INTERNATIONAL MULTIDISCIPLINARY JOURNAL FOR RESEARCH & DEVELOPMENT
SJIF 2019: 5.222 2020: 5.552 2021: 5.637 2022:5.479 2023:6.563
eISSN 2394-6334 https://www.ijmrd.in/index.php/imjrd Volume 10, issue 11 (2023)

CAUSES OF PULMONARY FIBROSIS

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Annotation: Pulmonary fibrosis is a lung disease that occurs when lung tissue becomes damaged and scarred. This thickened, stiff tissue makes it more difficult for your lungs to work properly. As pulmonary fibrosis worsens, you become progressively more short of breath.

Key words: Lung damage, blood, fibrosis, pulmonary fibrosis,symptom.

The scarring associated with pulmonary fibrosis can be caused by a multitude of factors. But in most cases, doctors can't pinpoint what's causing the problem. When a cause can't be found, the condition is termed idiopathic pulmonary fibrosis.

The lung damage caused by pulmonary fibrosis can't be repaired, but medications and therapies can sometimes help ease symptoms and improve quality of life. For some people, a lung transplant might be appropriate.

Signs and symptoms of pulmonary fibrosis may include:

- Shortness of breath (dyspnea)
- A dry cough
- Fatigue
- Unexplained weight loss
- Aching muscles and joints
- Widening and rounding of the tips of the fingers or toes (clubbing)

The course of pulmonary fibrosis — and the severity of symptoms — can vary considerably from person to person. Some people become ill very quickly with severe disease. Others have moderate symptoms that worsen more slowly, over months or years.

Some people may experience a rapid worsening of their symptoms (acute exacerbation), such as severe shortness of breath, that may last for several days to weeks. People who have acute exacerbations may be placed on a mechanical ventilator. Doctors may also prescribe antibiotics, corticosteroid medications or other medications to treat an acute exacerbation.

Pulmonary fibrosis scars and thickens the tissue around and between the air sacs (alveoli) in your lungs. This makes it more difficult for oxygen to pass into your bloodstream. The damage can be caused by many different factors — including long-term exposure to certain toxins, certain medical conditions, radiation therapy and some medications.

Long-term exposure to a number of toxins and pollutants can damage your lungs. These include:

- Silica dust
- Asbestos fibers

- Hard metal dusts
- Coal dust
- Grain dust
- Bird and animal droppings

Some people who receive radiation therapy for lung or breast cancer show signs of lung damage months or sometimes years after the initial treatment. The severity of the damage may depend on:

- How much of the lung was exposed to radiation
- The total amount of radiation administered
- Whether chemotherapy also was used
- The presence of underlying lung disease

Medications

Many drugs can damage your lungs, especially medications such as:

- Chemotherapy drugs. Drugs designed to kill cancer cells, such as methotrexate (Trexall, Otrexup, others) and cyclophosphamide, can also damage lung tissue.
- Heart medications. Some drugs used to treat irregular heartbeats, such as amiodarone (Cordarone, Nexterone, Pacerone), may harm lung tissue.
- Some antibiotics. Antibiotics such as nitrofurantoin (Macrobid, Macrodantin, others) or ethambutol can cause lung damage.
- Anti-inflammatory drugs. Certain anti-inflammatory drugs such as rituximab (Rituxan) or sulfasalazine (Azulfidine) can cause lung damage.

Many substances and conditions can lead to pulmonary fibrosis. Even so, in most cases, the cause is never found. Pulmonary fibrosis with no known cause is called idiopathic pulmonary fibrosis.

Researchers have several theories about what might trigger idiopathic pulmonary fibrosis, including viruses and exposure to tobacco smoke. Also, some forms of idiopathic pulmonary fibrosis run in families, and heredity may play a role in idiopathic pulmonary fibrosis.

Many people with idiopathic pulmonary fibrosis may also have gastroesophageal reflux disease (GERD) — a condition that occurs when acid from your stomach flows back into your esophagus. Ongoing research is evaluating if GERD may be a risk factor for idiopathic pulmonary fibrosis, or if GERD may lead to a more rapid progression of the condition. However, more research is needed to determine the association between idiopathic pulmonary fibrosis and **GERD.**

Risk factors

Factors that make you more susceptible to pulmonary fibrosis include:

Age. Although pulmonary fibrosis has been diagnosed in children and infants, the disorder is much more likely to affect middle-aged and older adults.

Sex. Idiopathic pulmonary fibrosis is more likely to affect men than women.

Smoking. Far more smokers and former smokers develop pulmonary fibrosis than do people who have never smoked. Pulmonary fibrosis can occur in patients with emphysema.

Certain occupations. You have an increased risk of developing pulmonary fibrosis if you work in mining, farming or construction or if you're exposed to pollutants known to damage your lungs.

Cancer treatments. Having radiation treatments to your chest or using certain chemotherapy drugs can increase your risk of pulmonary fibrosis.

Genetic factors. Some types of pulmonary fibrosis run in families, and genetic factors may be a component.

Complications

High blood pressure in your lungs (pulmonary hypertension). Unlike systemic high blood pressure, this condition affects only the arteries in your lungs. It begins when the smallest arteries and capillaries are compressed by scar tissue, causing increased resistance to blood flow in your lungs.This in turn raises pressure within the pulmonary arteries and the lower right heart chamber (right ventricle). Some forms of pulmonary hypertension are serious illnesses that become progressively worse and are sometimes fatal.Right-sided heart failure (cor pulmonale). This serious condition occurs when your heart's lower right chamber (ventricle) has to pump harder than usual to move blood through partially blocked pulmonary arteries.Respiratory failure. This is often the last stage of chronic lung disease. It occurs when blood oxygen levels fall dangerously low.Lung cancer. Long-standing pulmonary fibrosis also increases your risk of developing lung cancer.Lung complications. As pulmonary fibrosis progresses, it may lead to complications such as blood clots in the lungs, a collapsed lung or lung infections.

To diagnose your condition, your doctor may review your medical and family history, discuss your signs and symptoms, review any exposure you've had to dusts, gases and chemicals, and conduct a physical exam. During the physical exam, your doctor will use a stethoscope to listen carefully to your lungs while you breathe. He or she may also suggest one or more of the following tests.

If other tests haven't diagnosed the condition, doctors may need to remove a small amount of lung tissue (biopsy). The biopsy is then examined in a laboratory to diagnose pulmonary fibrosis or rule out other conditions. The tissue sample may be obtained in one of these ways:

Bronchoscopy. In this procedure, your doctor removes very small tissue samples — generally no larger than the head of a pin — using a small, flexible tube (bronchoscope) that's passed through your mouth or nose into your lungs. The tissue samples are sometimes too small for an accurate diagnosis. The biopsy may also be used to rule out other conditions.The risks of bronchoscopy are generally minor and might include a temporary sore throat or discomfort in your nose from the passage of the bronchoscope. However, serious complications can include bleeding or a deflated lung.During bronchoscopy, your doctor may conduct an additional procedure called bronchoalveolar lavage. In this procedure, your doctor injects salt water through a bronchoscope into a section of your lung, and then immediately suctions it out. The solution that's withdrawn contains cells from your air sacs.Although bronchoalveolar lavage samples a larger area of the

lung than other procedures do, it may not provide enough information to diagnose pulmonary fibrosis. It might also be used to rule out other conditions.

Surgical biopsy. Although a surgical biopsy is more invasive and has potential complications, it may be the only way to obtain a large enough tissue sample to make an accurate diagnosis. This procedure may be done as a minimally invasive surgery, called video-assisted thoracoscopic surgery (VATS), or as an open surgery (thoracotomy).

During VATS, your surgeon inserts surgical instruments and a small camera through two or three small incisions between your ribs. The camera allows your surgeon to view your lungs on a video monitor while removing tissue samples from your lungs. This procedure is performed after you've been given a general anesthetic, so you'll be asleep during the procedure.

During open surgery (thoracotomy), a surgeon removes a lung sample through an incision in the chest between your ribs. The procedure takes place after you've been given a general anesthetic.

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