

# Pulmonary fibrosis overview



## What is fibrosis of the lungs?

**Pulmonary fibrosis of the lungs occurs when the lung tissue becomes damaged, resulting in scarring and thickening of lung tissues.**

- Thickened, stiff tissue makes it more difficult for the lungs to work properly.
- As pulmonary fibrosis worsens, it becomes progressively harder to breathe, causing shortness of breath.
- Lung damage caused by pulmonary fibrosis cannot be repaired.
- Medications and therapies can sometimes help ease symptoms and improve quality of life.

## Causes

Damage can be caused by many different factors.



### Environmental

- Silica dust
- Asbestos dust
- Hard metal dust
- Coal dust
- Grain dust
- Bird and animal droppings



### Radiation

Severity of lung damage may depend on:

- How much of lung exposed
- Total amount of radiation administered
- Whether chemotherapy also used
- Presence of underlying lung disease



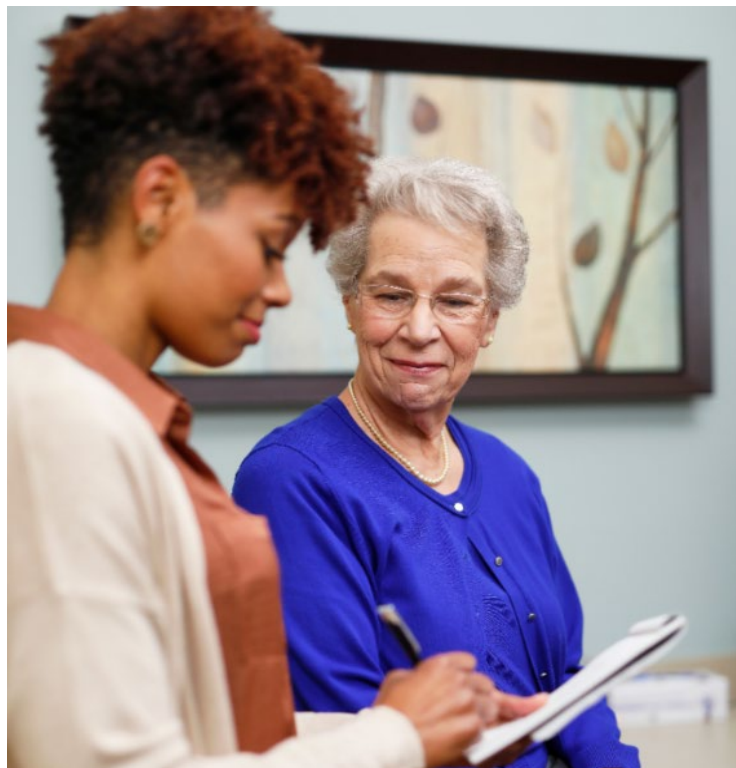
### Medical conditions

- Dermatomyositis
- Polymyositis
- Mixed connective tissue disease
- Systemic lupus erythematosus
- Rheumatoid arthritis
- Sarcoidosis
- Scleroderma
- Pneumonia



### Medications

- Chemo drugs (e.g., methotrexate, cyclophosphamide)
- Cardiac drugs (e.g., amiodarone)
- Some antibiotics (e.g., Macrobid®, Macrodantin®, ethambutol)
- Anti-inflammatory drugs (e.g., Rituxan®, Azulfidine™)



## Signs and symptoms

Signs and symptoms of pulmonary fibrosis may include:

- Shortness of breath (dyspnea)
- 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 per patient. Some become ill very quickly with severe disease, while others have moderate symptoms that worsen more slowly over months or years.

Some patients may experience a rapid worsening of their symptoms (acute exacerbation), such as severe shortness of breath, that may last for several days to weeks.

Those who have acute exacerbations may be placed on a mechanical ventilator. Prescribed antibiotics, corticosteroid medications or other medications are used to treat an acute exacerbation.

## Office note documentation – best practices

**Subjective** – Document current patient symptoms of pulmonary fibrosis (e.g., dyspnea, dry cough, fatigue, aching muscles and joints, unexplained weight loss).

**Objective** – Include:

- Related physical examination findings (e.g., fine, dry, inspiratory crackles in both lung bases, widening and rounding of tips of fingers or toes known as “clubbing”)
- Results of related diagnostic tests (e.g., chest X-ray, Computed tomography scan, echocardiogram)
- Results of any lung function tests (e.g., pulmonary function testing, pulse oximetry, exercise stress test)
- Tissue and cell analysis (e.g., bronchoscopy, bronchoalveolar lavage, surgical biopsy)

**Assessment/final diagnostic statement**

- Describe each final diagnosis to the highest level of specificity.
- Clearly link pulmonary fibrosis to the underlying cause, if known.

**Plan of treatment** – Document a clear and concise treatment plan.

- Clearly link the pulmonary fibrosis to medications being used to treat the condition.
- Include oxygen therapy, as appropriate.
- Note details of planned diagnostic testing and specialist referrals (e.g., pulmonologist).
- Date of next appointment

ICD-10-CM is a statistical classification; it is not a substitute for a healthcare provider’s final diagnostic statement. It is the provider’s responsibility to provide legible, clear, concise and complete documentation of each final diagnosis described to the highest level of specificity, which is then translated to a code for reporting purposes. It is not appropriate for providers to simply list a code number or select a code number from a list of codes in place of a written final diagnosis.

For electronic health records (EHRs) that insert diagnosis codes: The provider’s final statement of diagnosis should classify in ICD-10-CM to the EHR-inserted diagnosis code with description. Avoid mismatches between the two.

## Documentation tips for pulmonary fibrosis

Abbreviations

- Limit or avoid altogether
- Best practice: The initial notation of a diagnosis should be spelled out in full, with the abbreviation in parentheses.
- The diagnosis should be spelled out in full in the final assessment.

Terms of uncertainty

- For a confirmed diagnosis of a pulmonary fibrosis, do not use descriptors that imply uncertainty (such as “probable,” “apparently,” “likely” or “consistent with”).
- Do not document suspected pulmonary fibrosis as if the diagnosis is confirmed. Rather, document the signs and symptoms in the absence of a confirmed diagnosis.

Current versus historical

- Do not describe current pulmonary fibrosis as “history of.” In diagnosis coding, the phrase “history of” means the condition is historical and no longer exists as a current problem.

## Coding pulmonary fibrosis

The ICD-10-CM classification provides many different codes to represent the many different types and causes of pulmonary fibrosis.

Pulmonary fibrosis is coded from **Chapter 10, Diseases of the Respiratory System**, and is classified under category **J84 Other interstitial pulmonary diseases**.

The categories, subcategories and codes in the J84 section have multiple inclusions, exclusions and instructional notes that must be carefully reviewed and applied as indicated based on the medical record documentation.

Steps to ensure accurate and specific diagnosis code assignment:

1. Review the entire medical record to verify the condition remains a current problem.
2. Note the exact diagnosis description documented in the medical record.
3. Search the alphabetic index for that specific diagnosis description and the corresponding code.
4. Verify the code in the tabular list, carefully following all instructional notes, as applicable.