



ATS Patient Voices 5

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, and 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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“Medicine is learned by the bedside and not in the classroom.” –Sir William Osler

Lessons learned from patients can leave impressions as lasting as those of mentors and teachers. The patient perspective is paramount to our work at the American Thoracic Society. Since 2001, the ATS Public Advisory Roundtable (PAR), representing the patient voice of the Society, has helped to bring patients and families to the forefront of all programming and activities.

This focus is perhaps most evident at the annual ATS International Conference where PAR facilitates patient programs, such as the Meet-the-Expert patient and family forum, PAR Symposium, and several dozen scientific sessions that feature patient speakers, creating a unique communication experience for all the attendees.

Now in its fifth edition, Patient Voices highlights the stories of patients who have presented at past ATS International Conferences. You will hear stories of pulmonary disorders such as COPD, lung cancer, pulmonary hypertension, hypersensitivity pneumonitis, alpha-1 antitrypsin disease, idiopathic pulmonary fibrosis, and others.

Many patients have struggled all their lives with disease, and oftentimes they experience life-threatening conditions. Their stories confront great pain, fear, and grief. Through the vulnerability of these narratives we are able to shed light on pulmonary diseases and help inspire the many millions of people grappling with lung disease all over the world.

Together with patients and their families we will seek cures, and the opportunities for discovery have never been greater. We have the tools to unlock the mysteries of lung disease and advance pulmonary health through innovative research, clinical care, advocacy, education, and training.

Today’s and tomorrow’s leaders will translate respiratory discoveries and will do so in collaboration with patients. Now everyone, including a patient, is able to participate in the activities of the Society and join the ATS as a member.

Our commitment to research, in particular to deserving early career investigators, continues to grow through the ATS Foundation Research Program, in partnership with PAR.

We look forward to continued inclusion of the patient and family perspective in our work and hope that this booklet is valuable to clinicians seeking their viewpoints. More importantly, we hope that the booklet empowers patients and their stories to be told.

We sincerely appreciate the efforts of the ATS Public Advisory Roundtable, as well as the voices whose courage and wisdom have made this fifth edition possible.



A stylized, handwritten signature of David Gozal in black ink.

David Gozal MD, MBA
ATS President
2016-2017



A handwritten signature of Stephen C. Crane in black ink.

Stephen C. Crane, PhD, MPH
ATS Executive Director

Introduction

I write to you as a caregiver to my dear sister, Alex, who was diagnosed with pulmonary hypertension, and as the chair of the American Thoracic Society Public Advisory Roundtable (ATS PAR), which is solely dedicated to the integration of the patient perspective into clinical care.

ATS PAR is one of the only patient-centered groups in the United States that is a direct part of a medical membership association. It is known for its unique ability to respond to patient feedback and mobilize stakeholders to improve patient care. ATS PAR bridges the gap between patients and clinicians to create opportunities that open doors to collaboration and partnerships, and increase our understanding of lung diseases.

We build advocacy and raise awareness of respiratory health on a national level. To date, ATS PAR–affiliated member organizations have supported the ATS Foundation with more than \$5 million in research funding. ATS PAR leverages the power of the patient voice in our collective race for innovative treatments, therapies, and cures.

Thanks to the ATS PAR, patients share their disease-related experiences at annual ATS International Conferences, delivering the power of the patient voice to thousands of pulmonary, critical care, and sleep medicine researchers. More than a dozen patients attended ATS 2016 in San Francisco, California, to speak about their reality of living with lung disease.

The pages that follow are the firsthand accounts of patients who bravely shared their stories to large medical audiences to put a “face” to these diseases—to show the “heart and soul” behind the lungs and the conditions. In these intimate stories of resilience, may we find hope and strength.



Debbie Drell
Chair, ATS PAR

Sandra Rock

IDIOPATHIC
PULMONARY FIBROSIS



“I can’t dance with Jim without using oxygen, so I put it in a backpack and strap it on my back. Swing dancing is a little tough!”



I would like you to try something for me. Think of five of your friends or family members. Now imagine all of them passing away in the same year. Would you be heartbroken? This is what it's been like for me for the last 15 years, except I lose approximately 10 support group members a year to a devastating disease that has no cure.

Idiopathic pulmonary fibrosis robs you of the ability to transport oxygen so you lose energy, stamina and the ability to do everyday activities. I used to work, shop, travel, swim, maintain my house, do laundry, but now I can't work, and the rest I do on a very limited basis.

In pulmonary rehabilitation, I learned how to climb stairs using proper breathing technique, how to breathe and bend over to empty the dishwasher or dryer, and how to get in and out of bed or the car. These are all things you never think about when you can breathe normally.

I started using oxygen at night because my breathing was so slow I wasn't getting enough oxygen when I slept. It then transitioned to at night and during exercise. Now it's at night, during exercise, or any kind of exertion. Next it will be 24/7.

IPF affects the entire family. My daughters are constantly telling me to stop and rest. I can't run and play with my granddaughters. Jim, my loving husband of 45 years, has now become my caregiver. I can't dance with Jim without using oxygen, so I put it in a backpack and strap it on my back. Swing dancing is a little tough!

Darlene, my wonderful rehab therapist, introduced me to another IPF patient Kathy, and we started a support group in 2002. Unfortunately, Kathy passed away from her illness four months after we started the group.

Three years ago I had to give up the support group because of my health. I'm lucky, though, and for some reason I've lived longer than predicted. Most patients are expected to live three to five years after diagnosis.

Sandra Rock

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. thoracic.org/patients

When I ask other patients how IPF has changed their lives, answers include loss of income or loss of intimacy, inability to play sports, dance, do yard work, garden, go camping or traveling. Many people with IPF are also learning to live with oxygen and the fatigue that comes from a continuous cough. It's like your whole world being turned upside down.

One member whose husband has been on the transplant list for months says their strategy is not to plan ahead as they don't know what the future brings. They live each day as best they can, enjoying the simple pleasures in life. For me, it's a good meal or an evening with family and friends. Every extra day is precious.

Last year two medicines were approved by the Food & Drug Administration to help stop the progression of IPF. For many patients living with the disease and unable to take the drug or ineligible for a transplant, there's only anticipation of future research catching up to them, such as new medicines to stop the scarring and possibly even reverse it.

Trina Massey Davis

SARCOIDOSIS



*“Sarcoidosis may be a snowflake disease,
but living with it hits like an avalanche.
We need your help to dig us out.”*

My first flare occurred in my last trimester, in 1990. A cough led me to an ER visit, where I was given a prescription for cough medicine. Aside from the cough, I did not have any other symptoms that indicated a cold. I gave birth, and my cough subsided and soon after dissipated. Two years later, days after giving birth to my second child, I had a repeat episode. My journey began, initially with appointments for periodic shortness of breath. I was prescribed antibiotics and several different inhalers. The diagnosis? Bronchitis, and later asthma.

In 1997, my symptoms multiplied. Having done all he knew to do, my primary doctor referred me to a host of specialists—a hematologist, rheumatologist, ophthalmologist, neurologist. I was growing impatient. Then in February 1999 my health declined almost overnight. One morning I woke up to start my day. I got out of bed to walk, and everything around was dark. I couldn't breathe, there was a burning sensation. When I stepped outside, the cold winter air felt like it was attacking my lungs. I went back inside and found myself shuffling between the bed and the couch across the room.

I scheduled the first available appointment with a pulmonologist and saw one the next day. A scheduled bronchoscopy was performed, and results came back sarcoidosis, of the right upper lobe of the lung. During a flare I was prescribed medications. Their side effects included: weight gain, sleeplessness, change in temperament, full moon face, loss of hair, acne, cotton mouth, and a metallic taste anytime I ate or drank. Two years of a misdiagnosis had taken its toll, and the lack of coordinated communication with my many health care providers complicated the diagnosis.

In 2013 I woke up to an accelerated heart rate. I drove to the ER. Immediately my heart rate dropped below 50 beats per minute, and hospice was called. According to my EKG, my heart was being affected. In two years, I was seen by three different cardiologists.

SARCOIDOSIS



Sarcoidosis is an inflammatory disease that most commonly involves the lungs but can impact any organ in the body including the eyes, nervous system, liver and heart. For some patients, the disease is mild and for others it may be progressive and devastating. The cause of sarcoidosis remains unknown. However, experts suspect that sarcoidosis develops when a patient's immune system overreacts to some type of exposure—occupational, environmental or infectious—and small clumps of inflammatory cells called granulomas are formed and deposited in affected organs.

Learn more from ATS Public Advisory Roundtable member the Foundation for Sarcoidosis Research. stopsarcoidosis.org

In 2014 results from pulmonary function tests were persistently problematic and led me to take six weeks off from work. During this time, my HR department challenged what my specialist wrote on my medical leave documents, which stated “extrinsic, chronic severe asthma and sarcoidosis.” The date for my return to work was based on the outcome of my follow up appointment. I filed a complaint with Equal Employment Opportunity Commission and found myself battling sarcoidosis with a system that seemed to be working against me. Neither of the EEOC representatives assigned to my case had ever heard of the disease, and that added to my stress level. When I exhausted all my accrual leave, I had no choice but to be removed from the company payroll.

Every day I wake up exhausted. And with each episode I can't help but think, is this the one that is going to take my life?

There is a great need for research into the cause, cure, and treatment of sarcoidosis. Moreover, the medical profession needs better education so they can detect it earlier and diagnose it properly. Sarcoidosis may be a snowflake disease, but living with it hits like an avalanche. We need your help to dig us out.

Jacqueline Ploudre

SARCOIDOSIS



“People think you are faking illness or just enjoy complaining. What they really don’t realize is we are actually faking being well.”



Five years ago, while vacuuming I experienced a severe chest pain unlike anything I'd ever felt before. I instantly knew something was wrong. I saw my primary care physician the next morning, and after some tests that evening I received word that they had found granulomas, or nodules, in my lungs behind my thyroid and pancreas.

X-rays would suggest lymphoma or sarcoidosis. My first thought was, Oh my God I have cancer, and I'm going to die. Three months of tests and a lung biopsy later, it was confirmed that I had pulmonary sarcoidosis.

The disease hit me like a ton of bricks. For eight months my life consisted of work and bed. I had a chest pain that literally felt like cinder blocks were being dropped on my chest. A shortness of breath would only allow me to walk a short distance, and now that I have breathing issues, I have panic attacks when I'm in situations that compromise my space and air.

I became depressed and withdrew from my daily activities. I was put on anti-depressants and a steroid. The weight gain, hair loss, lack of sleep, and moodiness, wore me down.

As a wife and mother of two young boys I needed to take back my life. At the recommendation of the St. Louis City health commissioner, I met with a close friend who practiced holistic medicine. I got off the drugs and tried the holistic route. I also became gluten free, cut pork from my diet, increased fruits, vegetables and water intake, and I started walking. Finally, I was beginning to feel better and have not allowed myself to mentally go back down that road.

Regardless of how sick I feel, I remain upbeat.

Since I have sarcoidosis I always hear "You don't look sick." People think you are faking illness or just enjoy complaining. What they really don't realize is we are actually faking being well, and this takes real talent when deep down you really feel like hell.

Jacqueline Ploudre

Sarcoidosis is an inflammatory disease that most commonly involves the lungs but can impact any organ in the body including the eyes, nervous system, liver and heart. For some patients, the disease is mild and for others it may be progressive and devastating. The cause of sarcoidosis remains unknown. However, experts suspect that sarcoidosis develops when a patient's immune system overreacts to some type of exposure—occupational, environmental or infectious—and small clumps of inflammatory cells called granulomas are formed and deposited in affected organs.

Learn more from ATS Public Advisory Roundtable member the Foundation for Sarcoidosis Research. stopsarcoidosis.org

This disease needs to get on the radar. We need help, we need awareness, we need research, we need better medicines, we need easier conclusive testing. We need sarcoidosis clinics and doctors that understand the disease. We need a community fighting for cures with the same tenacity that we, as patients, lead our lives.

There are sarcoidosis warriors out there, and in spite of our illness we are fiercely working to make life more bearable for all who suffer from this disease.

Pamela Popp

SEPSIS/CRITICAL CARE



*“You relearn how to sign your name,
to understand your history, to remember
those that you love and who have loved
you. It all takes time.”*

The journey begins the day that you awake in the ICU. As you struggle to breathe, to take assessment of your body, your sense of self and of time—it becomes apparent that your life is forever changed. You can choose to succumb or you can take on this new journey, where your every move will be tested each and every day. The new path requires identifying your limitations and adapting. Finding resources is key, including physicians who understand what happens after the body reboots during septic shock. It takes the balance of 22 prescription medications, a wound vac, home administered IV medications, and nothing can remedy the chronic feeling of weakness, disorientation, and overwhelming loss.

My septic shock experience arose from a post-operative infection. Upon my return to the ED, providers managed endless symptoms with focused responses: a transfusion for the anemia, oxygen for the shortness of breath, a CT scan for the disorientation, a return to the OR in a search for the infection. While they addressed the symptoms, they missed the big picture: that I was quickly going into septic shock and would ultimately lose consciousness and begin organ failure. Eight days later I awoke in ICU to encounter physicians who were astounded at my survival, and together we began to learn what really happened.

Sepsis does not end when the patient leaves the hospital.

Six years later, I am fully functioning—I am working, can enjoy my hobbies, have rediscovered my passion for life. But during the six years I struggled with extensive memory loss, chronic fatigue, waves of disorientation, reactions to medication, and PTSD. It has taken a strategic physician to wean me from prescriptions, a nurturing psychologist to heal the emotional impact, and my own fierce determination to get this far.

SEPSIS



The word sepsis comes from the Greek meaning “decay” or “to putrefy.” In medical terms, sepsis is defined as either “the presence of pathogenic organisms or their toxins in the blood and tissues” or “the poisoned condition resulting from the presence of pathogens or their toxins as in septicemia.” Patients are given a diagnosis of sepsis when they develop clinical signs of infections or systemic inflammation; sepsis is not diagnosed based on the location of the infection or by the name of the causative microbe. Signs and symptoms include abnormalities of:

- Body temperature
- Heart rate
- Respiratory rate
- White blood cell count

Forty percent of patients diagnosed with severe sepsis do not survive. Until a cure for sepsis is found, early detection is the surest hope for survival.

Source: Sepsis Alliance. “Definition of Sepsis.” sepsisalliance.org

You don't forget the nightmares, the memory of struggling to breathe, the feeling of isolation, panic, and pain. You relearn how to sign your name, to understand your history, to remember those that you love and who have loved you. It all takes time.

Post-sepsis syndrome does not share the recognition of other catastrophic conditions. There are no ribbons, no fundraisers, no resources to address the psychological devastation, hair loss, memory loss, and financial strain of hundreds of thousands in medical bills.

Post-sepsis patients find themselves, and their families, struggling every day to understand what happened, why it happened, and most importantly, when things will get better. For a lot of patients, that last question can overtake them on their weakest of days, when the will to survive comes into jeopardy.

Sometimes the sense that nothing will return to normal, that an identity is lost, that they have failed somewhere in life and are now paying for it, is overwhelming. According to the CDC, there are 1.4 million sepsis survivors in the U.S.—all of whom trudge through the journey alone, looking for hope even as they stagger forward.

I am grateful that clarity has been provided in my own treatment, and I wish the same for fellow survivors.

Mary Kitlowski & Rebekah Giannakos

PRIMARY CILIARY DYSKINESIA



“My sister, Rebekah, and I both had constant runny noses and ear infections, and I had a persistent cough.”



My goal is to participate in a race in every state. In October 2014 I founded Running On Air to raise awareness of lung diseases, including the rare genetic disorder primary ciliary dyskinesia, which my sister and I have been diagnosed with since we were teens. With an FEV1 of 40 percent, I get through each race wearing a portable oxygen concentrator on my back.

The road to advocacy for PCD started at a young age. My sister, Rebekah, and I both had constant runny noses and ear infections, and I had a persistent cough. Our parents took us to see an otolaryngologist. We both had a series of surgeries to remove our tonsils and adenoids, and then had tubes inserted in our ears from the ages of 4-12. Rebekah developed pneumonia at the ages of three and nine. At the age of five I started seeing an allergist and receiving regular allergy shots. Rebekah soon followed with allergy shots at age 11. After rounds of tests, we were given the diagnosis of severe allergies and allergic rhinitis. But despite treatment, the coughing and congestion only seemed to get worse.

As a teenager I started having coughing fits at night. My mom would eventually wake me and make hot tea. At first these episodes were sporadic, every couple of months or so.

When I was 17 I started a part-time job working for my allergist. I was coughing so much that patients made jokes like, “You should find a good allergist to help you with that cough.” My allergist tried different medications, but nothing helped.

At the same time, my dad insisted that I get a chest X-ray. It showed that I had bronchiectasis. All of my doctors were surprised. My allergist said to me “Every time you come in to see me I ask you if you’re coughing more than normal.” I replied, “I’m not coughing more than normal. This is how much I always cough.” I was referred to a pulmonologist and began testing for the cause. Initial tests were inconclusive.

Mary Kitlowski/Rebekah Giannakos

Primary ciliary dyskinesia (PCD) is an inherited disorder of motile (moving) cilia. PCD is also sometimes referred to as Kartagener syndrome (PCD with situs inversus) or immotile cilia syndrome.

A person with PCD experiences chronic, recurrent infections in the lungs, ears and sinuses due to the loss of ciliary activity in those areas.

PCD is a genetic disorder, meaning it is inherited from one’s parents and cannot be acquired from the environment. PCD is most often passed in what is called an ‘autosomal recessive’ pattern of inheritance, in which the disease is only expressed when a child inherits two copies of a mutated gene—one from each parent.

Other conditions that may be associated with PCD are:

- Situs Inversus and Organ Orientation
- Polysplenia
- Congenital heart defects
- Hydrocephalus
- Fertility Issues

Learn more from ATS Public Advisory Roundtable member the Primary Ciliary Dyskinesia Foundation. pcdfoundation.org

Rebekah was 14 when she developed a cough. My parents wanted her to have a chest X-ray, too, and were told there was no way we could both have bronchiectasis. My parents kept insisting. A different specialist diagnosed her with primary ciliary dyskinesia, and eventually I was diagnosed as well.

Rebekah has been hospitalized numerous times for pneumonia starting in high school, had a peg tube inserted due to weight loss, and about two years ago received a double lung transplant. At one point her lung function declined to the point of using oxygen full time. In 2009 and 2012 she and her husband had a girl and a boy, respectively, after four rounds of in vitro fertilization. Currently, she is back working part time and has the energy and lung capacity to keep up with her two active kids.

I am married and choose not to have children of my own. Periodically I get lung infections, which are treated with IV antibiotics. The coughing fits haven't gone away. Sometimes they keep me from sleeping in my own bed, but I manage them and other obstacles.

I continue to race. At first I could only run for 30 seconds, but I didn't give up. I still can't run a whole mile, but by running and walking I've completed over 15 races from one mile up to half marathons. I believe that what we do to raise awareness does lead to better lives—and one day cures for lung diseases.

Karen Erickson

ALPHA-1 ANTITRYSPIN DEFICIENCY



“My doctor asked ‘What matters most to you?’, making it clear that I had to take a lead role in defining and reaching my success.”



Not a day goes by when I don't think back to two things: my donor and the gift he was willing to give, and the two-year prognosis that I was given 16 years ago. Both have stopped me in my tracks and both have also motivated me to continue this battle full force.

I've been on a long journey with Alpha-1 antitrypsin deficiency, and it's taken me from searching for a diagnosis to forging forward after receiving a bilateral lung transplant. At many points along the way, my partnership with providers (or lack thereof) has played a huge role in my success.

In the late '90s I was running a business, completing a degree, and training like crazy for triathlons. I was in a great place, but I started to notice that I was becoming more and more breathless. I had to rest during my busy days, the gains at the gym were becoming smaller and smaller until they were non-existent and I was always exhausted. I chalked it up to getting older and just a part of my busy lifestyle. But my symptoms worsened.

My doctors kept telling me the same thing. "Look at you sweetheart, you're fine. It's just exercise-induced asthma." Or, "You are just anxious about all the stuff you have going on. Here are some inhalers...take them in an emergency." Over the next year, I heard the same response over and over again. I was never given any other advice, testing or treatment plan, nor did I push the issue further. I came to expect it as my new normal.

Everything changed the day I was given my first pulmonary function test. The technician left the room, and the doctor came in. She told me I had the lungs of an 83 year-old and gave me a diagnosis of COPD. I was 33. Rather than digging in a bit more, she recommended that I stop smoking. I let her know that I wasn't a smoker. That statement was dismissed, and I left the office with a smoking cessation prescription.

I used and abused my inhalers to get me through my days and made frequent trips to urgent care, along my routes to and from work. On one of my trips in due

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. 2014. thoracic.org/patients/patient-resources/resources/alpha-1-antitrypsin.pdf

to breathlessness, a physician assistant asked if she could test me for a rare genetic disorder—although she was confident that I didn't have it. Blood was drawn, and I went about my business. Next thing I knew she called me into the office to discuss the test results.

The next morning the conversation went like this, “You have a genetic disease called Alpha-1 antitrypsin deficiency. Your body is basically eating your own lungs. There is no cure for it, but with treatment we think we can get you two years.” The doctor went on to talk about referrals to a pulmonologist and other necessary steps, but I didn't hear a thing. My head was stuck on the 730 days I had left—my friends and family, my dogs, my job, everything...

I did see a pulmonologist, and he was excited to see me. He had only heard of Alpha-1 and was told never to expect to come across a patient. He admittedly knew very little about the disorder but managed to get me on a monthly augmentation therapy.

Uncharacteristically for me, I did not follow up with any research of my own. The news had rattled me enough, and I figured I had 730 days to live.... I distanced myself so much that I left for Ireland shortly after starting therapy.

It was there that everything changed again—but this time for the better. After months of infusing in a local hospital with no questions asked, a doctor came in to start my IV and asked what I was doing. I told him that I had AATD and it was replacement. I said “they” gave me two years to live. He laughed and quickly put that notion to bed. He let me know that if I died in the next two years, it wouldn't likely be from AATD. This doctor was doing research with an Alpha-1 expert at Royal University. My weekly visits soon became a spectacle. The team assured me that with proper exercise, nutrition, and compliance to medications, I could crush those 730 days.

The conversation turned me around. I headed back to the U.S. and hit the ground

running. I read everything I could, found a new doctor, brought materials to her, and we reviewed all the research updates we could find. I became involved with the Alpha-1 Foundation and met world-class AATD specialists. Several patient advocates had the same “two year” prognosis and had shattered it many times over.

My doctor asked “What matters most to you?,” making it clear that I had to take a lead role in defining and reaching my success. With answer in hand, we worked out a treatment and follow-up plan, and as my disease progressed I was referred to a transplant center. I worked very closely with them, and we were a team. On March 26, 2013, I received the gift of life from a young man taken from his family too soon.

On the flip side of transplant, my team and I continue our regular and open communication. We use phones, emails, apps, electronic health record platforms, and meet face to face. They still cheer and motivate me, and they make me feel I am never alone in this battle.

Ellen Egbert

CHRONIC OBSTRUCTIVE
PULMONARY DISEASE



“Participation was an important part of rehab class. I vividly remember all of us blowing pinwheels while learning about pursed lip breathing.”



I used to tease a friend about some of her Southern colloquialisms. One of these was “usta could,” as in “I usta could do such and such, but I can’t anymore.” Ten years ago, I found shortness of breath filling my life with “usta could’s.” I blamed my weight, age, and arthritis.

I’m grateful to the PFT technician who told me to ask my doctor for a rehab referral. I’m now grateful to my first pulmonologist for making the referral and for prescribing the liquid oxygen that makes it relatively easy for me to continue to enjoy an active life. But I was not feeling particularly grateful at the time.

I was told I had severe emphysema, required round-the-clock supplemental oxygen, would have to forego badly needed hip revision surgery because I couldn’t undergo general anesthesia, and that flying to visit friends and family in New Jersey was no longer an option.

I went home, turned on my computer, looked up “emphysema life expectancy” and read that I had just four years of decreasingly productive life ahead of me. I wasn’t thrilled.

Although I found other information on the Internet that was informative and more encouraging, it was with trepidation that I attended my first pulmonary rehab session, leaning on a cane, uncomfortable with a cannula, and struggling to push, pull, and otherwise maneuver an E canister. I was immediately put to ease by the two primary instructors, one a highly qualified nurse practitioner and the other an experienced respiratory therapist. Our sessions were split between classroom activities and exercise.

We were a small but diverse group of patients. Some of us, like me, had tried to research COPD or had an inkling of information from our doctors. Others remained completely in the dark. Some of us were anxious to learn; others, perhaps in denial,

COPD is a disease of the airways and lung tissue that causes difficulty with the transfer of oxygen into the bloodstream and with moving air in and out of the lungs. It is an inflammatory process that breaks down the fragile air sacs in the lungs and obstructs the airways with mucus and constriction of the muscles surrounding the airways. The inflammatory process affects not only the lungs, but also the entire body and can result in unintentional weight loss and muscle wasting.

- COPD is the third leading cause of death in the U.S.
- It is estimated that every four minutes an individual dies of COPD.
- COPD kills more women than men each year.
- An estimated 12 million adults have COPD and another 12 million are thought to be undiagnosed or developing COPD

*Learn more: ATS Patient Information Series.
“Chronic Obstructive Pulmonary Disease (COPD)”
New York, NY: AmericanThoracic Society.
[thoracic.org/patients/patient-resources/resources/
copd-intro.pdf](http://thoracic.org/patients/patient-resources/resources/copd-intro.pdf).*

were reluctant. Our knowledgeable, inventive teachers were patient as they shared flip charts, demonstrated techniques, and encouraged questions and discussions. Instruction continued in the adjacent gym, where we were introduced to exercise equipment, taught to manage and measure our oxygen levels, and were applauded for our efforts and improvements.

Participation was an important part of rehab class. Group discussion not only taught us to share our problems and offer suggestions for resolving them, but also instilled the confidence to solve our own problems and develop self-management techniques. I vividly remember all of us blowing pinwheels while learning about pursed lip breathing. Since my house is full of stairs, I especially appreciated the day the respiratory therapist took us to a stairwell. That instruction helped us climb stairs and San Francisco hills.

For those leading pulmonary rehab groups, I urge care. When “experts” are brought in to teach specific topics yet are not familiar with the style and content of class, they can lose attention, use technical terminology, or talk down to the class during lecture.

I hope emphasis will be placed on the patient/doctor relationship and the right to both ask questions, and demand answers. Insurance was not discussed in my rehab program but since coverage, particularly Medicare, for oxygen is becoming more stringent I hope it will be a standard rehab topic so that patients are made aware of their options.

Pulmonary rehab helped me discover a happy and busy life with COPD. I also found a new pulmonologist who supported my wishes by approving air travel to the east coast and hip surgery using local anesthesia.

Instead of “usta could’s,” I now have an unending supply of “still can’s.”

Debbie Breslawksy

NONTUBERCULOUS
MYCOBACTERIA (NTM)



“The feeling of isolation is devastating. You look to speak with people in the same situation, but where do you find them? A support group would be nice, but none exist. After years of chatting up everyone you know, you find enough people to start your own group.”



You're sailing along in life. You've launched the kids, are about to have some serious fun with your husband, travel, go on adventures, then bam! Cough, cough, cough. Fatigue sets in. And after many attempts at getting a correct diagnosis from far too many doctors you come to grips with having a disease you've never heard of, and people, including most of your doctors, haven't either. The diagnosis: non tuberculous mycobacteria (NTM), a highly resistant mycobacterial infection in the lungs that will stay with you for the rest of your life.

You look OK. Your friends don't understand why you're acting like a wimp. Exhaustion overcomes you. You don't have the energy to do the things you love.

You go to the pharmacy to fill a barrage of prescriptions, an antibiotic cocktail (wish it were another kind of cocktail) and you start coughing. And with that cough, you're able to clear an entire section of Costco. You explain you're not contagious, but people don't believe you. You hide your face in shame.

You used to sing. You used to run. You used to ski. Now you stay home most of the time because you are on three or more strong oral antibiotics, and sometimes inhaled and IV antibiotics, two infusions per day for months at a time. Your gut is majorly annoyed, and so are you. Dining out isn't fun anymore. Your appetite is gone.

So many little things we take for granted! Attending a concert or big party? I don't think so. You now avoid crowds for fear that your weakened immune system is susceptible to catching cold, or even worse, pneumonia. Who doesn't like a hot, steamy shower? A relaxing Jacuzzi? A visit to Miami or Hawaii? All vacations to humid climates are out! Not a good idea to be sitting under a mister at an outdoor café in summertime or hanging out in the produce section of your favorite supermarket because the spray from the misters are filled with NTM. And the NTM-infused aerosolized mist is looking for a place to park, most likely in your lungs.

Debbie Breslawsky

- NTM infection causes the bronchial tubes to fill with mucus and, over time, may lead to dilation and scarring of the tubes.
- Types of Nontuberculous Mycobacteria include: mycobacterium avium complex, mycobacterium kansasii, mycobacterium abscessus, mycobacterium chelonae, mycobacterium intracellulare, mycobacterium fortuitum.
- Common symptoms of NTM include coughing, night sweats and fever, loss of weight, loss of appetite, and lack of energy.

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

To help clear the lungs, you have an array of machines and gadgets which can take up several hours to administer each day: the lung shaker, aka the “vest,” bronco- dilator. These are your new best friends. They are to be used every day for the rest of your life for airway clearance.

You exercise when you can to keep your lungs operating as well as possible. Is all this working? Sometimes you think you are doing a whole lot of huffing and puffing for nothing.

After a few years, you might catch a break for a few months. Yippee, a drug holiday! And if you are lucky enough to get a negative culture, the celebration is short-lived because it will soon be positive. Your lungs are so compromised.

The feeling of isolation is devastating. You look to speak with people in the same situation, but where do you find them? A support group would be nice, but none exist. After years of chatting up everyone you know, you find enough people to start your own group in New York City. The support is invaluable, so you start another group in the dry climate of California where you have moved part time to escape the humidity and to breathe better. Then another group in Connecticut. Patients find you online. Networking helps you find doctors who specialize in NTM, discover treatment options, receive feedback from others in the same boat. You speak from experience about CT scans, spirometries, PFTs, and on and on and on.

Together, you make the best of a bad situation. You put on your game faces when you are feeling sick and keep pushing yourselves.

Some of us can look forward to a lobe or two of our lungs having to be removed. Scary. Yes, the procedure has become more simplified using a video-assisted surgery. However, there’s nothing simple about it if you’re the patient.

We have lost too many friends to this dreaded disease, and many of us fear the same fate for ourselves. More than anything, we need to raise the level of provider education, in medical schools and beyond. We need research to determine genetic predisposition, effective treatments with fewer side effects. Even better—a CURE—or prevention.

Susceptibility to NTM is inherited. If not for ourselves, we must succeed for our future generations.

Brenda Clark

SARCOIDOSIS



“The news nearly knocked me out of my chair. I read it over and over again and several times aloud to myself. I was very scared and also angry. I would have appreciated a phone call or in-person consultation.”



Like many patients diagnosed with sarcoidosis, I sought to find out as much as I could about the disease, scouring the Internet and reading a great deal of literature from my health care providers. It was disappointing to find out the cause was unknown and that there was no cure—yet.

I was age 44 at the time, in June 2008. My ophthalmologist was the first to make the diagnosis. I needed treatment for inflammation, which I had in one eye, and that quickly progressed to pain and blurred vision. On the day we reviewed my chest X-rays, I received a second diagnosis of iritis. I jokingly asked my doctor if this was arthritis of the eye.

I began a treatment of steroid drops to relieve the inflammation and return to normal eye sight. The iritis flared up roughly six months later in the other eye. So far I have not had another recurrence.

I learned that symptoms could appear and disappear throughout my lifetime and what really stuck was that the microscopic lumps in my lungs could appear in almost any body organ. In four years this happened.

When presented to the emergency room late one Sunday evening in excruciating pain I assumed I had a ruptured appendix or kidney stone. The ER physician sent me for abdomen and pelvis CTs without contrast, and those later revealed I was suffering from bilateral kidney stones. This was a first for him—he said he never had a patient appear with bilateral kidney stones. The nurse who came with an injection for the pain asked if I had any children. When I asked her why she said that this pain was comparable to childbirth. My partner, who was with me in the ER, reminded me of a quote from the comedian Carol Burnett who once said giving birth is like taking your lower lip and pulling it over your head. Things would not end so lightheartedly.

Sarcoidosis is an inflammatory disease that most commonly involves the lungs but can impact any organ in the body including the eyes, nervous system, liver and heart. For some patients, the disease is mild and for others it may be progressive and devastating. The cause of sarcoidosis remains unknown. However, experts suspect that sarcoidosis develops when a patient's immune system overreacts to some type of exposure—occupational, environmental or infectious—and small clumps of inflammatory cells called granulomas are formed and deposited in affected organs.

*Learn more from ATS Public Advisory Roundtable
member the Foundation for Sarcoidosis Research.
stopsarcoidosis.org*

The next business day, I received the following email from my primary care physician:

“Dear Ms. Clark, I reviewed your recent CT abdomen scan with the radiologist and it shows abnormally increased lymph nodes in your abdomen which needs further evaluation by another scan and a biopsy to rule out other abnormalities such as cancer. I have sent the referral to surgery and radiology for these studies.”

The news nearly knocked me out of my chair. I read it over and over again and several times aloud to myself. I was very scared and also angry. I would have appreciated a phone call or in-person consultation.

That August I went in for my surgical consult visit and was fortunate to have a compassionate physician, who must have read the anxiety on my face the moment we met. She went over my options and in the 45 minutes she spent with me, she was extremely thorough. She believed it was best to get an MRI with contrast as soon as possible because the previous CT previously was without contrast, and she said this would help with surgical planning. It wasn't until years later when I went back and read my medical record for the first time, that I recognized the significance of this visit with my surgeon. Abnormal lymph nodes were everywhere, and there were varying sizes from two to four centimeters in my upper abdomen, intestine, diaphragm, chest, and esophagus. Thankfully, the biopsy came back negative. No cancer. Conditions and findings were found to be consistent with the history of sarcoidosis.

While this entire process took over five weeks to determine, it felt more like five months. Today I suffer from fatigue, skin rashes, and other various conditions likely related to the disease. Though it helps to stay active with organizations like the Foundation for Sarcoidosis Research, and I appreciate being able to share my personal and medical history with you.

Kathy Przywara

ASTHMA



“My daughter has allergic and exercise-induced asthma. It’s painful as a parent to watch her struggle during sports. But with proper management, pretreatment and persistence, she was able to become a second degree black belt.”

I have asthma. I mainly have allergic asthma, but I also get flares with upper respiratory infections. It is cough-variant asthma, which means I cough instead of wheeze. This makes it harder to tell the start of a flare since my peak flow readings don't drop.

I was diagnosed as an adult. The first time I was given a prescription for a rescue inhaler was after returning from a trip to Alaska with severe bronchitis. There was no mention of asthma. It was typical for me to get at least one bout of bronchitis each year.

I continued to have bronchitis regularly for several more years. I wasn't told I had asthma until I was pregnant. My asthma was worse with my second pregnancy. That was the first time I was started on a preventative inhaler. I really didn't want to be on medications while pregnant, but the doctor assured me that breathing was important for the baby. My asthma has continued to get worse and become more persistent, as have my nasal allergies, which are my biggest trigger.

I likely had undiagnosed asthma as a child. I remember having difficulties in gym class when running. I would also cough until I was gagging every spring during fitness testing. I was always coughing as a kid, but allergy testing came back negative, so the doctor dismissed my symptoms. Asthma definitely affects my daily life, I have to be careful with pets and avoid triggers like pollen. Often I feel like I'm holding others back.

Both of my children have asthma. My son has mostly outgrown his asthma, but it has transformed into chronic sinus issues. My daughter has allergic and exercise-induced asthma. It's painful as a parent to watch her struggle during sports. But with proper management, pretreatment and persistence, she was able to become a second degree black belt. I remember one time she was sparring and having asthma issues. She kept coughing, which meant she couldn't focus, and her sparring partner landed a punch and I remember thinking, "she just got sucker-punched by asthma."

ASTHMA



Asthma is a chronic disease that swells the airways, or breathing tubes, of your lungs. This swelling (inflammation) causes the airways to make thick, sticky secretions called mucus, and it causes the muscles in and around your airways to get very tight or constrict, which makes it very hard for you to get air into and out of your lungs.

Asthma can be caused by genetics, allergies, respiratory infections, and irritants such as:

- Molds and dust
- Exhaust fumes from vehicles
- Chemicals in garden sprays
- Strong odors from paint, perfumes, colognes, hair spray, deodorants, and cleaning products
- Tobacco smoke
- Weather changes
- Stress or exercise
- Medications
- Sulfites in foods such as dried fruits, wine, and beer

Learn more: ATS Patient Information Series. "What is Asthma?" New York, NY: American Thoracic Society, 2013. thoracic.org/patientsg

Jenny Lefferts

LYMPHANGIOLEIOMYOMATOSIS (LAM)



“My biggest fear was that I had three children to raise. I immediately got online and searched for facilities that understood this disease.”



I'll never forget driving to the emergency room on a Saturday afternoon in April 2011. I hadn't been feeling well, and I was pretty certain I had pneumonia and didn't want to wait until Monday to start on a course of penicillin. The ER wasn't busy so the nurse told me to hop in a wheel chair and he would escort me to radiology. We laughed the whole way because I didn't feel it was necessary. The laughing stopped when my X-ray indicated a partial pneumothorax (collapse) of my left lung. I thought they were kidding. I started to laugh but this time they didn't join in. I said that I'd deal with it on Monday because I had stuff to do, and they explained that I was not allowed to leave the hospital and that they were immediately going to insert a Heimlich valve into my chest to inflate the lung and admit me.

Most people at risk for a pneumothorax are usually very tall, thin men. Since I am a middle-aged, 5'3" female it was strongly suggested upon discharge that I follow up with a doctor and have a high resolution CT scan with contrast. I complied and after my scan I followed up with the ordering physician. He simply stated that there was a long word, 24 characters to be exact, at the end of the report that he had never heard of—but not to worry! Although he did suggest that I should avoid any rock climbing or scuba diving plans in the future.

When I got home I searched the word Lymphangioleiomyomatosis (LAM). Wikipedia stated that I had a rare, incurable, and fatal lung disease with a life expectancy of 8 to 10 years after diagnosis. I was scared, angry, and upset. My biggest fear was that I had three children to raise. I immediately got online and searched for facilities that understood this disease. My location was a blessing as I quickly found out that the nearby Stanford Hospital had a LAM program that was studying the disease. I called them immediately and the staff there invited me to a LAM support group that was being held the following weekend.

Lymphangioleiomyomatosis (LAM) is a progressive cystic lung disease typically manifesting in women of reproductive age. LAM can be either sporadic or associated with tuberous sclerosis complex (TSC). LAM involves smooth muscle proliferation that contributes to parenchymal cysts formation in the lungs. While LAM is considered an interstitial lung disease, clinically, it is essentially a cystic lung disease and shares significant physiological features of emphysema including bilateral multiple cysts and airflow obstruction.

- Symptoms may include shortness of breath, collapsed lung, chest pain, cough, fatigue
- Women with LAM may be misdiagnosed with asthma, emphysema, or bronchitis.
- Median survival in patients with LAM has varied from 10 to 30 years.

Learn more from ATS Public Advisory Roundtable member The LAM Foundation. thelamfoundation.org

What happened next was nothing more than a miracle. LAM, a rare disease, with approximately 1500 documented cases globally, had formed a foundation in 1995. A mother of a LAM patient, Sue Byrnes and Dr. Frank McCormack founded the foundation together, and it is a driving force in finding effective treatments and hopefully one day a cure for the disease.

A representative called a LAM liaison for my region, and she met with me in person and told me about an NIH study. By September 2011 I was in Bethesda, Maryland being followed by one of the top physicians focused solely on the disease. I have been back three times.

Since my diagnosis in May 2011 lymphangiomyomatosis has an official ICD-10 code (diagnosis code), the approval of the use of Siralumus as the first ever drug to help treat the disease, and continued funding at the NIH.

My disease is progressing slowly. I am one of the fortunate ones. I still participate at the NIH and also follow up with my provider at UCSE. But if it hadn't been for the LAM Foundation I don't know how long I would have suffered as there was so little known about this disease.

I hope to give back however possible. One breath at a time.

New Patient Resource: Asthma Today

Download Now!

Available at thoracic.org
Featuring highlights from ATS 2016



Save the Date: Lung Disease Week

Each year, the American Thoracic Society Public Advisory Roundtable presents [Lung Disease Week at the ATS](#), a series of weeks that focus on specific lung disorders for which ATS PAR member organizations provide support and guidance to patients and their families.

Find links to [information for patients and experts](#), including disease definitions, clinical trial updates, support group information, ongoing legislative efforts, patient stories, testimonials, interviews, videos, and photos.

Attend live events or watch and listen online to webinars with experts in disease research and clinical care presented by ATS PAR partners.



Join the Society-wide initiative at
thoracic.org/patients/lung-disease-week/.

Get the Facts! ATS Patient Information Series

The American Thoracic Society's Patient Information Series features **free** downloadable fliers that describe lung diseases, treatments, and tests in patient-friendly terms.

Topics include:

- Asthma
- COPD
- Critical Illness
- Lung Problems and the Environment or Work
- Lung Problems in Babies, Children, Teens
- Lung Problems from Bacteria, Virus, Molds, Fungi
- Lung Cancer
- Lung Problems that are Uncommon or Rare
- Lung Problems that are Seasonal
- Lung Problems and Smoking
- Sleep Problems
- Tobacco Series
- Surgery and Transplantation for Lung Problems
- Tests, Procedures and Monitoring for Lung Problems

Browse the entire selection of Patient Information Series fliers at thoracic.org/patients.



Meet the Experts

Each year, the American Thoracic Society Public Advisory Roundtable (ATS PAR) holds its patient-focused Meet-the-Experts forum as part of the larger ATS International Conference. This free event is open to lung and airway disease patients and their families. Attendees learn the latest research, clinical trials, and clinical care, and network with other individuals who share their experiences with lung diseases.



More than 20 expert speakers are usually available, as well as a number of breakout sessions to give patients and families a chance to interact with prominent pulmonologists and experts in critical care and sleep medicine. Lunch, oxygen, and parking is provided free of charge.

To learn more, contact Mr. Courtney White at cwhite@thoracic.org.



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Previous Voices

Patients and speakers featured in *ATS Patient Voices 4*.



“Pumped full of fluids, I was so swollen that my wedding rings would not come off. My husband of 41 years began the arduous process of cutting them off. You can imagine his pain and sorrow with this difficult task.”

-Millie Camp
Acute Respiratory Distress Syndrome



“This is my new normal. With a positive mindset and the strong support of family and friends, I will continue to take life one shot at a time.”

-Kirk Mease
Sarcoidosis



“The scars on my body and feet make it tricky to put on socks, and my mom gets frustrated when we change them a lot before the bus gets here. But I’m thankful to be alive.”

-Annie Costello
Oxygen Therapy



“It was like the disease fueled a new fire to live, and I wanted to approach every moment with genuine intention. I rode my bicycle across the U.S., stood atop the tallest peaks of Alaska, and climbed the faces of El Capitan in Yosemite.”

Jon Bernhard
Nontuberculous Mycobacterial Disease
(NTM)

View all past editions and many more patient resources at: thoracic.org/patients.

ATS Patient Voices 5



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