

# A Snapshot of IPF Care in Europe



An EU-IPFF benchmarking report on access to  
Idiopathic Pulmonary Fibrosis (IPF) care in Europe

# Foreword

## from the EU-IPFF president



National IPF patient organisations have capitalised on this growing political focus, joining hands to drive policy change together.

The European Federation on Idiopathic Pulmonary Fibrosis & Related Disorders Federation (EU-IPFF) was established in 2016 to improve the care and the quality of life of IPF patients across the EU. Already we have developed the first European IPF Patient Charter and successfully encouraged the adoption of a Written Declaration by the European Parliament.

This 2018 EU-IPFF Benchmarking Report on Access to IPF Care in Europe is another important accomplishment for our community. The comparison of care standards identifies best practices to be encouraged and gaps to be filled, and also proposed concrete solutions to improve patients' quality of life. The resulting conclusions will propel policy asks to ensure a high level of quality care across Member States. Ultimately, we hope that this Report can inform decision-making at every level, to eliminate the inequalities in access to specialised care that people with idiopathic pulmonary fibrosis still experience today.

A handwritten signature in blue ink, which appears to read "Carlos Lines Millán".

**Carlos Lines Millán**  
EU-IPFF President  
November 2018

Idiopathic Pulmonary Fibrosis (IPF) is a rare, long-term, progressive disease that affects the lungs. Most people have never heard of it. But for those affected, the symptoms are debilitating: a gradual decline in lung function leads to reduced oxygenation of the blood, brain and other vital organs. Without treatment, many patients only survive two to five years after diagnosis.

This makes recent pharmaceutical investment in this area a game-changer. IPF patients have a choice of treatment options. It has been enabled by the emergence of a European legislative framework supporting work on rare diseases. This began in 1999 with a European Regulation on orphan medicines that incentivised manufacturers to develop drugs. It has grown to include Council Conclusions released in 2009 that encouraged Member States to adopt a national plan or strategy for rare diseases, as well as the 2011 Directive on patients' rights in cross-border healthcare that established the European Reference Networks for rare diseases.

The content of this Benchmarking Report represents the views of the authors only and it is their sole responsibility.

This Report is not intended as an exhaustive or scientific review of the care and management of IPF in Europe. The interviews on which this Report is based have provided a useful insight into some issues affecting a selection of IPF patients and their families, as well as identifying some discrepancies in IPF care within and among some European countries.

Boehringer Ingelheim International GmbH and F. Hoffmann-La Roche LTD financially supported the Benchmarking Report.

Recognising that this Benchmarking Report only represents a snapshot of the IPF landscape in thirteen EU countries, the EU-IPFF welcomes feedback. For more information or to share any concerns or comments you may have, please contact: [secretariat@eu-ipff.org](mailto:secretariat@eu-ipff.org)

Powered by Weber Shandwick.

Last updated: May 2019

# Rationale

With a view to driving evidence-based policies, the European Idiopathic Pulmonary Fibrosis & Related Disorders Federation (EU-IPFF) has commissioned this Benchmarking Report to measure how those European countries represented by its members are performing on different areas of IPF care and management. The Report highlights best practices and identifies gaps where urgent policy action is needed.

The Benchmarking Report aims to:

- **Collect quality information about IPF patient care in EU-IPFF member countries in order to compare the situation of patients between countries;**
- **Identify gaps in IPF care across EU-IPFF member countries;**
- **Identify best practices in IPF care across Europe;**
- **Make recommendations for solutions that could improve IPF care and the overall quality of life of IPF patients**

# Methodology



A qualitative study was designed to collect data from 16 patient organisations from 13 Member States.<sup>1</sup> A structured questionnaire developed in collaboration with medical experts and patient representatives was used for phone interviews and included both closed and open-ended questions focused on the following areas:

- **Information about the disease;**
- **Diagnosis and access to specialised care;**
- **Access to pharmacological and non-pharmacological treatment;**
- **Access to palliative care and end-of-life care.**

EU-IPFF member organisations and partners were invited to take part in phone interviews pending written consent. Each EU-IPFF member organisation had the possibility to identify the most suitable representative for the 60-90 minute interview.<sup>2</sup>

The results of the interview fed into a scorecard illustrating country performances according to specific indicators. The results have been analysed using a points-based system: each answer has been assigned a colour code based on the level of progress and implementation. A detailed matrix was developed to support the codification process.

**The colours represent:**

-  **No existence of policy or development;**
-  **Policy or development exists but implementation has not started yet;**
-  **Policy or development exists and is fully implemented;**
-  **No information available.**

The codified answers were shared with interviewees and medical experts for approval and final validation. The findings of each interview have been used to develop a European scorecard that visually compares country performances against specific indicators.

**Acronyms used in the snapshot:**

**Idiopathic Pulmonary Fibrosis (IPF):** a rare, progressive and ultimately fatal pulmonary disease that affects the fragile tissue in the lungs. The cause of IPF is unknown and there is no cure.

**Interstitial Lung Diseases (ILDs):** a group of disorders characterised by progressive scarring of the lung tissue between and within the air sacs. IPF is an ILD.

**ILD Specialised Centres or Centres of Expertise:** centres for the management and care of rare disease patients, designated at a national level by each EU Member State. ILD/IPF centres of expertise specialise in ILDs and they aim to provide IPF patients (among others) with the highest standards of care around timely diagnosis, appropriate treatment, and follow-up.

<sup>1</sup>Austria, Belgium, Bulgaria, France, Germany, Greece, Ireland, Italy, The Netherlands, Poland, Spain, United Kingdom and Hungary. The questionnaire was translated by a translation agency in the following languages: German, Spanish, French and Italian.

<sup>2</sup>The majority of the interviews were conducted in English and four interviews were conducted in the native language spoken by some patient representatives (Italian, French and Spanish). Four interviewees preferred to respond to the questionnaire in writing.



# Highlights

The Benchmarking Report outlines the current state of IPF care and management in Europe, identifying best-performing countries along with challenges that demand greater political attention and an immediate response.



International guidelines on IPF<sup>3</sup>, validated by the relevant international respiratory medical societies, including the European Respiratory Society, are available. Only a handful of countries have developed or are developing national guidelines, or are on track to implement such guidelines<sup>4</sup>.



IPF patients lack access to reliable information. In most countries, information and self-management materials are either limited or not fully integrated into the care process. Newly diagnosed IPF patients do not always receive any supporting information from specialised centres; in the majority of the cases, it is up to patient support groups to provide these.



Across all countries, respondents reported that prompt diagnosis is challenging. Patient representatives noted slow referral periods, with extended gaps between the initial suspicion of IPF and a confirmed diagnosis. Delayed diagnosis or misdiagnosis due to unclear signs and symptoms are major obstacles within the diagnostic process.



EMA-approved pharmacological treatments for IPF are available and reimbursed with no out-of-pocket costs for patients in all thirteen countries surveyed. However, timely referral to specialised centres, a prerequisite for receiving a prescription for treatment, remains a major challenge. Many respondents deemed the 'wait-and-watch' approach to treatment to be a barrier.



A number of interviewees in different countries also reported access to non-pharmacological treatment (including oxygen therapy, pulmonary rehabilitation and lung transplantation) as being either restrictively available or, to some extent, unavailable. When available, it is not always fully covered or reimbursed, resulting in out-of-pocket expenses for patients.



Access to palliative care and end-of-life care is extremely fragmented. Some countries have established palliative care centres and teams, while others are only beginning to provide this kind of support.

“As an IPF patient, I was invited to join a team of expert healthcare professionals to review the German Guidelines on IPF. I strongly felt that I could contribute to this, providing input based on my personal experience. I am particularly satisfied because the concepts of timely access to treatment upon diagnosis, exceptional use of biopsy upon joint doctor-patient decision, and timely referral to a specialised ILD centre were included into the guidelines. I am glad that the patient’s voice was heard during such a process. Patients, alongside with medical experts, are the ones who know best what they need.”

**A patient from Germany**

<sup>3</sup> Available at <https://www.ncbi.nlm.nih.gov/pmc/articles/PMC5450933/>

<sup>4</sup> The following countries have developed national guidelines: Germany, United Kingdom, Ireland. The following countries are in the process of developing national guidelines: Poland, Austria.

# Diagnosis and access to specialised care



## Key challenge

Early diagnosis remains the main challenge across all countries surveyed.

International guidelines on the diagnosis of IPF<sup>5</sup> were updated in 2018. But they have not been fully implemented in all countries surveyed. Diagnostic procedures are generally standardised; however, the majority of countries surveyed do not have nationwide IPF diagnostic and care pathways. Instead, these are often established by each expert centre and can therefore vary from region to region.

General practitioners have a limited knowledge of or training in recognising IPF. The symptoms are therefore sometimes confused with those of other lung diseases. Additionally, genetic testing is not widely performed, not even when another family member has IPF. This further complicates the diagnostic process.

Timely access to specialised centres and multidisciplinary care has been flagged as a factor that can make a real difference for patients. Countries where patients have access to multidisciplinary teams perform better in the diagnostic process, resulting in a better experience for patients. Yet the composition of multidisciplinary teams varies greatly. Many

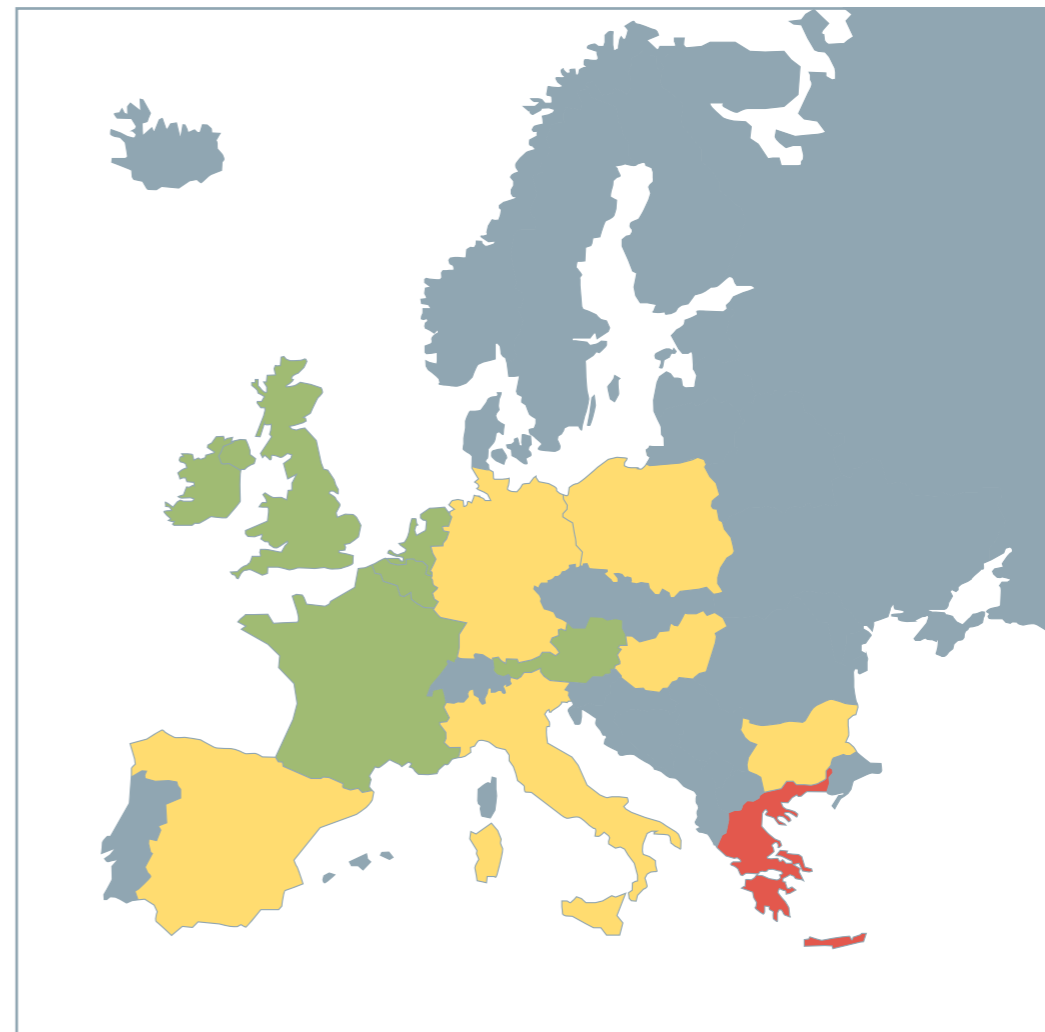
countries, such as Bulgaria, Italy and Spain, report shortages or a lack of recognition for specialist lung nurses, who play a crucial role in supporting patients and carers. By contrast, when specialist nurses are available to accompany patients in dealing with their disease every day, as is the case in the UK, positive effects are reported.

*“Much remains to be done to improve IPF diagnosis in France. While I was diagnosed fairly quickly, the same cannot be said of my brother. He was misdiagnosed for years, not knowing what his condition was. It is only when I found out that I had IPF and after he shared this information with his doctor that he finally received an accurate diagnosis. We have to do more to ensure that people can be diagnosed in a more timely manner.”*

*A patient from France*

*“Communication with the respiratory physician and with members of the multidisciplinary team is essential in order to get quality information about how to manage IPF in everyday life. After my husband received his IPF diagnosis, we were really worried about what to do next and I did not know how best to support him. In those cases, it has been really essential to interact with the multidisciplinary team and to exchange information with fellow patients in the support group.”*

*A carer from Austria*



<sup>5</sup> Available at: <https://www.atsjournals.org/doi/abs/10.1164/rccm.201807-1255ST>

# Access to pharmacological treatment



## Key challenge

Access to treatment in a timely manner following diagnosis is a priority for IPF patients.

There are currently no curative pharmacological treatments for people living with IPF. However, two antifibrotic drugs that can slow down disease progression have been approved by the European Medicines Agency. Reimbursement criteria for these drugs differ between countries, but none of the thirteen countries surveyed reported out-of-pocket costs for patients. However, this is not representative of the wider situation across the EU, since access to and reimbursement of EMA-approved drugs remains a barrier in countries not represented in the EU-IPFF (e.g. Malta).

Despite the theoretical availability of EMA-approved treatments, they are sometimes delayed in practice. This can be due to delayed diagnosis, clinical inertia (“wait-and-watch”), national prescription rules, or point-of-care treatment dispensation practices. Overcoming these challenges is vital, since timely access to treatment increases the ability to slow disease progression.

Even when IPF has been initially diagnosed, the “wait-and-watch” attitude is still reported. Some respiratory physicians prefer to monitor

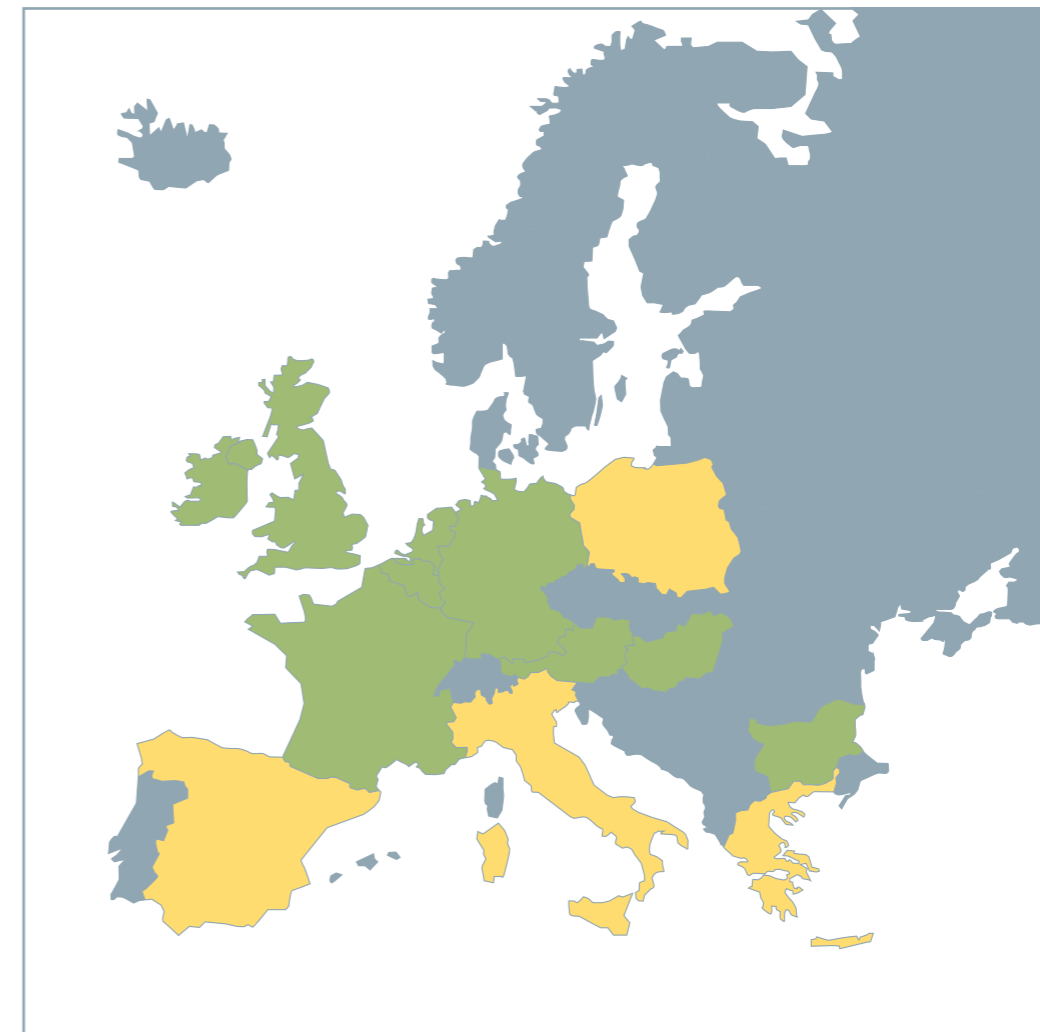
the evolution of the disease, postponing treatment. However, the unpredictable nature of IPF means that a patient’s condition can worsen rapidly: something that early adoption of antifibrotic treatment could mitigate.

Prescription rules are another significant hurdle. In most countries surveyed the prescription of pharmacological treatment is restricted to specialised ILD centres. However, in Austria, France, Germany, Ireland and Spain respiratory physicians can also prescribe treatment. Only in Ireland can general practitioners (GPs) prescribe antifibrotic treatments (with the renewal of prescriptions made by a respiratory physician).

Finally, treatment dispensation and the geographical distribution of centres present additional obstacles. In some countries (Belgium, Italy, Poland and Spain), treatment is only provided through hospital pharmacies or specialised centres. In Greece, only state pharmacies are allowed to dispense pharmacological treatment – but frequent shortages can result in long wait times for patients.

In other countries (Austria, Bulgaria, France, Germany, Hungary, Ireland, and The Netherlands), treatment can be dispensed by community pharmacies. This should make it easier to access treatment; however, not all community pharmacies have on-site availability of antifibrotic drugs, meaning patients have to order their treatment first before they can collect it. In the UK, community pharmacies offer home delivery, which speeds up access to prescribed treatment.

While, in principle, patients in all EU-IPFF member countries have access to and are reimbursed for antifibrotic treatments, it is clear that there are lingering disparities in the prescription and distribution processes between and within countries. To ensure equal access to treatment, these must be addressed.



# Access to non-pharmacological treatment



## Key challenge

Access to non-pharmacological treatment options is not homogeneous across Europe.

Non-pharmacological treatment options, such as lung transplantation, pulmonary rehabilitation, oxygen therapy and psychological support, are essential for holistic IPF care. Lung transplantation is currently the only curative form of treatment but it is far from ideal; access is limited, there are intrinsic risks associated with the procedure, and of course, there is a limited availability of organs. Oxygen therapy and pulmonary rehabilitation play an essential role in managing IPF but major inequalities persist as regards to patient access. Psychological support is often seen as a 'nice-to-have', with few hospitals or centres of expertise able to provide such support.

Lung transplantation is the only curative treatment available but it is only a viable option for a small number of IPF patients. There are many barriers to transplantation, such as the patient's health condition and age, the limited availability of viable organs, and long waiting times. There are specific limitations in some countries, such as in Poland, where there are a limited number of transplantation centres, or in Bulgaria and in Greece, which both lack lung transplantation centres – meaning patients must travel abroad for transplants.

Oxygen therapy at home or in hospital is a common therapeutic option for IPF patients. However, disparities exist between EU-IPFF Member States. These include: limited access (in Greece, it is only available in the hospital); delayed access; reimbursement issues (in Bulgaria and Poland, oxygen therapy at home is only partially reimbursed or not at all); or regional differences (in Ireland reimbursement is not standardised and depends on regional health budgets; in Italy, different regions provide different supply valves for the oxygen concentrators, affecting patients' in-country mobility).

As far as pulmonary rehabilitation is concerned, many respondents reported limited availability and reimbursement. In some countries, pulmonary rehabilitation programmes are not specifically dedicated to IPF patients. In Greece, for instance, IPF patients do not have access to such programmes – or if available, they are only reimbursed when administered in hospital. Both Bulgaria and Spain mentioned time limitations as their main barrier – in Spain rehabilitation is provided for four months, but it is only provided for ten days in Bulgaria.

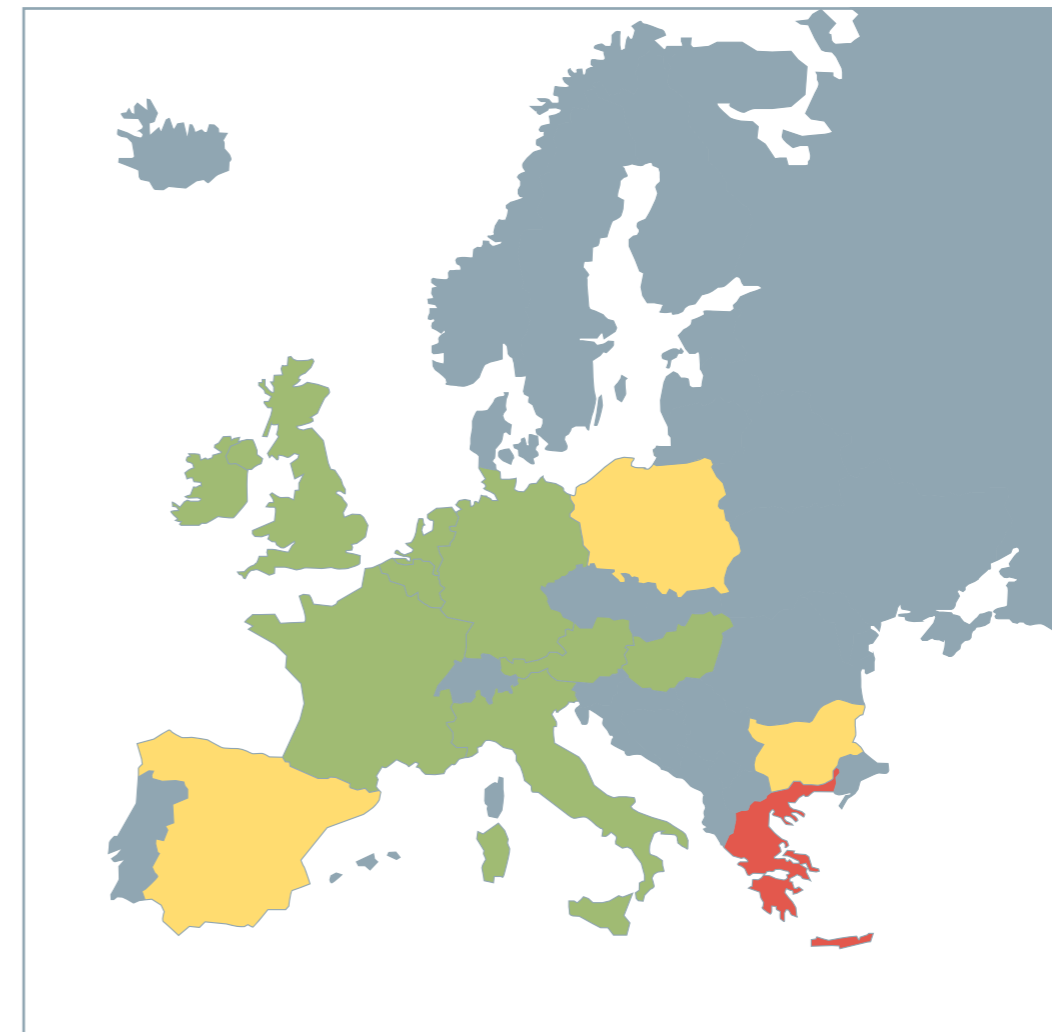
Only some countries (Belgium, France,

Germany, Ireland, UK) provide pulmonary rehabilitation at an ambulatory level, with very few countries having pulmonary rehabilitation centres either in-hospital or outside hospital at patients' disposal.

The majority of IPF patients reported a need for access to psychological and emotional support for themselves, for their families and for their carers. Access to psychological support is currently only available at a local level, through patient support groups. However, this is not systematically available and patients do not always know how to find help.

“The Spanish healthcare system greatly supports transplant programmes. In fact, in the last few years Spain has been recognised as a world leader in lung transplantation, and IPF is the most frequent reason for lung transplant.”

*A healthcare professional from Spain*



# Access to palliative care and end-of-life care



## Key challenge

Palliative care is not offered systematically to European IPF patients.

Access to palliative care and end-of-life care is the most fragmented area across Europe. In principle, it should play a key role in holistic IPF management; however, it is not sufficiently embedded in IPF care and it is not available or reimbursed in all the countries surveyed.

Many respondents explained that palliative care is often confused with end-of-life care, instead of being understood as symptom control and management. This further hinders access, since patients may be deterred from asking for palliative care if they believe it corresponds to end-of-life care. Many respondents also reported that existing end-of-life care programmes are usually focused on cancer patients, with no specific programmes in place for IPF patients.

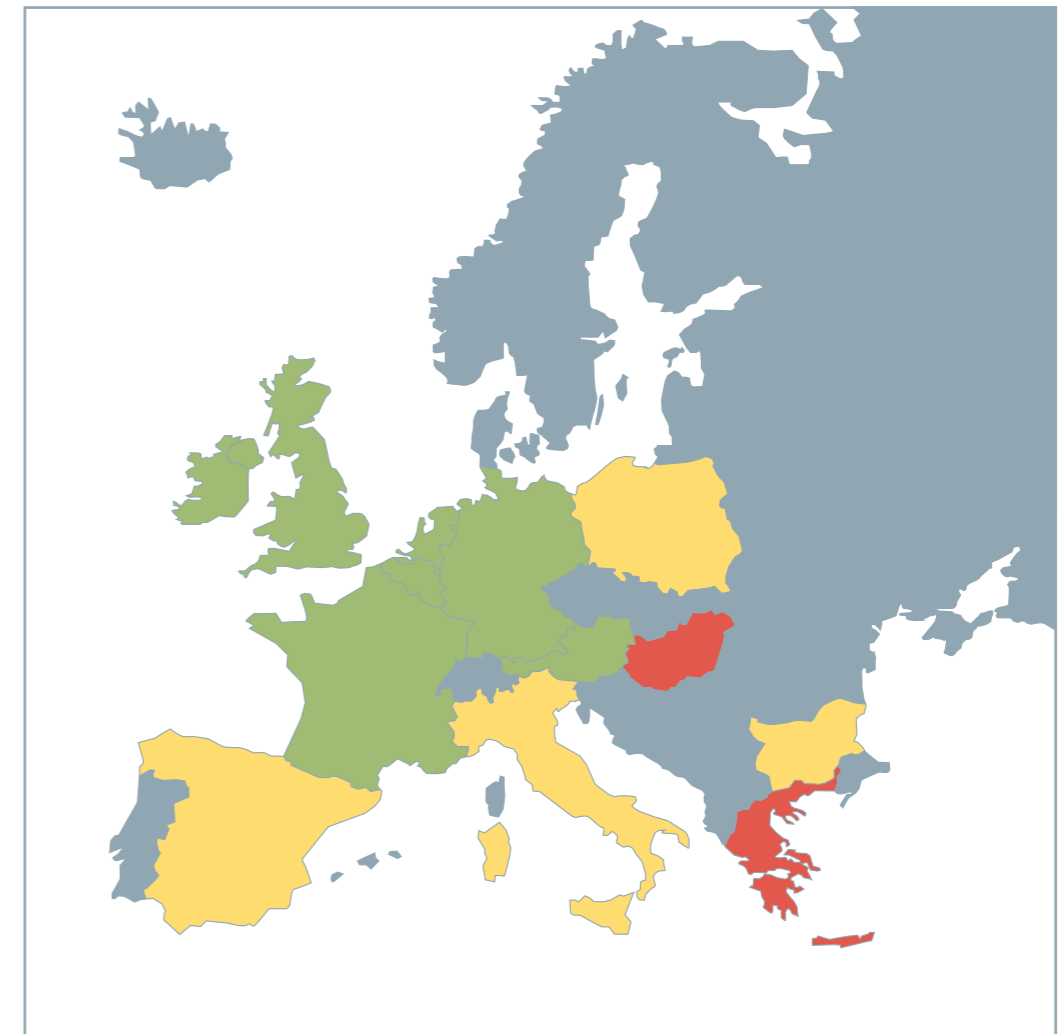
In countries where palliative care is offered, patients and their families are involved in decisions about this type of treatment. And when available, palliative care is usually delivered in hospitals and clinics with palliative care units. In some countries (e.g. UK) patients can choose to receive palliative care at home, but only Austria reported the availability of mobile palliative care teams.

Home hospice care is not widely available in Europe; in some countries, such as Bulgaria, Italy and Poland, only private hospices exist. Such costs are not reimbursed, which can create a significant financial burden for patients and their families.

All survey respondents highlighted the importance of a holistic approach to palliative care as part of IPF treatment plans.

*“In countries where the the organisation of healthcare services is decentralised, patients often report inequalities in access. This is particularly true in relation to oxygen therapy and palliative care. My hope is that the recommendations of patient and family organisations will be considered to ensure fair and equal access to quality care and support for IPF patients and caregivers across Europe.”*

*A patient representative from Italy*





# Conclusions

Many people living with a rare disease face significant challenges, from diagnostic delays and lack of available treatments, to difficulty in accessing appropriate health services. Patients and their families feel isolated, unsupported and economically curtailed.

This Report reviewed best practices across thirteen EU countries, identifying existing gaps in the care and management of IPF. Although the results may not reflect the wider European situation, they do provide important insights into health inequalities and areas for improvement.

The findings show an alarming variation in access to specialised care across countries. Not one of the countries surveyed was excelling on all indicators. However, some countries did display best practices that could be replicated elsewhere.

To enable transformation in IPF care and management, action must be taken in all countries, starting with the implementation of existing guidelines to help standardise processes and address health inequalities.

Greater awareness of IPF signs and symptoms among general practitioners is needed to address delays in diagnosis and increase referral to specialist care. Rapid referral can improve diagnosis and timely access to

treatment, which can slow disease progression. Addressing the disconnect between primary and secondary care, implementing more efficient treatment dispensing practices and ensuring that available treatments are delivered to the right patient in a timely manner could significantly improve the patient experience. Increasing access to multidisciplinary teams, including specialist nurses, is vital to challenge clinical inertia and improve patients' quality of life.

The Report showed that too many barriers exist in access to non-pharmacological treatment including lung transplantation, supplemental oxygen, pulmonary rehabilitation, and palliative care. Long waiting lists and lack of donor organ availability represent some of the main hindrances to lung transplantation. Oxygen therapy and pulmonary rehabilitation can make a positive impact but action must be taken to make it more available, accessible and affordable. Similarly, palliative care should be destigmatised and better embedded into IPF treatment plans. Finally, for patients living with IPF, access to psychological support is fundamental and must be offered systematically and holistically.

*"A rare disease like IPF has been unnoticed for years, but this Benchmarking Report represents a good opportunity to shed light on such a forgotten disease. The burden of the disease is not only on the patient himself, but also on the family and the society as a whole. The indirect costs of IPF and its implications on quality of life are significant and cannot be ignored."*

*A patient representative from Italy*



# Call to action

Across the countries surveyed, there is a clear need to improve the diagnostic, treatment and care pathway for IPF patients and to address fragmentation of IPF care, which is creating huge health inequalities between and within countries. To support evidence-based policy-making, the EU-IPFF have identified recommendations that address the main barriers previously outlined in the Report.

While healthcare delivery remains a Member State competence, there is a role for the EU to play in sharing best practices and pooling resources, to the benefit of all European IPF patients. The European Reference Networks, in particular, represent an important avenue for the improvement of all areas mentioned by this Report.

The upcoming 2019-2024 European parliamentary term represents an opportunity for future EU leaders to promote radical policy change that can improve healthcare services and outcomes. We call upon current and future European policy-makers to ensure that:



Rare diseases remain a priority in the future European Commission Agenda.



Funding is earmarked for rare diseases in the future Horizon Europe framework, in order to reach the ambitious target of developing 1.000 new therapies for rare diseases by 2027, including a cure for IPF.<sup>6</sup>



Greater support is given to the integration and sustainability of established European Reference Networks for screening at-risk groups, expediting diagnosis and encouraging standardisation of rare disease care (including access to non-pharmacological treatment).



Opportunities for rare disease patient groups to collaborate and capacity-build are encouraged through future funding programmes, such as the European Social Fund Plus and the European Joint Programme on Rare Diseases.

<sup>6</sup> As set by the International Rare Diseases Research Consortium (IRDiRC) - <http://www.irdirc.org/about-us/vision-goals/>

## At a national level, national and regional governments can improve the delivery of healthcare services by focusing on:



Raising awareness of IPF amongst healthcare professionals and primary care practitioners to improve the recognition of early signs and symptoms.



Ensuring the implementation of IPF guidelines within the country.



Supporting collaboration with relevant European Reference Networks and networking excellence at national level, to better connect primary and secondary care and foster early diagnosis and timely access to treatment.



Promoting multi-disciplinary care and recognition of specialised nurses at national level.



Recognising the burden and financial impact of IPF on patients and their families, and ensure access to non-pharmacological treatment, especially oxygen therapy.



Providing access to palliative care to all IPF patients.



Encouraging the development of local support groups, given their role in providing patients with information and peer support.

The EU-IPFF is committed to working with all stakeholders to ensure that action on rare diseases remains a priority area at European and national level and support exchange of knowledge to guarantee that people with IPF have access to fair, equal and continuous quality care regardless of where they live in Europe.

# The European Scorecard

INDICATORS	COUNTRIES												
	Austria	Belgium	Bulgaria	France	Germany	Greece	Hungary	Ireland	Italy	The Netherlands	Poland	Spain	United Kingdom
GUIDELINES													
Availability of IPF guidelines (national and international)	Green	Green	Yellow	Green	Green	Red	Green	Green	Yellow	Green	Green	Yellow	Green
Implementation of IPF guidelines	Green	Green	Yellow	Yellow	Green	Red	Green	Green	Yellow	Green	Green	Red	Yellow
Assessment of IPF care pathways	Red	Yellow	Green	Grey	Red	Red	Green	Green	Yellow	Grey	Red	Red	Green
DIAGNOSIS AND ACCESS TO SPECIALISED CARE													
Assessment of diagnostic procedure	Green	Green	Green	Green	Green	Green	Green	Green	Green	Green	Green	Yellow	Green
Assessment of standardisation of IPF diagnosis	Green	Green	Grey	Grey	Green	Red	Yellow	Grey	Yellow	Green	Grey	Yellow	Green
Availability of information materials	Yellow	Yellow	Yellow	Yellow	Yellow	Yellow	Yellow	Yellow	Yellow	Yellow	Yellow	Yellow	Yellow
Assessment of self-management education tools	Red	Red	Yellow	Yellow	Yellow	Red	Red	Green	Yellow	Yellow	Yellow	Red	Yellow
Assessment of distribution and activities of specialised centres	Green	Green	Yellow	Green	Yellow	Yellow	Grey	Green	Yellow	Green	Yellow	Yellow	Yellow
Availability of and access to multidisciplinary team	Green	Green	Yellow	Green	Green	Red	Grey	Green	Green	Green	Yellow	Red	Green
ACCESS TO PHARMACOLOGICAL TREATMENT													
Availability of pharmacological treatment	Green	Green	Green	Green	Green	Green	Green	Green	Green	Green	Green	Green	Green
Reimbursement of pharmacological treatment	Green	Green	Green	Green	Green	Green	Green	Yellow	Green	Green	Green	Green	Green
Assessment of distribution of prescription centres	Green	Yellow	Green	Green	Green	Yellow	Green	Green	Yellow	Green	Yellow	Yellow	Yellow
Assessment of distribution of dispensation centres	Green	Green	Green	Green	Green	Yellow	Green	Green	Yellow	Green	Yellow	Yellow	Green
ACCESS TO NON PHARMACOLOGICAL TREATMENT													
Availability of and access to oxygen therapy in the hospital	Green	Green	Yellow	Green	Green	Yellow	Green	Green	Green	Green	Green	Green	Green
Reimbursement of oxygen therapy in the hospital	Green	Green	Green	Green	Green	Yellow	Green	Green	Green	Green	Green	Green	Green
Availability of and access to oxygen therapy at home	Green	Green	Green	Green	Green	Red	Green	Green	Green	Green	Yellow	Green	Green
Reimbursement of oxygen therapy at home	Green	Green	Red	Green	Green	Red	Yellow	Yellow	Green	Green	Yellow	Green	Green
Availability of and access to pulmonary rehabilitation programmes	Green	Green	Yellow	Green	Green	Red	Green	Green	Yellow	Green	Yellow	Yellow	Yellow
Coverage of pulmonary rehabilitation programme	Green	Yellow	Green	Green	Green	Red	Green	Green	Yellow	Green	Yellow	Yellow	Green
Availability of lung transplants	Green	Green	Red	Green	Green	Red	Green	Green	Green	Green	Yellow	Green	Green
Availability of and accessibility to information on clinical trials	Green	Green	Green	Green	Green	Red	Green	Green	Yellow	Green	Green	Green	Green
ACCESS TO PALLIATIVE CARE AND END-OF-LIFE CARE													
Availability of and access to palliative care	Green	Green	Green	Green	Green	Red	Red	Green	Yellow	Green	Yellow	Yellow	Yellow
Coverage of palliative care	Green	Green	Red	Green	Green	Red	Grey	Green	Yellow	Green	Yellow	Green	Green
Availability of palliative care centres and mobile teams	Green	Green	Grey	Green	Green	Red	Grey	Green	Red	Green	Green	Yellow	Green
Inclusivity of decision-making process for patients and families in palliative care	Green	Green	Grey	Green	Green	Red	Grey	Green	Yellow	Green	Green	Yellow	Yellow
Availability of and access to home hospice	Yellow	Yellow	Yellow	Yellow	Green	Red	Red	Yellow	Red	Green	Yellow	Yellow	Yellow

# IPF National Scorecards



# Austria

## IPF Scorecard

### General introduction and guidelines

According to a national survey conducted in Austria, national guidelines are currently in preparation. In the meantime, Austria implements ATS/ERS/JRS/ALAT international IPF guidelines.

In 2011, the Austrian Health Ministry created a national coordination centre for rare diseases ([Nationale Koordinationsstelle für seltene Erkrankungen - NKSE](#)). The objective of this centre is to improve care for people living with rare diseases. Its current priority is the coordination of Austrian hospitals participating in European Reference Networks, and the establishment of reference centres and care pathways for rare diseases.

In addition, in 2014, the Austrian government issued an action plan for rare diseases called [NAP.se](#). A condensed five-page document can be found [here](#). The action plan has nine concrete actions which aim to be worked on until 2018. An update is expected in 2019.

### Diagnosis and access to specialised care



**Key challenge:** Although diagnosis is considered standardised in clinics caring for IPF patients, timely referral and late diagnosis remain a challenge.

There are no official ILDs specialised centres in Austria but some centres have expertise in ILDs and work with multidisciplinary teams. In 2015, the [Ministry of Health](#) said they would prioritise the establishment of centres that are specialised in rare diseases. A list of six disease areas was prioritised; lung fibrosis was not part of this list. The specialised rare diseases centres to be established in 2018/2019 include: dermatology, facial deformalities, bone diseases, paediatric diseases, growth impairment, epileptic diseases and movement disabilities.

In clinics caring for IPF patients, the diagnostic procedure has been reported to be standardised. International IPF guidelines are properly implemented and multidisciplinary discussions take place to support diagnosis. Upon diagnosis, information materials are available for patients, most of the time through local patient support groups.

### Access to non-pharmacological treatment



**Key challenge:** IPF patients do not have access to specialised ILD nurses and there are no pulmonary rehabilitation programmes specifically designed for IPF patients.

Non-pharmacological treatment options are available to IPF patients in Austria and the costs associated with these treatments are reimbursed. Oxygen therapy at home or in hospital is available to patients upon prescription by the treating clinics or the respiratory physician. Pulmonary rehabilitation programmes are also available in hospital, but they do not necessarily take into account the specific needs of IPF patients. Lung transplantation is an option, however the patient's age, general health condition and the risks associated with transplantation could represent serious barriers. IPF patients have access to clinical trials.

### Access to palliative care and end-of-life care



**Key challenge:** The important role that palliative care can play in ensuring a better quality of life for IPF patients is not fully recognised.

Palliative care is available to IPF patients in Austria and it is normally delivered in clinics with palliative care units; mobile palliative care teams are also an option. However, long waiting times for hospice care have been reported.

In the overall patient journey, the respiratory physician, the multidisciplinary team and the patient groups are the ones providing relevant support to IPF patients and their families. Additional psychological and emotional support for patients and their carers would be needed.

### Access to pharmacological treatment



**Key challenge:** The 'wait and watch' approach has been flagged as one of the main barriers to prompt access to pharmacological treatment.

EMA-approved pharmacological treatments for IPF are available and reimbursed with no out-of-pocket costs for patients.

The pharmacological treatments can be prescribed by the clinics following IPF patients and/or by a respiratory physician; they are dispensed by community pharmacies. Nevertheless, the lack of disease awareness results in treatment access delays.

### Patient statement

*"In Austria there are gaps in the availability of non-pharmacological care. In particular, there is no satisfactory IPF home care service and IPF patients do not have access to a specialised ILD nurse. In some rehabilitation centres too little attention is paid to the specific needs of IPF patients."*

*Unfortunately, there are hardly any educational tools for patients on IPF self-management. Local patient support groups and healthcare professionals tackle these issues and provide support to patients and their families."*

**Günther Wanke, Lungenfibrose Forum-Austria**



# Belgium IPF Scorecard

## General introduction and guidelines

The ATS/ERS/JRS/ALAT international IPF guidelines are implemented in specialised centres in Belgium but there are no national IPF guidelines. In Belgium there also is a rare disease [plan](#) which was published in 2013.

**Key challenge:** Delayed diagnosis and timely referral remain major challenges in Belgium. Recognising early signs and symptoms of IPF is considered to be essential to accelerate patient referral to a respiratory specialist.

## Diagnosis and access to specialised care



Seven centers in Belgium have the general rare disease entry, but this is not on the level of the different diseases.

Diagnostic procedures for IPF are standardised within ILDCentres, where IPF care pathways are in place and follow-up is performed. The ILDCentres work with different satellite centres in a hub and spoke model but there is no structured cooperation with primary care.

IPF guidelines are properly implemented in the ILDCentres and multidisciplinary discussions take place. Upon diagnosis, informative materials are available to patients, usually thanks to local patient support groups.

## Access to pharmacological treatment



**Key challenge:** In the patients' opinion, changing the prescription rules by allowing general practitioners to prescribe IPF treatments could improve access.

EMA-approved pharmacological treatments for IPF are available and reimbursed with no out-of-pocket costs for Belgian patients. The pharmacological treatments are prescribed and dispensed by the ILDCentres (at this moment only 3 centres comply with the criteria to prescribe).

## Access to non-pharmacological treatment



**Key challenge:** Lung transplantation is an option for IPF patients but the evaluation and weight attributed to some parameters can vary between the different centres.

Non-pharmacological treatment options are available for IPF patients in Belgium and costs are reimbursed. Oxygen therapy at home or in hospital are available to patients upon doctor prescription. Pulmonary rehabilitation programmes are also available in hospital but only 60% of the costs are reimbursed by the health insurance (additional costs might be covered by a complementary health insurance).

Although lung transplantation can be offered to IPF patients, there are big differences in how ILDCentres assess and weigh criteria to offer lung transplantation. IPF patients have access to clinical trials.

## Access to palliative care and end-of-life care



**Key challenge:** The government does not sufficiently support the provision of IPF care in ILDCentres. The ILDCentres should be empowered to provide the necessary support to improve quality of palliative care for IPF patients.

Palliative care is available to IPF patients in Belgium. It is normally delivered in hospital, at home or in specialised centres. As highlighted in a [2017 report for the European Institute of Bioethics](#), Belgium is ranked quite high for its performance in and availability of palliative care. The main palliative care hurdles are that it is often proposed too little and too late, it should be more embedded in traditional care and home availability of palliative care should be reinforced.

In the overall patient journey, the respiratory physician, the specialist nurse together with other members of the multidisciplinary team (such as the psychologist and the assistant nurse) are the ones providing relevant support to the IPF patients and their families. Patient groups also play an important role when it comes to psychological and emotional support for patients and their carers.

## HCP statement

*"It is vital that primary care physicians listen for the initial signs and symptoms of IPF (especially the Velcro-like crackles) and refer patients to a respiratory specialist. Through a network of specialised doctors, even the more complex patients could be quickly referred to the ILDCentres."*

**Prof. Wim Wuyts, University Hospital Leuven**



# Bulgaria

## IPF Scorecard

### General introduction and guidelines

The ATS/ERS/JRS/ALAT international IPF guidelines are available in Bulgaria; however, they are not always correctly implemented in the Bulgarian centres. No national IPF guidelines are available.

IPF is included in the a [list of rare diseases](#). However, the disease file including information about the course of disease and its prognosis is still under development.

**Key challenge:** Delayed diagnosis and lack of communication between the general practitioner and the respiratory physician have been reported as the main challenges in Bulgaria. Timely referral to a respiratory specialist would ensure accurate diagnosis and faster access to treatment.

### Diagnosis and access to specialised care



In Bulgaria there are no specialised ILD centres but there are 6 hospitals specialised in lung diseases, which do not operate fully as specialised centers (the University Hospital of respiratory diseases "St. Sofia" - Sofia Medical University; the pulmonary department of the Military Hospital Sofia - Medical Faculty of Sofia University; the Acibadem City Clinic Tokuda Hospital in Sofia; the University Hospital "St. George"- Plovdiv Medical University; the University Hospital "St. Marina" - Medical University Varna; the University Hospital " G. Stranski" - Medical University Pleven).

IPF guidelines are only partially implemented in these hospitals. Although multidisciplinary teams (MDTs) MDTs are becoming more common in Bulgarian hospitals, MDTs for IPF care are available and fully in place in only one centre. There are no ILD nurses who can support patients in their daily lives. Upon diagnosis, information materials are available to patients, mainly provided by the local patient support groups.

**Key challenge:** Although both EMA-approved pharmacological treatments for IPF are approved and reimbursed, patients still experience some delays in access to treatment as the therapy is not started immediately after diagnosis.

### Access to pharmacological treatment



EMA-approved pharmacological treatments for IPF are available and reimbursed with no out-of-pockets costs for Bulgarian patients. The pharmacological treatments can be prescribed by five of the clinics specialised in lung diseases and they are dispensed by community pharmacies.

The current prescription rules might represent a barrier to access to pharmacological treatment; it has been reported that allowing general practitioners or respiratory physicians to prescribe treatment has the potential to improve the issue.

### Access to non-pharmacological treatment



**Key challenge:** There are currently no clinics in Bulgaria that can perform lung transplantation. However, the Bulgarian Ministry of Health has recently signed an agreement with a foreign clinic that can perform lung transplantation for Bulgarian patients. Access to pulmonary rehabilitation remains too limited.

Non-pharmacological treatment options are available to IPF patients in Bulgaria but the costs associated with these treatments are not always fully reimbursed. Oxygen therapy is available in hospital and at home but is only reimbursed in the case of the former.

Pulmonary rehabilitation programmes for patients with chronic lung diseases are available in hospital but no specific IPF programmes have been developed. The costs are covered only for a period of 10 days per year and any additional days are at the expense of the patient.

Lung transplantations are not performed in Bulgaria. IPF patients in Bulgaria have access to clinical trials.

### Access to palliative care and end-of-life care



**Key challenge:** Palliative care costs are not reimbursed for IPF patients.

Palliative care is available to IPF patients in Bulgaria but it is not reimbursed by the national healthcare system. Only private hospices provide this type of care and the costs can be high.

In the overall patient journey, the families and local support groups are the ones providing relevant support to IPF patients.

### Patient representative statement

*"Delayed diagnosis is a big issue in Bulgaria and it is mainly due to lack of knowledge of the disease at primary care level and late referrals to respiratory physicians. In addition, the lack of communication between specialised centres and the National Health Insurance Fund, which reimburses the treatment, causes further delays in access to pharmacological treatments . We at IPF Bulgaria, hope that this situation will improve soon so that access to pharmacological treatments for IPF patients will not be delayed unnecessarily."*

**Todor Mangarov, IPF Bulgaria**



# France

## IPF Scorecard

### General introduction and guidelines

The ATS/ERS/JRS/ALAT international IPF guidelines are implemented in specialised centres in France. In 2017, the network of rare diseases in France published national guidelines on the diagnosis and treatment of IPF (available [here](#)). OrphaLung, the network of rare pulmonary diseases centres, is accredited by the French Ministry of Health and [treats numerous rare pulmonary conditions](#), including IPF.

**Key challenge:** Timely referral has been reported as the main challenge in France. Recognising early signs and symptoms of IPF is considered to be essential to accelerate patient referral to a respiratory specialist.

### Diagnosis and access to specialised care



There are 23 specialised centres for rare pulmonary diseases in adults in France. They are integrated in the OrphaLung network (Network of rare pulmonary diseases centres). OrphaLung is part of RespiFil which, in addition to the centres involved in OrphaLung, brings together reference centres and competence centres working on paediatric rare pulmonary diseases and pulmonary hypertension.

This is very helpful for physicians to refer difficult cases of ILD; however not all patients need to be referred to these centres as some centres work well with primary care providers.

The diagnosis process is pretty much standardised, as the French IPF guidelines are properly implemented and multidisciplinary teams are available in the specialised centres. However, there is some heterogeneity in clinical practice and quality of care, as well as a certain lack of education and information, which if solved could lead to earlier diagnosis.

### Access to pharmacological treatment



**Key challenge:** Ensuring timely diagnosis can contribute to guaranteeing timely access to pharmacological treatment.

EMA-approved pharmacological treatments for IPF are available and reimbursed with no out-of-pocket costs to patients.

The pharmacological treatments can be prescribed by specialised centres and hospital respiratory physicians and are dispensed by community pharmacies (upon order placement).

### Access to non-pharmacological treatment



**Key challenge:** Although pulmonary rehabilitation is available, access is still limited by a number of factors (disparity of centres across the country, reimbursement issues).

Non-pharmacological treatment options are available to IPF patients in France and costs are reimbursed.

Oxygen therapy at home or in hospital are available to patients upon doctor prescription.

Pulmonary rehabilitation programmes are also available in hospital and at home; however rehabilitation needs to be initiated in the hospital. Some issues remain such as the geographical distribution and availability of outpatient rehabilitation centres across France as this is not well organised and not available everywhere. Reimbursement of pulmonary rehabilitation is available, if it has been initiated in long-term and rehabilitation care units.

Lung transplantation is an option for patients. However the age, the overall health status and the progression of the disease could represent serious barriers.

IPF patients in France have access to clinical trials through the OrphaLung network.

### Access to palliative care and end-of-life care



**Key challenge:** Although palliative care is available, patients have expressed the need for more support around palliative care and pain management.

Palliative care is available to IPF patients and it is normally delivered in hospitals or in specialised palliative care centres. However, palliative care is mainly dedicated to oncology patients, hence there is limited access for patients with IPF.

However not all patients have access to it in a timely manner. A recent survey conducted with IPF and PF patients showed that 18% of surveyed patients expressed the need for more support around palliative care and pain management.

In the overall patient journey, the respiratory physician and the specialist nurse are the ones providing relevant support to the IPF patients and their families. Patient groups also play an important role when it comes to providing psychological and emotional support to patients and their carers.

### HCP statement

*“Many patients and primary care providers are not aware of the OrphaLung network of ILD centres. We hear stories of patients being referred to the cardiologist first, then to a pulmonologist before finally coming to the expert centre. It feels like a relief for patients to finally have an accurate diagnosis, with detailed information given to them with regards to what to expect from treatment and prognosis.”*

**Vincent Cottin, CHU Lyon**





# Germany

## IPF Scorecard

### General introduction and guidelines

Germany has national IPF guidelines and recognizes international guidelines (ATS/ERS/JRS/ALAT). The German national guidelines were published in the medical journal [Pneumologie](#) in 2003 and were [updated](#) in 2017 to include new treatment therapies. The guidelines were also [translated and updated](#) into English in 2018. The national guidelines were developed under the umbrella initiative, [Deutschen Gesellschaft für Pneumologie und Beatmungsmedizin – The German Association for Pneumology and Airway-medicine](#).

IPF is included in the [national list of rare diseases](#). In addition, since 2012 Germany has been collecting and analysing data on the burden of IPF across the country through an expert-driven IPF registry called INSIGHTS-IPF (results are available [here](#) - log-in information required). An ILD registry from in and out-patient facilities - the [EXCITING registry](#) – is also available.

**Key challenge:** The lack of general IPF awareness is leading to delayed specialist referrals and consequently delays timely access to treatment.

### Diagnosis and access to specialised care



The national guidelines on IPF, including recommendations on diagnosis, have been updated in 2017 and are available [at this link](#). This document mainly discusses the various treatment options. Section 3 (pp. 85-90) of the original [German guidelines](#), published in 2013, provided an illustration of the diagnostic pathway (figure 3); it is now commonly used across Germany. Despite various resources, the referral to specialist care remains a key challenge and consequently delays access to treatment.

There are 25 specialised ILD centres across Germany, where multidisciplinary teams are in place. Upon diagnosis, informative materials are available for patients, mainly provided by the local patient support groups.

**Key challenge:** Early diagnosis and timely access to treatment remains the main challenge. Ensuring patients go to the general practitioner can be the first step towards early diagnosis.

### Access to pharmacological treatment



EMA approved pharmacological treatments for IPF are available and reimbursed with no out-of pockets costs for patients in Germany. The pharmacological treatments can be prescribed by the specialised centres, by the respiratory physician and by other physicians with experience in the diagnosis and treatment of IPF. The treatments are dispensed by community pharmacies.

### Access to non-pharmacological treatment



**Key challenge:** The costs of pulmonary rehabilitation are covered only when it is performed in hospital.

Non-pharmacological treatment options are available to IPF patients and costs are reimbursed. According to the German IPF guidelines, various non-pharmacological treatment options are recommended including: long-term oxygen therapy, lung transplantation and pulmonary rehabilitation. These therapy options are available in hospital and in some cases at home upon doctor's prescription. All therapy options are reimbursed if a doctor's prescription is available. Patients have mentioned that challenges persist if the patient requires pulmonary rehabilitation at home. Pulmonologists and specialised centres are also aware of ongoing clinical trials and they communicate this to patients when applicable.

### Access to palliative care and end-of-life care



**Key challenge:** Patients are not always aware of the potential benefits palliative care can have in the management of the disease and how it can improve their quality of life.

Palliative care is available to IPF patients and is normally reimbursed upon doctor's prescription. End-of-life-care is provided by hospices, palliative care facilities and family carers at home.

In the overall patient journey, the family, the respiratory physician and the patient support group are the ones providing relevant support to the IPF patients and their families. Patient groups also play an important role when it comes to providing psychological and emotional support for patients and their carers. Nevertheless, the lack of information and communication about options beyond pharmacological treatments may be a barrier for patients in Germany. Communication to patients and their families about palliative care in general and end-of-life care in particular is still influenced and made difficult by the general misconception of palliative care as medicines for dying patients.

### Patient statement

*“As a patient I feel like my doctor should explain to me the benefits of palliative care already at the early stage of the disease so that I would have more time to better understand how it can help me in my daily life, before it really becomes necessary.”*

*I also do not have a clear idea about how the end-of-life stage will look like and I feel there should be more communication about this because it is not only physically but also psychologically very challenging to deal with this disease.”*

**Klaus Geissler, Lungenfibrose e.V**



# Greece

## IPF Scorecard

### General introduction and guidelines

The ATS/ERS/JRS/ALAT 2018 international IPF guidelines are not always correctly implemented in Greece. The Hellenic Thoracic Society and the Scientific Society for Rare Diseases & Orphan Drugs organise a number of scientific activities for physicians throughout the year that help address some of the implementation issues.

The Greek Alliance of Rare Diseases refers to the ORPHANET's [list of rare diseases](#) among which IPF is included. However, an official list is not available.

### Diagnosis and access to specialised care



**Key challenge:** Getting a correct and timely diagnosis is very challenging as general practitioners are not always aware of the disease.

There are a few specialised centres in major Greek cities; however, there are no IPF patient pathways in place. Misdiagnosis and late diagnosis are very common in the country a multidisciplinary teams are not available to patients.

Upon diagnosis, information materials are available for patients; they are provided by the existing patient support group and the specialised centres.

**Key challenge:** Only state pharmacies are allowed to dispense pharmacological treatment, pending the approval of doctor's application by a government appointed committee, creating unnecessary delays in patient access.

### Access to pharmacological treatment



EMA-approved pharmacological treatments for IPF are available and reimbursed with no out-of pockets costs for patients. The pharmacological treatments can only be prescribed by physicians working in state or private hospitals. These physicians need to submit an application - including all the necessary examinations - to a government-approved committee and it might take up to 10-15 days in order to get an approval (due to the treatment's high cost). This can result in long waiting times for patients. There are also issues with point-of-care dispensing as the pharmacological treatment can only be dispensed by the national health scheme pharmacies.

Access to pharmacological treatments also differs from one city to another. In fact, in smaller cities, the distribution system is more efficient and less exhausting for the patient, who receives the pharmacological treatment at home by post. On the contrary, in larger cities patients experience long waiting times in order to get the treatment.

### Access to non-pharmacological treatment



**Key challenge:** Access to non-pharmacological treatments is very poor in Greece, in particular lung transplantation which is virtually not an option for Greek patients as there is no lung transplantation centre in the country.

Non-pharmacological treatment options are not always available and fully reimbursed for IPF patients in Greece. For example, to have access to liquid oxygen, patients need to pay 25% of the cost.

IPF patients do not have easy access to pulmonary rehabilitation programmes, since only a few hospitals provide this service. At the moment, there is no lung transplantation centre in Greece. Greek patients have to go to a lung transplantation centre abroad (usually in Austria). Given that there are not enough lung donors from Greece the majority of Greek candidates cannot receive a transplant, even abroad.

There are a few Greek centres participating in clinical trials but access to these trials seems to be problematic for patients.

### Access to palliative care and end-of-life care



**Key challenge:** Patients have limited access to palliative care.

Palliative care is offered only in a few hospitals in the country. Home hospice is not a viable option for patients.

Families and respiratory physicians are the ones providing relevant support to IPF patients. IPF has a huge impact on family members acting as the patient carer; there is a significant loss in terms of productivity at work for these relatives who would need to quit their job to take care of the patients.

### Patient statement

*"In addition to proper and effective medical and professional help, patients need the support of family, friends and patient organisations at both national and European level, to give us hope, and belief in a better future for IPF patients."*

**Anthony Antoniou, Hellenic Pulmonary Fibrosis Association**



# Hungary

## IPF Scorecard

### General introduction and guidelines

The ATS/ERS/JRS/ALAT international IPF guidelines are implemented throughout the country. Hungarian adaptation of the international guidelines is under development.

### Diagnosis and access to specialised care



**Key challenge:** The lack of standardised diagnostic process and delayed diagnosis remain the main challenges in Hungary.

There are 12 [specialised ILD centres](#) in Hungary. The diagnostic process is not standardised and varies depending on each centre's experience. Upon diagnosis, information materials are available for patients at the specialised centres and are provided by local patient support groups.

### Access to pharmacological treatment



**Key challenge:** Long waiting times to be referred to a specialised centre might hinder prompt access to pharmacological treatment for patients.

EMA-approved pharmacological treatments for IPF are available and [reimbursed](#) to patients. The pharmacological treatments can only be prescribed by ILD centres but can be dispensed by any community pharmacy.

### Access to non-pharmacological treatment



**Key challenge:** Supplemental oxygen for travelling is only available in liquid form and this reduces patients' longterm mobility .

Non-pharmacological treatment options are not always available and fully reimbursed to IPF patients in Hungary. Oxygen therapy is available in hospital, where it is fully reimbursed. Several forms of oxygen therapy at home are also reimbursed with the exception of mobile oxygen concentrators. Inpatient pulmonary rehabilitation is available to all IPF patients and is fully reimbursed. Hungarian patients can have access to [lung transplantation](#); however age and comorbidities may limit this access. IPF patients in Hungary have access to clinical trials.

### Access to palliative care and end-of-life care



**Key challenge:** Patients have limited access to palliative care.

[Palliative care](#) can be provided at home and at certain inpatient care facilities. It is considered a last resort option when other therapeutic options have been exhausted.

In the overall patient journey, the respiratory physician and specialist nurse are the ones providing relevant support to IPF patients.

### Patient statement

*“Unfortunately IPF is still an unknown rare disease. A significant effort is needed to raise awareness of IPF, especially among primary care providers, to make sure that patients are referred to the specialised centres. There is a lot to be done to increase IPF patients access to ambulatory oxygen and palliative care, and to ensure they receive adequate support for reduced mobility from the government.”*

**Maria Rigo, Magyar Tudofibrozis Egyesulet**





# Ireland IPF Scorecard

## General introduction and guidelines

The ATS/ERS/JRS/ALAT international IPF guidelines seem to be correctly implemented in Ireland. In August 2018, national guidelines were developed by the Irish Thoracic Society (ITS) and a Position Statement on the Management of Idiopathic Pulmonary Fibrosis (available [here](#)) was published. These guidelines are implemented in the Irish specialised ILD centres.

A [National Patient Charter for IPF](#) was developed by the Irish Lung Fibrosis Association (ILFA) and launched in 2015. It calls for early and accurate diagnosis, access to treatment and support services for patients.

In addition, the Irish Thoracic Society created an Interstitial Lung Group that set up a National Patient Registry to record the incidence of and clinical data on patients with IPF in Ireland. This spans all the hospital groups with most specialised centres actively participating.

**Key challenge:** Early diagnosis remains the main challenge. More awareness of IPF is needed, especially at a primary care level to recognise the features of IPF and stimulate referrals.

## Diagnosis and access to specialised care



Diagnostic procedures for IPF are standardised and IPF guidelines seem to be well implemented within the centres with specialist interest in ILD, where multidisciplinary teams are in place (an IPF multidisciplinary care pathway is included in the ITS statement on IPF). These centres are well distributed across the country and they work in collaboration with other hospitals regionally and with primary care providers.

Upon diagnosis, information leaflets and printed materials are available for patients from some healthcare professionals and ILFA. IPF self-management is also considered an integral part of IPF care.

**Key challenge:** Ensuring patients are referred in a timely manner to an ILD specialised center to facilitate prompt access to pharmacological treatment remains a concern.

## Access to pharmacological treatment



The two EMA-approved pharmacological treatments for IPF are available and reimbursed with no out-of pockets costs to patients in Ireland (only if a patient has a medical card). The pharmacological treatments can be prescribed by respiratory physicians and the prescription can be renewed by general practitioners. The antifibrotic treatments are dispensed by community pharmacies.

## Access to non-pharmacological treatment



**Key challenge:** Pulmonary rehabilitation programmes are available but only in some hospitals, and they do not specifically target IPF patients

Non-pharmacological treatment options are available to IPF patients and costs are reimbursed (only if a patient has a medical card).

Oxygen therapy at home or in hospital is available to patients with a doctor's prescription. Oxygen therapy for patients admitted to hospital is provided for free while the cost of home oxygen is fully reimbursed only if a patient has a medical card. Partial reimbursement to cover the increased electricity costs associated with using oxygen therapy is not standardised and it depends on regional/local health budgets. Pulmonary rehabilitation programmes are also available in some hospitals and in the community. However, it appears that at present the majority of pulmonary rehabilitation services target patients with Chronic Obstructive Pulmonary Disease (COPD), therefore there is a clear need to expand resources to include more patients with IPF.

Lung transplantation and clinical trials are also available to IPF patients.

## Access to palliative care and end-of-life care



**Key challenge:** IPF patients have access to home hospice but waiting times vary across the country.

Palliative care is available to IPF patients across all settings (community, hospital and hospice) and it is fully reimbursed.

In the overall patient journey, the family, the respiratory physicians, the nurses, the patient organisation and the support groups provide relevant support to IPF patients and their families.

## Patient representative statement

*"While it appears that Ireland is performing well in some indicators, IPF care could be better. National healthcare policies, education and awareness activities and healthcare investment are needed to improve the IPF clinical care pathway and to ensure patients have access to an early and accurate diagnosis and treatments, and that healthcare professionals are optimally resourced. ILFA and the Irish Thoracic Society will continue to lobby for improved resources and an ILD clinical care programme."*

**Nicola Cassidy, Irish Lung Fibrosis Association**



# Italy IPF Scorecard

## General introduction and guidelines

A [Position Paper](#) on the diagnosis and management of IPF was adopted in 2015 by the Italian Association of Hospital Pulmonologists (Associazione Italiana Pneumologi Ospedalieri, AIPO) and the Italian Medical Respiratory Society (Società Italiana Medicina Respiratoria, SIMeR). As the organisation and delivery of healthcare services are a competence of the Italian Regions, regional IPF care pathways are not available in all regions.

The very last version of the 2018 ATS/ERS/JRS/ALAT international IPF guidelines has not been translated, and these guidelines are not always correctly implemented across all the specialised centres.

The Italian Ministry of Health regularly updates a [List of Rare Diseases](#) that are covered by the National Health System. IPF was included in the List in 2017.

**Key challenge:** Delayed diagnosis and late referral to the specialised centres have been reported as the main challenges in Italy.

## Diagnosis and access to specialised care



The diagnostic procedure is not standardised and it might differ between regions, leading to different quality of care. Specialised ILD centres are available across the entire territory and are listed in the [IPF Centres Guide](#) (Observatory of Rare Diseases, Osservatorio Malattie Rare). Multidisciplinary discussions take place in these centres with the objective of achieving a correct diagnosis. [Late diagnosis](#) remains an issue as not all IPF patients are referred to the specialised centres in a timely manner. Although they are critical support system for patients and carers, there are shortages and lack of recognition of [specialist ILD nurses](#). Upon diagnosis, information materials are available for patients, mainly provided by the local patient support groups.

**Key challenge:** Prescribing centers are identified at a regional level and the number of centers might differ a lot from one region to another one ranging from 1 or 2 centers per region to 20 centers per region.

## Access to pharmacological treatment



EMA-approved IPF pharmacological treatments for are available and reimbursed with no out-of pockets costs to Italian patients. The pharmacological treatments are prescribed by specialist respiratory physicians (as allowed by the regional Health System) and treatment is only provided through hospital pharmacies.

## Access to non-pharmacological treatment



**Key challenge:** Inequalities persist between regions when it comes to access to non-pharmacological treatments, in particular re. oxygen therapy (different supply valves that can limit patient mobility) and pulmonary rehabilitation (only reimbursed in hospital)

Non-pharmacological treatment options are not always available to IPF patients in Italy and the costs associated with these treatments are not always fully reimbursed.

Oxygen therapy is available and reimbursed both in hospital and at home. The main issue is that [different regions provide different supply valves](#) for the oxygen concentrators affecting patients' in-country mobility.

Pulmonary rehabilitation – when available – is only reimbursed when it is performed in hospital. Private centres providing rehabilitation exist but the patients have to bear all the costs. Huge discrepancies among the regions have also been reported, with the biggest difference between the North and the South of the country.

Lung transplantations are performed in Italy but [donor availability and long waiting times](#) (up to two years) can represent serious barriers.

IPF patients have access to clinical trials although retrieving information about it might be difficult at times.

**Key challenge:** Palliative care is not always available for IPF patients in Italy

## Access to palliative care and end-of-life care



Palliative care is [not always available](#) to IPF patients in Italy but when it is made available, it is covered by the National Healthcare System. Priority is given to terminaly ill patients and access to patients with other diseases might be very difficult.

For end-of-life care, only private hospices are available and the costs can be quite high.

In the overall patient journey, the respiratory physician, the family and the local support groups are the ones providing relevant support to IPF patients.

## HCP statement

*“Delayed diagnosis and late referral to the specialised centres are two of the main issues affecting IPF patients in Italy. Patients with suspected IPF should promptly be referred to an ILD specialised centre. Through a team of different professionals who are expert in the field, the patient could possibly avoid invasive procedures and have a correct diagnosis, which is the starting point for the proper management of IPF and a better quality of life.”*

**Elisabetta Balestro, University of Padova**



# Poland

## IPF Scorecard

### General introduction and guidelines

The ATS/ERS/JRS/ALAT international IPF guidelines are implemented in Poland and Polish national guidelines are in development. However, there are no IPF care pathways. There is no official list of rare diseases.

**Key challenge:** There are no specialised ILD centres in Poland.

### Diagnosis and access to specialised care



There are no official ILD centres in Poland but respiratory physicians are available in hospital. Multidisciplinary teams are also not widely available and in some hospitals there are no ILD nurses who can support patients with the daily management of the disease.

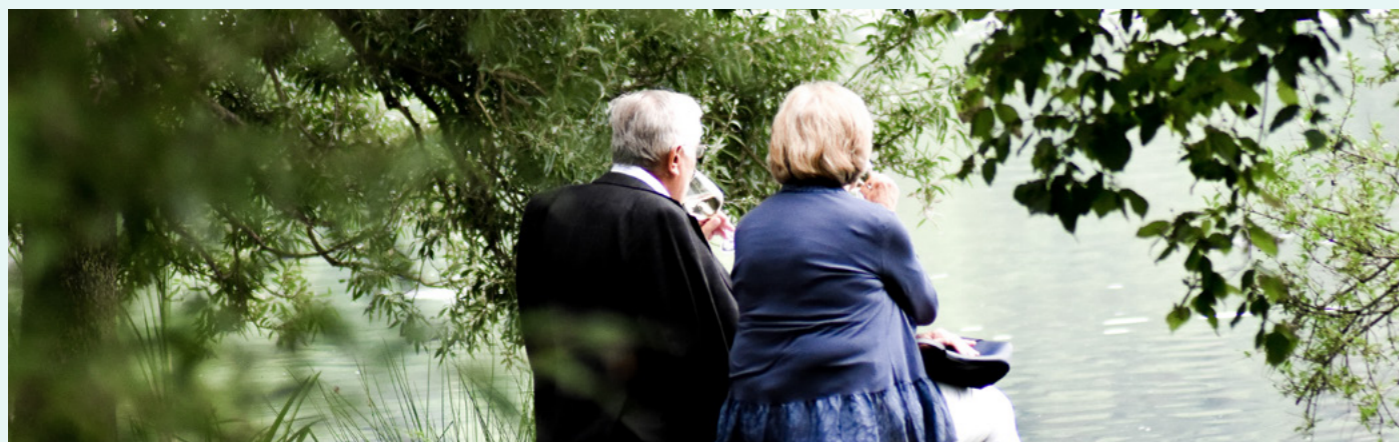
Upon diagnosis, information materials are available for patients, usually provided by the patient support groups

### Access to pharmacological treatment



**Key challenge:** Only some hospitals are allowed to prescribe the pharmacological treatments, and those who can have a set reimbursement capacity which can hinder access to treatment.

EMA-approved pharmacological treatments for IPF are available and reimbursed with no out-of-pocket costs to patients. However, in some of the biggest hospitals, where the number of patients exceeds the reimbursement capacity, there is a waiting list to get pharmacological treatment. The pharmacological treatments can be prescribed by respiratory physicians in hospitals that are authorised to prescribe them; the treatment is only provided through hospital pharmacies.



### Access to non-pharmacological treatment



**Key challenge:** Access to non-pharmacological treatments is not optimal in Poland. There are limitations and long waiting times to access oxygen therapy, pulmonary rehabilitation and lung transplantation.

Non-pharmacological treatment options are available to IPF patients in Poland but the costs associated with these treatments are not always fully reimbursed. Oxygen therapy is reimbursed when delivered in the hospital and at home (via home oxygen concentrators) but the waiting times can be very long; portable oxygen supply devices are not reimbursed. Pulmonary rehabilitation programmes are available only in hospitals and the cost is covered by the national healthcare system; however waiting times might be long. Pulmonary rehabilitation is also delivered privately but it is not reimbursed. There are very few transplantation centres in Poland which means that although lung transplantation is available to IPF patients, in reality waiting times are very long and access is limited. IPF patients in Poland have access to clinical trials.

### Access to palliative care and end-of-life care



**Key challenge:** There is no dedicated palliative care service targeted to IPF patients.

Palliative care is mainly delivered to oncology patients and it is reimbursed by the national healthcare system when it is performed in hospital and at home. Both private and public hospices exist.

In the overall patient journey, the respiratory physician and the local patient support groups are the ones providing relevant support to IPF patients.

### HCP statement

*“Although both antifibrotic drugs are available to Polish patients, resources and funding are still not sufficient to provide them right after diagnosis. The lack of officially-recognised specialised ILD centres is also an issue that affects timely diagnosis and access to treatment. Additional issues are insufficient oxygen supply and poor access to rehabilitation centres, given the long waiting lists. On a positive side, over the past few years both healthcare professionals and patients have acquired significant knowledge about the disease.”*

**Katarzyna Lewandowska,**  
**National Tuberculosis and Lung Diseases Research Institute Warsaw**



# Spain

## IPF Scorecard

### General introduction and guidelines

Spain follows the ATS/ERS/JRS/ALAT international IPF guidelines and has developed [national guidelines](#) for diagnosis and treatment of IPF, which have been updated in 2017. Promoting the harmonised implementation of IPF guidelines all over Spain remains a priority.

Spain has a "[National Strategy for Rare Diseases](#)"; but it does not include an implementation plan with dedicated budget. The central government has allocated some funding for the research and development of medicines for rare diseases.

**Key challenge:** Existing ILD units lack formal recognition as reference centres and are not available in all regions, creating inequalities between patients, and delays in referral and diagnosis of IPF.

### Diagnosis and access to specialised care



Not all Spanish regions have an ILD Unit with standardised procedures for IPF diagnosis and care. A peculiarity of the Spanish healthcare system is that the existing specialised ILD Units lack governmental accreditation (named CSUR), preventing them from participating as 'full members' in the European Rare Networks for rare diseases (such as ERN-LUNG).

This situation creates access inequity amongst Spanish patients. This [fragmented approach](#) also applies to referral procedures from primary care to specialised centres.

Raising awareness of IPF among radiologists and primary care physicians remains a priority in order to improve timely diagnosis and access to specialised care.

**Key challenge:** Inequality in access to pharmacological treatment amongst Spanish regions is a major challenge as different regional governments may allocate different budgets to rare diseases.

### Access to pharmacological treatment



The two EMA-approved treatments are available and fully reimbursed in Spain. As budget for orphan drugs is limited, the progression of the disease also plays a role when it comes to granting access to pharmacological treatment, thus creating delays and limiting access.

The pharmacological treatments can be prescribed by respiratory physicians and are only dispensed by hospital pharmacies.

### Access to non-pharmacological treatment



**Key challenge:** Access to pulmonary rehabilitation is too limited, geographically and in time.

When it comes to non-pharmacological treatments, [oxygen therapy](#), both outpatient and inpatient, is reimbursed, under the condition that it is prescribed by a respiratory physician.

Pulmonary rehabilitation is mainly offered to COPD (Chronic obstructive pulmonary disease) patients, and it is open to IPF patients only in a few centres. Even when available, pulmonary rehabilitation is fully reimbursed only for a period of 4 months.

The [Spanish transplant programme](#) is the best supported procedure in the Spanish healthcare system.

### Access to palliative care and end-of-life care



**Key challenge:** Lack of connection between ILD units and palliative care teams means that IPF patients have very unequal access to palliative and end-of-life support.

There are inequalities in access to [palliative and end-of-life care](#) across the Spanish regions, since there is no official circuit between ILD Units and palliative care teams. Access is granted on the basis of individual initiatives of some ILD Units.

The patient's right to die at home is granted at the initiative of individual centres'.

### Patient representative statement

*"In Spain it is necessary to improve early diagnosis of IPF. It is also necessary to create specific Multidisciplinary Centres that would allow an adequate follow-up and treatment to those suffering from the disease."*

**Carlos Lines Millán,**  
**Asociación de Familiares y Enfermos de Fibrosis Pulmonar Idiopática**



# The Netherlands IPF Scorecard

## General introduction and guidelines

In The Netherlands, there is no official list of rare diseases. Experts refer to the Orphan list of rare diseases, which includes IPF.

The ATS/ERS/JRS/ALAT international IPF guidelines are correctly implemented. The Dutch Pulmonologists Society has adopted a position paper that endorses most of the recommendations from the international guidelines. The document is not available in the public domain.

**Key challenge:** Late diagnosis has been reported as the main challenge. Raising awareness of IPF among general practitioners is considered to be one of the first steps towards timely diagnosis.

There are three officially acknowledged ILD/IPF specialised centres in The Netherlands: the Erasmus University Medical Centre in Rotterdam (ILD/IPF), the St. Antonius Ziekenhuis in Nieuwegein (ILD/IPF) and the Onze Lieve Vrouwen Gasthuis in Amsterdam (IPF). Furthermore, a network of treatment centres working in close collaboration with the abovementioned specialised centres exists. It aims to provide expert care for patients closer to their home. This network connects with the specialised centres through multidisciplinary team meetings taking place via teleconference.

In all these centres, IPF guidelines are properly implemented and the diagnostic procedure for IPF has also been reported to be standardised. In 2018 a [vision document](#) describing the current status of care and unmet needs of IPF patients was developed by the Erasmus Medical Center in Rotterdam with the support of Longfibrose Patiëntenvereniging, the Dutch IPF patient group. On the basis of this document, there are ongoing projects aimed at aligning IPF care pathways in The Netherlands.

Upon diagnosis, information materials provided by the local patient support groups and by the specialised centres are available to patients.

**Key challenge:** Late referral to specialised centres has been flagged as one of the main barriers to prompt access to pharmacological treatment.

EMA-approved pharmacological treatments for IPF are available and reimbursed with no out-of-pocket costs to patients.

The pharmacological treatments can be prescribed by the specialised centres and they are dispensed by the community pharmacies.

## Diagnosis and access to specialised care



## Access to pharmacological treatment



## Access to non-pharmacological treatment



**Key challenge:** Pulmonary rehabilitation programmes have been developed but are not always widely available across the country.

Non-pharmacological treatment options are available to IPF patients in The Netherlands and the costs associated with these treatments are reimbursed.

Oxygen therapy at home and in hospital is available to patients and is fully covered by the national healthcare system. Pulmonary rehabilitation programmes have been developed but are not always widely available across the country.

Lung transplantation is an option for patients, however the general health condition could represent a barrier to this procedure. IPF patients in The Netherlands have access to clinical trials.

## Access to palliative care and end-of-life care



**Key challenge:** The delivery of palliative care is not always optimal as there is a lack of awareness on the needs of IPF patients. Additionally, palliative care is too often confused with end-of-life care, without necessarily looking at optimising quality of life.

Palliative care is available and normally delivered either in the hospital or in local settings. However lack of awareness of palliative care among IPF patients, as well as confusion with end-of-life care means that it is not optimally organised and delivered.

In the overall patient journey, the respiratory physician, the ILD specialised nurse and the patient support groups are the ones providing support to the IPF patients and their families.

## Patient representative statement

*“Late diagnosis remain the main issue in the Netherlands. Increasing knowledge and improving understanding of the disease among the general population and primary care providers are two of the main goals of the Dutch IPF patient organization. This can facilitate early diagnosis and ultimately contribute to better access to pharmacological treatment.”*

*“Patient support groups play a key role in supporting the patients and their families after diagnosis; we believe this is very important for the everyday management of the disease and to improve the quality of life of patients living with IPF.”*

**Harry Van Den Haak, Longfibrose Patiëntenvereniging**





# United Kingdom IPF Scorecard

## General introduction and guidelines

The ATS/ERS/JRS/ALAT international IPF guidelines are available and the following national guidelines are also in place:

- The [National Institute For Health And Care Excellence \(NICE\)](#) clinical guidelines for diagnosis and management of IPF;
- [NICE quality standards](#) for IPF, which include chapters on diagnosis, the need for specialist nurses, oxygen therapy, pulmonary rehabilitation and palliative care;
- [National patient pathways](#) developed by NICE, specifically for IPF are also available and include a detailed flowchart on the patient care pathway, from diagnosis to management.

It is important to note that NICE guidelines were established in the UK to provide health service managers and commissioners of the NHS services guidelines for advice, prevention, diagnosis, treatment and longer-term management of diseases. The availability of guidelines is exemplary in the UK, however there is no current data on the follow-through of HCPs.

The National Health Service (NHS) recognises IPF as a rare disease requiring specialist commissioned healthcare services; however, the incidence and the prevalence of IPF is now increasing in the UK. IPF is also mentioned in [The British Thoracic Society Interstitial Lung Disease Registry Programme](#).

In addition, the United Kingdom has developed a strategy to for rare diseases as well as an [implementation plan](#) – both endorsed by the Department of Health.

**Key challenge:** IPF patients are not always being referred to specialised centres by general hospitals in a timely manner.

## Diagnosis and access to specialised care



The diagnostic process for IPF is standardised in the specialised centres, where there is access to a specialist multidisciplinary team. Currently, there are 23 specialist-prescribing centres in England, located in all major cities. However, in Wales, Scotland and Northern Ireland a slightly different structure exists where there are regional hospitals with ILD expertise. ILD specialist nurses are available to support and advise patients on the everyday management of the disease, representing a significant advantage for patients. Delayed referrals from primary to secondary or on to specialist ILD care are still problematic and lead to misdiagnosis, delays in diagnosis and access to treatment. Upon diagnosis, informative materials are available for patients; these are provided by clinical nurse specialists and patient support groups.

**Key challenge:** Patients with FVC>80% are currently excluded from treatment based upon NICE guidance giving rise to inequalities in access to pharmacological treatment in the UK.

## Access to pharmacological treatment



EMA-approved pharmacological treatments for IPF are available in the UK and NICE national guidelines are in place for pharmacological treatment:

- [NICE guidelines TA054: Pirfenidone for treating idiopathic pulmonary fibrosis](#);
- [NICE guidelines TA379: Nintedanib for treating idiopathic pulmonary fibrosis](#).

The pharmacological treatments are reimbursed with no out-of pockets costs for patients who are eligible to receive anti-fibrotic therapy. They can be prescribed by the specialised centres and can be delivered directly to the patient's home. Of note, in England, NICE limits access to the anti-fibrotic therapies. Patients whose FVC is outside of the treatment range are currently not eligible to receive NHS funded anti-fibrotic therapy.

## Access to non-pharmacological treatment



**Key challenge:** Pulmonary rehabilitation programmes are available but remain generic and not specifically adapted to the needs of IPF patients.

Non-pharmacological treatment options are available to IPF patients in the UK. Oxygen is delivered to the patient's home and pulmonary rehabilitation is predominantly provided in the patient's local hospital or at ambulatory level. However, pulmonary rehabilitation programmes are rarely specifically tailored to people with pulmonary fibrosis as they are generic programmes given to all patients with lung disease.

Non-pharmacological treatments are provided by the NHS, therefore there are no costs for patients other than travelling from home to hospital. Lung transplantation and clinical trials (which are usually run from the specialised centres) are also available to IPF patients meeting the designated criteria.

## Access to palliative care and end-of-life care



**Key challenge:** Patients may need to proactively ask for palliative and supportive care as it is not always embedded into the IPF treatment plan.

Palliative care is available to IPF patients at no additional cost. In the UK there are national NICE guidelines in place for palliative and end-of-life care ([Quality standard \[QS13\]](#)). Patients can choose to receive palliative care in their preferred place of care, supported by specialist palliative care nurses and physicians. However, timely and local access to specialist palliative care is variable across the country.

In the overall patient journey, the family, respiratory physician, specialist nurses and patient groups are the primary support system for IPF patients and their families.

## HCP statement

*“There is a great need for standardised patient pathways to enable earlier diagnosis and timely referral to specialised ILD centres. The rate that IPF progresses is very variable and unpredictable. There are no biomarkers that are currently able to identify those people at risk of more rapidly progressive disease and who may benefit from earlier treatment interventions. It is hoped that an improved understanding of the genetics and disease pathogenesis will lead to a precision medicine approach whereby disease modifying treatments are tailored to an individual patient according to their specific genes and molecular signature.”*

**Helen Parfrey, Papworth Hospital**

# Acknowledgements

The EU-IPFF would like to thank all the patients, carers and healthcare professionals who contributed to this Report by sharing their professional and personal experiences. In particular, we would like to thank the following patient organisations:

**LOT Austria (AT)**

**Lungenfibrose Forum Austria (AT)**

**Association Belge contre la Fibrose Pulmonaire Idiopathique (BE)**

**Belgische vereniging voor longfibrose (BE)**

**IPF Bulgaria (BG)**

**Association Pierre Enjalran Fibrose Pulmonaire Idiopathique (FR)**

**Lungenfibrose e.V. (DE)**

**Hellenic Pulmonary Fibrosis Association (GR)**

**Magyar Tüdőfibrozis Egyesület (HU)**

**Irish Lung Fibrosis Association (IE)**

**AMA Fuori dal Buio (IT)**

**FIMARP (IT)**

**Longfibrose Patiëntenvereniging (NL)**

**IPF Polish Society (PL)**

**Asociación de Familiares y Enfermos de Fibrosis Pulmonar Idiopática (ES)**

**Action for Pulmonary Fibrosis (UK)**

**British Lung Foundation (UK)**

**Pulmonary Fibrosis Trust (UK)**

The EU-IPFF also wants to thank all its members and the members of the Scientific Advisory Board for their help in contributing to this Benchmarking Report and for their invaluable comments on earlier drafts.

The EU-IPFF is also grateful to Boehringer Ingelheim International GmbH and F. Hoffmann-La Roche LTD who have financially supported the development of this Benchmarking Report.

# Limitations

## For the purpose of this Benchmarking Report:

- No differentiation was made between the severity of IPF (moderate, mild and severe cases);
- The sample of the study represents an inherent bias since it was limited to the EU-IPFF member countries (Austria, Belgium, Bulgaria, France, Germany, Greece, Ireland, Italy, The Netherlands, Poland, Spain, United Kingdom plus Hungary).
- Perceptions from patient groups located in countries not represented by the EU-IPFF are not covered by this Report;
- The responses provided by patient groups are appropriate to the best of their knowledge, however they might be stemming from personal experiences or from experiences of other patients in each association;
- Some questions that were asked during the interviews have not been included in this short Report as not enough information on certain indicators was provided by the interviewees or responses varied too broadly and did not allow for comparison;
- Some patient representatives participated in phone interviews; for health-related reasons or for language-related reasons, some patient representatives filled in a written questionnaire instead;
- The replies were validated by the members of the EU-IPFF Scientific Advisory Board; however, the majority of them works in specialised ILD centres, which could affect their perception of IPF care in their country;
- The answers to the phone interviews were codified by interviewers; an interview guide was developed to minimise the personal bias but cannot fully eliminate it;
- When possible, the interviews were conducted in the native language of the respondent and then translated into English;
- The same colour code used to assess different countries does not necessarily mirror the same level of progression.
- A third party supplier translated the IPF national scorecard into the various national languages. Therefore, there may be slight differences from the original report in English.
- Each member organisation had the opportunity to edit their national scorecard, which may result in differences from the scorecards in English.



The European Idiopathic Pulmonary Fibrosis and Related Disorders Federation (EU-IPFF) brings together eighteen European national patient associations from thirteen European countries. Its mission is to serve as the trusted resource for the IPF community by raising awareness, providing disease education, advancing care, and funding research. EU-IPFF collaborates with physicians, medical organisations, people with IPF, caregivers and policy-makers throughout Europe.

For further information, please visit [www.eu-ipff.org](http://www.eu-ipff.org).