



ATS Patient Voices 2

A publication from the American Thoracic Society Public Advisory Roundtable.

ATS Patient Voices is published by the American Thoracic Society Public Advisory Roundtable (ATS PAR). Since 2001, ATS PAR has been a core component of the Society and a mutually beneficial partnership wherein organizations that represent persons affected by respiratory diseases, illnesses requiring critical care, sleep-related disorders collaborate with the ATS to advance their shared educational, research, patient care, and advocacy goals.

The ATS strives to improve health worldwide by advancing research, clinical care, and public health in respiratory disease, critical illness, and sleep disorders. The roots of the ATS reach back to 1905, when a small group of physicians and researchers began sharing information about tuberculosis. Since then, it has grown into an international society with more than 15,000 members.



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PULMONARY • CRITICAL CARE • SLEEP

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Disclaimer

This publication includes stories of lung disease patients as told to the American Thoracic Society by the patients or their representatives. The views expressed in these stories do not reflect those of the ATS. The ATS makes no claim as to the efficacy of treatments, veracity of diagnoses, or competency of any physician or medical institution referenced herein.

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The American Thoracic Society (ATS) has long held the inclusion of the patient perspective as a core component of its mission. For more than a decade, the ATS Public Advisory Roundtable (PAR), which represents the patient voice of the Society, has played an invaluable role in helping the organization shape its policies to keep families and patients as a central focus of all ATS programs and activities.

Nowhere has this been more evident than at the annual ATS International Conference where PAR has facilitated patient programs such as the **Breathing Better with the ATS** patient and family forum, the **Meet-the-Expert** public forum, the **PAR Symposium** and the many patient speakers integrated throughout the scientific session curriculum.

Patient Voices, now in its second edition, highlights the stories of some of the patients who have spoken at past ATS International Conferences on their experiences with pulmonary disorders such as COPD, lung cancer, pulmonary hypertension, hypersensitivity pneumonitis, alpha-1 antitrypsin disease, and idiopathic pulmonary fibrosis, among others. These brave patients, many of whom have had lifelong struggles with their diseases, put faces and voices to these oftentimes life-threatening conditions. Their stories serve as an inspiration to many others who have pulmonary diseases. They illustrate that a full life can go on after diagnosis, and that patient voices will be heard.

Dr. Finn's focus on equality of access to health and health care emphasizes the importance of addressing both population and individual measures of health status and well-being. A major thrust of Dr. Finn's presidency in 2013-14 is to explore ways of enhancing what the ATS does for patients and their families. The ATS has redoubled its efforts to forge new alliances with patients and patient advocacy

organizations at the national and grassroots levels on the issues of disease awareness, public education, and advocacy. The ATS has also opened up its membership criteria—now anyone, including a patient, is able to join and participate in the activities of the Society. The ATS continues its commitment to funding cutting edge research through the many grants awarded to deserving investigators by the ATS Foundation Research Program in partnership with PAR. This booklet is another manifestation of these efforts to strengthen the relationship between patients, their families, and the ATS.

We salute the ATS Public Advisory Roundtable as well as the patients who have given talks at the ATS International Conference that have inspired us and made this booklet possible. We hope that this booklet will be valuable to clinicians who are seeking the patient perspective and to other patients and their families. The ATS will continue its firm commitment to working with patients and its PAR members on advocacy, research, and educational issues. We look forward to continued inclusion of the patient perspective in the work of the Society as we progress toward cures for many lung and airway diseases.



Patricia W. Finn
Patricia W. Finn, MD
ATS President 2013-14



Stephen C. Crane
Stephen C. Crane, PhD, MPH
ATS Executive Director

It started out as a realization that though patients are central to what pulmonologists and other medical professionals do, there needed to be a way in which patients and their advocates could interact directly with the Society. There needed to be an effective way to communicate patient needs and a way for physicians to understand their perspective; thus the American Thoracic Society Public Advisory Roundtable (ATS PAR) was born.

Many of the founding members of ATS PAR were patients, family members, and advocates who understood the real needs of patients and the lung diseases from which they suffered. The original ATS PAR members articulated those needs and communicated them to the ATS, and bridged the gap between patients and physicians. The implementation of PAR not only created opportunities to strengthen medical care, but also opened the door to collaboration and partnership to increase understanding of lung diseases and to lead efforts to fund treatments and cures.

Now, more than ten years later, ATS PAR is still one of the only patient-represented committees within a medical membership association. ATS PAR remains a vital part of the ATS and is held in high esteem within the organization. ATS PAR is known for its unique ability to respond to patient needs and mobilize efforts to improve patient care, increase research efforts in lung disease, and build advocacy and awareness of lung disease and lung health on a national level.

Additionally, to date, PAR-affiliated member organizations have supported the ATS Foundation research grant program with more than \$6 million in funding for innovative medical research in lung disease. The Chair of ATS PAR is a standing member of the ATS Board of Directors, with a direct line of communication with ATS leadership.

For the past several years, ATS PAR has had the privilege of assigning patient speakers to medical sessions at the ATS International Conference, a scientific meeting of respiratory professionals including clinicians and researchers. The patients share their personal stories with ATS members, giving them an up close and personal look into the lives and experiences of patients with lung disease. These compelling patient stories provide the important and central “patient voice” for the conference attendees and allow research and innovation to move forward with passion while never losing sight of patients. Never before has a medical association elevated patients to such a visible position within its organization. ATS PAR is truly “the patient voice of the ATS.”



Regina Vidaver
Regina Vidaver, PhD
Chair, ATS PAR



Larry Hoffman

Alpha-1 Antitrypsin Deficiency

“I had the normal “why me” thoughts. But I look back at that now as a blessing since I finally knew what was causing these symptoms.”

When I was born with jaundice, they did not know how to treat it properly. Five others in my pediatric unit died, but somehow I pulled through. I now know that other alpha-1 antitrypsin patients were born with liver issues as well. All that was 62 years ago, and since then, I have been affected by breathlessness in many ways.

In my younger days, I was last in running races, wrestling, and football. I was told it was because I was out of shape or too chubby. I would be constantly out of breath whenever I had to do something physical. I developed a way to hide these breathless moments by yawning.

I married my wife 38 years ago, and we had a son and daughter. During these years I had two careers. I spent 17 years driving a semi-trailer, delivering and picking up construction materials, including coal tar pitch, fiberglass, and hot asphalt. I was also exposed to constant diesel fumes. Then I was self employed as a remodeling contractor, which exposed me to lead paints, old plaster dust, roofing materials, drywall, and fiberglass. I wore protection over the years, which was to some benefit, but I was still exposed to many hazards, and I even smoked cigarettes for a period of time.

That exposure likely contributed to my shortness of breath and diminished lung capacity. Some days I found myself gasping for air. Sometimes, I felt like I was breathing through a cocktail straw. Had I known about Alpha 1 early on, I would have made some lifestyle changes.

Once I hit 50, things got worse. Daily activities began to get harder to accomplish with winter bronchitis bouts. I was always running out of energy and getting short of breath. In the winter of 2009, I was on a hunting trip in Nebraska and ended up so

ALPHA-1 ANTITRYPSIN DEFICIENCY

Alpha-1 antitrypsin deficiency is an inherited form of emphysema. Patients with the disease do not have enough of a protein called alpha-1 antitrypsin (AAT) in their blood. This protein is made in the liver, and it protects the lungs so they can work normally. Without enough AAT, the lungs can become damaged by emphysema. It also can cause liver damage. There is no cure for Alpha-1 antitrypsin deficiency.

Testing is recommended for those who have:

- Obstructive lung disease.
- Family history of alpha-1.
- Early-onset emphysema (younger than 45 years old).
- Emphysema without an obvious risk factor.
- Emphysema that is worse at the bottom of the lungs.
- Chronic asthma (in adolescents and adults).
- Recurrent pneumonia or bronchitis.
- Unexplained liver disease.

Learn more: ATS Patient Education Series. "What is Alpha-1 Antitrypsin Deficiency?" New York, NY. 2006. patients.thoracic.org

sick and short of breath that I had to be assisted in walking. A respiratory specialist diagnosed me with alpha-1 at 58 years old. I had the normal “why me” thoughts. But I look back at that now as a blessing since I finally knew what was causing these symptoms.

Winters have become harder, as I’m becoming more affected by the cold air. I must keep my face covered outside when the temperature is low. I also have to bring up phlegm in the morning along with a deafening cough which is not a favorite household sound. I can pretty much walk comfortably on flat ground. But climbing stairs, hills, and quick starts are hard. I’m on disability, and I felt like I didn’t deserve the help in the beginning. But I can no longer do a lot of the remodeling tasks.

At certain times, I need my wife to help me do things I never would have asked years ago. At first, this gave me thoughts of being less, but I know that she is the best caregiver for me. My family is also aware of the extra help I need, and we work together. I am learning to accept my limits.

Larry Hoffman was a patient speaker at the ATS 2013 International Conference in Philadelphia.



Addison Kelly

CHILDHOOD ILD

By Mary Kelly, Addison's mother

“The nurses shared other families’ similar experiences, which helped to guide me in this non-typical life.”

When I was 26 weeks pregnant with my second child, Addison, my husband and I were told our baby had a mediastinal teratoma and was in severe fetal hydrops. She (and I) underwent fetal surgery at the Children’s Hospital of Philadelphia Center for Fetal Diagnosis and Treatment. The tumor was removed, but Addison was born at 27 weeks and intubated. She spent nine months in the NICU, received treatment for kidney failure, sepsis, and necrotizing enterocolitis, to name a few. She also went through surgeries such as bilateral diaphragm plication, tracheostomy, and gtube placement. One of her doctors told us she was the sickest baby in the NICU.

Addison’s physician remained positive and realistic, and all of the doctors treated my husband and me as part of the team. They included us when talking about their plan and helped us appreciate the small gains Addison made—one hour, day, and month at a time.

When Addison was four months old, failed extubation and further testing revealed both sides of her diaphragm were paralyzed. One phrenic nerve was severed and the other was damaged by scar tissue. I will never forget the day I was told that my daughter needed a tracheostomy for long-term ventilatory support. I cried. How was Addison going to play with her brother, friends, or simply just be a little kid? The CHOP nurses shared other families’ similar experiences, which helped to guide me in this non-typical life.

With a miniature NICU in your home, your world becomes very small. As supportive as my community was, I still felt isolated and lonely with a technology dependent child at home. Simple tasks like a trip to the grocery store became overly

CHILDHOOD INTERSTITIAL LUNG DISEASE (chILD)

The word “interstitial” refers to the tissues that surround the air sacs (alveoli) in the lung and airways (breathing tubes). Interstitial diseases can make lung function and oxygen levels low. Childhood Interstitial Lung Disease (chILD) is a group of rare lung diseases found in infants, children and teens. Adults can have ILD too, but their diseases differ from children’s.

Types of chILD have included:

- Bronchiolitis Obliterans
- Chronic Bronchiolitis
- Connective tissue associated lung disease
- Cryptogenic Organizing Pneumonia (COP)
- Alveolar Capillary Dysplasia
- Hypersensitivity Pneumonitis
- Lung growth abnormalities
- Neuroendocrine Hyperplasia of Infancy (NEHI) or Persistent Tachypnea of Infancy
- Pulmonary Interstitial Glycogenosis (PIG)
- Surfactant dysfunction mutations

Learn more: ATS Patient Education Series. “What is Interstitial Lung Disease in Children?” New York, NY. 2010. patients.thoracic.org

burdensome. And then there was trying to maintain some sort of normalcy for our son, who was two when Addison came home from the hospital. We did not want him to be afraid of his sister, but we needed him to understand that he could not pull on her trach, gtube, or vent tubing. We had an accidental decannulation at one point, when he used her vent tubing as a leash! But I loved the fact that they were laughing and playing together just like any other siblings would. We were being “normal.”

When Addison was learning to walk, my husband built a stand for the ventilator on wheels. Eventually, Addison started pushing it herself, which gave her freedom and independence. Also, we hired a physical therapist who helped start a ballet class for children with special needs, a speech therapist who was one of very few to also offer feeding therapy, and an occupational therapist. All were key in her development.

When Addison was nearly two years old, we slowly started weaning her from the ventilator, and she got to the point when she only needed support at night. Our life has simplified, as we are now down to the Bipap machine and pulse ox. Thank you to all of the teams who gave us the confidence to establish our own “normal,” which has helped us raise an energetic, smart, and thriving 6-year-old girl!

Mary Kelly spoke at ATS 2013 International Conference in Philadelphia.



Patsy Menig

CHRONIC OBSTRUCTIVE
PULMONARY DISORDER
(COPD)

“Those of us with COPD either make adjustments to our lifestyle or lose our quality of life.”

I'm 64, and it's important that you understand that when I was growing up, the women in cigarette commercials were beautiful. One floated down the river in a canoe with a floral dress, handsome man by her side, waterfall in the background, and smoked like it was the best thing in the world. I couldn't wait to be old enough to smoke! My addiction to cigarettes began at the age of 18.

In 2000, I was diagnosed with chronic obstructive pulmonary disorder (COPD), which I attribute to many years of smoking cigarettes.

I have been married to my husband, Joe, for 41 years, and I am the proud mother of three daughters, one son, and three granddaughters. I'm an avid gardener, love to be outside, and enjoy nature very much. But, gradually, I noticed it was difficult to lug a wheelbarrow full of mulch or replant perennials. Thankfully, I had children who were eager to help with these chores so I was still able to enjoy my gardens.

Those of us with COPD either make adjustments to our lifestyle or lose our quality of life. So, container plantings became my main focus. I could come home from the nursery, sit on a bench, and create beautiful combinations of plants for the patio.

Following a hospital stay for pneumonia in 2004, my family doctor referred me to Abington Hospital in Abington, Pennsylvania. He reviewed my CAT scans and pulmonary function test results with me and mentioned a colleague of his at Temple University Hospital who was considering candidates for Lung Volume Reduction Surgery (LVRS). I researched the procedure online, and it seemed a bit frightening—but so was not being able to breathe. I decided to take the chance.

CHRONIC OBSTRUCTIVE PULMONARY DISEASE (COPD)

Chronic obstructive pulmonary disease (COPD) is an umbrella term for conditions, including chronic bronchitis and emphysema, which impede the flow of air in the bronchi and trachea.

- COPD is caused by a mixture of chronic inflammation causing narrowing of the small airways (i.e., obstructive bronchiolitis) and destruction of the walls of the air sacs of the lungs. This causes hyperinflation in the lungs and difficulty exhaling and inhaling (i.e., emphysema). The relative contribution of this process varies among individuals.
- COPD is increasingly associated with other diseases, such as pneumonia, hypertension, heart failure, forms of heart disease, lung vascular disease, cancer, osteoporosis, and depression.

Learn more: American Thoracic Society. Breathing in America: Diseases, Progress, and Hope. New York, NY. 2010. thoracic.org/education/breathing-in-america.

I met with the thoracic surgeon who drew a picture of the lungs before and after the surgery to make sure I understood the process, which I found helpful. As they say, a picture is worth a thousand words!

My LVRS took place in late November 2005, and I was able to return to work in April 2006 following three months in a pulmonary rehab program, in which I'd exercise on my own. I found my breathing became much easier than before the surgery. I have done two things of which I'm proud and that would not have been possible prior to surgery: I climbed to the top of Barnegat Light Lighthouse in New Jersey with my granddaughter Aubrey on Valentine's Day 2007, and I nearly hiked to the top of Hawk Mountain in Pennsylvania with my daughter Alison and her husband in 2009.

I just finished my second year in a clinical trial for a new drug. I'm pleased to make this small contribution for future patients who have COPD as well as the physicians who treat them.

Patsy Menig was a patient speaker at the ATS 2013 International Conference in Philadelphia.



Vijai Sharma

CHRONIC OBSTRUCTIVE
PULMONARY DISORDER
(COPD)

“With the love and prayer in my heart, here is my message to all my COPD friends: regardless of the severity of your COPD, never give up!”

I was diagnosed with emphysema in 1994 at age 53. Based on the pulmonary function test readings, my pulmonologist said that my lungs looked as though they were those of a 76-year-old man. It was quite a come down for someone who was used to hearing, “You look like you are not over 40,” to be told, “Your lungs look like you are 76.” My doctor also told me that emphysema is a horrible, progressive, and irreversible disease. It was a diagnostic shock all right!

My usual optimistic attitude surrendered to pessimism. Even though I was a clinical psychologist and helped others with emotional troubles, in the first 12 to 18 months, I experienced high emotional stress that often bordered on clinical anxiety and depression. My use of medication escalated. I went from taking two to three puffs twice a day from each Atrovent and Albuterol inhaler, to taking these puffs four times a day or more if needed. Just moving about the house would render me breathless. Exercise of even mild intensity seemed out of question.

One Sunday afternoon, two longtime friends dropped by my home. They were husband and wife, both respiratory therapists. They asked me to consider lung rehabilitation. However, there was no pulmonary rehabilitation program anywhere nearby. So, I decided to create my own rehabilitation program.

I slowly increased the duration of my walking, and selected specific exercises to strengthen my arms, legs, abdomen, and chest. I focused on making my diaphragm and ribcage more flexible and strong. I began to do yoga postures and breathing exercises. I lifted light weights, and I joined a gym. It was a tough fight to get a handle on shortness of breath, excessive fatigue, and the psychological demons of worries, fears, and hopelessness.

Vijai Sharma

CHRONIC OBSTRUCTIVE PULMONARY DISEASE (COPD)

The most important risk factor for COPD in the United States is cigarette smoking. Other factors, including occupational or environmental exposures to dusts, gases, vapors, biomass smoke, malnutrition, early life infections, recurrent respiratory infections, genetic predisposition, increased airways responsiveness, and asthma may be important in many individuals.

- The best known genetic risk factor for COPD is alpha-1 antitrypsin deficiency. Alpha-1 antitrypsin is a special protein that protects the lungs from enzymes known as proteases.
- COPD is diagnosed using a medical device called a spirometer, which measures air volume and flow, the main components of common clinical breathing tests.

Learn more: American Thoracic Society. Breathing in America: Diseases, Progress, and Hope. New York, NY. 2010. thoracic.org/education/breathing-in-america

When I first walked on the treadmill, I couldn't last longer than three minutes. So, my goal became breaking the three minute barrier. I learned to pace the treadmill speed with my breathing. I also learned to maintain abdominal breathing with active exhalation during the entire time of the exercise. Today—on a good day—I can stay on treadmill for 30 to 45 minutes!

Sticking doggedly to my personal rehabilitation program for the past 15 years has been the best thing I have ever done for myself. Yogic postures and breathing techniques and a variety of other exercises have helped me a great deal in maintaining my lung function, increasing my exercise capacity, and managing my stress level.

With the love and prayer in my heart, here is my message to all my COPD friends: regardless of the severity of your COPD, never give up! Continue trying to improve the capacity you have. You may not see a change in your spirometry numbers, but you can learn to breathe more efficiently and correctly. You can hope to be able to do more with the breathing capacity you have!

Vijai Sharma, PhD, was a patient speaker at the ATS 2012 International Conference in San Francisco.



Lynn Markwell

HYPERSENSITIVITY
PNEUMONITIS

“I entered into a pulmonary rehabilitation program later that year. My life completely changed. I was surrounded by other people with lung diseases—I was no longer isolated!”

In August 2003, I noticed I was short of breath and sometimes had a dry, hacking cough during my five-mile walks. By the next year, the symptoms worsened, and I'd have to bend over to cough and catch my breath while my heart raced. Afterwards, I would collapse into bed for a half hour then drag myself to work.

I was the head of a highly regarded classical music program at a school, which included music education as part of their core curriculum, and I stayed in my office more than usual. I couldn't climb the ninety stairs that I used to take at a full run. My stamina was greatly reduced and, while producing programs, I sweated a lot if they involved physical work. It was embarrassing.

My coughing fits became so uncomfortable for my walking partner that she refused to walk with me until I saw a doctor. I was diagnosed with exercise induced asthma, prescribed medication, and sent on my way. It didn't help, and I grew weaker over the summer. I couldn't get enough air into my lungs. I couldn't inhale. I couldn't yawn deeply. I was exhausted and spending a lot of time on the couch. I was panting in my sleep.

One parent at my school was also my endocrinologist at University of California, San Francisco. I was referred to the chest clinic and within minutes, I was diagnosed as having interstitial lung disease. My first DLCO upon arrival was 7.7. In January 2005, I had a VAT lung biopsy, which revealed the culprit: hypersensitivity pneumonitis.

Later that year, I was declared disabled. I was 52 years old, and it was a shock to leave my job—my six-days-a-week, 14-hour-days job—with no warning. I was at my peak earnings in a position that I loved, and suddenly it all stopped. My husband of

HYPERSENSITIVITY PNEUMONITIS (HP)

This interstitial lung disease is caused by an immune response to an inhaled antigen of organic material from bacterial, fungal, plant, or animal proteins. Initial exposure results in sensitization in which the body forms antibodies to these antigens. Repeated exposure results in inflammation. If the exposure is not halted, it can permanently damage the lung.

- HP can be categorized as acute, subacute, and chronic.
- Some individuals may have a genetic predisposition.
- Treatment consists of removing the source of the exposure and eradicating any residual antigens to prevent re-exposure. Systemic steroids are often used if the individual is severely ill or removal of the trigger is insufficient.
- Those at high risk include farmers, bird hobbyists, sandblasters, miners, tunnelers, millers, and potters.

Learn more: American Thoracic Society. Breathing in America: Diseases, Progress, and Hope. New York, NY. 2010. thoracic.org/education/breathing-in-america

40 years and I survived because we always lived below our means, had no debt, and money in the bank. We adjusted our lifestyle to a single income. I was thankful that our son was away at college and didn't have to deal with the illness on a day-to-day basis. This disease tears families apart, but we are committed to each other to the very end.

I entered into a pulmonary rehabilitation program later that year. My life completely changed. I was surrounded by other people with lung diseases—I was no longer isolated! I had social and emotional support all while developing muscles. I learned so much, including energy conservation, anxiety management, and pursed-lipped and diaphragmatic breathing.

For the past eight years, I have worked out five days a week in hospital rehabs and in my garden on Saturdays. I now practice power yoga every week, and I believe this long-term and consistent workout routine has delayed the need for lung transplants.

My DLCO has fluctuated, but I'm happy to report that after a six-week run of high dosages of prednisone, my unadjusted DLCO was 15.36. That's the life of living with an interstitial lung disease.

Lynn Markwell was a patient speaker at the ATS 2012 International Conference in San Francisco.



Connell Rodden

IDOPATHIC PULMONARY
FIBROSIS

“When I was first diagnosed, I felt like time was on our side. Unfortunately, time is running out for me and many others who wait (not always patiently) for new lungs.”

In January 2008, I was diagnosed with idiopathic pulmonary fibrosis. My nurse practitioner heard “crackles” in my lungs while I was being examined. The first thought was pneumonia. She sent me to have a chest x-ray, and later she called to say that I had better see a pulmonologist as she suspected something much worse. IPF is a chronic, progressive disease that usually results in an average survival rate of three to five years. To know that you have a terminal disease with no cure or treatment is devastating.

Because there is no cure or treatment for IPF, the only option is lung transplantation, and I was recently listed for transplant by my doctor at Temple University Hospital who I have been with for five years. I participated in several studies and clinical trials through Temple Health to explore both the effectiveness of trial medication and to track the progression of the disease. I would do most anything to assist in finding a cure or treatment for IPF. I certainly appreciate the efforts of all the doctors, scientists, and medical personnel and hope someday they will find a treatment and eventually a cure.

Initially, I was able to function relatively well—resting more than usual—but I was able to accomplish normal tasks and hobbies. As the disease and my symptoms worsened, I had to give up many hobbies and interests. I could no longer hike the trails to go hunting with my buddies. Fishing became a chore, as I no longer had the strength, breath, and endurance to reel in the big ones.

These days, I can’t I swim and snorkel with my youngest daughter (one of our favorite vacation activities), and sightseeing and most outdoor hobbies have become

IDIOPATHIC PULMONARY FIBROSIS

Pulmonary fibrosis (PF) describes a group of lung diseases in which thickening of the walls of the air sacs (called alveoli) caused by scarring (fibrosis). Scarring in alveoli prevents oxygen from passing into blood vessels. This can result in coughing, shortness of breath, fatigue, and low blood oxygen levels. The scarring also makes the lungs “stiff” and difficult to inflate, which means they hold less air than normal lungs.

- The diagnosis is idiopathic pulmonary fibrosis (IPF) when the cause of the PF is unknown.
- The amount of scarring can increase with time, making the lung even stiffer, further limiting its filling capacity, and limiting the ability of oxygen to pass through air sac walls.

Learn more: ATS Patient Education Series. “Idiopathic Pulmonary Fibrosis (IPF)” New York, NY. 2011. patients.thoracic.org

a thing of the past. I need oxygen 24/7. Even going to dinner with the family and playing with my seven-year-old grandson is no longer easy, as I experience shortness of breath. It is a horrible and frightening experience not being able to breathe.

When I was first diagnosed, I felt like time was on our side. The feeling was, “Five years is a long time, anything can happen.” I thought, “They are finding cures for many diseases—why not IPF?” Unfortunately, time is running out for me and many others who wait (not always patiently) for new lungs.

Connell Rodden was a patient speaker at the ATS 2013 International Conference in Philadelphia.



Elayne Klein

LUNG CANCER

“When my eyes filled with tears, I realized that he had given me my first chance to verbalize and focus on the emotional and physical strain of having lung cancer.”

My journey with lung cancer began seven years ago when I caught the flu from my four-year-old granddaughter during an epidemic in California. I underwent radiological tests, and although I had already recovered from the cough, the repeat CT scans showed that a 1.7 centimeter mass was in my lung. A biopsy was performed showing non-small cell adenocarcinoma. I thought they had mistaken me for another patient. Then I curled into a ball in confusion and fear.

My thoracic surgeon performed a right thoracotomy with lower lobectomy and a mediastinal lymph node dissection. He soon informed me that the cancer had spread to my lymph nodes and therefore I had stage 3A lung cancer. I looked wide eyed at my husband and asked, “Was I just given a death sentence?” I immediately thought about my youngest daughter’s future wedding. I wanted to be there. I wanted to be there to see her children born. I wasn’t finished living yet! I was angry.

Soon after, I met with my oncologist who gave me the option of choosing between two chemo treatments. I chose the slightly more effective one which meant that I would lose my hair. She also explained the frightening survival statistics. Fortunately, the follow-up appointment with my pulmonary doctor helped me look at the diagnosis more positively. “Forget the bad statistics,” he said. “You could be in the percentage of people who survive. We will help you get through this.” He simply spent time with me. He asked me, “How are you doing?” When my eyes filled with tears, I realized that he had given me my first chance to verbalize and focus on the emotional and physical strain of having lung cancer.

LUNG CANCER

Lung cancer is an abnormal, uncontrolled growth of cells within the lung. It’s the leading cause of cancer deaths in the United States, and it will cause more deaths this year than cancer of the breast, prostate, colon, liver, or kidney and melanoma combined.

- Lung cancers are generally divided into two major types, small cell lung cancer and non-small cell lung cancer. Small cell lung cancer tends to grow more quickly than non-small cell lung cancer.
- Causes of lung cancer include:
 - Smoking
 - Second-hand smoke
 - Radon or asbestos
 - Genetic factors
 - Air pollution

Learn more: ATS Patient Education Series. “What is Lung Cancer?” New York, NY. 2008. patients.thoracic.org

Later, I started 33 cycles of radiation. I had decided to empower myself by experimenting with green protein, energy drinks. I listened to meditation CDs, and I went for Reiki healing sessions. Fortunately, I found the National Lung Cancer Partnership, and their wonderful website of encouraging Stories of Strength written by survivors of all stages of lung cancer. The NLCP also provides funding for lung cancer research and offers a ton of helpful information. It was important for me to make lists about what I wanted to accomplish, like having fun (imagine that!) and walking along the Delaware River with my faithful Labrador retriever. My wish right now for all cancer patients is for a comprehensive post treatment plan to help us all adjust to our lives in the best possible way.

At present, I have a metastasis in my opposite lung, considered indolent. On a positive note, my original tumor was found to be EML4-ALK translocation—that's a good thing. My doctor and I have chosen not to treat the nodules or get a biopsy unless I develop more symptoms or there is a truly meaningful treatment down the road that will give me an extended quality of life.

When we received news of the cancer recurrence, my husband and I made the decision to follow our dream to be near our children, and so we moved across the country to California. I am now affiliated with Stanford Medical Center, and my hope is that there will soon be research that will help me stay alive.

Elayne Klein was a patient speaker at the ATS 2013 International Conference in Philadelphia.



Sara Whitlock

LUNG CANCER

“I lost my job, my hair, and perhaps worst of all, my ability to be much of what I thought defined me as a person—my ability to be a wife to my husband or a mother to my children.”

In October 2010, I was diagnosed with Stage 4 Lung Cancer.

We sat with the medical and radiation oncologists as they laid out the extent of my disease and our treatment options. I'm not sure how much I really heard during those first days. I mostly remember the drawing my radiation oncologist did with all the areas of cancer represented by red circles all through my chest. I remember telling my children. I can recall my family and friends' stricken faces when we told them the news. I also remember lying in bed in the early morning hours, and how my husband and I clutched each other and wept.

My family traveled to our home that first weekend and gathered around me, laid their hands on me and on one another, and we prayed for wisdom and for healing. I remember feeling my little brother's arms shake as he prayed for me and how his tears splashed onto my face and joined my own.

We decided to ignore the dismal statistics, and my team at the Wright-Patterson Air Force Base Medical Center and the Cleveland Clinic developed a very aggressive treatment approach. I began chemo on my 15th wedding anniversary. I lost my job, my hair, and perhaps worst of all, my ability to be much of what I thought defined me as a person—my ability to be a wife to my husband or a mother to my children. I went from running three miles several times a week to being unable to walk up a short flight of stairs.

We followed six cycles of chemo with six weeks of radiation to my chest. When a node in my abdomen seemed resistant to treatment, a biopsy revealed that it was the same cancer as in my lungs. We decided to start treatment with Alimta every 21 days, and the results of my PET scan showed no uptake anywhere, and the decision

Sara Whitlock

LUNG CANCER

Treatments for lung cancer include surgery, chemotherapy, radiation therapy, and targeted drug therapy. Tests for lung cancer include:

- Chest x-ray to identify “spots” that could be cancerous.
- Chest CT scan to identify and further characterize “spots” that could be cancerous.
- Positron emission tomography (PET) scan to characterize the metabolic activity of “spots” and to identify potentially cancerous cells in lymph nodes and other tissues.
- Sputum (or mucus from your lungs) tests to check for cancer cells.
- Bronchoscopy to obtain lower respiratory specimens to check for cancer cells in the lung(s).
- Lung biopsy to check for cancer cells in the lung(s).
- Thoracentesis (sampling fluid from the pleural space around the lung) to check for cancer cells in the space around the lung(s).

Learn more: ATS Patient Education Series. “What is Lung Cancer?” New York, NY. 2008. patients.thoracic.org

was made to radiate my abdomen for good measure. We continued the Alimta, and I scheduled another PET scan.

I remember the anxiety waiting for my oncologist to call with the results. I sat on the couch and planned my funeral. I couldn't help my mind from going in those dark places. And ten minutes later, the phone rang.

“Hello?”

“You remain in complete remission.”

“WHAT?!”

“I said that you remain in complete remission.”

At that point, words fail me. The tears start. “We'll talk more later,” he says gently, and hangs up.

My husband walks in and I fall into his arms with relief. He holds me up—like he always does—and we are so thankful. Later, we have our Lenten devotional, and my husband says a prayer that we will remember every day the miracle that has taken place in our own house.

So far, I've tested negative for all the genetic mutations discovered. But I am confident that your hard work will result in more discoveries and more treatment options for me. More time with my family. More time with my friends. More time—those are such sweet words.

Sara Whitlock was a patient speaker at the ATS 2012 International Conference in San Francisco.



Tina Silks

PULMONARY ARTERIAL
HYPERTENSION

“If it wasn’t for research, I would not be here today. We, however, need more research done.”

I was diagnosed with pulmonary arterial hypertension in February 1995. It was not an easy journey to diagnosis. I started getting ill at the age of 34. At first, my monthly cycle stopped, and then the fluid started coming on in my lower extremities and abdomen. I went to the emergency room. The doctor said I looked pregnant put a Doppler radar on my belly. He told me that the fast heartbeat was a baby. I was the mother of two boys—I did not feel pregnant. But he insisted and sent me home. After two-and-a-half months, I went back to the ER because I couldn't walk up a flight of stairs without sitting down a couple of times. I insisted they do blood work to prove I wasn't pregnant, and they finally agreed with me.

After four more months of painful testing, I was given an appointment with the medical specialties doctor. He sat me down and told me the outcome: I had PAH, a poor prognosis with maybe three years to live “if I was lucky.” The pressure in my lungs was around 130 millimeters.

In August 1995, I started taking Flolan and Lasix with Potassium Chloride, which was the only medication available to me at that time. I was so sick by then that I was living in a hospital bed in my living room and using a wheelchair. I stopped walking due to skeletal pain from the medication. I was unaware I could be seen by a pain management team.

Two years went by until I finally spoke up to my doctor. He prescribed a cocktail of medication to relieve most of the pain, and I started physical therapy. After a couple of years, I started feeling better and getting out of the house. My pressure was down to 38 mm. By this time more medications were available to PAH patients. He

PULMONARY HYPERTENSION

Pulmonary hypertension (PH) is high blood pressure in the arteries going to the lung. If it persists or becomes very high, the right ventricle of the heart, which supplies blood to the pulmonary arteries, is unable to pump effectively, and the person experiences symptoms that include shortness of breath, loss of energy, and edema, which is a sign of right heart failure.

- PH is a general term that means that the blood pressure on the right side of your heart is too high, but does not explain why it is high. Causes include chronic lung disease (e.g., chronic obstructive pulmonary disease, interstitial lung disease, etc.), heart disease, and blood clots in the lung.
- Pulmonary arterial hypertension (PAH), is a specific type of PH that occurs when the blood vessels in the lung are directly diseased and become thick and narrow.

Learn more:

ATS Patient Education Series. “Pulmonary Arterial Hypertension” New York, NY. 2013. patients.thoracic.org

American Thoracic Society. Breathing in America: Diseases, Progress, and Hope. New York, NY. 2010. thoracic.org/education/breathing-in-america

wanted to start titrating the Flolan down and put me on Tracleer. It went smooth and I transitioned with no problems.

I then went into a double-blind study for Tyvaso. It was FDA approved and added to my medication list, along with Adcirca. My pressures are maintaining at 45 millimeters, and I'm very active. I even started a support group in my area, and I am a peer network mentor. If it wasn't for research, I would not be here today. We, however, need more research done.

Too many patients are losing their lives to PAH. It has to stop. We need more options! Please help us fight for our lives and the possibility of a cure.

Tina Silks was a patient speaker at the ATS 2012 International Conference in San Francisco.

Previous Voices

Patients and speakers featured in the first edition of *ATS Patient Voices*.

“On my first day with my prostheses, I only walked 10 feet. However, the next day I walked 168 feet, then 468 feet and I continued to walk farther each day.”

–Jennifer Ludwin
Sepsis

“It’s been hard for me to accept. Yet, I have reason to hope. The puzzle pieces for my diseases are coming together.”

–Nicole Seefeldt
LAM and Tuberos Sclerosis Complex

“My pulmonologist is very active in our support group. It has opened his eyes and changed the way he handles his patients.”

–Rodney K. Reese
Sarcoidosis

“I was very fortunate to travel through life with a twin, and we are tremendously grateful to be alive and have never-ending gratitude to our organ donors.”

–Isabel Stenzel Byrnes
Cystic Fibrosis

“I cherish my life now more than ever before, and I wouldn’t trade it in for a ‘normal’ life for all the money in the world.”

–Len Geiger
Chronic Obstructive
Pulmonary Disease

Previous Voices

Patients and speakers featured in the first edition of *ATS Patient Voices*.

“Doctors urged my family to consider removing me from the ventilator. It was a ‘quality of life issue’ since I would likely ‘never breathe on my own again’ if I lived.”

–*Eileen Rubin*
Acute Respiratory Distress Syndrome

“As my disease progressed, requirements increased as did the complications.”

–*Beth Mittelstadt*
Pulmonary Fibrosis

“Thanks to the PH community, I’ve survived a 1.2-mile gauntlet swim, a 56-mile bike ride in 90 degree heat, a 13.1-mile trail jog, and all 70.3 miles in the Orlando, Florida, IronMan contest.”

–*Robert Ngo*
Pulmonary Hypertension

“Kids of all ages notice that Nora is wearing oxygen. They pretty quickly realize that they have to watch out for the tube, and often the older kids will try to keep it from getting stuck on anything.”

–*Claire A. McCormack, Nora’s mother*
Pulmonary Hypertension

“It’s not just about the cure—it’s about improving the quality of life for patients until there is a cure.”

–*Ashley Holley*
Sickle Cell

Visit <http://patients.thoracic.org/par/patient-voices/index.php>.

Previous Voices

Patients and speakers featured in the first edition of *ATS Patient Voices*.

“During treatment, I kept working. I was teaching undergraduates and trying to run as much as possible. I was not going to take this lying down.”

–*Maki Inada*
Lung Cancer

“I made the nurse take a picture of me taking my first pill. For me, it was a grand moment. It was the moment I went from a gloomy certainty about what my future held, to a blissful land of the unknown.”

–*Heather Kirkwood*
Hermansky-Pudlak Syndrome

“Most people breathe these bacteria and fungi in and simply breathe or cough them back out, but they were making a home in my lungs.”

–*Geoff Burkhart*
Nontuberculous
Mycobacteria

“I began to dread going to bed because I knew I’d have to be strapped up to the CPAP”

–*Peter Helm*
Obstructive Sleep Apnea

“Going back to work that fall was a nightmare. I began to get sick almost at once. The same tightness, wheezing, asthma attacks increased and I wasn’t able to take care of my son’s needs because I was so sick.”

–*Laura Steves*
Work-Exacerbated Asthma

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ATS Patient Voices 2



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A publication from the American Thoracic Society Public Advisory Roundtable.