特發性肺纖維化

以及其可能如何影響

您

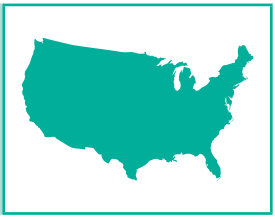
IPF = 特發性肺纖維化。

解密 IPF

IPF 是一種罕見、嚴重且進行性的肺部疾病，使病人的呼吸隨時間變得更加困難。因為 IPF 的臨床症狀和其他心肺疾病的症狀相似，例如慢性阻塞性肺病 (COPD)、鬱血性心臟衰竭 (CHF) 和哮喘/氣喘，所以醫生可能難以診斷 IPF。

每年有大約 **50,000** 例新增 IPF 病人

在美國，IPF 影響多達 **132,000** 人



IPF 是稱為間質性肺部疾病（interstitial lung diseases, 簡稱 ILD）之大型症候群的其中之一。 ILD不同於哮喘/氣喘和 COPD，它是一種*限制型*疾病。瘢痕會導致肺部硬化，使呼吸時肺部擴張更加困難。

定義 IPF

特發性
(Idiopathic)
是指疾病成因未知

肺部
(Pulmonary)
與肺部有關

纖維化
(Fibrosis)
指瘢痕

換言之，某些未知來源的物質導致肺部產生疤痕。

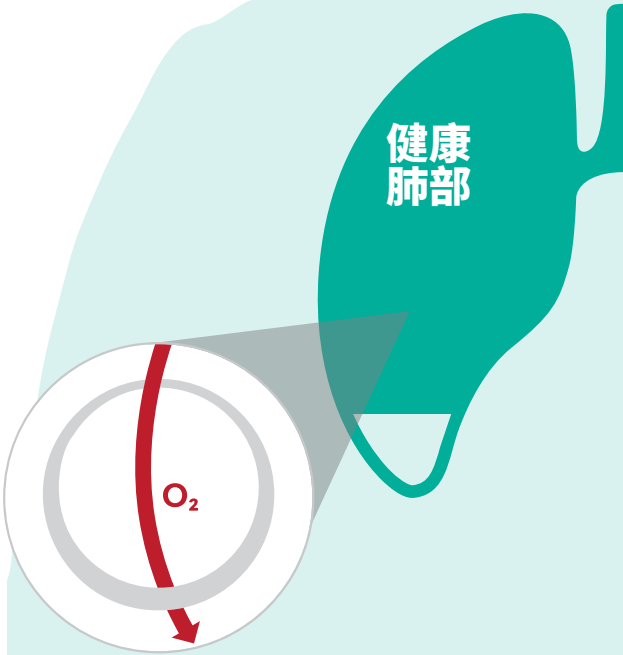
IPF 是一種進行性的肺部疾病，會隨時間惡化，所以越早開始治療越好。

請向您的醫師生問關於 IPF 的現有治療方法

您的努力工作的肺部

正常呼吸

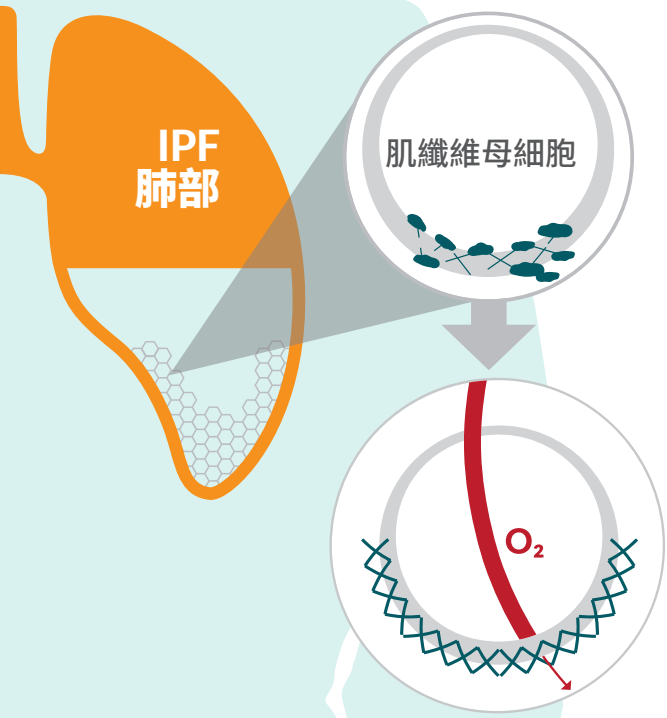
- 在健康的肺部，空氣從您的口鼻腔向下進入您的肺， 再進入稱為肺泡 (alveoli) 的小氣囊
- 您吸氣時，空氣中的氧氣（O_2）會通過肺泡壁進入動脈血管。您呼氣時，廢氣通過靜脈血管進入肺泡並最終排出您的身體



IPF 和您的肺部

瘢痕堆積

- IPF 病人身體的修復過程會對肺泡壁和周圍肺部組織中的小傷害產生過度反應
- 稱為肌纖維母細胞的特殊細胞 (myofibroblasts) 會產生過多瘢痕組織， 並在肺泡和血管壁之間堆積

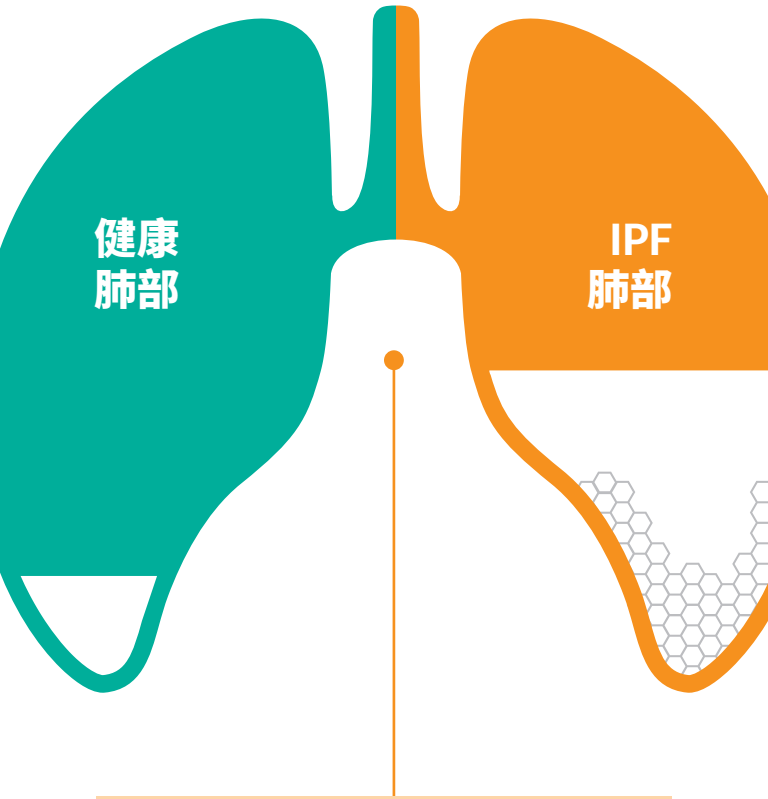


用 IPF 的肺呼吸

- 瘢痕組織會干擾氧氣流入血液中。如 IPF 肺部圖所示，當瘢痕惡化，受損的肺泡會形成囊腫。這些囊腫在一起會使肺形成蜂窩狀

瘢痕組織造成肺功能加速喪失

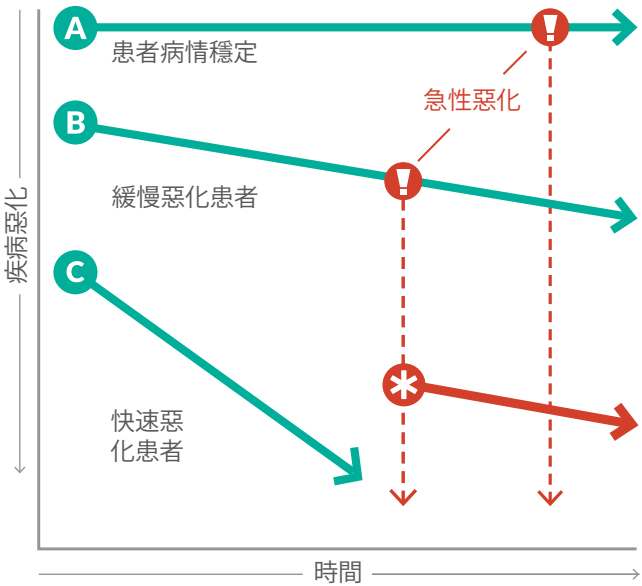
沒有肺部疾病的人隨著年齡的增長，每年大約自然喪失 **30 至 65 毫升**的肺功能。



IPF 患者因為瘢痕組織在肺部的堆積，每年可能喪失的肺功能是肺部健康人的 **3 至 4 倍**，每年大約喪失 150 至 200 毫升的肺功能。

IPF 在所有患者中的進程

IPF 的惡化速度無法預測。有些人可數年保持穩定，而其他人可能惡化的速度更快。



- A 患者病情穩定，惡化緩慢
- B 緩慢但穩定的惡化
- C 快速惡化
- ! 急性惡化
- * 急性惡化後肺功能突降

急性惡化加快病情惡化

急性惡化可能導致肺功能快速下降：

- 特徵是症狀嚴重且突然惡化
- 可能在病程的任何時間點發生
- 有時可能是 IPF 的第一個指徵
- 成因仍未知

每四個 IPF 患者中有一個可能會在 3 年內發生急性惡化

急性惡化期間會發生什麼

如果您出現以下一種或多種症狀，請立即聯繫您的醫生:

 呼吸變得更困難	 咳嗽更嚴重
 發燒或有類流感症狀	 嚴重時，自行呼吸困難

急性惡化不只是「當天不舒服」還可能導致住院。您的醫生將確定您是否曾發生急性惡化。

診斷 IPF

風險因子

雖然 IPF 的成因還不確定，但是某些因素已知可以刺激肺部並增加罹患 IPF 的風險。這些風險因素包括:

GERD （胃酸反流），或經常胃灼燒（GERD 疾病令胃酸逆流至喉嚨）。有些人可能會吸入微量的胃酸，而這可能會損壞肺部。GERD 與 IPF 的關係仍需確定

基因
遺傳因素可能加大患病風險

病毒
包括愛潑斯坦巴爾、 A 型流感、C 型肝炎、人類免疫缺乏病毒 (HIV) 和皰疹病毒

環境因素
包括長期接觸石棉、無機塵土（如二氧化矽）或有機物質（如細菌和動物蛋白質）

吸菸

IPF 的徵象與症狀

IPF 患者的一些常見症狀包括:

呼吸短促
在活動後，甚至稍後休息時；快速、淺表呼吸

乾咳、猛烈咳嗽
沒有改善

未嘗試減重，但**體重逐漸下降**

疲倦或只是感覺不適，包括肌肉和關節疼痛

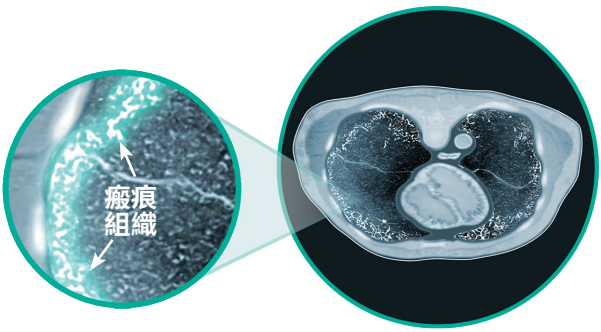
杵狀指
手指或腳趾末端變寬和變圓

IPF 的檢測

許多檢測可用於識別和監測 IPF。HRCT 通常用來單獨診斷 IPF，但是確診也可能需要肺部活體切片。

HRCT

與 IPF 相關的瘢痕組織可能會使肺部受損的部位看起來呈蜂窩狀。醫生使用高分辨率電腦斷層掃描（即 HRCT）在肺部尋找這些蜂窩狀部位。通常單獨進行此項檢測即可診斷 IPF。



肺部切片

如果在 HRCT 後仍不確定，醫生可用肺部切片以確認診斷。 做肺部活組織切片時，醫生會收集少量肺部組織檢體， 並在實驗室檢測這些組織檢體。

以下是診斷過程中進行的其他檢測:

肺功能檢測

肺功能檢測可測量您肺部的運作情況。使用肺量計時，您盡可能深吸一口氣後用力吐氣。這可測量您肺部可吸入的空氣量，以及您呼氣的速度。也可以使用其他方法檢測您的肺活量。

脈搏血氧飽和度分析儀

一台小型感測器會連接到您的手指或耳朵上以監測您的血氧度。

如果您有關於肺功能檢測 (PFT) 和最大肺活量 (FVC) 的問題，請諮詢您的醫生。

積極面對 IPF 確診後的生活

您可採取積極措施應對 IPF 對您的肺部及生活方式的影響。

建立團隊

有您的朋友、親人和醫生做您抗擊 IPF 的隊友對您的精神和身體健康很重要。

尋找支援團體

支援團體會提供關懷、疾病資訊和關於保持健康方式的提示和建議

「這些網站皆提供 IPF 的資訊。您知道的越多，您就會獲益越多。」

—Evelyn N., San Antonio, NM
2015 年確診

這些網站提供有價值的 IPF 資訊

pulmonaryfibrosis.org
patientslikeme.com
lung.org
lungsandyou.com
inspire.com
clinicaltrials.gov

如果您需要諮詢與 IPF 有關的更多資訊或是需要更多的參考資料
請致電 1-844-IPF-ANDU
(1-844-473-2638)
或造訪網址：lungsandyou.com

要積極主動！

立即詢問您的醫生
有關可用的
IPF 治療

Lungs & You™

IPF 資訊 • 支持 • 激勵

造訪網址：lungsandyou.com 或
致電 1-844-IPF-ANDU
(1-844-473-2638)

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IPF
AND HOW IT MAY AFFECT YOU

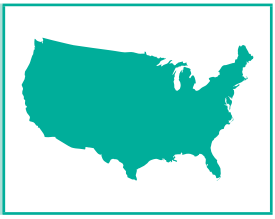
IPF=idiopathic pulmonary fibrosis.

Demystifying IPF

IPF is a rare, serious, and progressive lung disease that makes it harder to breathe over time. IPF can be difficult for doctors to diagnose because its symptoms are similar to other heart and lung diseases, like chronic obstructive pulmonary disease (COPD), congestive heart failure (CHF), and asthma.

About **50,000** new people are found to have IPF each year

IPF affects up to **132,000** people in the United States



IPF is one of a large group of conditions called interstitial lung diseases, or ILD. Unlike asthma and COPD, ILDs are *restrictive* conditions. Scarring causes the lungs to stiffen, making it harder for them to expand while breathing.

Defining IPF

Idiopathic
(ID-ee-oh-PATH-ik)
means that the cause of the disease is unknown

Pulmonary
(PUHL-mon-air-ee)
refers to the lungs

Fibrosis
(fye-BRO-sis)
means scarring

In other words, something of unknown origin is causing the lungs to become scarred.

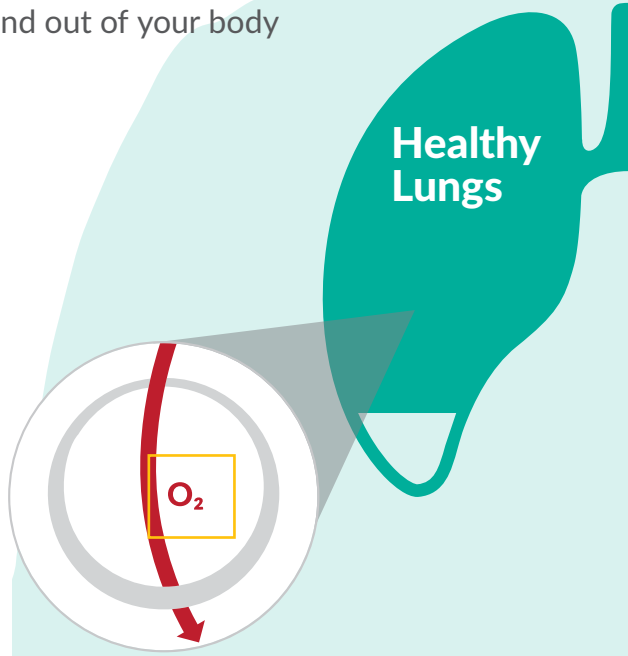
IPF is a progressive lung disease that advances over time, so the earlier that treatment begins, the better.

Ask your doctor about available treatments for IPF today

Your hardworking lungs

Normal breathing

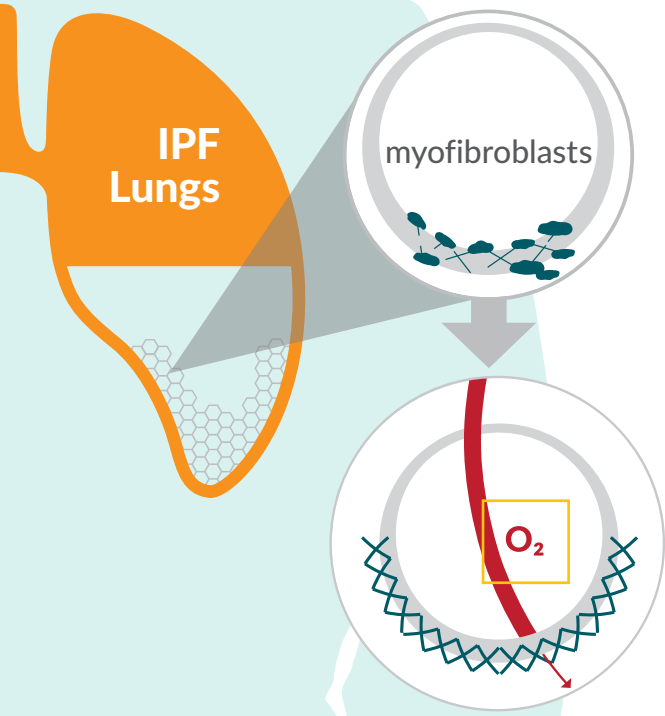
- In healthy lungs, air passes through your mouth and down to your lungs into tiny air sacs called alveoli (al-VEE-oh-lye)
- When you inhale, the oxygen (O_2) in the air passes through the walls of the alveoli into the bloodstream. When you exhale, waste gasses pass from the bloodstream into the alveoli and out of your body



IPF and your lungs

Scars build up

- In IPF, the body's repair processes overreact to tiny injuries in the walls of the alveoli and surrounding lung tissue
- Special cells called myofibroblasts (MYE-oh-FYE-bro-blasts) produce too much scar tissue, which builds up between the walls of the alveoli and the blood vessels

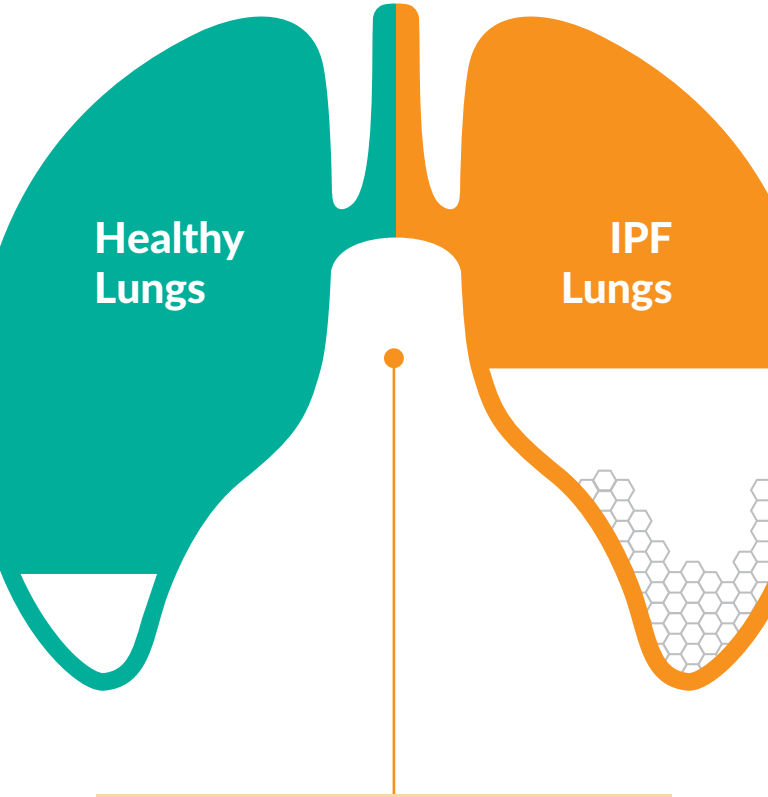


Breathing with IPF

- The scar tissue interferes with the flow of oxygen into the blood. As shown in the IPF lung, when scarring worsens, damaged alveoli form cysts. Together, these cysts form a honeycomb pattern in the lung

Scar tissue causes an increased loss of lung function

People without lung disease naturally lose lung function as they age, usually between **30 to 65 mL per year.**

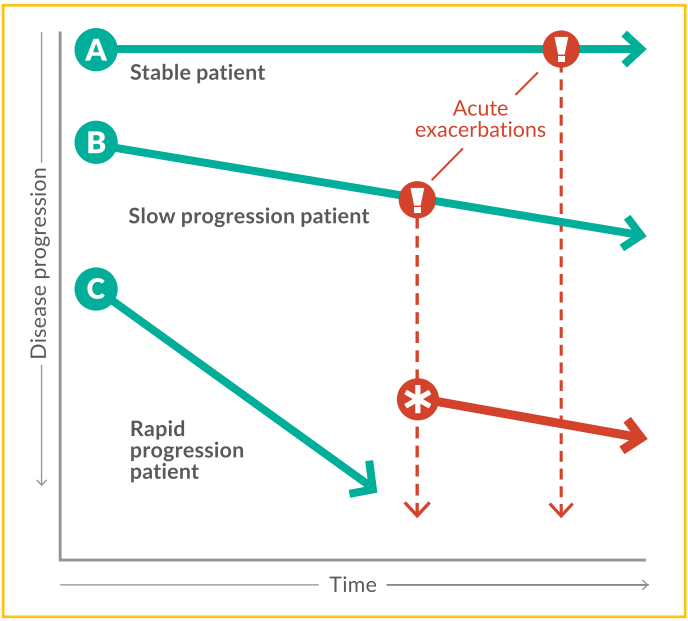


Patients with IPF may lose **3 to 4 times** more lung function, or 150 to 200 mL per year.

This is due to the accumulation of scar tissue in the lungs.

IPF progresses in all patients

It is not possible to predict how quickly IPF will progress. Some people remain stable for years, while others get worse more rapidly.



- A — Stable disease in which there is very little progression
- B — Slow-but-steady progression
- C — Rapid progression
- ! — Acute exacerbation
- * — Sudden drop in lung function following acute exacerbation

Acute exacerbations accelerate disease progression

Acute exacerbations are very serious because they can lead to a rapid decline in lung function:

- Characterized by a serious and sudden worsening of symptoms
- May occur at any point in progression
- May sometimes be the first indicator of IPF
- Cause is still unknown

1 in 4 people with IPF may suffer an acute exacerbation over a 3-year period

What happens during an acute exacerbation

If you have one or more of the following symptoms, call your doctor immediately:

 Breathing becomes more difficult	 Worsening cough
 Fever or flu-like symptoms	 In severe episodes, trouble breathing on your own

More than just “having a bad day,” an acute exacerbation may lead to hospitalization. Your doctor will determine if you’ve had an acute exacerbation.

Diagnosing IPF

Risk factors

Although doctors are not sure what causes IPF, they do know that some things can irritate the lungs and may increase a person's risk for IPF. These risk factors include:

GERD (gastroesophageal reflux disease), or frequent heartburn—With GERD, stomach acid backs up into the throat. Some people may breathe in tiny drops of this acid which may damage the lungs. The relationship of GERD to IPF still needs to be determined

Genes
Genetic factors may contribute to disease risk

Some viruses
including Epstein-Barr, influenza A, hepatitis C, HIV, and herpes virus

Environmental factors
including long periods of exposure to asbestos, inorganic dusts like silica, or organic matter like bacteria and animal proteins

Cigarette smoking

IPF signs and symptoms

Some of the common symptoms that patients with IPF describe include:

Shortness of breath at first after activity, but later even when at rest; fast, shallow breathing

Dry, hacking cough that does not get better

Gradual weight loss when not trying to lose weight

Tiredness or just not feeling well, including aching muscles and joints

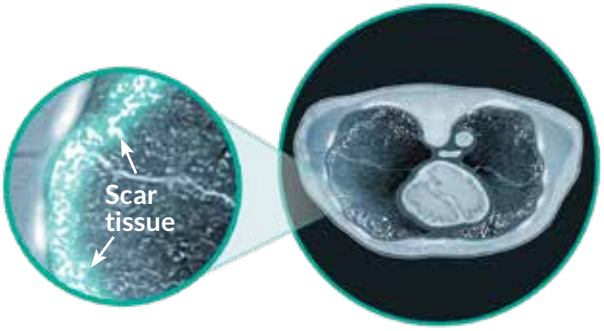
Clubbing—the widening and rounding of the tips of the fingers or toes

Testing for IPF

A number of tests are used to identify and monitor IPF. Diagnosis is usually achieved with HRCT alone, but lung biopsy may also be needed.

HRCT

The scar tissue associated with IPF may make the damaged areas of the lungs look like honeycombs. Doctors look for these honeycomb patterns using a high-resolution computed tomography scan, or HRCT. Usually this test alone can diagnose IPF.



Lung biopsy

If the doctor is still not sure after the HRCT, he or she may order a lung biopsy to confirm the diagnosis. During a lung biopsy, the doctor collects small samples of lung tissue which are examined in a laboratory.

Here are some other tests that will be performed during the diagnosis process:

Lung function tests

Lung or pulmonary function tests measure how well your lungs work. Spirometry is a test where you take your deepest breath and blow as hard and long as possible into a device. This measures how much air your lungs can hold and how fast you can exhale. Other tests may be used.

Pulse oximetry

A small sensor is attached to your finger or ear. It measures the oxygen in your blood.

Ask your doctor about your pulmonary function tests (PFTs) and forced vital capacity (FVC) reading.

Moving forward with IPF

There are positive steps you can take to deal with how IPF affects your lungs and your lifestyle.

Build a team

Having friends, relatives, and doctors who care about you is important to your mental and physical health

Find a support group

A support group offers a caring environment, disease information, and tips and advice on how to stay healthy

“The upside to all of this has been education. The more you know, the better off you are.”

—Evelyn N., San Antonio, NM
Diagnosed in 2015

These websites have valuable information on IPF

pulmonaryfibrosis.org
patientslikeme.com
lung.org
lungsandyou.com
inspire.com
clinicaltrials.gov

To speak to someone about IPF and request more educational materials, **call 1-844-IPF-ANDU (1-844-473-2638) or visit lungsandyou.com**

BE PROACTIVE!

Ask your doctor about available treatments for IPF today

Lungs & You™

IPF INFORMATION • SUPPORT • INSPIRATION

Visit lungsandyou.com or call 1-844-IPF-ANDU (1-844-473-2638)

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SUBMISSION ID:	1855595
DATE OF TRANSLATION:	23-Apr-2020
ELECTRONIC FILE NAME:	PC-US-106012 Unbranded IPF Patient Brochure - October 2018 Update
SOURCE PROJECT CODE:	PC-US-106012
TARGET PROJECT CODE:	PC-US-113656
SOURCE LANGUAGE:	English
TARGET LANGUAGE:	Traditional Chinese (Taiwan)
TRANSPERFECT JOB ID:	US0632382

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