Westies Leading the Way for Innovations in Lung Fibrosis

Dogs Needed for Innovative Comparative Research Effort at University of Minnesota By Teresa Barnes

hen the Westie Foundation of America (WFA) held its first meeting on Westies and Lung Fibrosis in 2007, few veterinary researchers were focused on lung fibrosis in the lungs of Westies and virtually no human researchers were aware the lung disease that accounted for more than 40,000 deaths in the U.S. each year, the same number of deaths attributed to breast cancer in humans, was also an increasingly common cause of death in the West Highland White Terrier.

Until that time, human researchers were largely unaware of the naturally occurring disease in domestic animals. Most Pulmonary Fibrosis (PF) research in the lab is carried out with the use of mouse models that are induced with fibrosis

because the disease doesn't naturally occur in the species.

After the seminal meeting, human researchers began to pay attention to PF in Westies (also known as Westie Lung Disease) and veterinary researchers expanded their reach in the disease efforts.

Though studying dogs in lung fibrosis was a new concept, the idea of studying canines with naturally occurring disease is common in cancer research efforts. In fact, the National Cancer Institute (NCI) has an entire division devoted to comparative research efforts comparing canine cancers to human cancers and making strides towards therapies.

A paper published in the Annals of the American Thoracic Society in the winter of 2013 laid out the importance of the Westie in the future of lung fibrosis research and was a call to action to the human research community, paving the way for another meeting that would move the science forward even faster.

In the spring of 2014, the WFA convened the Fibrosis Across Species meeting, with the help of generous individual donors and corporate sponsors, the meeting brought together veterinary scientists and human lung fibrosis experts from around the globe. They met in Louisville, Kentucky, by no accident, and the same week as the Kentucky Derby.

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Attention was given to PF in horses, particularly race horses, as well as dogs, including the Westie, and other domestic animals.

Stemming from the Fibrosis Across Species meeting are efforts that involve collaborations between veterinarian and human PF researchers. One of those is a new collaboration at the University of Minnesota between researchers who first started their collaborative research by attending the meeting, though they had worked in the same university for years.

Ned Patterson, DVM, PhD, and Peter Bitterman, MD are now working closely to better understand the Westies' risk of PF as well as their potential role in the discovery of new

> therapies – therapies that could not only save Westies from the deadly lung disease but also save humans.

The work of the veterinarian and pulmonary research is focused on the complex cell circuitry that leads to the lung scarring or fibrosis.

"This important research is a direct result of efforts by veterinary scientists and human PF scientists working together to reach a common goal," said Kay McGuire, DVM, a veterinarian and WFA board member. "We are pleased the WFA and Westies could lead the way in this effort."

The Minnesota team hopes to accelerate the pipeline of new agents by comparing the genetics and scarring signaling circuitry in Westies and humans, looking for abnormalities shared by both. The idea is that genes and signaling steps present in both human and Westie PF are very likely to represent the essential steps in the lung scarring process, and represent good targets for new drugs.

"At the University of Minnesota Veterinary Medical Center, we have been seeing Westies with and without pulmonary fibrosis for our studies with Dr. Bitterman at the medical school," said Patterson. "We are also looking at the genetics in Westies themselves in search of susceptibility genes to hopefully help future breeding decisions."

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The work is currently in progress using lung samples generously provided by human patients at the time of biopsy, transplant or their demise; and by Westie patients at the time of euthanasia for intractable shortness of breath.

Westie owner Dena Zimmer has an appreciation for the collaboration of the two experts and the commitment of the WFA to support research efforts in PF.

Zimmer has seen firsthand the work they are doing. Her two Westies (sisters), Sally and Suzi, were both diagnosed with PF and were subsequently the first dogs recruited for the Patterson-Bitterman research study at the University of Minnesota.

In humans, familial or genetically-related PF cases are relatively common and make up at least one fifth of PF cases.



Sally in CT scan.



Suzi in CT scan.

In Westies, it is not yet clear what percentage of cases are familial.

Though not happy to hear both of her dogs were ill with the deadly lung disease, Zimmer was excited her dogs were asked to be part of the innovative research. "[It] is sort of cool I think. They are very calm dogs, so they didn't have to be sedated for any of the tests, including the CT scan, and so their photos are being used to show folks that many Westies don't have to be sedated to do this very short (2-3 min) scan," Zimmer said. "I was very impressed with both doctors and enjoyed and learned from them on that and one more visit "

Sally developed heart issues, which are not uncommon co-morbid conditions with PF, and, as a result, Zimmer was forced to put her down in April. She donated, by necropsy, Sally to the research study



Suzi is on left and Sally on right)

so that Patterson and Bitterman could continue to learn from her. Suzi remains in the study and continues to fight PF.

"I am so encouraged by this study and the compassion shown to participating Westies and their owners. Dr. Bitterman and Dr. Patterson are examples of the high caliber of researchers necessary to overcome PF," said Bebe Pinter, President of the WFA.

Meanwhile, the research is moving forward and more Westies are needed for the Patterson-Bitterman study.

"While we always would like more study participants, there has already been more interest and dogs participating than we expected. Bringing in affected dogs to our hospital has also helped symptomatic treatment of the affected dogs in collaboration with our cardiologists and rehabilitation specialists," Patterson said. "For the study, we just need two blood samples from unaffected dogs eight years or older, and the blood samples plus chest x-rays, and an un-sedated CT for affected dogs of which all costs are covered by the study."

Please contact Dr. Patterson at patte037@umn.edu with any questions about the study and mention this article.

Zimmer, along with other Westie supporters, is an administrator for a relatively new Facebook group created as a forum for discussion of Westie Lung Disease/Pulmonary Fibrosis in the U.S. Much of the discussions on the page address dogs' disease symptoms, current treatments and studies that are underway. To join the group, visit: https://www.facebook.com/groups/327689790763155/.