Why do we get breathless?
Idiopathic pulmonary fibrosis

Idiopathic pulmonary fibrosis is a condition in which air sacs which forms the linings or walls of air sacs/alveoli get stiffened and it becomes harder to breathe. It is one of the common reasons of breathlessness in people more than 50 years of age.

Symptoms and signs:
The most common symptoms of idiopathic pulmonary fibrosis are chronic dry cough over many weeks to months and sometimes years and exertional breathlessness. It is initially present on exertion and as it progresses, can be present at rest.

These symptoms can be quite significant and can affect daily activities. As it is a progressive disease, it will affect the functioning of heart causing leg swelling and worsening of breathlessness.

Causes:
There are no definitive reasons of development of pulmonary fibrosis. We know that people patients with the known family history of pulmonary fibrosis or with smoking history or have significant dust exposure are at increased risk of idiopathic pulmonary fibrosis.

Investigations:
Blood tests are needed to exclude other causes of pulmonary fibrosis. CT scan is performed to determine extent and pattern of pulmonary fibrosis of the lungs.

Breathing tests are performed to assess the functioning of the lungs. It measures the size of lungs and diffusion capacity of lungs. It provides baseline severity but also helps with progression of the disease over time.

Treatment of idiopathic pulmonary fibrosis:
It is a progressive disease which means it will get worse over time (months and sometimes years). We cannot stop the progression of the pulmonary fibrosis altogether but we can slow it down, sometime quite significantly with medication which are called antifibrotic medicines. These include pirfenidone and nintedanib.

Patients who suffer from reflux disease benefit from anti-reflux medication. Patients should be offered pulmonary rehabilitation which helps preserve general well-being and improve pulmonary functions.

If a patient develops heart failure due to pulmonary fibrosis, they may require diuretic therapy.

If the disease progresses despite all those options and patient is fit enough for lung transplant, it can be considered if available.

Acute exacerbation of idiopathic pulmonary fibrosis:
Sometime pulmonary fibrosis get can flare up causing significant breathlessness or respiratory failure. Many such cases can be related to chest infections including viral infections. If the patients develop significant respiratory failure, they need to be admitted to a hospital and managed under care of a qualified pulmonologist.

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