Understanding pulmonary fibrosis

- Pulmonary fibrosis (PF) is a condition in which the tissue inside and between the air sacs in the lungs becomes scarred, thickened, and stiff.
- The scarring is called fibrosis.
- The scarring and stiffness make it more difficult for oxygen to pass through the walls of the air sacs into the bloodstream.
- The scarring makes it hard for a person to breathe.
- Vital organs, such as the heart and brain, may not get enough oxygen to work properly.

Causes of pulmonary fibrosis

PF can result from a number of causes including:

- Genetics
  - Familial pulmonary fibrosis (FPF)
  - Hermansky-Pudlak Syndrome (HPS)
- Exposure
  - Job-related and environmental toxins and pollutants
  - Radiation treatments for cancer
  - Certain medications
  - Viral and bacterial infections
- An immunologic disorder
  - Lupus, rheumatoid arthritis, sarcoidosis, and scleroderma, etc.
- No known cause.
  - PF with no known cause is called idiopathic pulmonary fibrosis, or IPF.

Your diagnosis is thought to be:

\[\text{__________________________}\]
Treating your pulmonary fibrosis

Some treatments may help relieve the symptoms of PF, and help keep you active and healthy, such as:

- Medications
- Oxygen therapy
- Pulmonary rehabilitation
- Lung transplantation
- Preventative measures, such as flu and pneumonia vaccines

Some people with PF benefit from treatment with medications. Your doctor might discuss some of the following treatments with you:

**Nintedanib** is an anti-fibrotic therapy that is approved to treat patients with IPF in the US. In clinical trials, nintedanib has been shown to slow the loss of lung function in mild-to-moderate IPF.

**Pirfenidone** is an anti-fibrotic therapy that is approved to treat patients with IPF in the US, EU, Asia, and Canada. In clinical trials, pirfenidone has been shown to slow the progression of mild-to-moderate IPF.

**Prednisone** is an anti-inflammatory therapy that can help some people with inflammation in the lungs.

The treatment option(s) that you and your healthcare provider have chosen are:

**Your follow-up testing and appointments**

Please return for a follow-up appointment in:

[ ] month(s)

In addition, the following tests or procedures have been recommended for you:

- Pulmonary (lung) function test (PFT)
- Arterial blood test (ABG)
- Cardiopulmonary exercise test (CPET)
- Six-minute walk test (6MWT)
- High-resolution computed tomography (HRCT) of the chest
- Blood tests
- Lung biopsy
- Echocardiogram (ECHO)
- Esophagram
- Rheumatology consultation
- Cardiology consultation
- Lung transplant consultation
- Other consultation
- Referral to a Pulmonary Fibrosis Foundation Care Center

**Lifestyle changes for you to live better with pulmonary fibrosis**

To stay as healthy as possible, here are some steps you can take:

- Stay active and keep in shape
- Eat well and maintain a healthy body weight
- Get plenty of rest
- Stop tobacco use and avoid secondhand smoke
- Learn and practice relaxation techniques
- Join a support group
- Try to keep a positive attitude

**SUPPORT FOR YOU FROM THE PULMONARY FIBROSIS FOUNDATION**

The mission of the Pulmonary Fibrosis Foundation (PFF) is to serve as the trusted resource for the pulmonary fibrosis (PF) community by raising awareness, providing disease education, and funding research.

To learn more about how the PFF can help support you, contact the PFF Patient Communication Center at 844.TalkPFF (844.825.5733) or pcc@pulmonaryfibrosis.org, or visit the PFF online at www.pulmonaryfibrosis.org.

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