

## Idiopathic Pulmonary Fibrosis

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The interstitial space refers to the area in the lung between the alveolar epithelium and capillary endothelium. Interstitial lung disease is a broad category of inflammatory and fibrotic pulmonary diseases involving primarily the interstitial space. Idiopathic pulmonary fibrosis (IPF), previously known as cryptogenic fibrosing alveolitis, is a form of interstitial lung disease that has been recognized in dogs, cats and humans and is characterized by inflammation and fibrosis of the pulmonary interstitium and alveolus. Histologic features of IPF are termed unusual interstitial pneumonia and include interstitial fibrosis with foci of fibroblasts or myofibroblasts, metaplasia of alveolar epithelium and interstitial smooth muscle hyperplasia. The diagnosis and treatment of IPF can be difficult since permanent loss of pulmonary function has often occurred before clinical signs are recognized. This condition is well characterized in humans and is associated with a poor prognosis regardless of treatment.

### IDIOPATHIC PULMONARY FIBROSIS IN CATS

Idiopathic pulmonary fibrosis is a relatively uncommon condition in cats. The mean age of cats with IPF is 8 years with no apparent breed or sex predilection. Cats with IPF most commonly present for respiratory distress or cough and the duration of these signs is typically less than 6 months. Physical examination findings include tachypnea, increased inspiratory or mixed inspiratory and expiratory effort, and adventitious lung sounds. Generally, results of clinicopathologic and infectious disease assessment are nonspecific in nature. Mild neutrophilic inflammation may be noted on cytologic analysis of bronchoalveolar lavage fluid in some cats. Radiographic findings include dense patchy or diffuse interstitial, bronchial or alveolar infiltrates. Definitive diagnosis is based on histologic evaluation of lung tissue. A variety of treatments have been proposed for the treatment of IPF in cats including corticosteroids and bronchodilators. Currently, there is no published evidence that any treatment has a positive or negative impact on the clinical outcome. Prognosis for cats with IPF is poor to grave. In one retrospective study, more than 50% of cats with IPF died or were euthanatized within weeks of diagnosis while only 30% survived for a year or more after diagnosis.

## IDIOPATHIC PULMONARY FIBROSIS IN DOGS

Idiopathic pulmonary fibrosis is an uncommon condition in dogs. Most dogs with IPF are middle aged to older with no sex predilection. While IPF is considered rare in the general canine population, the condition appears to be prevalent in terrier breeds, especially West Highland white terriers. Clinical signs can include coughing, tachypnea, dyspnea and cyanosis. Inspiratory crackles are the most common physical examination finding in dogs with IPF. Clinicopathologic findings are usually nonspecific. Some dogs will have evidence of concurrent chronic bronchitis on bronchoalveolar lavage fluid analysis. Radiographically, generalized interstitial or bronchointerstitial pattern are common findings while patchy alveolar infiltrates are identified less frequently. Diffuse ground-glass opacities in all lung lobes on high resolution CT is supportive of the diagnosis of IPF in dogs. Definitive diagnosis is based on histologic evaluation of lung tissue. As in cats, there is currently no effective treatment for IPF in dogs. A combination of corticosteroids and bronchodilators has been anecdotally recommended. Similarly, there is little information pertaining to prognosis in dogs with IPF. However, median survival of 15.5 months (range 3-41 months) has been reported in one study of West Highland white terriers with IPF.

References are available upon request.

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