

# Life with IPF

## Newly diagnosed with IPF

If you are newly diagnosed with IPF you probably have numerous thoughts and questions running through your head. Now is the time to look for information and guidance from your doctor and treatment team and for support from family and friends to help you on your journey with IPF:

- Find out as much as possible about the disease and your therapy options from your doctor and treatment team. Have a look at our [treatment](#) pages to see what options you might have.
- Equally as important, seek out support from friends, family and patient groups. Although IPF is a relatively rare disease, there are many societies and foundations for patients with IPF. Have a look at our [resources](#) page to find the patient association that's right for you.
- Think about which changes that you can make to your lifestyle and surroundings to reduce symptoms and improve your quality of life. Some suggestions are in the table below:

### Lifestyle Factors

There are a number of different changes that you can make to your lifestyle that may help reduce your symptoms and improve your quality of life:

- If you still smoke, there is no better time than now to quit. Although this may be easier said than done, your doctor will be able to guide you through the process.
- Get active and stay active – both physically and mentally. As IPF progresses, physical activity will become more and more difficult. It is important to do what you can and rest when needed. Staying mentally sharp will help for the later phases.
- Like many illnesses, for patients with IPF it is important to maintain a healthy diet. Diets low in sodium, fat (particularly saturated fat), and added sugar, but high in whole grains and protein are beneficial for patients with IPF.
- In addition to being active, it is also important to get your rest, relax and avoid stress. Healthy sleep patterns and relaxation techniques are beneficial. As the disease progresses, everyday activities will become more exhausting. Take your time and take a break when you need it. Listen to your body.
- You should be cautious travelling to high-altitude destinations because the low oxygen levels in the air may cause increased breathlessness.

## Your Surroundings

In addition to lifestyle changes, you may find it helpful to look for ways to make your day-to-day life easier on your lungs by making a few adjustments to your home/surroundings:

Take steps to improve indoor air quality and limit irritation to your lungs by:

- Changing or installing air filters
- Limiting the use of woodstoves and fireplaces
- Removing dust regularly and promptly
- Limiting the use of harmful cleaning products which leave fumes lingering in the air
- Hindering mycobacterial growth, these are particularly found in bathrooms/showers
- Limiting pet dander, especially in the bedroom
- Avoiding aerosols
- Avoiding using toiletries with strong fragrances
- Avoiding fine, airborne powders such as flour and wood dust

## Living with IPF

If you are living with IPF you know that daily activities that once seemed trivial are now a challenge. However there are ways to lessen the burden of IPF on your daily life.

Although there is no cure for IPF, treatment does have the possibility to relieve symptoms and slow the progression of scarring (fibrosis). Some patients with IPF benefit from non-pharmacological treatments such as oxygen therapy and pulmonary rehabilitation. For selected patients, a lung transplant may be an option. See our [treatment](#) pages for a more detailed overview of the treatment options currently available.

After being diagnosed with IPF, it is important to maintain your overall health and avoid concomitant illnesses. Talk to your doctor about ways to avoid catching seasonal colds/flu and how you can reduce your risk once those bugs start going around. Keep yourself up-to-date on recommended vaccinations and immunisations.

## Travelling with IPF

Whether you are planning to visit a familiar place or thinking of exploring a new destination, you may be concerned about how your new or progressing condition will affect your travels. IPF shouldn't automatically cut your holiday hopes short. Talk to your specialist or other health care providers about your plans – he or she may be able to help you realise them. Here you will find some helpful information to get the conversation started, as well as a general guide of what to expect in the planning process and travelling phase.

# Choosing a destination

When choosing a destination, it is important to keep the following in mind:

## Climate



## Air Quality



## Terrain



## Elevation



## Mode of Transportation



### Climate

Although there aren't any guidelines or recommendations concerning climate- and weather-related conditions for patients with IPF, you should consider what type of climate or weather patterns you are comfortable in before choosing where to go and when. Depending on your individual level of sensitivity, humidity, or lack thereof, and extreme temperatures could cause discomfort.

### Air Quality

Air quality is an important factor for everyone, however, for those with respiratory conditions, air quality plays an even greater role in their health and well-being. A recent study has shown an association between high levels of two common air quality indicators, ozone (O<sub>3</sub>) and nitrogen dioxide (NO<sub>2</sub>), and acute exacerbations in patients with IPF.<sup>1</sup> Although the study looked at an exposure period of six weeks, looking into local air quality levels is worth considering for anyone with a respiratory illness. The World Health Organisation provides data on ambient air pollution for major cities around the world. In addition, many areas provide data on their air conditions and pollution levels on regional websites.

Visit the WHO's Global Health Observatory page to view world-wide interactive pollution maps.

### Terrain

Considering the terrain of your destination is not limited to outdoor, nature activities. Cities can also be hilly and have varying degrees of walkability and wheelchair-friendliness. It might be worth looking into the overall terrain of the place you are planning to visit whether it is a nature-focused holiday or a city trip. You should also consider what sites you plan visit and look into whether they offer options for tourists with limited mobility such as lifts/escalators versus stairs and the availability and accessibility of public transport.

### Elevation

Travelling to high elevations may require patients with IPF to use supplemental oxygen or to increase their oxygen flow if already on oxygen therapy at or near sea level. Depending on the severity of your condition, your doctor may do a pre-travel evaluation to determine whether you need oxygen therapy at your destination or if monitoring your oxygen levels and physical status once you arrive to your high-altitude destination is sufficient.<sup>2</sup>

## Mode of Transportation

Choosing how you will get to your destination is an important part of the planning process. If you choose to fly, there may be more planning involved and some arrangements will need to be made far in advance. Read more about airplane requirements concerning in-flight oxygen here. Depending on your mobility, you may also need to make special arrangements when travelling by train or bus.

## Medications – Taking medications when you travel

Observing the points below will help you with the logistics of transporting medications as well as to aid the efficacy of your medications while travelling.

**Packaging:** Medications should be stored and transported in their original containers and be accompanied by the original labelling/information leaflets – This is important for security checks as well as to inform medical personnel what medications you are currently on should you need medical assistance.

**Prescriptions:** For prescription drugs it is indispensable to check if you have enough medication for your stay before starting your travel. Keep in mind that it could be necessary to get prescription refills at any point during your time away from home. For this reason, keep a copy of the prescription with you – this is also important for security checks.

**Timing:** Remember, medications are usually taken based on lapsed time and not necessarily administered based on the specific time of day. Travellers who take medication according to a strict timetable should consider the time difference between their destination and home. When travelling across time zones, it is important to seek medical advice and consult your physician prior to departure. Bring your flight schedule and information on time zone changes and your doctor can help you plan a proper medication adjustment.<sup>3</sup>

Mode of Transportation specific precautions:

**When flying** – Keep medications and prescriptions in your carry-on for two reasons: luggage can be lost and it is also possible that the temperature in the luggage carrier is too extreme for your medication.

**Travelling by bus** – You may want to consider keeping temperature-sensitive medications in the cabin with you as temperatures in the cargo hold of tour buses may vary depending on the climate/weather along your route and at the destination.

**By car** – Don't leave temperature-sensitive medications in the car for long periods of time since the interior temperature in cars can vary greatly – become very hot when left standing in direct sunlight or become very cold when left outside in certain climates.

# Flying – Do I need in-flight oxygen?

Aircraft cabins are not pressurised to sea level, but rather to altitudes of up to approximately 2,450 meters (8,000 ft). If you need oxygen therapy for your daily activities, you will need your oxygen in-flight as well, and may need to increase the flow during the flight. If you are not currently on oxygen therapy, it's a good idea to have your doctor assess whether you will need in-flight oxygen if you are planning to fly. Read more about airplane requirements concerning in-flight oxygen here.

## Fit-to-fly Assessments

If you have an underlying condition, some airlines require you to provide a statement from your doctor indicating you are "fit-to-fly". In addition to the information in your medical record and your shared travel plans, your doctor may perform one of the following assessments before issuing a letter to the airline.\*

\*Please note, regardless of your performance on the following assessment(s), medical history and the certification from your doctor, airlines have the right to deny you from flying if the captain or airline medical department/adviser feels your condition poses a threat to the aircraft, other passengers, or crew, or requires special medical attention during the flight, or if your condition may be worsened by the flight.

### Walking Test

One of the simplest assessments doctors use as an indicator for one's fitness to travel is known as the "walking test". The patient is asked to walk 50 meters (or yards), or climb one flight of stairs. If he or she is unable to complete the task due to breathlessness or any other respiratory symptoms, the doctor will probably recommend the use of in-flight oxygen.<sup>4</sup>

### Hypoxic Challenge Test (HCT) or Hypoxia Altitude Simulation Test (HAST)

The hypoxic challenge test (HCT), also known as the hypoxia altitude simulation test (HAST), is a more involved assessment of one's ability to cope with decreased oxygen. As the name indicates, the oxygen levels at high altitude (approximately 8000 ft, or 2438 m) are simulated and the patient's physiological response is recorded.

Before the test, the patient's partial pressure of arterial oxygen ( $\text{PaO}_2$ ) is measured. During the test, the patient breathes 15.1% oxygen, balanced with nitrogen, for 20 minutes. Then the patient's  $\text{PaO}_2$  is measured again. If the  $\text{PaO}_2$  has fallen below a certain level (usually below 50 mmHg), the respiratory specialist will recommend in-flight oxygen.<sup>5</sup>

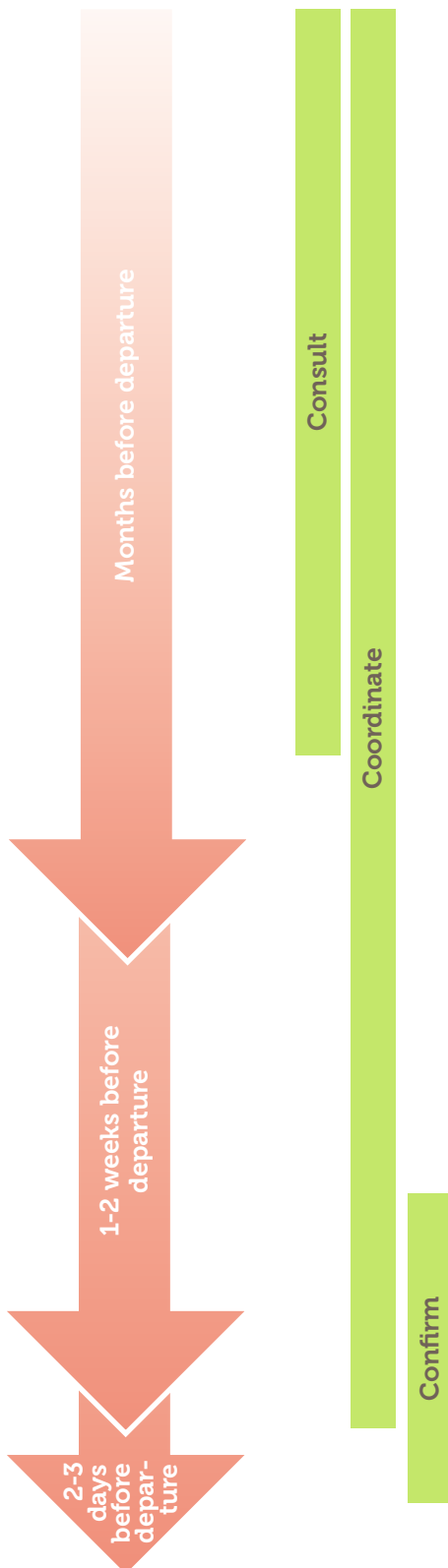
## Flying with oxygen

If your doctor recommends the use of in-flight oxygen, there are few organisational matters you need to take care of before your flight – some of which need to be done months in advance and even before booking airline tickets.

Each airline has their own specific rules and policies regarding in-flight oxygen supply. The European Lung Foundation has a nicely organised "Airline Index" which lists all major airlines and their oxygen policies as well as contact information.

Some airlines allow, or even require, passengers to provide their own in-flight oxygen supply. Check with the Federal Aviation Administration for a list of approved portable oxygen concentrators.

## Checklist for plane travel



### Before planning your trip...

- Talk to your doctor about your travel plans and any concerns you may have
- If you are not currently on oxygen therapy, your doctor will need to determine whether you are candidate for in-flight oxygen

### Before booking your airline tickets...

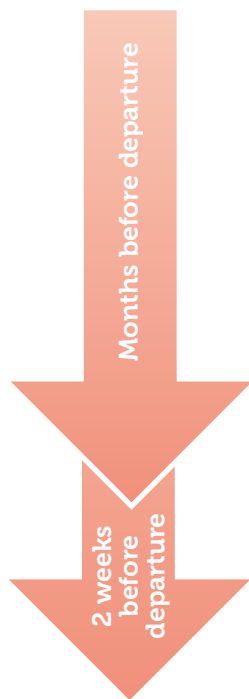
If you need in-flight oxygen, check the airline's oxygen policy and "fit-to-fly" requirements

- Does the airline supply in-flight oxygen, or are you required/allowed to bring your own portable oxygen supply?
- What type of oxygen delivery system does the airline offer on board?
- Available flow rate settings?
- Mask or nasal cannula?
- What are the costs?
- Information regarding battery supply/in-flight electrical supply
- What are the policies regarding use of oxygen during take-off and landing?
- Complete and submit the airline's MEDIF form.<sup>6</sup>
  - One portion will be completed by you, the other must be completed by your doctor
  - Only after the airline reviews the completed form and determines you are eligible to fly, the ticket reservation will be finalised
- Contact the airport(s) you will be using to arrange for assistance to/from the gate
- Check with your insurance provider about your coverage while travelling – additional traveller's insurance may be beneficial or necessary

### Before enjoying your get-away...

- Make sure you have an adequate supply of all prescription medicines as well as new prescriptions for refills. Please note that you need to check the local situation since not every medication is available in every country.
- Contact the airline to confirm all necessary precautions and special arrangements will be realised on your flight.
- Contact the airport to confirm the status of your planned assistance.

## Checklist for car, bus or train travel



### Before planning your trip...

- Talk to your doctor about your travel plans and any concerns you may have regarding
  - Climate
  - Air quality
  - Terrain
  - Altitudes
  - Mode of transportation

### Before booking your journey...

- Check with your insurance provider about your coverage while travelling – additional traveller's insurance may be beneficial or necessary

### Before enjoying your get-away...

- Make sure you have an adequate supply of all prescription medicines as well as new prescriptions for refills. Please note, that not all medication is available in all countries around the world. Check out before!

## Vaccinations

As vaccination requirements and recommendations vary depending on the travel destination, it is important to talk to your pulmonary specialist about the recommended vaccines for your particular travel itinerary. He or she may even want to consult with a travel medicine specialist to gain further insight.

Regardless of destination, it is recommended that patients with IPF stay up to date with yearly influenza and anti-pneumococcal vaccines.<sup>7-9</sup>

## Precautions

### General information on travelling with a serious, lifestyle-limiting disease

#### Doctor's notes & other certifications needed for travel

You should obtain a note from your doctor if you are planning to travel abroad and have or require the following items.<sup>10</sup>

- Any special medications, such as controlled substances or investigational drugs; this might also be worthwhile for your IPF medication
- Needles, syringes
- Metal implants
- Large quantities (>100 mL) of liquids (generally, this only applies if you are planning to fly)

For air travel, you may also be required to provide a note from your doctor stating you are "fit to fly". See the section on "Fit-to-Fly Assessments" above for more information.

#### Insurance

Before you go on holiday, check with your insurance company to verify what medical treatment/procedures are covered while travelling away from home. Be sure to ask<sup>11</sup>:

- If there are any specific activities that are not covered.
- What documentation is required for reimbursement when receiving care away from home/out of network?
- If pre-authorisation or second opinions are required for any medical procedures. If so, which ones?
- If repatriation is covered by your insurance.

Based on your current medical coverage, you may want to consider travel health insurance.

Even if your insurance covers out-of-country/out-of-network medical care, you may be required to pay for the treatment upfront and file for reimbursement once you return home. Be sure to get copies of all bills and receipts, and if possible records of the procedures performed and be familiar with your insurance plan's reimbursement policy.

In addition to medical coverage, it might be worth considering purchasing general travel insurance which will minimise your financial losses if you are suddenly unable to travel due to health reasons.



## Choosing peace

IPF patients experiencing discomfort may be given palliative care, which is designed to relieve pain and improve the patient's quality of life, through physical, psychological, social and spiritual approaches. At the later stages of the disease, you may receive hospice care, which allows dying patients peace and comfort and also offers support for families.

Patients with a chronic and terminal illness like IPF may be subject to a wide variety of emotions that may seem to cycle or come in waves until an equilibrium is reached. Psychologists have proposed that the modern grieving process, which patients with a terminal illness and their families face together, may have 5 linear steps:

Crisis	In the beginning, the diagnosis is presented, the shock takes over and the daily lives of the patient and the patient's family are turned upside down.
Unity	The family comes together to provide support for the patient: managing treatment and other support strategies, and helping organise insurance and legal matters.
Upheaval	The ongoing treatment and life with IPF is a trying time for the whole family. Even though patience may at times wear thin, it is important for everyone to calmly and constructively voice their concerns and feelings with one another.
Resolution	As health deteriorates and the patient and family face saying goodbye to one another, the time comes for gathering thoughts, resolving issues, and supporting one another.
Renewal	A long process which begins at the funeral of the patient in which the family slowly but surely adjust to their great loss.

## References

1. Johansson KA, et al. Acute exacerbation of idiopathic pulmonary fibrosis associated with air pollution exposure. *Eur Respir J* 2014;43:1124–1131.
2. Luks AM. Do lung disease patients need supplemental oxygen at high altitude? *High Alt Med Biol* 2009;10:321–327.
3. WHO I World Health Organization, Chapter 2: Mode of travel: health considerations. Available at: <http://www.who.int/ith/ITH2010chapter2.pdf>.
4. Shand D. The assessment of fitness to travel. *Occup Med Oxf Engl* 2000;50:566–571.
5. Mohr LC. Hypoxia during air travel in adults with pulmonary disease. *Am J Med Sci* 2008;335:71–79.
6. IATA I International Air Transport Association - Medical Manual. Available at: <http://www.iata.org/whatwedo/safety/health/Documents/medical-manual-2013.pdf>.
7. Cottin V, et al. Diagnosis and management of idiopathic pulmonary fibrosis: French practical guidelines. *Eur Respir Rev Off J Eur Respir Soc* 2014;23:193–214.
8. NHS Choices: Pulmonary fibrosis (idiopathic) - Treatment. Available at: <http://www.nhs.uk/Conditions/pulmonary-fibrosis/Pages/Treatment.aspx>. Accessed February 13, 2015.
9. Medscape: Idiopathic Pulmonary Fibrosis Treatment & Management. Available at: <http://emedicine.medscape.com/article/301226-treatment>. Accessed February 13, 2015.
10. Ahmedzai S, et al. Managing passengers with stable respiratory disease planning air travel: British Thoracic Society recommendations. *Thorax* 2011;66 Suppl 1:i1–30.
11. Travel Insurance, Travel Health Insurance, & Medical Evacuation Insurance - Chapter 2 - 2014 Yellow Book | Travelers' Health | CDC. Available at: <http://wwwnc.cdc.gov/travel/yellowbook/2014/chapter-2-the-pre-travel-consultation/travel-insurance-travel-health-insurance-and-medical-evacuation-insurance>. Accessed February 13, 2015.