



# Research Strategy

2022 – 2025



Action for  
Pulmonary Fibrosis



**Our vision** is to find a cure for pulmonary fibrosis so that everyone affected by the disease has a better future.





# Introduction

APF is a patient driven organisation and our community is at the heart of everything we do. Our mission is to provide support to families, raise awareness, campaign and educate to improve access to the highest standard of care. We are dedicated to funding high quality, high impact pulmonary fibrosis research.

**APF funds research that has the potential either to:**

- a) contribute towards the discovery of an effective treatment or cure, or
- b) accelerate change which will enable our community to live well for longer.

We are able to invest in research because of the dedicated fundraising activities by patients, caregivers, family and friends: from marathons to coffee mornings, selling crafts to singing songs, and making donations in memory of loved ones. We are also indebted to the generosity of several philanthropic trusts and funds.

The Research Strategy sits beneath the **APF Strategy** and is fundamental to meeting the key strategic aims of the charity.





# What is pulmonary fibrosis?

**Pulmonary fibrosis is an umbrella term for a range of progressive lung diseases, which cause stiffening and scarring of the lungs.**

The lung scarring eventually prevents the transfer of oxygen into the bloodstream. Key symptoms of pulmonary fibrosis include breathlessness and chronic cough. On physical examination, Velcro-like crackles are likely to be heard and clubbing of fingers may be observed. The key symptoms of pulmonary fibrosis can be confused with other respiratory disorders, such as chronic obstructive pulmonary disorder (COPD), asthma and bronchiectasis, and these alternative diagnoses need to be ruled out.

Pulmonary fibrosis affects around 70,000 people in the UK. Idiopathic pulmonary fibrosis is the most common form and affects around 30,000 people in the UK. Other forms of pulmonary fibrosis include: familial pulmonary fibrosis; those associated with autoimmune diseases (such as rheumatoid arthritis and systemic sclerosis); toxicity caused by some medications; exposure to inorganic substances (such as asbestos and coal dust); hypersensitivity pneumonitis caused by exposure to organic dusts and fibres (such as bird feathers and mould found on hay, straw and grain); and sarcoidosis.



# 70,000+

people in the UK are affected by pulmonary fibrosis



# Why is research into pulmonary fibrosis needed?

Tragically, many forms of pulmonary fibrosis have a poor prognosis and a devastating impact on a patient's quality of life.

As the disease progresses, patients experience increasing levels of breathlessness, fatigue and muscle weakening, which can make everyday activities difficult to achieve. People living with pulmonary fibrosis often require oxygen therapy and, towards the end of their lives, many patients need this continuously. Idiopathic pulmonary fibrosis is the most aggressive form of the disease. Life expectancy is just three to five years following diagnosis and around 6,000 people die from the disease every year in the UK. Treatment options for pulmonary fibrosis are very limited and only a handful of patients are eligible for lung transplantation.

Pulmonary fibrosis has a life changing impact on all aspects of life for patients' and their families. By listening to their lived experiences, we know that investment in research is desperately needed to provide hope for a better future.



# What our community tell us...

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*“Within the time span of about an hour I went from being somebody with a cough and shortness of breath to somebody with a terminal illness for which there is no cure and no indication of how long I can expect to live.”*

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*“I’m thankful there is a medicine I can take, but I find the side effects quite difficult to manage.”*

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*“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.”*

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*“I am fully aware, as we all are, that there is no happy ending to IPF.”*

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*“Some days I just find it all a bit overwhelming. I wish there were more I could do to control my symptoms.”*

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*“It’s so worrying not knowing whether my kids will be affected by PF too. What should they be doing to avoid getting it?”*

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

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*“My mum was doing OK, but suddenly got really sick – they call it an acute exacerbation. We were lucky she pulled through but it was a really frightening time.”*



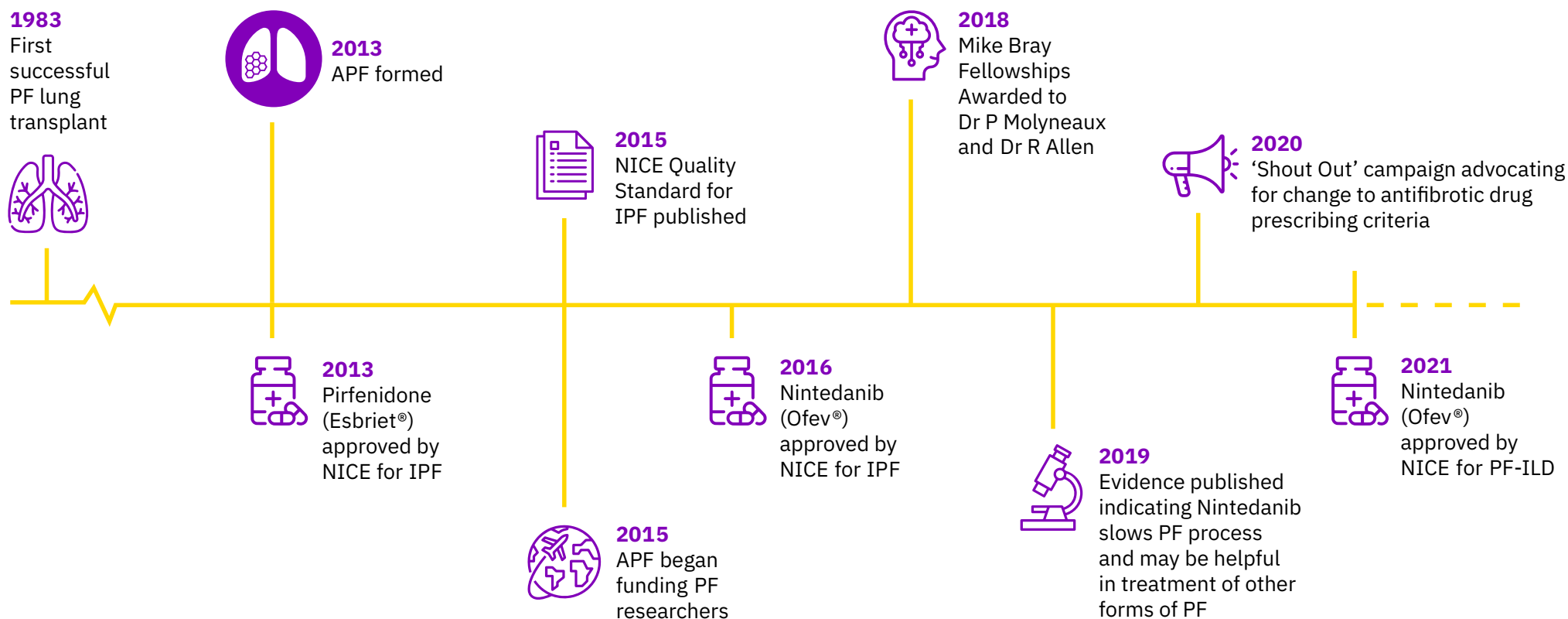
## How has our understanding of pulmonary fibrosis **evolved** in the last 10 years?

Our understanding and ability to treat pulmonary fibrosis is continually changing and there have been many advances over the last 10 years:

- Opportunities for drug discovery have accelerated and the number of clinical trials for treatments targeting pulmonary fibrosis have dramatically increased. This has led to two antifibrotic medications (nintedanib and pirfenidone) being approved by NICE which slow disease progression.
- Theories around the mechanisms of fibrosis continue to evolve, with an emphasis on the role of recurrent or chronic injury to the alveoli, genetic predisposition, and dysfunctional repair processes leading to structural damage of the lungs.
- Computational and technological advances have led to improvements in imaging, genetic understanding, and diagnostics.
- The potential benefit of ambulatory oxygen and pulmonary rehabilitation for patients with pulmonary fibrosis is better understood, which in turn has led to increased availability of these interventions.



# Research evolution





# Our research

APF was set up as a charity in 2013 by a group of patients, family members and healthcare professionals who were acutely aware of the need to increase understanding of the disease, support patients and families, and raise vital funds for research.

As APF has grown, we have been able to invest in high quality, high impact research. Since 2015, we have funded scientists to share their research at conferences around the world, raising the profile of pulmonary fibrosis research and enabling collaborations.

In 2018, we invested over £580,000 in research when we awarded our first Mike Bray Fellowships to Dr Richard Allen, University of Leicester, and Dr Phil Molyneaux, Imperial College London. Their research aims to increase understanding of the causes of pulmonary fibrosis and support the development of new treatments. Dr Molyneaux's research focuses on the role of bacteria in disease and investigates whether taking prophylactic antibiotics can slow disease progression.

Dr Allen's research focuses on the genetics of pulmonary fibrosis, which provides insight into the different chemical and biological pathways involved in the process of fibrosis. Their important research has the potential to support the development of targeted treatments.





# Where are we now and **where do we want to be?**

Despite medical advances, the prognosis for patients diagnosed with pulmonary fibrosis remains very poor.

Investment in research is desperately needed to enable people living with pulmonary fibrosis, their carers and families, to experience a better quality of life and have more time together. This research strategy lays the foundations for the expansion of our research programme, and the impact we aspire to make for our community. By listening to patients, carers, health care professionals and academics within our community, we have identified four key areas of unmet needs. Our research priorities are informed by these areas of unmet need.





## 1. Pulmonary fibrosis can be difficult to diagnose.

- There can be a marked delay between the onset of symptoms and diagnosis.
- Patients are often diagnosed when the disease has progressed to a later stage.
- 54% of patients with idiopathic pulmonary fibrosis who we surveyed waited over 6 months for a diagnosis, 20% waited over two years for a diagnosis and 35% were misdiagnosed.

➤ We will fund research which enables people living with pulmonary fibrosis to be diagnosed early and accurately.



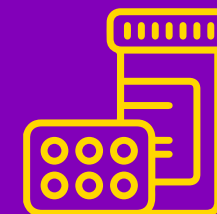
*Including, but not limited to:*

- Development of technologies that support early diagnosis and intervention.
- Identification of early indicators of disease.
- Understanding of familial risk and genetics.

## 2. Treatment options are limited.

- Current anti-fibrotic medication slows but does not stop the progression of the disease.
- Not all patients respond well to the available treatment options.
- Treatment side effects can be difficult to manage and prohibitive for some patients.
- Patients report many unmanaged symptoms including breathlessness and chronic cough.
- Oxygen therapy is challenging to manage both physically and practically.
- Acute exacerbations are very difficult to manage.

➤ We will fund research which leads to the development of effective treatments or a cure.



*Including, but not limited to:*

- Identifying drug targets.
- Developing interventions that can be used at the pre-fibrosis stage.
- Development of treatments that stop progressive fibrosis.
- Development of treatments that reverse the fibrotic process.
- Effective symptom control.
- Prevention and management of acute exacerbations.

### 3. Pulmonary fibrosis continues to have a devastating impact on people's lives.

- Patients' quality of life is severely affected by the symptoms of pulmonary fibrosis, including breathlessness, chronic cough, and fatigue.
- Patients, their carers and families experience a negative impact on their wellbeing, including psychological, physical and financial stress.
- Access to appropriate palliative and end-of-life care can be very limited.

#### > We will fund research which enables patients, carers and families to live well for longer.



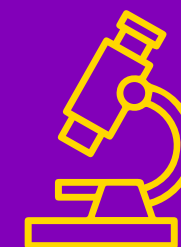
*Including, but not limited to:*

- Interventions which lead to improved quality of life and wellbeing.
- Greater understanding of the burden of disease on patients, carers and families.
- Technology and treatments that enable patients to participate as fully as possible in daily activities.
- Greater understanding of beneficial lifestyle changes.

### 4. There is still a lot we do not know about pulmonary fibrosis.

- The lung scarring process is not fully understood.
- The progression of the disease varies. It is not possible to predict which patients will deteriorate rapidly and those who remain stable, or know why.
- Patients with a diagnosis of idiopathic pulmonary fibrosis do not know what caused the disease.

#### > We will fund research that improves our understanding of the biological basis of pulmonary fibrosis.



*Including, but not limited to:*

- Understanding the basic mechanisms and progression of the disease, including prediction.
- Broadening our understanding of all forms of pulmonary fibrosis.
- Identifying at risk populations and risk factors (location, ethnicity, hobbies, employment, comorbidities).



## At APF, we understand how important it is to accurately identify the priority areas of unmet need within our community.

As part of our on-going review of research priorities, APF has partnered with Imperial College London and the **James Lind Alliance (JLA)** to carry out a Priority Setting Partnership for pulmonary fibrosis. The partnership brought together patients, carers and clinicians to jointly agree the Top 10 research priorities for progressive pulmonary fibrosis (PPF). We welcome applications that address one or more of these priorities.

### JLA Top 10 Research Priorities:

1. How can the diagnosis of PPF be improved in terms of accuracy and the time taken (screening programme, early signs and symptoms that could be detected in primary care, blood markers, imaging, biopsy, artificial intelligence, etc.)?
2. Can new treatments other than pirfenidone and nintedanib slow, halt or reverse the progression of PPF?
3. What can be done to improve the speed and accuracy of PPF diagnosis in primary care (e.g. training, integration of case-based studies in GP training, awareness campaigns)?
4. What is the best time for drug and non-drug interventions (pulmonary rehab, oxygen therapy, psychological support) to start to preserve quality and length of life for patients with PPF?
5. What are the best ways (drug, non-drug and aids) to treat cough in PPF?
6. Would early treatment delay progression, lung function decline, and improve survival in PPF?
7. Which therapies will improve survival in PPF?
8. What treatments (drug, non-drug and aids) can reduce breathlessness and phlegm production in PPF?
9. To what extent do different interventions (pulmonary rehab, oxygen therapy, psychological support) impact length of life in patients with PPF?
10. Can new treatments for PPF be developed with reduced side effects? Does how the drug is delivered (e.g. oral, nebulised, through a vein) affect potential side effects of the drug in PPF?



# What types of research grants do we offer?

**Our vision is to find a cure for pulmonary fibrosis so that everyone affected by the disease has a better future. We are dedicated to funding high quality research which makes a meaningful difference to the APF community, both now and in the future.**

**We offer a broad range of funding options so that we can:**

- Invest in research across the pulmonary fibrosis field, for example, from improving our understanding of genetics to developing evidence-based mental health support for patients and caregivers.
- Build capacity in pulmonary fibrosis research by supporting researchers throughout their career, in particular early to mid-career.
- Support research at all stages of development, from nurturing novel ideas to enabling established work to continue.
- Ensure that the impact of our investment will be seen both in the short and medium to longer term.
- Respond flexibly to the funding needs of applicants. We are happy to part-fund or completely fund a project, and where appropriate, can cover the cost of staff, consumables and equipment.

APF sees the huge value in funding large scale projects, such as bespoke registries and specialist interstitial lung disease (ILD) research and treatment centres. Such large-scale projects are beyond our funding scope for this strategic period. However, we will support others, collaborate, and advocate for these resources to benefit our community. APF is open to working with collaborative partners and joint funding research projects.

> Please contact us at: [research@actionpf.org](mailto:research@actionpf.org)



## Travel & Attendance Awards

**up to £1000**

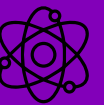
Supporting patients, carers and early career researchers to attend conferences and workshops to learn, collaborate and network.



## Satellite Symposium Grant

**up to £5000**

This grant aims to raise the profile of pulmonary fibrosis research, provide opportunities for education and facilitate collaboration.



## PhD Studentship

**up to £150,000 / up to 4 years**

Providing the opportunity for graduates to continue their studies in pulmonary fibrosis. Funding awarded to academic institution.



## Mike Bray Fellowship

**up to £300,000 / 3 years**

Enabling postdoctoral researchers and early to mid career academics to progress their research in pulmonary fibrosis.



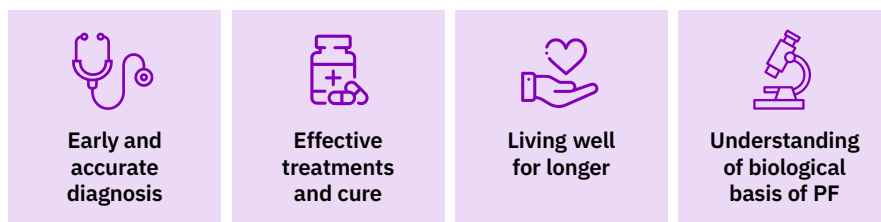


# How do we choose which research we are going to fund?

**APF will only fund research that has the potential either to:**

- a) contribute towards the discovery of an effective treatment or cure, or
- b) accelerate change which will enable our community to live well for longer.

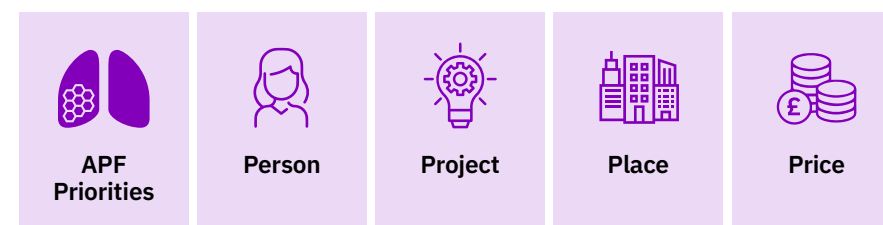
All research must have the potential to meet one or more of the prioritised areas of unmet need and applications for funding that address the JLA Top 10 are particularly welcomed:



It is essential that all research applicants engage directly with the pulmonary fibrosis community. Where appropriate, patient and caregiver involvement should begin at the priority setting stage, and continue throughout the development and delivery of research, including sharing findings. APF can support researchers to involve people affected by pulmonary fibrosis in their work.

All applications are assessed in relation to the four APF priorities for research, the person, project, place, and price.

**Different funding calls may target one or more of the following identified priority areas:**



APF only provides funding to UK establishments, although UK grant applicants may have international collaborators.

Recommendations for funding are made by our Research Review Panel (RRP). The RRP is an independent panel of experts, including scientists, patients and caregivers, who provide impartial advice to the Board of Trustees. The RRP sits up to three times a year to consider new applications.

**> Further information about the RRP [can be found here.](#)**

APF is guided by the Association of Medical Research Charities (AMRC). All applications are assessed in accordance with their principles of peer review: accountability, balance, independence, rotation and impartiality.

**> Further information on grant application procedures [can be found here.](#)**

# How do we support research outside of awarding grants?

**APF has a culture of collaboration and support that goes beyond financial investment. We are committed to helping create an environment where researchers can thrive, and expertise can develop. Our community of patients, caregivers, world leading academics and healthcare professionals provide a valuable resource for researchers.**

APF communicates about research within our community and through our wider networks, raising the profile and understanding of pulmonary fibrosis research. We foster collaboration between researchers and enable researchers to form meaningful relationships with people affected by pulmonary fibrosis. We are able to make important contributions to research we do not fund through patient and public involvement (PPI) opportunities.

APF and our community provide insight into the patient and carer experience throughout the research cycle. We work directly with individual researchers, laboratories, and as part of nationwide collaborations. We support research across all levels, from PhD studentships, to clinical trials, and appraisals for drug marketing authorisation.

APF has an advocacy role in ensuring relevant findings guide best practice and inform quality standards of care. We campaign to ensure patients living with pulmonary fibrosis throughout the UK have equal access to the best possible care and support. We want more people living with pulmonary fibrosis in England, Wales, Scotland and Northern Ireland to have the opportunity to participate in clinical trials, and this is an area of growth for the charity.





# What is the impact of our involvement in research?

APF's involvement in research ensures that the needs and experiences of people affected by pulmonary fibrosis are prioritised. Our ability to fund high quality, high impact research means that developments in the field can be made more rapidly.

As a community, we have the potential to transform the understanding, diagnosis and treatment of pulmonary fibrosis and enable patients, their carers and families to live well for longer.

Together, we will find a cure for pulmonary fibrosis.



If you would like any  
further information or  
if you have any questions  
please contact:

[research@actionpf.org](mailto:research@actionpf.org)



**Action for  
Pulmonary Fibrosis**

This research strategy will be formally reviewed every three years. Amendments will be made to this document as appropriate, and in response to new information.

Registered Charity 1152399 in England and Wales. Registered Charity SC050992 in Scotland.



[www.actionpf.org](http://www.actionpf.org)