Section 11

How will the disease progress?
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IPF progression varies from person to person

It’s important to remember that the progression of IPF can vary significantly from patient to patient. It is impossible to predict exactly how rapidly IPF will progress for you.

Slow progression
Most people with IPF experience a slow, but steady, worsening of their disease. In you have IPF that is progressing slowly, you could have the symptoms for a long time before being diagnosed and then your disease could still progress relatively slowly.

Stable progression
Some people with IPF remain stable.

Rapid progression
Some people with IPF suffer a fast decline to death. This is called rapid progression.

Acute exacerbations
A minority of patients may experience unpredictable acute (sudden and short-term) worsening of their disease, called acute exacerbations. An event like this may be fatal or may leave a person with IPF with substantially worsened disease.

If it is not possible to identify the cause of this acute respiratory decline, the term ‘idiopathic acute exacerbation of IPF’ is sometimes used, where idiopathic refers to the unknown.

An acute exacerbation can happen at any time while someone has IPF. Sometimes a person is not diagnosed with IPF until their first acute exacerbation.

The main risk factor for acute exacerbation of IPF is advanced disease.

Figure 11: How IPF progresses over time (adapted from: [21])
The danger of an acute exacerbation, makes regular monitoring by your doctor essential so they can:
• track how the disease is progressing;
• see how well you are responding to treatment; and
• decide the next steps that should be taken.

As part of your regular monitoring, you will have lung function tests to monitor the progression of your lung disease. [32]

**Other conditions IPF patients might have (co-morbidities)**

IPF can be a debilitating condition as on top of the adverse effects of pulmonary fibrosis, most IPF patients have other, associated conditions. When you have more than one condition at the same time, it is called a co-morbidity. If you have co-morbidities alongside your IPF, they can have a negative effect on the quality of your life and your prognosis. This is why when doctors are managing your IPF they also need to identify and treat any co-morbidities you may develop. [39]

These co-morbid conditions can affect the lungs (pulmonary comorbidities) or other parts of your body (non-pulmonary comorbidities).

**Pulmonary co-morbidities**

Pulmonary co-morbidities include:
• pulmonary hypertension;
• emphysema;
• venous thromboembolism;
• chronic obstructive pulmonary disease (COPD); and
• lung cancer. [40][41]

**Non-pulmonary co-morbidities**

Non-pulmonary co-morbidities include:
• coronary artery disease;
• congestive heart failure;
• sleep-disordered breathing;
• gastro-oesophageal reflux disease (GERD); and
• anxiety or depression.

To find the full definitions of co-morbidities, please visit [http://erj.ersjournals.com/content/46/4/1113](http://erj.ersjournals.com/content/46/4/1113)