

Jeff Goldstein

PULMONARY FIBROSIS



“Lung transplant should be more than just about surviving. It should also be about living and giving.”

I am a double lung transplant recipient, with chronic myelogenous leukemia that is currently in remission. I have undergone approximately four dozen Moh's surgeries, four courses of radiation therapy, a complete right auriclectomy with removal of lymph nodes and partial parotid gland, and tumor surgery with removal of left side lymph nodes and all of my parotid gland.

One humid summer evening in Miami, I collapsed in my driveway while dragging trash to the curb. I couldn't catch my breath and thought, "Will my wife realize I am not in the house and get to me in time to save me?" I was finally able to control my breathing and thus began my relationship with IPF, idiopathic pulmonary fibrosis.

How could I have a diagnosis of "end stage lung disease?" When I think back I realize there were symptoms I didn't recognize but rationalized as being out of shape, overworked, or tired from not getting enough sleep. This is the *modis operendi* of IPF, and in fact of many of the fibrotic lung diseases. Unless you are able to detect it early by some serendipitous opportunity, you don't know it or feel it until there is significant lung damage.

I was transplanted, just barely in time, on July 6, 2003. My transplant went well, and I considered myself fortunate. It took almost a year and a half to reach my new normal. It was grueling, exhausting, and often uncomfortable. I was grateful, but I felt guilty. I saw others whose transplants didn't work and were rejected within days. Often those patients' bodies were so weak that they couldn't recover or hold on much longer. And for some, their only hope for life—new lungs—didn't come in time. Why has it worked for me? Did I really deserve my new lungs?

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Pulmonary fibrosis describes a group of lung diseases in which thickening of the walls of the air sacs (called alveoli), caused by scarring, can result in cough, shortness of breath, fatigue and low blood oxygen levels. It can be caused by an identifiable irritation to the lungs, but in many cases the cause is unknown, which is described as idiopathic pulmonary fibrosis (IPF). Idiopathic means there is no known cause at this time. Symptoms include:

- Dry cough or shortness of breath
- Abnormal breath sounds—crackles (like Velcro) can be heard by your health care provider when you take a deep breath
- The ends of your fingers and/or toes have changed to a club shape (called "clubbing")

Learn more: *ATS Patient Information Series. "Idiopathic Pulmonary Fibrosis (IPF)"* New York, NY: American Thoracic Society 2015. thoracic.org/patients/patient-resources/resources/idiopathic-pulmonary-fibrosis.pdf

Perhaps there wasn't an answer. Then a timely letter from my donor's family relieved me of that burden of guilt. I knew the statistics all too well: 65 percent of transplant recipients do not survive the first year, 50 percent of those do not survive to year three and of those that do, 50 percent do not survive to year five. I decided that my transplant was a gift, and I adopted a line from my favorite movie, *Shawshank Redemption*, "Get busy living!"

My brother is an important part of my story. In April 2012, Dan was diagnosed with "likely" IPF. What followed were regular visits to a local pulmonologist to monitor his health status. In December 2013, he was diagnosed with IPF and non-small cell lung cancer. We struggled heavily with the diagnosis.

There was nothing we could do to keep Dan comfortable. He underwent chemotherapy with some positive results, but he continued to suffer from pneumothoraces, and his health declined precipitously. On May 8, 2014, my brother, Dan, died.

Lung transplant can be a lifesaving and transforming experience for those fortunate enough to receive them. But lung transplant should be more than just about surviving. It should also be about living and giving.

Along with a group of other transplantees I started the national nonprofit Lung Transplant Foundation, of which I'm the current president. Our mission is to raise funds to promote research in post-transplant rejection and successful post-transplant adjustment. For other families, there's still hope.