation (fibrosis) in the lungs – resembling a honeycomb pattern – making the lungs less able to take in oxygen and ultimately leading to respiratory failure.[4]

WHAT TO LOOK OUT FOR

NICE guidance suggests clinicians should be aware of IPF when assessing patients with the following clinical symptoms:[3]

- AGE OVER 45 YEARS
- PERSISTENT BREATHLESSNESS ON EXERTION
- PERSISTENT COUGH
- BILATERAL INSPIRATORY CRACKLES WHEN LISTENING TO THE CHEST
- CLUBBING OF THE FINGERS
- NORMAL SPIROMETRY OR IMPAIRED SPIROMETRY USUALLY WITH A RESTRICTIVE PATTERN BUT SOMETIMES WITH AN OBSTRUCTIVE PATTERN

Perhaps surprisingly, though, there are currently 15,000 people affected by IPF in the UK with 5,000 expected to die within the next 12 months,[5] and the same number receiving a diagnosis every year[4] – making it one of the most common of the so-called rare diseases.

Need for early diagnosis

Clearly given patients' short life expectancy it is important the disease is diagnosed early so they can receive the necessary support to help them better manage their condition and make the most of the time they have left with family and friends.

However, there is no specific diagnostic tool for early recognition of IPF. And as the disease's primary symptoms of breathlessness and cough are common to a number of conditions, many people are initially misdiagnosed with asthma, COPD or cardiovascular disease.[6] This means patients commonly receive the wrong treatments.

So what can be done?

Patients don't receive the best level of care

NICE also requires that diagnosis is carried out by a specialist multidisciplinary team (MDT).[3] However, to access an MDT patients need to be referred by their GP. Worryingly, in a survey of 122 patients and carers carried out by the British Lung Foundation,[7] patients said they struggled to get a diagnosis, and often found themselves left alone to navigate their GP, local hospital and specialist centre for their care.

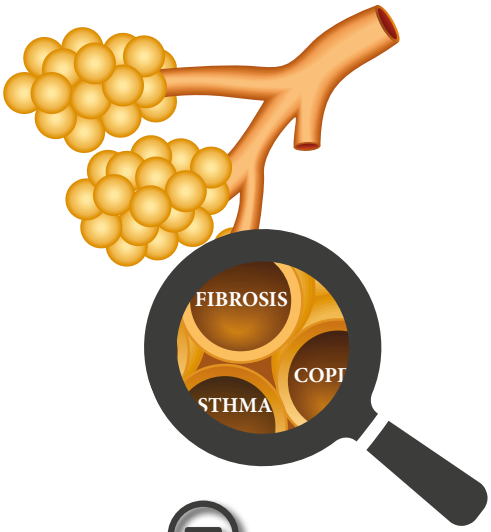
According to the survey, 30% of patients had to wait six months or more for a referral to a specialist, and 12% had to wait more than two years. And even after seeing a specialist, almost 40% reported waiting three months or longer before they were told they had IPF.

To help patients understand their condition and coordinate their care NICE recommends all patients should have access to an interstitial lung disease specialist nurse, but again, a significant proportion (36%) reported they had not been offered this service.

A similar picture was seen in the disease information patients receive. NICE recommends that both

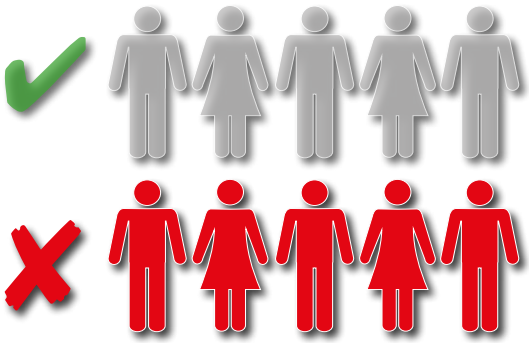
IPF: FACTS AND FIGURES

IPF is difficult to distinguish from other lung disorders



50%

are misdiagnosed



About

5,000

people are diagnosed in the UK **EVERY YEAR[4]**

Delays in diagnosis are frequent and have a **highly negative impact on patients' quality of life[9]**

verbal and written information should be provided to patients, but the survey showed that, in reality, 47% either didn't understand the verbal information they were given, or didn't have it explained at all; and almost half were given no written information.

As one respondent to the BLF survey[5] wrote:
"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 … I felt like I was being sent home to die."

Stop patients being marginalised

IPF disease has a devastating impact on the lives of both patients and families. Patients are often unfairly marginalised because of the social stigma attached to what has been thought to be a smoker's disease. And treatment, even for those patients who do find their way through the healthcare system, is currently limited and includes: oxygen therapy; pulmonary rehabilitation and lung transplantation – although the latter is extremely high risk and available to very few eligible patients.[7]

To ensure the best use of limited healthcare resources, and to allow people with IPF to manage their condition and enjoy a good quality of life for the limited time remaining to them, early and accurate diagnosis is essential.

With predictions that in five years' time there will be close to 20,000 hospital admissions every year from IPF in the UK, costing health services almost £20 million in hospital care alone[8], what is clear is that those providing IPF services need to follow NICE guidelines for patient care – and government and policymakers need to prioritise IPF as though it were a cancer.

Without this necessary patient support the NICE pathway recommends, more NHS money will continue to be spent inappropriately in this disease area and the burden on the healthcare system will grow exponentially with an ageing population.

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[8]V. Navaratnam, A. W. Fogarty, G. R, T. McKeever and R. B. Hubbard, "The Increasing Secondary Care Burden of Idiopathic Pulmonary Fibrosis: Hospital Admission Trends in England From 1998 to 2010," Chest Journal, vol. 143, no. 4, 2013. (Last accessed February 2016)
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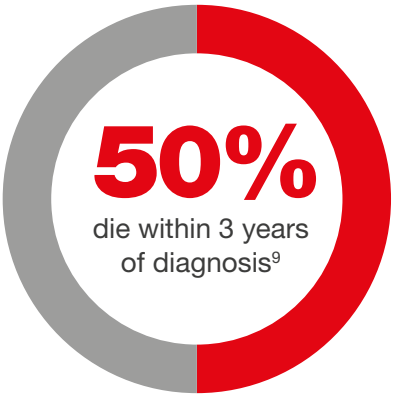
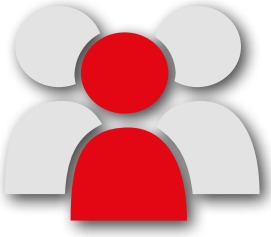
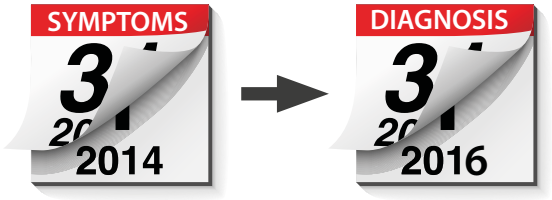
IPF: FACTS AND FIGURES



1 in 3
people are expected to die within the next 12 months
average life expectancy after diagnosis is 3 years[3]





Average time to diagnosis is
1–2 years
after onset of symptoms

IPF can be every bit as terrible as cancer, and often worse.

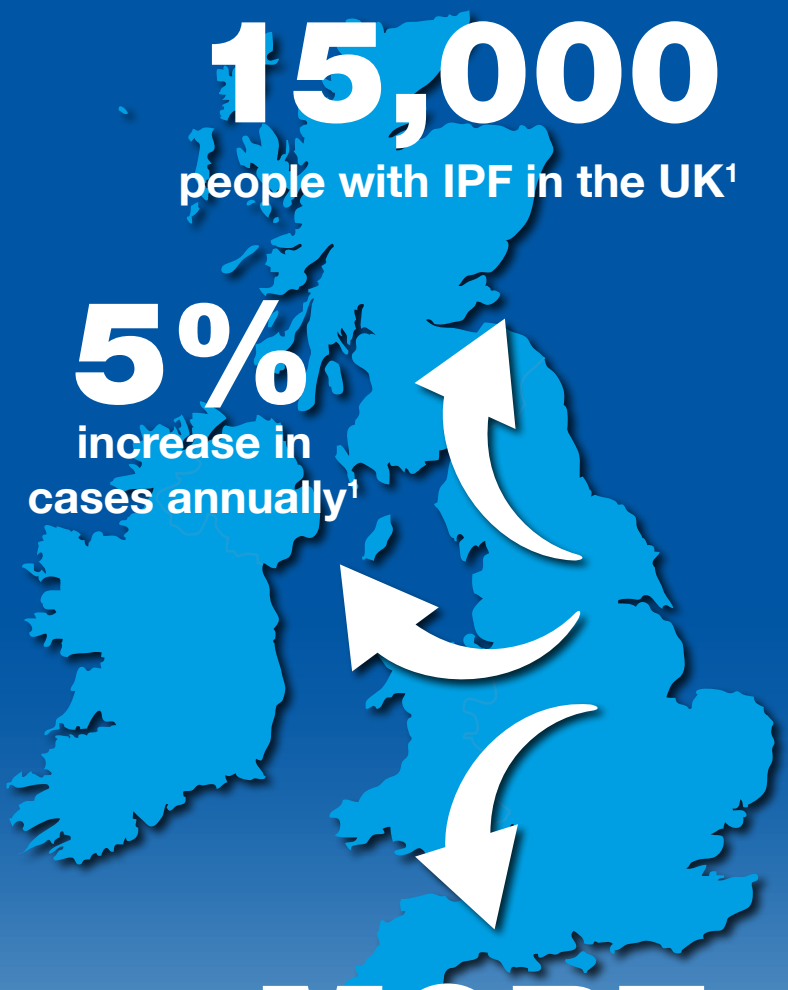
"It first started five years ago. ***I was always fit and active*** *– you need to be as a swimming teacher – but then I started becoming breathless when walking and,* ***over the next six months, I deteriorated so quickly I couldn't climb a flight of stairs.***

My GP originally suspected I had asthma. ***It took me four years to get to a person who actually understood what Idiopathic Pulmonary Fibrosis (IPF) is.***

The hardest thing for me now is that I can't work*. And my social life has changed immeasurably.* ***The isolation and loneliness I experience is very difficult to deal with****, but I try and deal with each day in a positive way.*

When I do tell people about my condition, they can't understand ***why more is not done to raise awareness of IPF****. If I had been diagnosed earlier,* ***I feel my life would be very different now."****

***Patient quotes are compiled from a number of interviews and sources**



Fifteen thousand people already have IPF in the UK alone, and the number of cases is increasing by 5% annually. In fact, more people now die every year from IPF than from leukaemia[1] - a condition that has benefited from greater awareness and research funding[2]

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The reality of IPF – A patient's insight

IPF is a fatal disease that's all the more difficult for patients due to the lack of awareness and investment in supporting people with the condition. Here **Tony Gowland**, IPF patient and ambassador for the charity Action for Pulmonary Fibrosis, highlights the reality of life with IPF and the seriousness of the current situation facing patients in the UK.

How would you describe the diagnosis process? As someone who had no previous knowledge of IPF, would you say it was straightforward?

It was probably late 2011 when I noticed I was coughing more than normal. I was cycling to work and back, reasonably fit and eating a healthy diet with the perfect body mass index. What could possibly go wrong? I started going to my GP with suspected chest infections and was given a variety of antibiotics over the next couple of months. Nothing changed and an equally inconclusive chest X-ray was followed by a CT scan, which resulted in my diagnosis of IPF. Later I realised I was quite fortunate that my diagnosis was made before I had experienced breathlessness. For the majority it seems that isn't the case.

How were you told you had IPF?

Any positives experienced up until the point of diagnosis were washed away by the manner in which the results were communicated to me. I was told in a reassuring voice that it wasn't cancer, or 'anything like that' – it was IPF – a condition I'd never heard of. My consultant at the time advised that if I wanted any details, the British Thoracic Society's website was very comprehensive. Only when I did as he suggested did I realise my diagnosis meant impending death – I took the bombshell full in the face, it was simply heartbreaking.

What has been your experience in accessing care or advice?

The lack of awareness among primary care physicians is legendary, but maybe we should be asking why? In 2014 there were 9,800 GP surgeries around the UK, during which time only 5,000 people were diagnosed with IPF. No GP ever diagnoses IPF, it's not their job – they see coughs and chest complaints that they either sort out or pass on to the next level. They're not given the proper information or tools to deal with and treat IPF patients and so the dedicated GP is effectively hung out to dry.

Where do you turn to for information about IPF?

Having IPF has sometimes struck me as if I'm in a foreign country, in trouble, aware of what I need but unable to make anyone understand.

Certain services are a real lifeline, having access to a specialist nurse for example is akin to having a translator – someone who can listen empathetically and then direct me towards the people or places that can help. Our concerns, fears and needs are understood only by our beloved carers and surprisingly few doctors and nurses.

Support groups and online chat forums are all that exist to give sufferers and carers alike the opportunity to share experiences and trade the empathy and compassion so necessary to help us navigate our way through what lies ahead. I find it worrying that vulnerable sufferers of this dreadful condition have to turn to social media for advice.

Do you think public awareness also has a role to play? For example, you don't hear much about IPF outside of the healthcare industry.
Yes, having IPF teaches you that you are not 'mainstream' as a health concern – according to the popularity stakes anyway. You are unlikely to meet anyone outside a medical establishment who has even heard of it.

Since my diagnosis almost four years ago, I have not once stumbled upon the words idiopathic pulmonary fibrosis being mentioned on TV, in the media or in general conversation.

The more IPF becomes a part of our collective consciousness, the more likely it is that things can change. That's just how the world works, sadly.

You mentioned cancer, which gets a lot of press. Do you feel that such coverage perhaps results in disproportionate awareness for certain illnesses?
It's frustrating in the extreme that I suffer from a disease that kills more people than leukaemia and yet I believe levels of awareness and funding for research is utterly out of balance.

"I find it worrying that vulnerable sufferers of this dreadful condition have to turn to social media for advice."

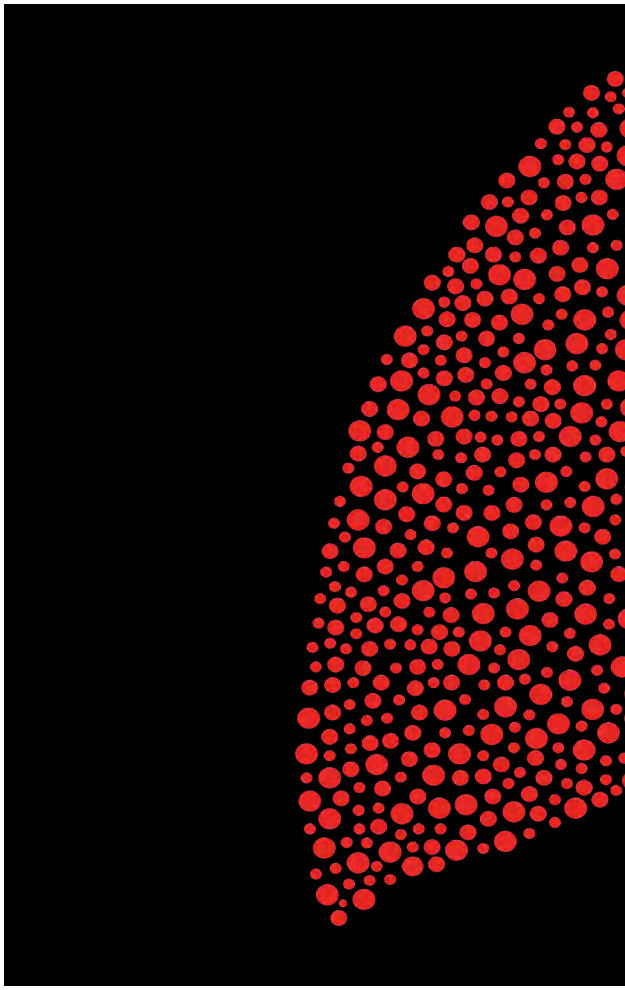
Does UK health policy regarding IPF funding and treatment need to change? What changes do you think would make the biggest difference?
There seem to me to be vast discrepancies in the quality and availability of care throughout the UK; yet as the NICE guidelines show, our basic healthcare needs are not that demanding. In the grand scale of things, the linchpin is a specialist nurse. Someone who understands not only the disease but our needs and the 'system' within which we find ourselves.

When those needs are fulfilled, they really help to alleviate suffering and allow us the chance to get the best we can from the limited time we are afforded. Surely the name of our disease shouldn't be the factor that determines the standard of our care?

We're not surprised when people have never heard the phrase "I'd rather have cancer". Sadly, here in 'IPF World' it's quite commonly used. We know why, it's time others did too.

For more information go to www.actionpulmonaryfibrosis.org

IPF: The unknown killer



Elizabeth Bray, Trustee and Charity Secretary at Action for Pulmonary Fibrosis; and **Ruth Sabella**, IPF Project Manager at the British Lung Foundation, reveal the issues currently facing IPF patients, and what can be done by the government and healthcare providers to improve quality of care and mortality rates.

Around 5,000 people are diagnosed with IPF in the UK each year, and that figure is climbing. Average life expectancy after diagnosis is three years, and more patients die each year from IPF than they do from leukaemia.

But the real issues exacerbating the problem for patients are the inconsistencies in quality of care, the struggle for diagnosis and the gaps in education – among public, policymakers and healthcare professionals alike.

What do you consider to be the main challenge in the UK in regard to IPF?

Elizabeth: Without a doubt, the main challenge is too little funding. Funding for research into IPF is inadequate, and limited access to specialist centres has resulted in variations in care for patients around the UK.[1]

Ruth: Funding is definitely the core issue, and I suspect that the root cause of this is a lack of awareness around the disease. Yet in stark contrast, more people die from IPF than from leukaemia every year.

"While improvements to IPF care are being made, there is still a disparity between best practice guidance and the reality for many patients when accessing services"

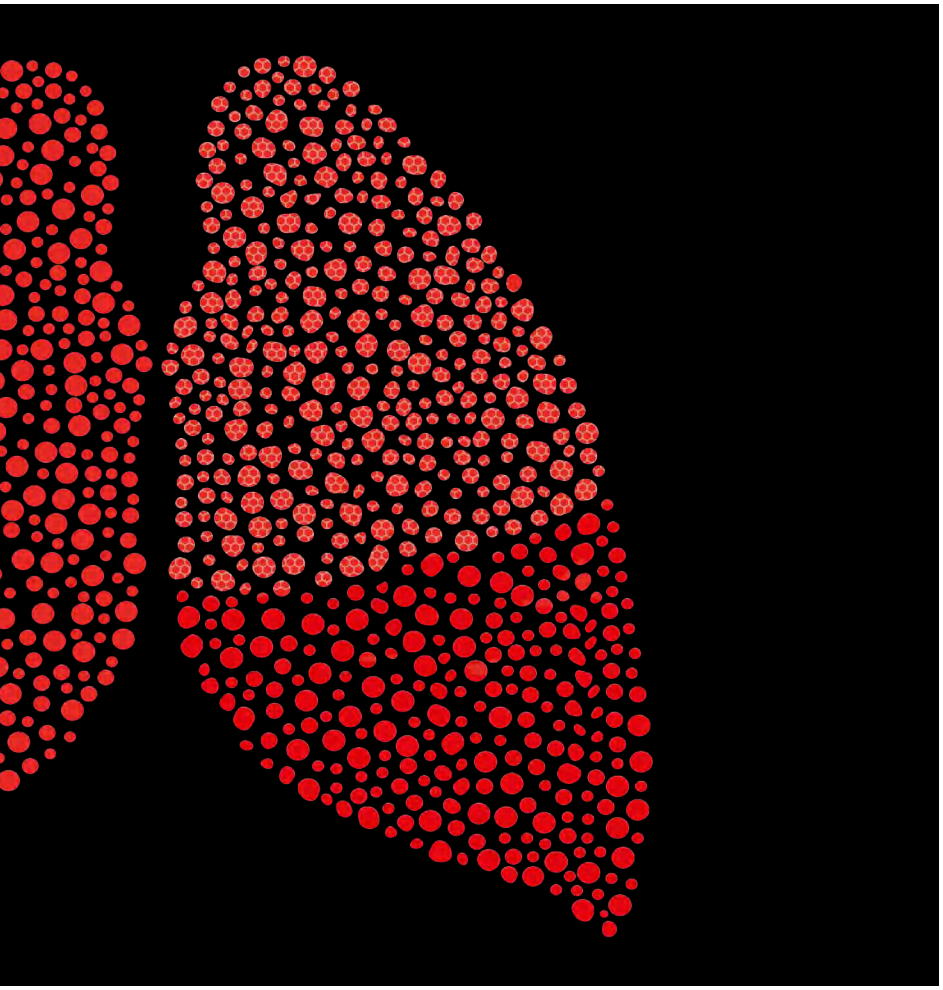
Ruth Sabella, British Lung Foundation

What impact has limited funding had on IPF treatment in the UK?

Elizabeth: There is very limited knowledge of the disease at primary care level, which often results in late diagnosis, and this in turn has a negative impact on the patient – especially now that drugs are available that can slow progression of the disease. Educating GPs about the symptoms of IPF will encourage referral for the appropriate testing to achieve diagnosis much earlier.

Ruth: I would also highlight the need for more ILD specialist nurses, who play a vital role in helping patients navigate their care, and for a better coordinated approach between specialist, local and palliative care. You need a specialist to diagnose IPF – and GPs will only see on average one IPF patient every seven years – so directing resources to educating the general public and GPs about breathlessness and alternative diagnoses, as well as increasing policymakers' and healthcare professionals' understanding of IPF is essential.

The BLF released a report earlier this year, *'Lost in the System: IPF Patient Experience in England'*, in which a direct quote from the carer of an IPF



patient sums up this situation succinctly:

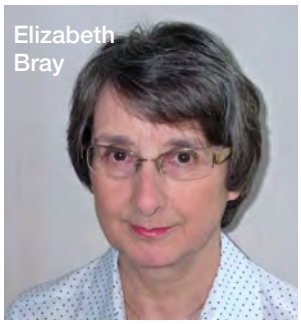
"There was a general lack of understanding of the condition's progression by GPs and others, and no services available to support patients and their families at the end of life. In my view, the health system let my mother, and us as a family, down badly at the end of her life, due to disjointed services, breakdowns in communication, and the fact that there was no one person to coordinate health and care services. It was truly a traumatic experience."[2]

What should be done in the UK to improve treatment and outcomes?

Ruth: The issue that needs most urgently addressing in the short term, and that would have the biggest impact, is improving access to seamless, well-integrated health and social care services – from diagnosis to end-of-life care.

An important research priority should be the creation of a UK biobank, with linked clinical data to support an interstitial lung disease (ILD) research network. This would allow a formal network of ILD researchers to obtain the samples they need to conduct research more rapidly.

Ruth Sabella



Elizabeth Bray

Elizabeth: While the UK is undoubtedly a world leader in the field of IPF, funding for IPF research in the UK is still far behind funding in the USA. As Ruth says, the UK could do much more to understand the biology of pulmonary fibrosis through generating biobanks and tissue sharing consortia.

In October 2015 Action for Pulmonary Fibrosis published a comprehensive report '*Working Together: Delivering a better future for patients with IPF*'.[1] This was the result of a comprehensive patient survey which showed that the UK could also do a lot more to improve the primary and secondary care interface to ensure that all patients are able to take part in pulmonary rehabilitation, get appropriate oxygen therapy and have contact with an ILD specialist nurse.

Is there anything you'd say the UK does well?

Ruth: The NICE guidelines published in 2013 and quality standards published in 2015 represent a good approach to treating and managing IPF, but while improvements to IPF care are being made, there is still a disparity between best practice guidance and the reality for many patients when accessing services.

We need to get to a point where all those providing IPF services are following the NICE guidance and pathway, where all patients are given Information Standard-approved information on IPF at the point of diagnosis, and all patients have access to a specialist ILD nurse to help them navigate their care.

Elizabeth: The UK performs well at research, comparatively. And when patients do receive treatment and care at specialist centres, as identified in our recent report, it is excellent. Additionally, the work that is done across the country by local support groups to provide accurate information and support to patients is vital. But we need to improve recognition of the disease in primary care, implement all NICE quality standards – especially oxygen therapy and pulmonary rehabilitation – and increase availability of anti-fibrotics to all patients with pulmonary fibrosis, not just those with a forced vital capacity (FVC) of 50%–80%.

In addition, we need to improve our screening of patients at risk of IPF and get better at identifying biomarkers that can identify and predict response to therapy. In the long term, we must find new ways to modify the disease with the ultimate aim of reversing fibrosis.

That will only come with increased funding, increased awareness and, of course, the involvement of the patient and the pharmaceutical industry.

What role can pharma and patients play in raising IPF awareness and improving the development of, and access to, treatment?

Ruth: As the research community, including pharmaceutical companies, enters a new era of clinical research activity on IPF, more patient involvement and partnership becomes pivotal. It is vital that pharmaceutical companies draw on the experience of patient groups to inform the development of their products. It's essential that patients are involved every step of the way in leading and informing research priorities. Only through collaborations in research design, study conduct and dissemination, can we ensure that patient-reported outcomes (PROs) are used as clinical endpoints.

Elizabeth: In the development of drugs it's crucial that pharma companies fully understand the experience of patients living with the disease. Patients need to be fully involved in all aspects of their treatment and care. Most healthcare considerations relate to improving survival but, for many patients, this is of secondary importance to improving quality of life. The development of personalised care is crucial to enabling patients to understand the disease and what they can do to ensure the best quality of life.

"Funding for research into IPF is inadequate, and limited access to specialist centres has resulted in broad variations in care for patients around the UK"

Elizabeth Bray, Action for Pulmonary Fibrosis

Finally, why should UK healthcare policy makers be aware of IPF?

Elizabeth: IPF is likely to become a considerable healthcare burden in the future. It is not rare, it is becoming more common and treatment is expensive. Patients suffer severely debilitating symptoms and survival time is very limited. Although classified as a rare disease, it is as common and kills more people than some cancers, but currently doesn't attract the attention or funding it needs to provide appropriate care management for all patients across the country.

There are at least 15,000 patients in the UK, and 13 patients die every day from IPF. There are three times as many people living with IPF than motor neurone disease (MND), but the age of people getting it is the same, the survival rates are the same (half of IPF patients are dead within three years of diagnosis) and, like MND, there is no cure.

Ruth: IPF is a lung disease with no known cause or cure. Every year in the UK around 5,000 people die because of IPF – more than the number of people dying each year from conditions such as leukaemia, ovarian cancer or kidney cancer.[2] It is predicted that in 2020 there will be close to 20,000 hospital admissions from IPF in the UK, costing health services almost £20 million.[3] The true cost, however, is likely to be much more: the estimate does not take into account expensive new drugs used to treat the condition or care outside of hospitals.

If IPF were a cancer, it would be the eighth most common cancer in the world. The average age at diagnosis is just above 70 and, shockingly, almost half of those affected by the disease will die within just three years from diagnosis. The burden of this disease on both the country's finances and on human lives, as well as the lack of prioritisation it receives in light of this, is evident.

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For more information go to www.actionpulmonaryfibrosis.org or www.blf.org.uk

Every year, more people die from IPF than leukaemia.

“***It started with a cough***. *Nothing major, just an irritating cough.* ***It wasn't until I'd had bronchitis four times over the space of three years that I was referred to a specialist respiratory centre.***

When I was diagnosed ***I had no idea what Idiopathic Pulmonary Fibrosis (IPF) was***. *It was hugely sobering when* ***I searched on the internet and found that survival times are just 2–3 years.***

In order to actively handle my condition, I was prescribed oxygen. *It's difficult to sleep because you get tangled up in the tubes - and I can't travel anymore -* ***but I'm a fighter and I want to live longer.***

I do wish more attention was given to this disease. *I'm on the lung transplant list, but it's a huge risk. I prefer to trust in the future and the hope that new research might bring;* ***you can't just stick your head in the sand and give up.***”*

*Patient quotes are compiled from a number of interviews and sources

Up to

50%

of all people with IPF are initially told they have other respiratory conditions[1]

Around 50% of people with IPF are initially told they have other respiratory conditions, including asthma and COPD,[1] leading to delays in referral to specialist care that can have a highly negative impact on a patient's quality of life.[2] Under-funding and under-recognition of IPF means more patients now die from this condition compared to leukaemia and ovarian cancer.[3]

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References

[1]Collard HR, Tino G, *et al*. Respir Med 2007;101:1350–4

[2]http://www.blf.org.uk/Page/IPF-Patient-differential-diagnosis-of-COPD (Accessed February 2016)

[3]http://www.bbc.com/news/health-29363887 (Accessed February 2016)

The sounds of IPF

Every year, more people die from idiopathic pulmonary fibrosis (IPF) than leukaemia. Here **Steven Wibberley**, Chief Operating Officer of the British Lung Foundation, and **Professor Klaus Dugi**, Medical Director and Managing Director at Boehringer Ingelheim UK & Ireland, say getting an early and accurate diagnosis is fundamental to managing this growing health burden.

Why is IPF so difficult to diagnose?

Klaus: Medical students are taught that frequent diseases are frequent and rare diseases are rare. If a patient presents to a GP with dry cough or breathlessness the natural assumption is that this could be a more frequently seen condition like asthma or COPD. This is a disease awareness issue because many physicians will not have heard of or feel comfortable with a diagnosis of IPF.

Steven: The number of patients being diagnosed with IPF is on the increase, so early and accurate diagnosis is critically important. Part of the problem is that the two main symptoms – a dry cough that doesn't seem to go away and breathlessness upon exertion – are also present in other lung conditions. If people have timely referral and an accurate diagnosis of IPF, they are more likely get the support and services they need, such as prompt access to good treatment, pulmonary rehabilitation and a lung transplant assessment. They also have a greater opportunity to tap into support from others with the same illness.

What tests do physicians perform to diagnose IPF?

Klaus: A physician will firstly take a full medical history, then perform a physical examination to listen to the patient's lung. This is very important because there are specific crackles you can sometimes hear in patients with IPF that are very different to the sound of asthma, for example. This will be followed by a spirometry test to determine whether the patient should be referred to a specialist for further investigation – such as a CT scan or, in rare cases, lung biopsy.

"With the number of people developing IPF and other interstitial lung diseases escalating in their thousands every single year, there is an urgent need for new diagnostic criteria and aggressive treatment of early disease"

Steven: The important point here is that a multidisciplinary team of health professionals meet together to make sure a diagnosis is accurate and correct, and to identify the best treatment support programme for the individual patient. It's a real worry that some patients aren't getting access to the specialist lung doctor, radiologist or nurse – all of whom need to be involved in these care decisions.

On top of this, misdiagnosis is also a concern. People will often say they were initially diagnosed with COPD – a far more common disease – and treated for that for several years before eventually being told they have IPF. The worry is that people are missing the opportunity to start the right treatment and will feel neglected by the system. That's why it's really important to get a correct and accurate diagnosis as promptly as possible.


Steven Wibberley (l) and Professor Klaus Dugi

How do patients respond to an IPF diagnosis?
Steven: When a patient is first diagnosed with IPF their initial reaction may be 'thank goodness it's not lung cancer'. However when they realise the prognosis can be worse, it can really hit the individual patient, their families and carers hard. The combination of psychological and physical effects can have a devastating impact – knowing they are living with a disease that might have some treatment but doesn't have a cure and will probably shorten their lives, alongside progressively becoming more out of breath.

Klaus: We shouldn't underestimate what a shock being diagnosed with IPF is and why it's important to give patients access to support networks and good quality information – to be able to share experiences – and for the family to understand both what to expect in the future and what they can do in the present to help their loved ones as much as they possibly can.

What can patients do to help themselves?
Steven: We would say to any patient with a respiratory condition that there are things they can do to manage their disease as best as they can – stopping smoking and taking exercise being key. Exercise may seem counterintuitive for people who are short of breath but helps to make the most of their lungs even when they are not functioning well. It is also important for patients to learn about the disease because being informed means they can have much better conversations with their healthcare professionals, ask better questions, and be involved in decisions about their care.

"We shouldn't underestimate what a shock being diagnosed with IPF is and why it's important to give patients access to support networks and good quality information"

Can patients with IPF go on the lung transplant list?
Steven: A transplant is only suitable for a very small number of patients – they have to be ill enough to need the transplant but healthy enough to survive what is a very major operation. We know in this country there are not always enough people who fill in their donor cards and go on the donor list. So a transplant is an opportunity for some patients, but not for enough.

Klaus: This is why research is so important and why it needs to be appropriately funded so we can extend patients' lives until they get to that very late stage that they may only be rescued by a lung transplantation. There's more research ongoing now compared to 10 or 15 years ago to better understand the disease and to find new therapies that will prolong survival, but there is so much more to learn.

What is being put in place to help medical professionals make those diagnoses earlier?
Steven: It's clearly a real challenge, particularly for health professionals working in primary care and general practice because they won't see many cases of IPF – perhaps one every few years. How might they diagnose or suspect IPF when day in, day out, they are seeing other respiratory problems?

Klaus: The plea that both Steven and I have to physicians is that if you feel someone is not your typical asthma or COPD patient – if you hear something or it doesn't respond to treatment - refer that patient to a specialist sooner rather than later. The same also applies to the patient themselves.

Steven: I agree. If you are being prescribed medication for COPD or asthma and your symptoms are not well managed – have a conversation with your GP and ask for a referral to a specialist. Getting that diagnosis as early as possible is absolutely key.

For more information, go to www.blf.org.uk or www.boehringer-ingelheim.co.uk

Half of all IPF patients are initially misdiagnosed

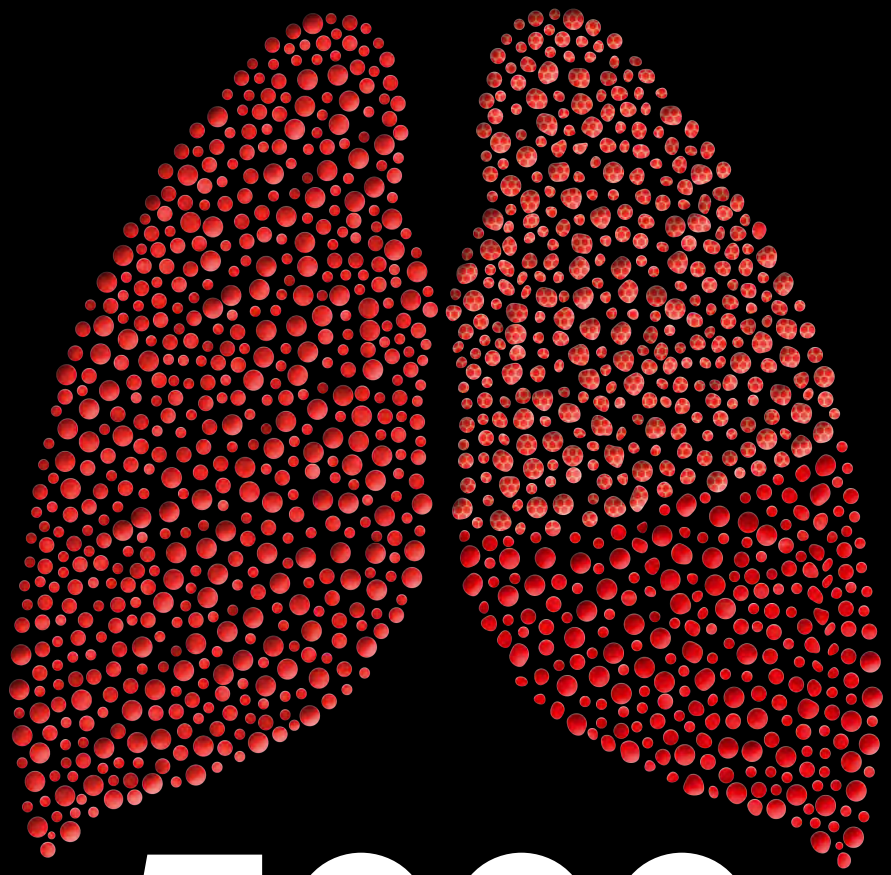
*"**I started to feel quite tired and breathless**, then the things I could once do easily suddenly became more difficult – like climbing the stairs and doing the housework.*

My GP initially prescribed inhalers, but they didn't have any effect. ***It wasn't until some months later that he heard some crackles on my lungs and referred me to a specialist.***

That's when they discovered the scarring and I was diagnosed with Idiopathic Pulmonary Fibrosis (IPF). ***I was told there is currently no cure, and that around half of people affected live no more than three years after diagnosis.***

If more people knew about IPF** and the signs to look out for then getting an early and accurate diagnosis would be more likely –* ***and we would have a better opportunity to start the right clinical management."*

*Patient quotes are compiled from a number of interviews and sources



5000

people will be diagnosed with IPF in the UK this year[1]

This year, 5,000 people will be diagnosed with IPF in the UK. There is no cure, and just half will live for more than 3 years after their diagnosis.[1] Up to 50% of patients are initially diagnosed with other respiratory conditions, including COPD and asthma.[2]

Boehringer Ingelheim

References

[1]https://www.blf.org.uk/Page/IPF-report-Lost-in-the-System (Accessed February 2016)

[2]http://www.blf.org.uk/Page/IPF-Patient-differential-diagnosis-of-COPD (Accessed February 2016)

Time to address rare lung disease

Lung disease in the UK will have a greater impact on mortality than some forms of cancer, warns **Professor Luca Richeldi**, consultant in respiratory medicine at Southampton University and global key opinion leader in IPF.

"Five thousand new cases of idiopathic pulmonary fibrosis (IPF) are diagnosed every single year, but with no known cause and a complex route to diagnosis this is just the tip of the iceberg," says Professor Luca Richeldi.

IPF belongs to a group of approximately 200 conditions known as orphan lung disease, which in many cases causes irreversible inflammation and scarring, loss of pulmonary function and ultimately respiratory failure. "Patients may initially be pleased they haven't been handed a diagnosis of lung cancer," he notes, "but often the prognosis is very similar, with an average survival rate of just three years."

"With the number of people developing IPF and other interstitial lung diseases escalating in their thousands every single year, there is an urgent need for new diagnostic criteria and aggressive treatment of early disease," he points out. But apart from certain areas of the country where specialist centres exist, patients risk going undiagnosed and may often receive an initial diagnosis of asthma or COPD – after which point their life expectancy has already been compromised.

Although the first ever treatments have now been approved for use in IPF, access is still a major challenge, he observes. "Here in the

IPF: THE MAIN ISSUES

- DIFFICULT, LATE DIAGNOSIS – WE CATCH THE DISEASE TOO LATE, WHEN OUTLOOK IS POOR AND CARE NEEDS ARE ALREADY SIGNIFICANT
- EXTREMELY LIMITED TREATMENT OPTIONS – WITH NONE THAT STOP THE DISEASE'S PROGRESSION
- NO KNOWN CAUSE – NO OPPORTUNITY FOR PREVENTION

UK we have drugs to slow the progression of IPF, which work in all phases of the disease. On top of this, we strongly advocate early diagnosis and medication – yet we can only treat patients who fall between the parameters of 50% to 80% lung function (forced vital capacity). This paradox of care means people with either early or advanced IPF – certainly a sizeable fraction of patients at diagnosis – will miss out on access to medicines," he warns.

Having drugs approved to treat IPF is the driver to building awareness of this inexorably progressive lung disease. "Giving greater exposure to this condition is vital," Richeldi explains. "It is a progressive disease with a severe prognosis and high mortality rate, which really needs to be a higher priority for health policymakers."

The UK is in a good position because of its strong primary care system – but as with any disease with low prevalence, physicians working in primary care need a greater level of awareness and education around IPF. "I'm a strong believer in lung sounds," Richeldi points out, because this condition is not only rare but also non-specific and easily missed, and chest X-rays and lung function tests are "ineffective".

So, being vigilant about listening to and recording the distinctive sounds of IPF – a similar sound to ripping Velcro apart – becomes critically important to ensuring both early and accurate diagnosis.

"With the number of people developing IPF and other interstitial lung diseases escalating in their thousands every single year, there is an urgent need for new diagnostic criteria and aggressive treatment of early disease"

Global learnings

Here in the UK there are a limited but growing number of specialist centres able to prescribe either of the two available treatments. "There are pros and cons to this versus other countries," Richeldi explains. "On the one hand access to medicines is currently more difficult in the UK, however in Germany and the USA where every pulmonologist is able to prescribe either of the two available treatments there may be a risk of over-treatment because of the complexity and specialism involved in diagnosing IPF.

The critical take-home message in this country is to be more inclusive and to strive for better engagement with primary care."

Patient-centric approach to R&D

Richeldi also believes that changing the foundations of how we look at drug development to include the patient and carer experience will lead to a step-change in improving the disease management pathway. "There is a huge need for patient-centred clinical research in IPF," he notes, "from trial design to discussion about the endpoint and how to better include patient reported outcomes."

However, until recently there has not been a huge amount of specific attention or investment in IPF – despite the major impact on patients, families and caregivers from both a physical and psychological perspective. But this looks set to change given the greater focus this field of research is now attracting.

"There is an exciting future ahead for the field of IPF with an approved approach to earlier diagnosis on the horizon and at least 20 molecules in the preclinical pipeline targeting a range of targets," Richeldi reveals. "150 years ago, the major killer globally was tuberculosis but a focus by industry and academia on combatting infectious, microbial and viral diseases has dramatically altered that healthcare landscape," he stresses.

Embracing, understanding and investing in new models of disease to combat the rising tide of progressive lung conditions would have an equally lasting and positive impact from the perspective of patient quality of life, total health burden and, of course, on the economy.

"My opinion is that the impact of fibrosis on human health has been underestimated so far and requires the same strength of approach as directed at TB and infections all those years ago. It's time we started taking this seriously," Richeldi stresses. "The government needs to sit up and take notice of people with rare and severe disease."

For more information go to www.southampton.ac.uk

A GOOD day in the life of an IPF patient

A TASTE OF FREEDOM

=

Have some energy

Be able to walk to the shops

Spend time with grandchildren

ALMOST FORGET IPF AND HAVE AN ORDINARY DAY

=

Less coughing

Talk with ease

Putting washing out

A BAD day in the life of an IPF patient

HOUSEBOUND

=

Isolation and loneliness

Hours to dwell on their situation, the inevitability

Tearful and depressed

STRUCK BY FATIGUE

=

Unable to stay awake or function without napping

Constant coughing hampering ability to talk

No energy as if life is ebbing away

Interestingly, not all patients have good and bad days as experienced in previous research; some patients we spoke to are constant with their condition

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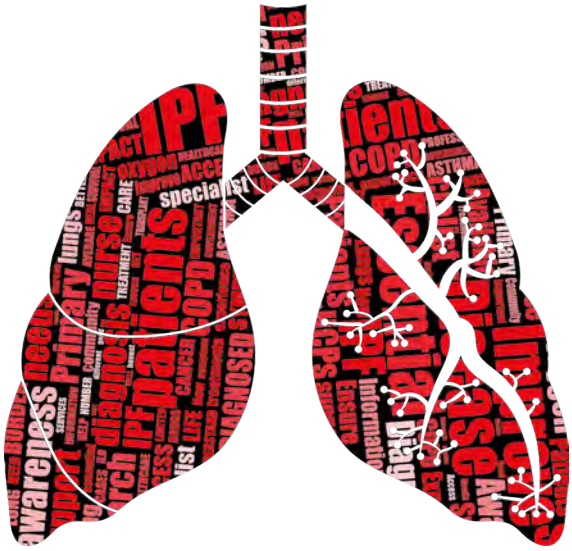
Boehringer Ingelheim data on file. DOF NIN16-01



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Action for Pulmonary Fibrosis

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British Lung Foundation

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