Canadian Pulmonary Fibrosis Foundation

Toward Exceptional Care:

A Canadian IPF Patient Charter

WHAT IS IDIOPATHIC PULMONARY FIBROSIS?

Idiopathic Pulmonary Fibrosis (IPF) is an irreversible lung disease that causes scarring in the lungs. The reason for the scarring is unknown. The scarring progressively gets worse, making it harder to breathe. Due to the progressive scarring of the lungs over time, the brain and other important organs do not receive enough oxygen, which ultimately leads to death. This is respiratory failure and is the leading cause of death in IPF.

Current IPF treatments include:

1. Medicines that can slow down disease progression
2. Pulmonary rehabilitation
3. Lung transplant

IPF has no cure, and diagnosis of IPF can take as long as 2 years. For many IPF patients, life expectancy without a lung transplant averages 2-5 years from the date of diagnosis, although some patients have lived for up to 20 years.

WHAT IS A PATIENT CHARTER?

A Patient Charter is a document which sets out a number of patient rights in an effort to create a national standard of care. A Charter makes recommendations to provincial governments, institutions and health care organizations to improve patients' overall quality of life, while simultaneously supporting efforts for targeted research to develop better long-term treatments, and to ultimately find a cure.

WHY IS A CHARTER IMPORTANT FOR IPF PATIENTS IN CANADA?

The Canadian Idiopathic Pulmonary Fibrosis Patient Charter is a way to call upon policymakers, health care providers and private payers to recognize IPF as a rare, rapidly lethal, chronic disease and to join in the campaign for greater awareness of this illness. The ultimate goal is to improve the overall quality and standards of care and ensure equal access to a continuum of supports for all Canadians affected by IPF.

The Charter was developed by the Canadian Pulmonary Fibrosis Foundation (CPFF) in close consultation with a national network of patients, caregivers, clinicians and health system leaders, many of whom came together in the spring of 2016 at a policy roundtable organized by the Institute of Health Economics (IHE).

For more information on the proceedings of the policy roundtable, please visit:

http://www.ihe.ca/research-programs/knowledge-transfer-dissemination/roundtables/ipfnf/ipfnf-docs

IDIOPATHIC PULMONARY FIBROSIS (IPF)

RARE, UNPREDICTABLE AND DIFFICULT TO DIAGNOSE

COULD THIS BE YOU?

WHAT IS IPF?

- **I** **IDIOPATHIC** REFERS TO AN UNKNOWN CAUSE
- **P** **PULMONARY** AFFECTS THE LUNGS
- **F** **FIBROSIS** MEANS SCARRING

MORE COMMON **IN MEN**

MOST PATIENTS ARE **OVER 50 YEARS** OF AGE

WHAT ARE THE SIGNS AND SYMPTOMS OF IPF?

COMMON SIGNS AND SYMPTOMS

- SHORTNESS OF BREATH "DYSPNEA"
- CHRONIC DRY COUGH
- A "CRACKLING" SOUND TO BREATHING (HEARD WITH STETHOSCOPE)
- FINGER CLUBBING
- BLUE TINGE TO LIPS, NAIL BEDS AND SKIN "CYANOSIS"

DIAGNOSIS OF IPF CAN BE CHALLENGING

ABOUT **50%** ARE INITIALLY MISDIAGNOSED

IPF SYMPTOMS ARE OFTEN SIMILAR TO OTHER DISEASES

	ASTHMA	COPD	HEART FAILURE	IPF
SHORTNESS OF BREATH (DYSPNEA)	○	○	○	●
CHEST TIGHTNESS	○			●
COUGH	○	○	○	●
FATIGUE			○	●

Diagnosis → Symptoms

AVERAGE TIME TO DIAGNOSIS IS **1 TO 2 YEARS** AFTER ONSET OF SYMPTOMS

COPD=chronic obstructive pulmonary disease

NORMAL LUNG VS. IPF LUNG

SCARRING/HONEYCOMBING IN THE LUNG RESTRICTS BREATHING AND OXYGEN EXCHANGE

IPF CAN BE TREATED

Medications are available that help slow IPF progression and preserve lung function for longer

WHAT ARE DOCTORS' GOALS FOR IPF TREATMENT?*

- Slowing disease progression
- Improving symptoms
- Sustaining quality of life

* Source: "Think of Everything Global Pulmonologist Survey", 2015.

PROGRESS IN TREATMENT IS BRINGING NEW HOPE TO IPF

QUESTIONS ABOUT IPF AND TREATMENTS? ASK YOUR DOCTOR TODAY!

CANADIAN PULMONARY FIBROSIS FOUNDATION

CANADIAN IPF PATIENT CHARTER

People with IPF and their families have the right to:

1. Equal levels of care across Canada based on the best standard of care currently available
2. Timely and accurate diagnosis and care, involving a skilled specialist and a multi-disciplinary care team
3. Seamless, well-integrated health and social care services, including timely access to ambulatory and personalized home oxygen services and appropriate IPF medications
4. High quality and accurate information about IPF, including full details of all treatments, clinical trials, support, service provisions and transplant options available to them
5. Improved, more accessible, compassionate palliative and end-of-life care when appropriate
6. Dedicated in-person or remote peer support networks for patients and their caregivers
7. Increased funding for IPF research to a level that reflects the considerable and growing impact of the disease in Canada
8. Effective and expanded clinical networks to ensure a coordinated approach to IPF management and seamless care between providers at all stages of the patient pathway
9. Swift access to specialist care and IPF tailored pulmonary rehabilitation programs, appropriate prioritization on transplant waiting lists, and prompt social care assessments and response for patients with exacerbations and rapid disease progression
10. Campaigns to improve public awareness and primary care recognition of the condition and its symptoms and encourage employers and service providers to better meet the needs of people with IPF

Canadian Pulmonary Fibrosis Foundation

Equal levels of care across Canada based on the best standard of care currently available.

At present, care and treatment for IPF patients is not equally available across Canada. Canadians in Vancouver, Calgary, Edmonton, Toronto, Hamilton, London, Kingston, Ottawa and Montreal have access to comprehensive Interstitial Lung Disease (ILD) units. However, patients living in Saskatchewan, Manitoba and Atlantic Canada – and in rural and remote areas of some provinces – do not have the same access to this specialized care. Moreover, not all respirologists have the same knowledge, experience and training in the treatment of IPF, which varies the level of treatment available to patients across the country. Creating a national standard of care will help ensure IPF sufferers are presented with the best possible treatment options at every stage of their disease.

Timely and accurate diagnosis and care, involving a skilled specialist and a multi-disciplinary care team.

Diagnosis of IPF can take up to 2 years, after which IPF patients need specialized care from various health care professionals. Standardizing protocols and diagnostic methods across Canada can facilitate faster, more accurate diagnosis of patients. As well, including IPF training in health care professionals' curricula and accreditation systems can help general practitioners and family physicians become more familiar with early signs and symptoms of IPF, allowing for faster referrals to multidisciplinary teams best suited to offer dedicated and personalized treatment to IPF patients.

Seamless, well-integrated health and social care services, including timely access to ambulatory and personalized home oxygen services and appropriate IPF medications.

In addition to highly trained physicians, many patients benefit from the involvement of specialized nurses, social workers, physiotherapists, and occupational therapists. Promoting and financing the development of these specialist multidisciplinary teams throughout every province and territory in Canada can ensure patients get the help they need to improve their overall quality of life.

Oxygen is essential for life. Current coverage and eligibility for ambulatory oxygen varies across the country, with some provinces requiring ongoing retests of oxygen saturation levels in order to continue treatment. Every IPF patient deserves equal opportunity to access ambulatory oxygen with a specialized respiratory team, which is crucial for patients' health and well-being and can ultimately enhance mobility, increase a patient's ability to exercise, and allow patients to continue their regular daily activities for as long as possible.

IPF has no cure, but some medications can slow down the rate of disease progression. In order to have nationwide, timely access to all IPF medications once Health Canada has approved a medication as safe and effective, we ask each Province and Territory to act swiftly to provide coverage.

High quality and accurate information about IPF, including full details of all treatments, clinical trials, support, service provisions and transplant options available to them.

IPF patients are at risk for rapid deterioration, which makes it vital to develop a national online information hub for patients and their families to have immediate access to various treatment options and services available to help manage the disease. The development of strategies and activities to empower patients by improving health literacy and the use of digital tools in health decision-making at both the provincial and pan-Canadian levels can aid in increasing patient choice.

Improved, more accessible, compassionate palliative and end-of-life care when appropriate.

IPF is a chronic, progressive disease that kills 3,500 Canadians every year. All people with IPF would benefit from improved and more accessible palliative care services. Because of this, Canadians should have equal access to palliative care services, including access to hospice nurses, home care and comfort measures for both symptom management and end-of-life care. Physicians should be aware of clear markers of disease progression to prompt a palliative referral at the appropriate time. Everyone in the specialized care team should acknowledge the psychological and emotional impact of IPF. The team should arrange palliative care using local services as much as possible to decrease stress for both patients and their caregivers.

Dedicated in-person or remote peer support networks for patients and their caregivers.

IPF has a heavy physical, psychological, social, and financial burden and it is important to develop a clear understanding of the disease's progression upon diagnosis. Anxiety is often associated with the disease as patients and their caregivers undergo enormous changes in their health that comes from living with IPF. Accessible support groups in the form of one-on-one or group meetings available in-person, by phone or on-line can help patients and their caregivers cope with the many stressors associated with the disease. Providing funding for caregivers is also important because more and more support will be needed over time to help IPF patients throughout their treatment and to perform daily activities. Keeping IPF patients living comfortably at home helps patients, their caregivers and the health systems charged with their care.

Increased funding for IPF research to a level that reflects the considerable and growing impact of the disease in Canada.

IPF has a death rate of 100 per cent, which is much higher than almost any other major disease. Yet, funding for research into the causes and cure of IPF continues to lag. Funding for IPF research needs more governmental support to help develop treatments and increase the level of care available for IPF sufferers. Governments across Canada should offer financial incentives to support promising research and help establish and maintain provincial IPF registries to aggregate national data to closely monitor the prevalence, detection and treatment outcomes of this fatal disease.

Effective and expanded clinical networks to ensure a coordinated approach to IPF management and seamless care between providers at all stages of the patient pathway.

As IPF progresses, patients require increased support from various care providers. A more integrated approach between health care providers is needed to ensure patients continue to receive the best and most appropriate care throughout their treatment. Leadership is required from health ministries, along with ILD units and local respirologists to ensure shared communication at all stages of the patient pathway. This should include the use of more eHealth technologies, such as web consultations with specialist care providers so patients can be treated locally in remote areas.

Swift access to specialist care and IPF tailored pulmonary rehabilitation programs, appropriate prioritization on transplant waiting lists, and prompt social care assessments and response for patients with exacerbations and rapid disease progression.

Being placed on waiting lists can be fatal for many IPF patients. Too often patients are placed in a queue for specialist appointments without prioritization based on their deteriorating condition. Triage nurses and clinic appointment personnel should be trained to recognize the clinical markers of risk in IPF patients with worsening symptoms. Caregivers should also be trained to recognize the symptoms of declining lung function to initiate appropriate steps for the patient to receive prompt care, including admission into a transplant unit.

Campaigns to improve public and primary care recognition of the condition and its symptoms and encourages employers and service providers to better meet the needs of people with IPF.

The rarity of IPF and lack of awareness campaigns has resulted in limited knowledge of the disease in the public and private sectors across Canada. Because of this, many employers and service providers fail to receive material on how to better serve and meet the needs of people with IPF. For example, many IPF patients have to travel with oxygen, have limited mobility, or experience increased symptoms with physical exertion. Because of this, people with IPF would benefit from exceptions from service providers for mandatory in-person appointments they may be unable to attend due to complications of their illness. An increased understanding of employers and service providers on the conditions that people with IPF live with can improve the overall quality of services received.

About The Canadian Pulmonary Fibrosis Foundation

The Canadian Pulmonary Fibrosis Foundation was created in 2009 to raise money to find causes and treatments for Pulmonary Fibrosis (PF), provide education and support for people affected by PF and to help answer non-medical questions frequently asked by those suffering with the disease. Our vision is to make lung transplants the **last** resort not the **only** resort for people with pulmonary fibrosis.

The CPFF is a not-for-profit charitable organization and a registered Canadian Charitable Foundation. Our mission is to raise funds to finance research to better understand and develop treatments to eventually find a cure for pulmonary fibrosis. From its creation in 2009 to September 2016 the CPFF has provided over $800,000 in research grants to leading institutions and researchers in Canada, including the University Health Network, University of Alberta's Lung Health Centre, St. Joseph's Hospital Foundation, Toronto General & Western Hospital Foundation and Vancouver General Hospital.

CPFF's key objectives in the battle against pulmonary fibrosis are to:

- Raise public awareness about this fatal disease
- Offer support to those affected by pulmonary fibrosis
- Serve as "the patient voice" for PF by representing Canadians affected by PF to government, health care professionals, media and the public
- Fund Canadian research into causes and treatments of PF

How you can join the fight against IPF

- Contact your local MPP, MLA or MNA to help raise awareness about IPF and the need for a national standard of care
- Donate to help find a cure at www.CPFF.ca
- Find more information and join the conversation about IPF at:

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