

# Action for Pulmonary Fibrosis

## Patient Survey Report 2018

Giving patients a voice



Action for  
Pulmonary  
Fibrosis



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# Foreword



*“ APF is determined to give patients a voice – and we will be taking our recommendations to NHS England, the Department of Health and Social Care and the UK Parliament. ”*

**Action for Pulmonary Fibrosis (APF) was founded in 2013 by patients, family members and specialists who had a vision for a better future for people living with the disease.**

We knew that people living with pulmonary fibrosis and their loved ones often struggled to find the right support. A dedicated national charity was needed. Since we started, we have put patient needs at the core of everything we do. This commitment continues today as we work with the whole pulmonary fibrosis community to improve outcomes for patients.

Our 2018 Patient Survey provides an opportunity to assess the quality of care for those living with the most common form of the disease, idiopathic pulmonary fibrosis, or IPF. It gives the most comprehensive picture to date of the care patients and their families receive and how this compares to national standards. The report recommends where improvements are needed. Although the focus is on IPF, many of the recommendations apply equally to other forms of pulmonary fibrosis.

IPF is a devastating and incurable interstitial lung disease, which affects 32,500 people in the UK. It kills 6,000 annually in the UK, with patients surviving on average only 3-4 years after diagnosis. This prognosis is worse than most major cancers. Patients become increasingly breathless, dependent on oxygen and disabled, and generally die from respiratory failure.

APF is determined to give patients a voice and will take our recommendations to the NHS, the Department of Health and Social Care and the UK Parliament.

There is often a temptation in reports of this kind to issue a series of very ambitious, and potentially undeliverable, recommendations. However, we know that the health service is already stretched and the dedicated staff within it are already trying hard to do the best they can for IPF patients. While the scope of this report is ambitious, the recommendations are practical and achievable, within a relatively short time frame. To realise these recommendations, everyone within the respiratory community has a role to play and we are committed to supporting this process.

APF looks forward to working with healthcare professionals, their networks, NHS providers and decision makers across the NHS to implement these recommendations and deliver a better future for people with IPF.

## **Steve Jones**

Chair, Action for Pulmonary Fibrosis

# Introduction

Three years ago, Action for Pulmonary Fibrosis (APF) launched its landmark 2015 Patient Survey Report<sup>4</sup>, which compared the experience of IPF patients in the UK against the system of care outlined in the NICE<sup>5</sup> Quality Standard. While we found some aspects of care were excellent, pointing to the hard work of healthcare professionals, we identified others where significant improvements were needed.

Since 2015, we have worked with health care professionals, patients and carers across the country to realise the recommendations, and we took the results to the UK Parliament. We were pleased to see that the update to NHS England's Service Specification for specialised interstitial lung disease (ILD) services<sup>6</sup> echoed many of our recommendations for more networked care, which is reflected in the latest NICE guidance.

Fast-forward three years to 2018 and we have carried out another Patient Survey to assess the standard of care for IPF patients. This 2018 report has provided an opportunity to assess how NHS care has changed since 2015 and to conduct more detailed analysis in key areas, including misdiagnosis and late diagnosis.

We were delighted that 776 patients from across the UK responded, more than double the number in 2015. Our analysis has resulted in some remarkable findings. Patients' **satisfaction** with their treatment **has fallen**, a third of **patients** still **lack access** to a specialist nurse and **only 63%** of individuals **feel 'well supported'** in managing their IPF.

It is clear there is still a long way to go to ensure equitable patient-led care for those living with IPF across the UK. We hope that the information in this report will be widely shared and used, to help drive improvements and bring us closer to this reality.

The methodology and limitations of the survey are outlined on page 9.

## What is IPF?

- Idiopathic pulmonary fibrosis (IPF) is a devastating and incurable lung disease. It causes scar tissue to build up around the small air sacs (alveoli) in the lungs, making it harder for oxygen to pass into the body where it is needed. The cause of this fibrosis is unknown. Life expectancy for an individual with IPF is around three years.
- In the UK, IPF is known to affect at least 32,500 people<sup>1</sup>. Each year 6,000 individuals die from the disease and the number of new cases diagnosed each year is increasing<sup>1</sup>.
- NICE outlines the key elements of care that people with IPF should expect in two documents: *Idiopathic Pulmonary Fibrosis in Adults: Diagnosis and Management* guideline<sup>2</sup>, and *Idiopathic Pulmonary Fibrosis in Adults, [QS79]*<sup>3</sup>, referred to in this report as the NICE Quality Standard.
- Currently, there are two anti-fibrotic drugs licensed for use in IPF in the UK: pirfenidone and nintedanib. While evidence suggests these medicines slow down the development of scar tissue in the lungs of people with IPF, access to the drugs is limited and they can have significant side effects.

## Key findings from the survey

- **35%** of patients are misdiagnosed and only **46%** are diagnosed correctly within six months of visiting their GP.
- Only **53%** of IPF patients have completed a course of pulmonary rehabilitation classes and only **14%** of patients receive pulmonary rehabilitation tailored to the needs of IPF patients.
- Almost a third of patients lack access to a specialist nurse. Those who have access do not always receive the full level of support they need.
- Some patients are still not being assessed for oxygen therapy, which would enable them to improve their mobility and take part in pulmonary rehabilitation courses.
- Only **63%** of patients feel well supported in managing their IPF, a drop from **76%** in 2015.

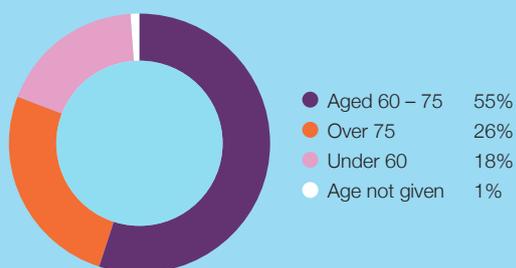
# The 2018 survey findings in detail

The NICE Quality Standard was developed to promote consistent care across the UK. It was first published in January 2015. Nearly four years later, the results of our 2018 patient survey point to a mixed picture of care across the UK. There are a number of areas where patients are unsatisfied with the quality of their care.

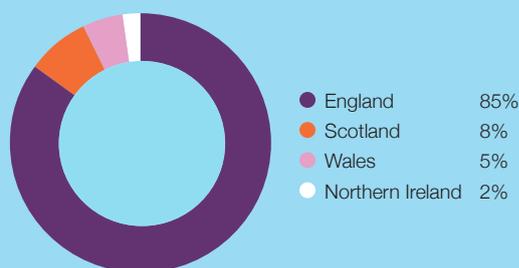
## Who responded?

A total of **776** people completed the survey, either online or by post.

### Respondents' age



### Respondents' location



These figures mirror the distribution of the UK population.

## Diagnosis

IPF is a challenging disease to diagnose. The main symptoms – a persistent cough and breathlessness, which get worse over time – can be difficult to distinguish from other respiratory conditions.

The NICE Quality Standard states that IPF should be diagnosed by a specialist multidisciplinary team (MDT), based on clinical features, lung function, radiology findings and pathology. The MDT should comprise a respiratory consultant, a thoracic radiologist, an ILD specialist nurse, and an MDT coordinator.

It is important that patients are diagnosed early, so they can quickly receive the support they need to help manage their condition. However, our findings indicate that misdiagnosis of IPF is common and correct diagnosis is often unacceptably slow.

- **Misdiagnosis:** 35% of respondents reported being misdiagnosed. The most common misdiagnoses were asthma, pneumonia, chronic obstructive pulmonary disease (COPD), and heart and digestive related conditions.
- **Slow diagnosis:** Only 46% of respondents report being diagnosed within six months of their first GP visit. For some 20% of patients it took more than two years to be correctly diagnosed.
- **Referral to hospitals:** 69% of patients were referred by their GP to a hospital (specialist or general) within six months of their first visit. 10% of patients were not referred for more than two years.

“ My dad was misdiagnosed for five years and was finally told he had IPF one week before he died. ”

“ More thought should be given to the psychological impact of being diagnosed with this devastating disease. ”

## Pulmonary rehabilitation

Pulmonary rehabilitation (also known as PR) is a short programme of classes which provides education and exercise for patients, so they are better able to manage their condition. The NICE Quality Standard calls for rehabilitation programmes to be tailored to specific respiratory conditions and suggests that this will contribute towards improved health, quality of life and exercise capacity.

“ My dad went on two 8-week PR courses but there was no mention of IPF, it was all about COPD. ”



## Regional differences

Our analysis shows that the quality of IPF care varies across the UK. Patients at specialist centres in England generally report high quality care and have good access to specialist nurses. The same is true in hospitals specialising in IPF in Scotland, Wales and Northern Ireland.

The quality of care in general hospitals, on the other hand, is reported to be variable. Some general hospitals provide very good care, sometimes by sharing care with a specialist centre. Others need improvement.

Some regions score highly on specific aspects of IPF care. For example, access to pulmonary rehabilitation is reported highest in East Anglia; misdiagnosis rates are lowest in the East Midlands; access to specialist nurses is reported to be highest in Yorkshire; and diagnosis is generally fastest in the East Midlands, Wales and Scotland.

However, overall the quality of care is patchy and no region scores above average on all metrics. There is a need for considerable improvement, if we are to achieve a consistent and equal level of IPF care across the country.

- **Pulmonary rehabilitation courses:** Only **53%** of respondents reported taking part in pulmonary rehabilitation, though this was up from **45%** in 2015. Only **27%** of those who took part (**14%** of all patients) were on courses specifically run for pulmonary fibrosis patients\*.



- Took part in pulmonary rehabilitation 53%
- On a course tailored to the specific needs of IPF patients 14%

- **Reasons for attending:** **87%** of those who attended wanted to better manage their symptoms; **74%** to increase their independence and **92%** to improve fitness.



- Wanted to improve symptoms 87%
- Increase independence 74%
- Improve fitness 92%

- **Positive impacts of PR:** **54%** of respondents said their symptoms improved after pulmonary rehabilitation; **69%** reported being more active; **62%** felt more in control of their condition, **57%** had a better quality of life, **51%** reported feeling less breathless and **44%** felt more independent.



“ I am currently fighting to get PR classes approved by my local CCG – it would apparently be easy if I had COPD! ”

\*This compares with 40% in 2015. The decrease may be because the 2018 survey includes more patients from general hospitals where specialised courses are generally not available.

## Oxygen therapy

Oxygen therapy is often important in the care of patients with advanced IPF. It helps them be more active and increases independence. It also makes it possible to take part in pulmonary rehabilitation classes.

NICE states that all patients should be assessed at regular intervals. We asked patients about their access to portable oxygen to use with exercise (ambulatory oxygen).

- **Oxygen assessment at diagnosis:** 72% of respondents reported being assessed for oxygen at diagnosis, up from 63% in 2015. 28% reported not being assessed.\*
- **Follow-up assessment:** 93% of patients reported being re-assessed for oxygen therapy at follow-up appointments. Only 7% were not re-assessed.
- **Currently receiving oxygen therapy:** 52% of respondents were currently receiving oxygen therapy; 48% were not.

“ Ambulatory oxygen is very important – it helps me keep active ”

“ I am on oxygen 24/7 but I still struggle with day-to-day activities. Going out is always a challenge because I worry about running out of oxygen ”



## Care coordination and access to a specialist nurse

Specialist ILD nurses are fundamental to good quality IPF care. As the NICE Quality Standard recognises, they are uniquely placed to ensure that patients, families and carers receive the information they require. Specialist nurses support patients at all stages of their journey to ensure they have timely access to the care they need. They are also trained to discuss, sensitively, prognosis, disease progression and life expectancy. The NICE Quality Standard states that care should be coordinated across all elements of the pathway, and those involved in providing the care.

Our report echoes the NICE Quality Standard. It shows that when patients have access to a specialist nurse they feel better informed about treatment options and more involved in discussions and decisions about their care.

- **Access to specialist nurse:** 71% of patients reported having access to a specialist nurse, while 29% respondents did not.
- **Value of specialist nurses:** Of those with access to a specialist nurse: 90% said they were available to speak to them between hospital visits; 81% said the nurse explained their treatment options clearly; 74% said they help them manage their symptoms; 66% said that their nurse provided them with written information and 52% said they provided support to their loved ones and carers.
- **Best point of contact:** When asked who was best placed to be single point of contact for their care, 53% said a specialist nurse, while 35% said a hospital doctor with specialist interest in IPF. Only 12% said either a GP, a non-specialist hospital doctor, a non-specialist nurse or another professional.

“ The support my father got was very good, especially from the specialist nurse who helped him to manage his condition ”

“ There should be more IPF specialist nurses available to support patients ”



\*Some patients may have been unaware that their arterial oxygen saturation was assessed, along with other blood tests, at diagnosis.

## Information on and access to treatment

The NICE Quality Standard states that patients with IPF should receive all the information and support they need throughout the care pathway. This includes information on investigations, diagnosis and management. Specialist nurses are well placed to provide such information.

Patients report:

- **At the time of diagnosis, 57%** of respondents felt their health care professionals explained their treatment options clearly, while **43%** did not.
- **Since being diagnosed, 73%** felt they, their family and carers had been as involved as much as they wanted in discussions and decisions about treatment options, while **27%** did not.
- **The majority of patients feel listened to: 77%** felt their views and opinions were listened to by healthcare professionals, **23%** did not; **72%** felt the goals they wanted to achieve through treatment were discussed with healthcare professionals while **28%** did not.
- **Treatment and quality of life:** only **46%** of respondents considered that treatment had improved their overall quality of life; **36%** considered it had not while **18%** did not know.

### Access to anti-fibrotic medicines is varied

**48%** of respondents had been prescribed one of the two therapies (Nintedanib **26%**; pirfenidone **22%**), **8%** had been prescribed both at different times and **44%** of respondents had not been prescribed either medication. Anti-fibrotic medicines can only be prescribed at prescribing centres, so patients at general hospitals may not be offered the medicines. This may be for clinical or other reasons. More needs to be done to improve access to antifibrotic therapies for IPF patients.

“ I cannot praise enough the support and treatment I've had from all the healthcare professionals from the time I became ill ”

“ After diagnosis, I have been left on my own to cope, with no help from anyone ”

## Management of care

Only 63% of patients feel well supported in managing their IPF, a drop from 76% in 2015. Although 73% of respondents felt they were as involved as they wanted to be in discussions and decisions about their treatment, 27% did not. The picture is mixed, but taken as a whole, results are worse than in 2015.

## The role of a support network

The importance of access to peer and carer support is emphasised in the NICE quality standard. This was confirmed in our survey.



 **54%** of respondents reported being a member of a support group

 **87%** of respondents attending told us that support groups provide mutual support

 **89%** agreed that support groups provide them with information to understand and manage their disease better

 **82%** agreed they provide social interaction, friendship and fun.

# Call to action for those caring for people with pulmonary fibrosis

Action for Pulmonary Fibrosis welcomes the inclusion of respiratory disease as a priority clinical area in NHS England's Ten-Year Plan, published in early 2019. Our goal is for interstitial lung disease, including pulmonary fibrosis, to be prioritised appropriately within the health service over the next ten years. A successful strategy will see the NICE Quality Standard implemented as standard for every patient with IPF in England. We look forward to discussing these plans to ensure they meet the needs of patients, as outlined in this report, while working with the NHS in Scotland, Wales and Northern Ireland to achieve the same in all parts of the UK.

However, while national level policy changes are critical for long-term outcomes, there are a number of clear recommendations emerging from the survey findings which can be acted on now. These are in line with the work of the Taskforce for Lung Health (of which APF is a member). We believe such actions will have an immediate impact on IPF patients, which is so important given the devastating and progressive nature of the disease.

These recommendations and measures of success are directed to NICE, the NHS and patient organisations. They cover the period of the Taskforce for Lung Health's 'National Five-Year Plan for Lung Health' (2019-2023), but we would expect to see significant progress by the midway point, in 2021.

## Recommendation 1

### The problem: late and inaccurate diagnosis

**Recommendation:** Create a clear patient pathway with services to ensure timely, accurate and confident diagnosis of all people with pulmonary fibrosis.

**Measures of success:**

- At least 92% of patients to start treatment within 18 weeks of the date a **GP referral** is booked, reducing to 12 weeks over the next five years.

There must be a clear pathway for respiratory patients to follow, which identifies all lung diseases early, offers timely diagnosis, and allows pulmonary fibrosis and other patients to get the treatment and support they need. Given the shortage of specialist interstitial lung disease staff and the tests needed to diagnose such diseases, our success measures are less ambitious than those for other respiratory diseases, proposed by the Taskforce on Lung Health. A key requirement will be for GPs and other health professionals to have a clear set of steps to take when a person presents with possible pulmonary fibrosis.

## Recommendation 2

### The problem: access to a specialist nurse

**Recommendation:** Increase access to ILD specialist nurses.

**Measures of success:**

- Increase from 71% to 85% in the proportion of patients reporting access to a specialist nurse.
- Over 90% of patients report they are able to speak to their specialist nurse between hospital visits, if required.
- Over 80% of patients report that specialist nurses explain treatment options clearly, advise them effectively on managing their symptoms and provide written information to help them manage the disease.

Specialist nurses are fundamental to good quality IPF care. There is an urgent need to ensure that all ILD specialist centres in England, and their equivalents in other nations, have adequate numbers of ILD specialist nurses, and respiratory nurses in general hospitals are provided with appropriate training in diagnosis and treatment of IPF. The number of ILD patients per specialist nurse should be determined and compared to lung cancer and other lung diseases, as a basis for appropriate workforce planning.

## Recommendation 3

### The problem: access to pulmonary rehabilitation

**Recommendation:** Improve IPF patients' access to pulmonary rehabilitation (PR) courses specifically run for pulmonary fibrosis patients.

**Measures of success:**

- Increase in the proportion of IPF patients who have completed a course of PR from 53% to 70%.
- Increase in the proportion of IPF patients who complete PR courses tailored to their specific needs from 14% to 25%.

Pulmonary rehabilitation is one of the most effective treatments for people living with IPF and other forms of pulmonary fibrosis. It is important for health care professionals to promote its benefits to patients and for clinical commissioning groups (CCGs) to prioritise funding of general and specific PR programmes.

## Recommendation 4

### The problem: access to pulmonary fibrosis support groups

**Recommendation:** Increase the number of pulmonary fibrosis support groups and provide better support to more isolated patients.

**Measures of success:**

- Over 80% of patients are sign-posted to a pulmonary fibrosis support group by their consultant or specialist nurse at the time of diagnosis.
- Increase from 70 to 100 in the number of support groups facilitated by Action for Pulmonary Fibrosis so that over 90% of patients in the UK live within an hour of a support group.
- Increase the effectiveness and coverage of the support groups and pilot new ways to provide support to isolated patients who are unable to attend support groups.

Support groups are an effective way of providing mutual support and friendship among patients and their families. They also empower patients by increasing access to information and knowledge about the disease. There is a need also to reach out to those patients who are unable to attend support groups because of mobility, remoteness and other reasons.

## Recommendation 5

### **The problem: the lack of new technologies to speed up diagnosis and support treatment**

**Recommendation:** Encourage research and development into new technologies to speed up diagnosis, support effective treatment and give early warning of acute exacerbations.

**Measures of success:**

- At least two new technologies in widespread use by GPs and hospitals, within five years, which are shown to speed up diagnosis, improve treatment and prevent hospital admissions for IPF and other forms of pulmonary fibrosis.

New technologies have a key role to play. They could include, for example, point of care tests for GPs to use in diagnosing IPF and other forms of pulmonary fibrosis more quickly; point of care tests to diagnose viral illnesses and give advanced warning of possible exacerbations; and use of Wearable Tech. It will be important to evaluate new technologies, to understand how the Tech interface, with ILD specialist nurses and teams, can predict and prevent hospital admissions.

## Methodology

The findings in this report are based upon evidence gathered in APF's 2018 Patient Survey. The postal and online survey ran from June to August and collected 776 responses from across the UK. All conclusions made in this report which compare the 2018 survey to its 2015 predecessor, or explore the relationship between two questions, are based upon an 85% statistical confidence level.

It should be noted that the survey in 2015 received significantly fewer responses – less than half the number who took part this year. In 2015, APF had only been established for two years, and much of our work was focussed on specialist centres. Since then, the number of support groups has expanded to nearly 70, many of which are based in general hospitals. We also have a more active social media presence, which broadens the reach of our organisation. The results of the 2018 survey are thus likely to be more representative of the IPF community as a whole.

While the survey was promoted online, a number of support group members completed the survey. However, 46% of the respondents were not a member of a support group, ensuring that the views of those who may currently not be receiving support from a peer-group are included within the findings and recommendations.

In the report, we have noted regional differences, but these should be treated cautiously because of the small number of responses in some regions. Comparisons with 2015 should also take account of the fact that a larger percentage of respondents in 2018 were patients at general hospitals.

In the instructions to respondents, it was made clear that the survey should be completed by patients or by somebody acting as a patient's main carer.

## The Story of Action for Pulmonary Fibrosis: Improving the quality of life for PF patients, now and in the future

APF was set up by a group of patients, family members and specialist clinicians with a clear vision: a world in which everyone living with pulmonary fibrosis has a better future. In the last five years APF has worked to support patients, raise awareness, educate primary healthcare professionals and fundraise for important research.

An important part of this work is expanding the number of patient support groups. We have assisted hospital staff and patients to establish local groups and there are now nearly 70 groups across the UK. They bring together patients and specialists to provide a supportive and informative environment in which to exchange coping skills and share experiences with others. For those patients and carers in need of additional information or advice, APF also offers a support line. Thanks to the enormous generosity and hard work of patients and their friends and family, APF is able to fund research each year devoted to pulmonary fibrosis.

APF is dedicated to working with the NHS and social care services to support all patients with IPF and other forms of pulmonary fibrosis and their carers. Through the extensive support group network, it is now recognised as a trusted voice of pulmonary fibrosis patients throughout the country.

Registered charity number 1152399 (England and Wales).



### References

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