# Terriers Join Fight Against a Killer Disease in Humans

## 'Westies' breed is also prone to pulmonary fibrosis, which has no cure

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*THURSDAY, Dec. 27 (HealthDay News)* -- A feisty breed of terrier could stop scientists from barking up the wrong tree as they research a deadly lung disease in humans.

The illness, called idiopathic pulmonary fibrosis (IPF), affects 128,000 Americans, is typically fatal within three years of diagnosis, and kills more than 40,000 people in the United States annually -- a death toll equivalent to that of breast cancer.

A fatal condition that looks remarkably like IPF also strikes the diminutive West Highland White terrier ("Westie"), however. And recently, medical scientists from the human and veterinarian worlds met for the first time to share information and pool resources against a mysterious killer.

"People may be a little startled at first to learn about this idea -- 'You're kidding me, you actually think there's promise in studying this dog to help my Dad with this disease?' And the answer is -- 'Yes'," said Mark Shreve, chief operating officer of the patient advocacy group Coalition for Pulmonary Fibrosis, based in San Jose, Calif.

West Highland White Terrier. Photo courtesy American Kennel Club

Because the Westie is so tightly bred, and because the illness progresses faster in dogs than humans, it is conceivable that dog-based research might yield valuable clues to the genetics or environmental factors that trigger pulmonary fibrosis in both species, experts explained.

"And if it transpires that it *is* the same disease, then obviously the options are limitless as to how we can look at information from dogs and use it to understand the disease in humans and vice versa," said Dr. Brendan Corcoran, director of the Hospital for Small Animals at the University of Edinburgh, Scotland, and a pioneer in researching pulmonary fibrosis in Westies.

According to Shreve, most people find it hard to believe that a disease like IPF even exists amid the wonders of modern medicine.

"We are dealing here with one of the few diseases left on the planet for which there are no proven causes and no treatments," he said.

Idiopathic pulmonary fibrosis occurs spontaneously, although certain factors -- such as smoking or exposure to airborne toxins -- do raise risks for the illness. "IPF is a

progressive scarring process in the lungs that gradually robs a person of the ability to breathe," Shreve explained. Some sort of signaling seems to go awry at the cellular level, he said, converting normal, expansive lung tissue into stiff, fibrotic scar tissue.

"Once it starts in patients with IPF, your body just never sends a signal to stop that scar tissue from being produced," Shreve said. "This scar tissue is obviously not lung tissue that is able to process oxygen."

There have so far been very few promising leads in discovering the root causes of IPF, said Dr. Jesse Roman, one of the country's leading researchers in the disease and a professor of medicine at Emory University in Atlanta.

"Studies do suggest very specific [cellular] pathways, and there's a number of molecules that everybody is tuned into," he said. "But how you block them and how they relate to what happens in humans, that's less clear."

So, scientists are turning to creative new ways of looking at IPF.

Cross-talk between scientists worldwide led to the first-ever summit on the disease that included both veterinary and human medical researchers. The meeting was held in October on the campus of Purdue University in West Lafayette, Ind., and was attended by Corcoran, Roman and others. It was sponsored by the Westie Foundation of America and the American Kennel Club (AKC) Canine Health Foundation.

Westies, which grow to just under a foot in length, are described by the AKC as "courageous and self-reliant, but friendly."

"They're a very popular pet because of their size and their nature," Corcoran said.

However, pulmonary fibrosis does pop up in the breed with regularity, first revealing itself as excessive panting and shortness of breath. The illness also tends to develop in the terriers' late middle-age (about eight or nine years), mimicking its typical onset in humans at about age 50 to 60.

Westies inevitably succumb to the lung fibrosis about a year and a half after their diagnosis, Corcoran said.

Still, "there's still the contentious issue of whether this is the same disease as occurs in humans," he said. The exact prevalence of the disease among Westies is also unclear, he added. That means the first aim of Westies-centered research will be epidemiological -- studying disease prevalence and gathering a core of dogs and their owners that researchers might follow going forward.

Getting postmortem samples of canine lung tissue will also be crucial to a better understanding of the causes of the disease, Corcoran said. But that has its own challenges, he added.

"Getting owners to volunteer their dogs for necropsy is always problematic," he said. In fact, it's often "harder in many instances to get lung pathology samples from dogs than it is from humans," Corcoran said.

"However, one of our plans is to try and build up a group of concerned owners who will volunteer to donate their dog when that day arrives. We've been having some discussions on that already with our colleagues in America," Corcoran said. "Hopefully, the more publicity that we get with this condition, the more we may get owners coming forward and volunteering their dogs for research."

Corcoran and the other experts said that a cure for IPF is definitely *not* around the corner -- the disease has been as

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tenacious in keeping its secrets as, well, a terrier.

But Westies may be just the foe in the fight against IPF requires. Corcoran pointed out that the dogs' tight breeding means genetic research could yield important clues. And their shorter lifespan -- a seventh of that of humans -- means scientists can watch the disease in "fast-forward," which might also speed research.

Westies are also free of certain confounding factors, such as smoking, that often muddle human research. "The dogs might turn out to have a very pure form of the disease that allows you to investigate the disease itself and not worry about other factors," Corcoran noted.

Given all of this, "why wouldn't you look at a Westie and research how the disease progresses?" said patient advocate Shreve.

"We think it's a very creative approach to trying to help out humans," he said, "and our patients don't really have the patience to hang around waiting for a miracle.

**More information**

To learn much more about IPF, visit the Coalition for Pulmonary Fibrosis.

SOURCES: *Brendan Corcoran, Ph.D., professor and director, Hospital for Small Animals, University of Edinburgh, Scotland; Jesse Roman, M.D., professor, medicine, and director, pulmonary allergy and CCM, Emory University, Atlanta; Mark Shreve, chief operating officer, Coalition for Pulmonary Fibrosis, San Jose, Calif.*

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