

Synopsis – Brendan Corcoran

Idiopathic pulmonary fibrosis (IPF) is a sporadically occurring respiratory disease of dogs and cats, but is more readily recognised in the West Highland white terrier. The pathology of IPF is poorly described in the dog and seems to resemble Non-specific Interstitial Pneumonitis (NSIP) which is at variance with the definite pathological description of Usual Interstitial Pneumonitis (UIP) applied to describe IPF in humans. The feline form of the disease in fact is more readily comparable to UIP, but is less common. Despite the pathological difference between dogs and humans the natural history of the disease shares many similarities including middle to old age onset, slow progression of clinical signs including coughing, dyspnoea and exercise intolerance with eventual respiratory failure and death. Typically dogs are first noted to be affected around 9 yrs. of age and will have a survival of 9-12 months (range 1-36). A cardinal sign of IPF in WHWTs is the presence of inspiratory crackles and that alone with signs of progressive chronic respiratory disease makes diagnosis likely. However the main differential diagnosis is chronic bronchitis which is best confirmed by bronchoscopy and identification of an inflammatory reaction on broncho-alveolar lavage. Routine haematology and biochemical profiles are normal, but many dogs are likely to be hypoxic, even if mucosal colour is normal. If cyanotic then that is a terminal sign.

Radiographic changes can vary depending on disease severity and are typically interstitial, but the best imaging modality is high resolution computed tomography (HRCT), and without confirmatory biopsy (not standard in companion animal medicine) this gives the best chance of a definitive diagnosis, especially if there is no evidence of chronic bronchitis. In the dog HRCT changes typically are dominated by ground glass opacity, typically associated more with NSIP than UIP, but other changes noted include traction bronchiectasis and parenchymal bands. There is limited information on changes in the cat. For treatment IPF is notorious as a disease with a very poor prognosis and limited response to medication. There are newer medications becoming available for human use (e.g. pirfenidone), but are not available for veterinary use yet. It is worth trying prednisolone to see if there is any response, and if none then to discontinue. There is no benefit in using bronchodilators. As up to 50% of dogs will have pulmonary arterial hypertension then treatment with sildenafil and pimobendan is worthwhile and can give some improvement in the respiratory signs. Weight control is also advisable. However, despite the progressive deterioration in respiratory function dogs appear content and will continue as best they can with normal activities. Only when quality of life is severely compromised, and definitely if cyanosis is noted, should euthanasia be carried out.