

PATIENTS, DOCTORS, AND RESEARCHERS FACE A  
TERRIFYING DISEASE TOGETHER | BY GAVIN JENKINS

# MUTUAL SUPPORT

Jerry French had never heard of idiopathic pulmonary fibrosis (IPF) when a doctor diagnosed him with the disease three years ago. It took French months to understand and accept what was happening: The interstitium—the space between alveoli and the bloodstream—in his lungs was thickening with scar tissue, making it difficult for oxygen to reach the capillaries. His lungs were stiff and couldn't properly inflate, diminishing his breathing capacity. It was going to get worse, and a lung transplant was the only cure.

For two years, French's internist had been treating him for a sinus infection. Despite a nagging cough, French, now 72, worked full-time in commercial contracting and construction. He played church-league softball and was active around his Clarksburg, W. Va., home. The statistics he and his wife learned from the pulmonologist were shocking: the median life span for IPF patients after diagnosis is three to five years.

PHOTOGRAPHY | TOM M. JOHNSON



The support group at the Simmons Center helps researchers as well as patients. LEFT: IPF patient Jerry French on his West Virginia property.



The idiopathic aspect—that no one knew the cause—was “devastating” to French. Genetic and environmental links have been found, but, French says, he wanted to put his “finger on what it was that did this.”

Like more than half of all IPF patients, French became depressed following the diagnosis. He sat in front of the television, obsessing over what would happen to his lungs and how it would feel. His wife, Katie French, encouraged him to get off the couch and do something. Anything.

He eventually listened and pulled out of

ence room. Nearly every patient breathed with the help of an oxygen tank. Most IPF patients are older than 50; the disease forces many to slow down. Some end up retiring early.

Jared Chiarchiaro’s keynote talk that day was called “Planning for Your Future with Your Loved Ones.”

Chiarchiaro, assistant professor of medicine, began his remarks with a question: “How many of you had never heard of IPF before you or your loved ones were diagnosed?”

Twenty-three hands shot into the air.

Chiarchiaro’s lecture focused on acknowl-

support group and other programs at the Simmons Center, says that the community atmosphere has inspired volunteerism among the patients. In 2003, Charles Ward, a 75-year-old retired firefighter, asked to talk to Lindell after a support group meeting. He was wondering, *After he died, could he donate his lungs for research?* Naftali Kaminski, then the center’s director, looked into it.

He learned that Ward’s lung had to be donated within six hours of his death. The timeframe would help doctors keep lung tissue in the best possible condition so they could

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the fog by attending IPF support group meetings hosted by the Dorothy P. and Richard P. Simmons Center for Interstitial Lung Disease at UPMC.

IPF can be terrifying. To address this, the Simmons Center started the support group program, among the nation’s first for IPF patients, in 2001.

A community of patients, their caregivers, and medical professionals has arisen from the gatherings. Today, the community not only supports patients and their families, but advances the work of University of Pittsburgh doctors and scientists, as well. Patient participation has yielded precious research resources, including the largest IPF tissue bank in the country.

The Simmons Center is named after a prominent Pittsburgh businessman and his wife. Following Dorothy Simmons’s death from IPF in January 2001, Richard Simmons donated \$5 million to the University of Pittsburgh to create the center; he stipulated that educating and supporting IPF patients be a priority along with finding a cause and a cure.

The monthly gathering is catered. Patients take turns talking about how they are doing. Then it starts to feel more like a college seminar, with physicians and other scientists explaining their research and fielding questions. At April’s meeting, 23 patients and caregivers sat around tables assembled in the shape of a horseshoe in a Scaife Hall confer-

encing death, talking to relatives about it, and planning for end-of-life care. The one patient who arrived alone said that he wished his wife had attended. “She still thinks I have 10 years,” he said to the group. “We’ve got to have this conversation.”

Other recent meetings have focused on clinical trials and stem cell therapy. Following each lecture, there is often an icebreaker. Once a year, the meeting includes a lab tour.

“Everyone there becomes family,” says Wesley Plietz, a retired postal worker from Glenshaw, Pa., who was diagnosed with IPF a decade ago. Plietz, 75, received a double-lung transplant seven years ago but still attends several meetings a year to share his story and give advice. After a lung transplant, patients have a 50 percent chance of living five years. Plietz tells people at the meetings that he has beaten the odds because he never forgets to take a rejection pill or vitamin. He jokes that he’s a pill peddler.

The meetings have even attracted patients who don’t have IPF. Skip Mortimer, 74, suffered from emphysema and bronchitis before a lung transplant six years ago. Mortimer, a Weirton, W. Va., resident, started attending the monthly meetings in Pittsburgh two years before the procedure because he’d heard that the group was supportive and active. He still attends and now gives a ride to a friend who has IPF.

Kathleen Lindell, PhD and RN research assistant professor of medicine who runs the

isolate live cells to study. After Ward died, Kaminski and Lindell created the country’s first rapid tissue (then known as “warm autopsy”) donation program for lungs. Rapid tissue donation was common with prostate cancer and neuromuscular diseases, but it had never been tried with lungs, says Lindell.

To date, 85 Simmons Center patients have participated, and French plans to donate after he dies. “If we’re working with researchers, why not do something more?” French says. “Because they can definitely do something more with the tissue.”

A number of labs have used Pitt’s banked lung tissue, as well as blood samples, in their research. In the 16 years since its founding, Simmons Center researchers—under the leadership of Kaminski and former medical director James Dauber, as well as current Simmons director Daniel Kass and medical director Kevin Gibson—have published 350 papers. Some of those are among the most cited in IPF literature.

One Pitt study, led by Ana Mora, associate professor of medicine, on scarred lung tissue samples revealed mitochondrial abnormalities in IPF patients. Mora’s team found that the abnormality was linked to aging and an enzyme deficiency that eventuates in fibrosis.

Kaminski and Gibson, a professor of medicine, discovered the first validated peripheral blood biomarkers for IPF. Such biomarkers should help doctors predict how IPF will progress in each case.





**Wesley Plietz had a double-lung transplant for IPF seven years ago; he's beaten the odds. After a lung transplant, patients have a 50 percent chance of living five years.**

That's important because an IPF patient's condition can plateau for years and then quickly deteriorate to acute exacerbation, which Kass, who is an associate professor of medicine, describes as falling off a cliff. Those patients might die within weeks.

Another investigation involving Simmons patients could change how doctors think about the disease. For years, the conventional wisdom was that IPF had nothing to do with inflammation. According to Kass, there was not a lot of inflammation evident in biopsies when pathologists looked under the microscope.

"So they thought it was a disease of scarring that never ends," Kass says. "And nothing appears to provoke that scar."

But Steven Duncan (Fel '85), a former associate professor at Pitt who is now at the University of Alabama at Birmingham (UAB),

thought the arguments against inflammation in IPF seemed specious, given how most fibrotic diseases have an inflammatory component.

Duncan suggested that IPF should be treated like an autoimmune disease, such as lupus or rheumatoid arthritis. Through the course of two years, Duncan and Michael Donahoe, Pitt professor of medicine and executive vice chair of clinical affairs, treated 10 Simmons Center patients who were hospitalized because of acute exacerbation. The doctors ordered plasma exchanges for the patients to deplete their bodies of autoantibodies. The work was supported by UPMC. Duncan says that, after one year, the new treatment was effective in more than 50 percent of cases; before this, patients did not respond significantly to any treatments.

"When it salvaged them, it was like a

miracle," Duncan says. "People who were critically ill would get up to leave the hospital days later using either low-flow oxygen or no oxygen."

Later this year, doctors at Pitt, UAB, Temple University, and Harvard University will begin a National Institutes of Health-funded multicenter trial using this method of treatment.

Simmons Center research associate John Sembrat, who contributed to the mitochondria study, says talking to patients at the monthly meetings keeps him motivated during those late nights in the lab.

"I always think: *How can you even breathe with a lung as damaged as that?* And then I get to meet these people, who are carrying around an oxygen tank. Every day, they're waiting and waiting for some kind of cure, some kind of help."

Mary Camphire, who was diagnosed with IPF two years ago, was at the April meeting with her husband, Jack. She called Chiarchiaro's talk the most difficult discussion she had attended and described her ride home to Edinboro, Pa., as "terrible." But after talking with her husband about the meeting for a couple of days, she was glad they'd gone.

"It opened my eyes," says Camphire, 75. "I guess you try to put things out of your mind when they're not in your face, and that brought it to the surface."

During the meeting, everyone shared what they hoped for most at the end of life. People wished to not be a burden on their loved ones and to have a painless death.

There were lighter moments, too, like when the couples shared what they did on their first dates. Jack Camphire had invited Mary to a jazz concert. However, she'd turned him down for dates so many times before that he hadn't bought the tickets. So he rushed to get them.

A couple of times, the mood in the room threatened to turn dour, and French turned it around.

At one particularly serious point in the discussion, he looked at his allies around the table and said, "We should be passing around a bottle of Crown [Royal]." Everyone laughed. ■