

IDIOPATHIC PULMONARY FIBROSIS

A seminar presented by: Dr. Brendan Corcoran
Sponsored by: Westie Foundation of America
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We were honored to have Dr. Brendan Corcoran from the Royal (Dick) School of Veterinary Studies at Edinburgh, Scotland as our speaker for the Montgomery County Educational Seminar. Dr. Brendan presented the history, current research and treatment of Idiopathic Pulmonary Fibrosis, known in Westie circles as "Westie Lung Disease", to an enthusiastic audience of breeders, pet owners, and veterinary staff. Information from his presentation is provided for the IMPRINT and for the Health Section of the W.H.W.T.C.A. web page.

Idiopathic Pulmonary Fibrosis is best described by analyzing the vocabulary utilized to describe the disease. Idiopathic, meaning from an unknown cause, Pulmonary, meaning the lung, and Fibrosis, meaning the formation of scar tissue makes up the analysis. Thus, in layman's terms: Lung scarring of an unknown cause. Clinicians often described the symptoms as "Cracklin Lungs."

Initial case histories, reported by Dr. Corcoran and his associates in 1999 in the United Kingdom, suggested that a "specific clinical entity consistent with chronic lung fibrosis occurs in specific breeds of terrier dogs." Since little data was available to confirm the radiographic and clinical findings in the canine, the study described a case of chronic pulmonary fibrosis in a terrier dog, where histopathological confirmation was possible and suggested that the condition might be analogous to idiopathic pulmonary fibrosis in humans. Utilizing this premise, Dr. Corcoran described the Interstitial Lung Disease Categories in dogs and cats based on the lung disease categories in humans. These categories were:

- Primary Lung Disease including: neoplasia respiratory infection hypersensitivity disorders
- Toxins
- Idiopathic Pulmonary Fibrosis

Dr. Corcoran emphasized, "despite the major accomplishments in the understanding of the pathogenesis of lung fibrosis, the diagnosis and management of pulmonary fibrosis continues to pose significant challenges." He also stated the following: "**The importance of Pulmonary Fibrosis in the West Highland White Terrier depends upon age.**" This suggests that younger dogs respond in a more positive manner to treatment, reflecting autoimmune conditions correlated with age.

The clinical features of the disease as experienced in the UK included:

Age of Onset

- Middle to old age
- Medium age of diagnosis 9 yrs. (range 4.5 - 13 years of age)

The Development and Prognosis

- Gradual onset
- Progressive deterioration
- Medium survival from clinical signs first being noted: 15.5 months (range 3-41 months)
- Medium survival from diagnosis: 7 months (range 3-41 months)

The Clinical signs of Idiopathic Pulmonary Fibrosis include the following:

- Dyspnea (labored breathing) tachypnoea
- Crackles heard in lungs upon examination
- Exercise intolerance
- Cough
- Otherwise presents as "normal"

Differential Diagnosis Considerations:

Pulmonary edema (an abnormal accumulation of fluid) associated with congestive heart failure.

- Chronic bronchitis
- Other extensive lung diseases

Diagnosis of Idiopathic Pulmonary Fibrosis should include the following:

- The characteristic clinical presentation
- The breed association
- The presence of Pulmonary crackles
- Use of Diagnostic Tests
 1. Imaging
 2. Bronchoscopy and airway cytology
 3. Pathology
 4. Blood gas analysis, haematology, biochemistry profile

Improvement is need in CT scanning technology. Tomography (HRCT) is beneficial in identifying the stage of the disease, but is very expensive. As with human diagnosis of IPF, canine diagnosis is difficult to identify. There are diffuse and patchy changes and variable findings, which may not improve diagnosis and may not affect therapy considerations. Disagreement about the classification of IPF in humans still abounds, but the degree of functional and structural changes caused by the disease correlates with the survival rate in

humans and canines. Therefore the therapy prescribed for humans and canines is extremely variable and can be addressed in these general categories:

Idiopathic Pulmonary Fibrosis Therapies:

- Anti-inflammatory therapy
 - Glucocorticosteroids
 - Azathioprine, Cyclophosphamide
- Additional therapy
 - Bronchodilators
 - Antibacterial therapy
 - Colchicines

Dr. Corcoran indicated that although treatments continue to improve, **"The response is variable, but usually poor, with inevitable progressive deterioration."**

At this time, the research into the causes of Idiopathic Pulmonary Fibrosis is SPECULATIVE. Again, turning to human research, some speculations are that the disease may be attributable to one or several of the following:

- Inflammatory reaction in the lungs
- Fibrotic reaction (We need to know WHY the lung scars so easily)
- Immune mediated
- Exposure to Toxins

Regardless of the cause, somehow the lungs get injured and disease begins. The question then becomes, **"Is this disease inherited"**? What is known is that it is a breed-associated disease, which suggests a genetic pool. In humans, there is a familial form, where family clusters have been affected. What is known on the human side is that both children and adults are affected. There is likely to be an autosomal dominant trait with penetrance. This means that the genes for the disease may be present, but do not necessarily manifest themselves. In order to determine the genetic basis of Idiopathic Pulmonary Fibrosis in canines and humans, there must be an accurate phenotype by which the affected individuals are identified. The use of molecular genetics and Microarray Technology to recognize the genes that are switched on in this disease is currently in the research stage and will affect drug protocols and gene therapy for both humans and canines.

According to Dr. Corcoran and his team of researchers, the future treatment of Idiopathic Pulmonary Fibrosis will be determined by the following:

- Improved Diagnostics including:
 1. Earlier Diagnosis
 2. High resolution CT scanning
 3. Lung biopsy
- Improved Therapy
 1. Controlled drug studies for the disease
 2. Adopt ideas from human medicine

- Epidemiology
 1. Determine the exact incidence and prevalence of the disease
 2. Conduct pedigree analysis to identify patterns of inheritance
- Molecular genetics

The West Highland White Terrier Club and the Westie Foundation of America thanks Dr. Corcoran for his excellent presentation. The Westie Foundation of America is currently working on funding for continued research in this area. Watch for information on how all Westie owners can support this important research that will benefit our Westies and the future of our breed, and all breeds that are affected by Idiopathic Pulmonary Fibrosis.