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Performing Genetic Studies and Other Pathogenesis-Based Investigations

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Description

Progressive scarring of the lung, also termed pulmonary fibrosis, has become the focus of many basic, translational and clinical investigations throughout the world. To date, this research has revealed much needed information about the epidemiology and pathogenesis of pulmonary fibrosing disorders, with particular attention to Idiopathic Pulmonary Fibrosis (IPF), the most common of the idiopathic interstitial pneumonias and the most devastating due to its poor prognosis.

However, despite many recent advances, only 2 so-called antifibrotic drugs are currently approved for the treatment of IPF; these drugs slow-down lung function decline, but do not improve the condition, and their role in other progressive fibrosing lung disorders remains unknown. Thus, much research is still needed to gain further insights into the pathogenesis of these disorders, to identify reliable diagnostic and prognostic biomarkers, and to develop effective and safe interventions that improve survival. If successful, this resea:\c:rch has the potential of positively affecting the natural course of related fibrosing disorders of the skin, kidney, heart, liver and other organs.

A major hindrance to progress in pulmonary fibrosis research is the lack of animal models capable of better resembling fibrosing lung disorders in humans and adequately predicting the efficacy of new interventions. Most animal models of pulmonary fibrosis available today require induction of lung injury by exogenous agents (bleomycin) and do not adequately model human disease, thereby raising questions about their utility in the quest for novel treatments. Even if animal models were able to duplicate most of the characteristics of human disease, such as the usual interstitial pneumonia or UIP histologic pattern found in IPF, it would be difficult to duplicate the genetic and environmental factors that contribute to disease development in humans. This, compounded by the anatomic and behavior differences between animals and humans, has prevented the development of a truly relevant model.

Interestingly, spontaneous progressive pulmonary fibrosis is not restricted to humans. In fact, this disorder has been recognized for over 2 decades in veterinary medicine in a variety of domestic animal species including cats, dogs and horses. Unfortunately, these disorders have received little attention in the biomedical community outside of veterinary medicine. Given that the affected species are long-lived animals that share a common environment with humans, they might represent relevant models of spontaneously occurring, progressive lung fibrosis. If so, investigating pulmonary fibrosis in these species could advance progress in this area.

Because of the potential of such approaches to accelerate discovery and to promote awareness, communication and collaboration regarding spontaneous progressive fibrosing lung disorders in mammals, the Westie Foundation of America (WFA) sponsored a 1-day meeting in October 2007 held in Lafayette, Indiana, USA. The WFA is the official breed association of the West Highland Terrier, a breed of dogs that is known to be afflicted with progressive lung fibrosis. This workshop brought together international physicians, veterinarians, pathologists, researchers and advocacy experts to discuss fibrotic lung disorders in humans and domestic animals. Afterward, a working group of the American Thoracic Society and participants of the initial workshop reported on the workshop findings and made the following recommendations: (1) Promote the conduction of detailed descriptive studies in affected domestic animals to define the clinical, imaging and pathologic presentation of pulmonary fibrosis in these species; (2) Emphasize the need for performing genetic studies and other pathogenesis-based investigations in naturally-occurring spontaneous models of pulmonary fibrosis to investigate the potential translation to IPF in humans as these models should provide more relevant tools to investigate the potential effectiveness of novel antifibrosis drugs in prehuman trials; (3) Emphasize the need for studies defining the anatomic and cellular differences in the lungs of different species for the adequate interpretation of discordant findings; (4) Stimulate the generation of suitable reagents to adequately test hypotheses in different species of animals; and (5) Promote the establishment of a consortium of interested centers and a central repository of clinical information and biologic specimens from naturally-occurring spontaneous models of lung fibrosis in domestic animals to enable further research that may benefit both physicians and veterinarians in their

efforts to adequately manage lung fibrosis in their patient populations.

In May 2014, a second meeting on comparative biology of pulmonary fibrosis was held in Louisville, Kentucky. As before, clinicians, researchers, veterinary doctors, pathologists and patient advocates came together to discuss the state of research in this field. The meeting was again endorsed by members of the working group on lung fibrosis of the assembly of respiratory cell and molecular biology of the American thoracic Society, and was supported by industry. The westie foundation, The Morris Animal Foundation and The AKC Canine Health Foundation. During the meeting, extensive discussions surrounded the limited progress made in the field since the first meeting. However, energized by the potential this field of investigation could have on understanding fibrosing lung disorders, the team powered through an ambitious agenda hoping to define a new path for such efforts. The proceedings of this meeting were not published; however, considering the perceived importance of the discussions held, a group of meeting organizers and participants came together to summarize its proceedings in this document.

The group discussion first focused on the fact that key clinical manifestations of pulmonary fibrosis are common in both humans and domestic animals. These similarities are best highlighted in recent publications showing that in canine IPF, for example, the disease is a chronic, progressive, interstitial lung disease affecting mainly middle-aged and old west highland white terriers.8 It is clinically characterized by exercise intolerance, restrictive dyspnea and coughing, and course crackles are present on lung auscultation. Abnormal blood gases and shortened “6-minute walk test” distance, a test that evaluates endurance and gas exchange capability, are common, and secondary pulmonary hypertension is not infrequent. These data emphasize the striking similarities in the clinical presentation of spontaneously occurring pulmonary fibrosis observed in humans and domestic animals as highlighted previously.