Editorial

Dear Sir /Madam,
This folder is designed to inform you about idiopathic pulmonary fibrosis (IPF), a complex and poorly-understood disease affecting both humans and dogs, essentially terriers and in particular, the West Highland White Terrier.
IPF is difficult to diagnose and hard to treat, and is currently associated with a poor prognosis.
We are seeking to gain knowledge and improve understanding of this disease, of unknown cause. Therefore the Faculty of Veterinary Medicine at the University of Liège (Belgium) has started a research programme, with the participation of veterinary partners and breeding clubs from several European countries.

The objectives of the programme include:
- improved accuracy of early diagnosis through the identification of blood biomarkers, (substances that can be measured in the blood)
- understanding the genetic background of IPF, with the hope of identifying possible carriers of the disease to enable their removal from breeding programmes, to decrease transmission of IPF to future canine generations
- investigation of the mechanisms leading to the development of the disease, hopefully contributing to the identification of new efficacious therapeutic targets

The success of this project relies on the participation of the largest possible number of affected and control animals. Affected animals are defined as dogs with confirmed disease (based on compatible clinical signs (see below) and adequate further examinations). Controls are healthy animals of more than 3 years of age, preferably with available pedigree, that are predisposed or not to the disease, and diagnosed as healthy, based on clinical examination and blood analysis at least, and preferably including thoracic CT scan examination. Owners of control animals will be informed about the results of clinical examination and blood tests. All samples for both patients and control dogs will be stored anonymously and any personal data collected will remain strictly confidential.
Should you require any additional information, please do not hesitate to contact us. We thank you in advance for your precious collaboration.

Faculté de médecine vétérinaire

? Are you an owner, breeder, or simly passionate about the West Highland White Terrier or another terrier breed?

? Are you keen to preserve the health of your animal and improve the quality of the breed?

You can help us. Please take part in the research programme on IPF; help us to better understand this disease in order to better fight it.

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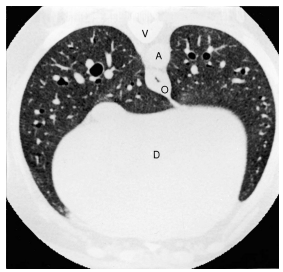
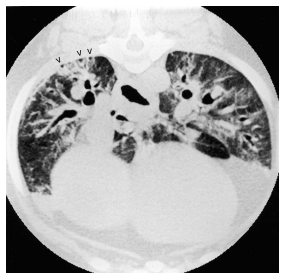
Idiopathic Pulmonary Fibrosis (IPF)

Information for owners





Université de Liège

Idiopathic Pulmonary fibrosis (IPF): a disease with many unanswered questions...

BACKGROUND

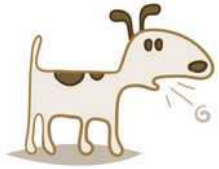
Idiopathic pulmonary fibrosis is a chronic and progressive disease characterized by development of scarring in the lungs. The lungs become thickened and stiff, and lose their ability to support oxygen transfer into the blood-stream.
In humans, the disease affects approximately 20 people per 100000. People over 60 are most frequently affected and mean survival time after diagnosis is between 3 and 5 years.
In dogs, the disease mainly affects middle-aged to older dogs of terrier breeds, and most commonly the West Highland White Terrier (WHWT). The Cairn Terrier, Scottish Terrier, Jack Russell Terrier, Bull Terrier, American Staffordshire Terrier, Yorkshire Terrier, Bichon, Shi-tzu.. are less predisposed but seem to be overrepresented as well.

CAUSES

To date, and despite extensive investigation, the cause of IPF remains unknown.
In humans, fibrosis seems to be initiated by an interaction of environmental factors (such as cigarette smoking, drugs, inhalation of viral, dust or heavy metal particles) and genetic predisposition.
In fact 10-15% of cases are considered to be familial, highly suggestive of a genetic predisposition, and may be due to specific gene mutations.
In dogs, a genetic component is strongly suspected since the diseases affects a single breed disproportionately, the West Highand White Terrier. However, genetic studies clearly identifying responsible genetic mutations in this breed are still lacking.

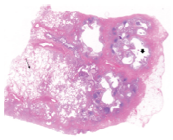
CLINICAL SIGNS

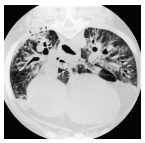
In humans, the main clinical signs include breathlessness aggravated by effort and presence of a dry cough. Generally, symptoms worsen progressively over months or years though in some people the disease remains stable for long periods or, conversely may worsen severely and rapidly.
In dogs the disease usually progresses slowly and clinical condition progressively deteriorates over months or years. The main clinical signs are exercise intolerance, cough, polypnoea or laborious breathing. Some dogs show episodes of syncope or cyanosis (blue-ish colouring of the tongue). On thoracic auscultation, diffuse harsh crackles can be heard in the majority of the cases; these are considered one of the hallmarks of the disease.



ADDITIONAL TESTING

In dogs, it is difficult to confirm a diagnosis of IPF. Firstly it needs to be distinguished from other chronic bronchopulmonary diseases (for example, chronic bronchitis) or from disease of cardiac origin. Therefore diagnostic tests may be performed including thoracic radiography, echocardiography, bronchoscopy and analysis of the bronchoalveolar lavage fluid (under anaesthesia when judged safe enough), non invasive pulmonary function tests such as blood gas analysis and the "6-min-walk-test". The most useful information can be gleaned from a thoracic CT scan. However, a definitive and accurate diagnosis requires histopathological examination of a small piece of lung tissue (lung biopsy). Since this procedure requires anaesthesia, is relatively invasive and expensive, it is not always performed. Lung biopsies may also be collected just after the animal's death.





TREATMENT

While there are currently no effective treatments for IPF, there are a variety of therapeutic options to help patients manage their condition and maintain their quality of life and routine activities.
In human medicine, Pirfenidone, a drug with antifibrotic and anti-inflammatory properties, has very recently been approved for use for human IPF. Its efficacy and safety in dogs remains questionable. Recent studies also show a beneficial role for N-acetylcysteine (NAC), a naturally occurring antioxidant molecule, on lung function in IPF patients. In humans, oxygen therapy is required in severe cases, or pending lung transplantation.
In dogs, lung transplantation is not available. Similar to the situation in humans, treatment essentially aims at minimizing clinical signs and optimizing quality of life of the dog. We hope to be able to propose novel molecules for the treatment of IPF patients in the near future since many new therapies are currently undergoing efficacy testing through clinical trials.

FOLLOW UP/MANAGEMENT

Both in human and canine medicine individual patient follow up is essential. Indeed, strategies to treat IPF are highly personalized, based upon the specific patient's medical history and other conditions (comorbities). A common complication in IPF dogs is the development of pulmonary arterial hypertension that leads to breathlessness and discomfort and may induce cardiac insufficiency.
It is therefore advised to have your pet checked on a 4-6 monthly basis, with repetition of the non-invasive testing procedures (echocardiography and lung function tests) to permit adaptation of the treatment. Here, at the faculty of veterinary medicine these examinations are offered within the framework of our clinical investigation programme.