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The Role of Idiopathic Pulmonary Fibrosis and Anti-Fibrotic Drugs

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Description

Systems biology is an emerging discipline that offers a novel approach to how biology is understood and researched. A global overview into this field is presented, together with examples of how systems biology can be specifically applied to the field of nutrition. Some of the most relevant current lines of research are covered, and those areas with special potential for future research are highlighted.

Idiopathic Pulmonary Fibrosis

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. Overall, the group remained impressed with the similarities observed in symptoms, lung examination, abnormalities in oxygenation and imaging studies, and outcomes when comparing humans and domestic animals with pulmonary fibrosis.

However, the differences observed in histopathology strongly argue against these being identical conditions. This prompted discussions regarding mechanisms of action and several presentations were devoted to this topic. To date, there is consensus that IPF and other forms of fibrosing lung disease are likely triggered by certain exposures in the setting of host genetics that render the lung epithelium susceptible to injury. In turn, epithelial cell injury leads to its dysfunction and the subsequent elicitation of intracellular pathways responsible for the overexpression of soluble pro fibrotic growth factors. Of these, transforming growth factor-β (TGFβ) is considered the most influential, but many other activated signals exert pro-fibrotic activity.

Interestingly, spontaneous progressive pulmonary fibrosis is not restricted to humans. In fact, this disorder has been recognized for over 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.

Anti-Fibrotic Drugs

However, despite many recent advances anti-fibrotic 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 fibro sing 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. A major hindrance to progress in pulmonary fibrosis research is the lack of animal models capable of better resembling fibro sing 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 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.

In addition to the above, the group also discussed that noted similarities between dogs and humans began to dissipate when evaluating the pulmonary histopathology findings present in these conditions. Because of the potential of such

approaches to accelerate discovery and to promote awareness, communication and collaboration regarding spontaneous progressive fibro sing 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. 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. 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 anti-fibrosis drugs in pre-human trials. Emphasize the need for studies defining the anatomic and cellular differences in the lungs of different species for the adequate interpretation of discordant findings. Stimulate the generation of suitable reagents to adequately test hypotheses in different species of animals. 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.