



ATS Patient Voices 3

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.



We help the world breathe®
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 (ATS 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 Meet-the-Expert patient and family forum, the PAR Symposium, and the many patient speakers integrated throughout the scientific session curriculum.

Patient Voices, now in its third 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 asthma, sleep apnea, cystic fibrosis, pulmonary hypertension, and 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.

Patients and their families are seeking cures, and the opportunities for discovery research have never been greater. We now have the tools to unlock the mysteries of lung disease. The ATS is advancing pulmonary health through innovative research, clinical care, advocacy, and training of tomorrow's leaders who will translate discoveries to patients. Indeed, the ATS partners with patients and patient advocacy organizations to find these cures, and on 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 ATS PAR. This booklet is another manifestation of these efforts to strengthen the relationship between patients, their families, and the ATS.

We greatly appreciate the efforts of 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 ATS 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.



Thomas Ferkol
Thomas Ferkol, MD
ATS President 2014-15



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

Because patients are central to what pulmonologists and other medical professionals do, the American Thoracic Society established the Public Advisory Roundtable (ATS PAR) in 2001 to enable patients and their advocates to interact directly with the ATS and help clinicians and researchers understand their perspectives.

ATS PAR is known for its unique ability to respond to patient needs, mobilize efforts to improve patient care, increase research efforts in lung disease, build advocacy and awareness of lung disease, and promote lung health on a national level. ATS PAR members are patients, family members, and advocates who understand the real needs of patients and the lung diseases from which they suffered. Following in the footsteps of the founders, they articulate those needs and help bridge the gap between patients and physicians.

Never before has a medical association elevated patient voices to a level of such an important function within its organization. ATS leadership has shown that ATS PAR remains a vital part of the organization and is held in high esteem. The ATS PAR chair is a standing member of the ATS Board of Directors, with a direct line of communication to ATS leadership.

The implementation of ATS PAR through the vision of Dr. Bill Martin, an ATS past president, not only created opportunities to strengthen medical care but also opened doors for collaboration and partnership. This in turn has helped to increase understanding of lung diseases and fund research that draws us closer to cures. To date, ATS PAR-affiliated member organizations have supported the ATS Foundation with more than \$5 million in funding for innovative and cutting edge medical research in lung disease.

For the past several years, ATS PAR has had the privilege of assigning patient speakers to medical sessions at the ATS International Conference, which draws approximately 12,000 pulmonary, critical care, and sleep medicine researchers and clinicians annually. These patients share their journeys through life with lung diseases that attendees are currently treating and researching.

Within these pages you will find many compelling patient stories that provided the “patient voice” during the International Conference and have inspired attendees to continue research and clinical care without losing sight of the patients.

It continues to be an honor and privilege for ATS PAR to be “the patient voice of the ATS.”



Gregory R. Porta
Chair, ATS PAR



Donna Bryson

Asthma

“My daughter had a great doctor with a great nurse. Involving the patient and caregiver as active participants is key.”

I am a wife, mom, friend, nurse, asthma educator for the American Lung Association, and I have severe persistent asthma. I was diagnosed nearly 60 years ago, and my earliest memories revolve around asthma: doctors, hospitals, oxygen tents, painful treatments, and that memory of struggling to breathe and the fear of that next time.

In high school, one of my dearest friends, Patti, who also had severe asthma, was found on the floor dead with her inhaler clutched in her hand. Until that moment, I believed the lie that was my mother's prayer: children don't die from asthma.

Then my asthma attack came. It was so swift that it didn't matter how hard I fought. I felt my lungs turn to stone, and air only moved in and out of my throat. I couldn't speak, or say "I love you" one last time. As the world closed in, became a cold gray cylinder, and finally a black speck, the last words I remember hearing were, "let's tube her."

I woke up, I got better, and my life with asthma went on.

Asthma continued to shape our lives. My son was two when he was hospitalized the first time with asthma. I sat by his bed and watched him endure the same things that I endured. I knew it was my fault and that I had done this to him—he had my asthma. I knew what life had in store for him, and my heart ached.

My oldest daughter also had asthma, which worsened in middle school. She was very athletic and played hi-intensity sports like basketball, soccer, and fast-pitch softball, so she was given a compact nebulizer. She was able to take Albuterol nebulizer treatments on the field or court. But when you need special treatment, it makes you different, which you do not want in middle and high school. She was very reluctant to come to the sidelines because she worried that it would be perceived as a "cop out" if she had to leave the game. I had to intervene and advocate for my daughter.

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. patients.thoracic.org

Also difficult was convincing the school that she must be allowed to carry her inhaler. Thank goodness that all 50 states now have self-carry laws, although the implementation of those laws may still need attention.

When the NIH Heart, Lung, and Blood Institute Guidelines appeared in 1989, and the use of inhaled corticosteroids was recommended, my world changed. My children and I improved dramatically. We still had exacerbations, but they were fewer and less severe.

When my third child was born, the medications were better and our understanding of the disease was improving. Her pediatric pulmonologist wasn't afraid to treat her with increased amounts of inhaled corticosteroids that she required for control. A nurse and I sat down and together, developed her first Asthma Action Plan, and discussed what I needed to know, why, and how to use the plan. Needless to say my daughter had a great doctor with a great nurse. Involving the patient and caregiver as active participants is key.

As my daughter got older and her care plan was updated, she was the one involved in decisions about her care and management of her health.

A school nurse introduced me to Open Airways for Schools, which teaches children about their asthma symptoms, medication, triggers, and warning signs, and gives them a chance to talk about how they feel. For the past 14 years, I have been working for the ALA, and I have had the opportunity to teach asthma self management. As a result, my confidence and skills in managing my asthma and my children's have grown tremendously.

Donna Bryson was a patient speaker at the ATS 2014 International Conference in San Diego.



Michelle Figueras

PULMONARY HYPERTENSION

“People who looked in worse shape than me would zoom up a flight of stairs with no problem. When I would try to keep up, I’d feel like I was ready to faint.”

Although I was diagnosed with idiopathic pulmonary arterial hypertension in 2013, I believe that I have had the condition for awhile. The first episode was in 2008. I went to the emergency room because I couldn't breathe and my ankles were swollen. I was diagnosed with "systemic" high blood pressure and prescribed diuretics to reduce the swelling. I started a low sodium diet and experienced shortness of breath and water retention if I binged on salty foods.

I was 26 years old and looked healthy, but I couldn't run, use an elliptical machine, or dance for more than a minute without feeling like I was going to pass out. People who looked in worse shape than me would zoom up a flight of stairs with no problem. When I would try to keep up, I'd feel like I was ready to faint.

By 2012, my fatigue had increased. I would sleep on weekends for 16 hours straight and I would still feel exhausted. My primary care physician diagnosed me with a single episode of major depression and prescribed anti-depressants.

By the next year, my symptoms worsened. The water retention became more frequent, and the shortness of breath became more evident when I retained water. On July 23, 2013, after a weekend of very poor eating decisions, my body decided it had had enough. My face was swollen, my abdomen felt full, and I was retaining fluid in both my legs. I couldn't even walk 10 to 15 feet without running out of breath. I went to urgent care, and the doctor heard a heart-murmur and told me that I needed to go to the emergency room so that they could rule out pulmonary hypertension. He was the first doctor who told me that it wasn't normal for a 30 year old to have ankle swelling and shortness of breath.

Michelle Figueras

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.

Learn more: ATS Patient Information Series. "Pulmonary Arterial Hypertension" New York, NY: American Thoracic Society 2013. patients.thoracic.org

I was transferred to University of California, Irvine Health, which performed my right heart catheterization and confirmed the diagnosis on July 23, 2014.

By searching the hashtags “pulmonary hypertension” on Facebook and Instagram, I found a local PH support group in Long Beach. I attended a meeting, and they urged me to look at the Pulmonary Hypertension Association website to find a specialist. Many recommended Ronald Oudiz, MD, at Harbor UCLA Medical Center.

During a PH support group meeting, speaker Joy Beckman, a pulmonary hypertension nurse who worked with Dr. Oudiz, discussed clinical trials. Since my insurance was such a pain to go through, I knew the only way I’d get treatment from him was through a clinical trial.

I qualified for AMBITION, a clinical trial for two drugs for that I was already prescribed to take after being discharged—Ambrisentan (Letaris) and Tadalafil (Adcirca)—which meant the risk of participating in the trial was non-existent. The study is meant to test the efficacy of single drug therapy versus combination drug therapy. I felt safe knowing that I would at least be getting some treatment.

Since being in the trail and being better educated about my condition, my quality of life has improved 100 percent. I can walk up two flights of stairs, do some moderate cardio exercise, and walk slight inclines without feeling short of breath. My water retention is under control and I understand my body a lot better.

The study is in its final phase, and I am curious to learn the results. It makes me feel good to know that I helped contribute to how PH will be treated in the future.

Michelle Figueras was a patient speaker at the ATS 2014 International Conference in San Diego.



Megan Gray

LYMPHANGIOMATOSIS

By Patti Perkins Gray, Megan's Mother
"After all we have been through, I look at Megan today and feel extremely blessed to still have her here with us."

Our journey with Lymphangiomas did not begin with any difficulty breathing. There was no chronic cough or cold that would not go away. In fact, Megan hardly ever had a cold. In the beginning it didn't look like a lung problem at all.

Megan was 15 months old in October 2009 when her collar bone broke for no apparent reason. Much testing followed. X-rays, MRI, and CT scans revealed cystic lesions in almost all of Megan's bones, her thyroid, abdomen, and in the soft tissue behind both of her collar bones reaching down into her chest. Finally, a biopsy of her thyroid provided the diagnosis of diffuse lymphangiomas.

Things were fairly uneventful until January 2012 when Megan got very ill with group A streptococcus (strep) bacterial infection in the blood. What began as a fever one evening quickly spiraled out of control. Within 24 hours our 3-year-old little girl was admitted to the pediatric ICU with sepsis and toxic shock syndrome, and soon after, all of her organs began to fail. It was a very long traumatic experience, and it is a miracle Megan survived at all.

For those first seven months in 2012, we were in the hospital more than at home. It was during this time that the pleural effusions began. Megan's disease had now progressed to her lungs. She required repeated chest tube placement to drain the fluid from around both lungs. With no FDA approved treatments for her disease, she was started on a combination of drugs (sirolimus, vincristine, and prednisone) that have been used off-label for lymphangiomas, alone, and in various combinations. After only a couple of weeks on this regimen the effusions stopped. Megan remained on this cocktail for more than a year until the side effects of the drugs got to be too much and we decided to take a break.

LYMPHANGIOMATOSIS

Lymphatic vessels reach every part of the body except the central nervous system (the brain and spinal cord). The lymphatic system has three main functions:

- Maintain fluid balance
- Defend the body against disease by producing lymphocytes
- Absorb fats and fat soluble vitamins (A, D, E, and K) from the small intestine and transport them to the blood, bypassing the liver

This condition is marked by the presence of cysts that result from an increase both in the size and number of thin-walled lymphatic channels that are abnormally interconnected and dilated. The deranged lymphatics tend to invade surrounding tissues and cause problems due to invasion and/or compression of adjacent structures.

*Source: Lymphangiomas and Gorham's Disease Alliance. "What Is Lymphangiomas?"
lgdalliance.org.*

In June 2014, Megan's pleural effusions came back with a vengeance. Three times in the span of five weeks, she had to be hospitalized and have chest tubes placed to drain the fluid from around her lungs. The sirolimus, vincristine, and prednisone regimen was restarted and seemed to be working, but when the effusion returned a third time, her doctors and I decided it was time to go ahead with pleurodesis, a surgical procedure to scar the lung to Megan's chest wall in hopes to stop effusions from returning. Five days after the procedure Megan was home and the surgery seems to have been a success. She has some shortness of breath but nothing compared to how it was with the effusions.

Lymphangiomatosis is so rare that most doctors have never heard of it and have no idea how to deal with the complications that it can cause. With that brings many fears and frustrations. It has been a blessing that I found the Lymphangiomatosis & Gorham's Disease Alliance (LGDA) and got in touch with others dealing with this disease. Megan has even met a couple of patients, which has helped her to know she isn't the only one.

After all we have been through, I look at Megan today and feel extremely blessed to still have her here with us. She is an inspiration to us all. I pray that one day this disease will be cured or at least better managed.



Will Headapohl

OBSTRUCTIVE SLEEP APNEA

“I still struggle with being tired and losing weight. But I am not grumpy like my dad was, and I’m negotiating life pretty well.”

Obstructive Sleep Apnea runs in my family. My father, sister, nephew, two teenage sons, and I have had it.

As a child, I was the spitting image of my father: foggy headed and a late bed wetter. Each morning, I heard him snore, but I didn't pay attention because I had to get myself focused. My twin sister was always chipper, but I felt like I had a perpetual hangover. I got Ds and Cs on my report card. Little did I know that OSA was causing my cognitive dysfunction and bedwetting.

During my early teens, my father developed Gastroesophageal reflux disease and then asthma. He took a lot of medications and became a huge grouch. As for me, I could not sit still in orchestra with what was most likely attention deficit hyperactivity disorder.

When my tonsils and adenoids were removed, I got smart, filled out physically, and began earning As and Bs—I could breathe again. My ADHD subsided, and I later finished two engineering degrees at Stanford University.

My father, on the other hand, was not doing so well, and snored like a freight train. He had an emergency triple bypass, and his snoring became epic, which made him persona non grata on hunting and ski trips. My mother was psychotic from lack of sleep, and she dreamt of killing my father—but not often, because she could not sleep. He then developed diabetes, prostate cancer, and ultimately passed away from pancreatic cancer, likely a complication from diabetes though he ate healthful foods.

I did well in my Silicon Valley career, but in my 40s, I began to suffer from lack of concentration, sleepiness, and hyperactive thoughts—my OSA had returned.

After a sleep study, I was diagnosed with mild apnea, and I tried oral appliances. Then I had major surgery through which my tongue was moved forward (the logic being

Will Headapohl

OBSTRUCTIVE SLEEP APNEA

A person with obstructive sleep apnea has times during sleep in which air cannot flow normally into the lungs. The block in airflow (obstruction) is usually caused by the collapse of the soft tissues in the back of the throat (upper airway) and tongue during sleep. Apnea means not breathing. OSA is more common in men, women after menopause, and people who are over the age of 65. OSA can also occur in children, as well as in people who are overweight, have large tonsils or adenoids, or certain types of jaw problems. Treatments include:

- Weight loss
- Avoiding alcohol for at least four hours before going to bed
- Strategies to help you sleep on your side
- Continuous Positive Airway Pressure (CPAP) device
- Oral devices
- Surgery

Learn more: ATS Patient Information Series. "What Is Obstructive Sleep Apnea In Adults?" New York, NY: American Thoracic Society 2009. patients.thoracic.org

it must need more room). The surgery was not a success; I learned what it was to be among the 30 percent with no cure. I struggled again until I met the founder of Resmed, who signed me up for a Continuous Positive Airway Pressure (CPAP) machine, and I have been a faithful user ever since.

I am not convinced CPAP therapy is perfect. Who wants to be tied to the electrical grid every night? I still struggle with being tired and losing weight. But I am not grumpy like my dad was, and I'm negotiating life pretty well.

As the board chair of the American Sleep Apnea Association, I work every day to help others find treatment. I am lucky because I've been treated, but 35 million Americans are going down the same path as my father. There has to be a better way.

So, how do we stomp out OSA and become apnea avengers and rid ourselves of this evil with asymmetrical force? The next big research idea is the Patient-Centered Outcomes Research Institute, which funds research that is "guided by patients, caregivers and the broader health care community." Many with OSA have become experts with knowledge to share, like myself, and through this we can combine 50,000 OSA patient-experts and improve our collective outcomes via patient-informed research.

Like many issues and diseases, it takes generations to sort them out and improve. My father suffered. I suffered less. I am hoping my nephew's son will avoid any of it.

Will Headapohl was a patient speaker at the ATS 2014 International Conference in San Diego.



Noël Holly

PULMONARY HYPERTENSION

“I went numb. I called my pastor’s wife and said that I was diagnosed with a rare and deadly disease. Driving home, the tears started.”

Prior to being diagnosed with Pulmonary Hypertension, I was someone who was relatively energetic. I was an office manager and executive assistant and was always on the go. I also stayed busy with gardening, teaching Sunday school, dancing, and keeping up my household.

I walked more to manage Type 2 diabetes, but I began to cough a lot. During a church retreat to the mountains in Idyllwild, I had a hard time breathing but chalked it up to being out of shape. Stairs became an issue for me, but I figured it was due to double pneumonia weakening my lungs years ago. It was a drier cough, and there were times when I was like a cat coughing up a fur ball.

Then came a morning when I got up for work but felt very weak. I thought I could go to the emergency room, get some oxygen, and go on to work. When I marched into the ER, I was given a bed, and they tested my oxygen saturation, which was in the low 80s. I was put onto oxygen, and they inserted an IV into my arm. I then underwent an MRI, CAT scan, and blood work.

The doctor informed me that I had massive blood clots in both lungs, and my initial response was, “Oh, get on with ya.” I thought blood clots only happened to smokers, and I didn’t smoke. I started to get rather indignant. But they insisted that I be admitted. Other tests followed, including a V/Q scan and ultrasound scan of my legs, and I had initially had a blood clot (deep vein thrombosis) in my leg that had travelled through the heart and into my lungs. It was a silent clot, as I never had redness, bruising, or pain.

I was given massive amounts of heparin and prescribed bed rest. After 10 days I was released. I continued to cough, and saw the doctor for follow up visits. As the year continued, the coughing worsened and it was still difficult to make it from my car to

PULMONARY HYPERTENSION

Because the different kinds of pulmonary hypertension are treated differently, it is important that your health care provider takes the time and orders the necessary tests to find out what kind of pulmonary hypertension you have. Tell your health care provider if you are having:

- Increased shortness of breath
- Dizziness
- Feeling like you might faint
- Fainting
- Chest pain
- Heart palpitations (feeling like your heart is racing or pounding)
- Increased swelling of your feet, legs or belly
- Your lips and/or fingers turn blue

*Learn more: ATS Patient Information Series.
“Pulmonary Arterial Hypertension” New York, NY:
American Thoracic Society 2013.
patients.thoracic.org*

front door without collapsing, and all other normal functions were impossible.

An echocardiogram revealed that I had PH. My doctor said it was rare, incurable, and that the life expectancy is three years. The Echo showed that both the left and right side of my heart was enlarged. I said, “Well, that was good for the Grinch—isn’t it good for me?” He sighed deeply, and referred me to a PH specialist at University of California, Los Angeles.

I went numb and called my pastor’s wife (a good friend) and said that I was diagnosed with a rare and deadly disease. Driving home, the tears started.

The PH specialist said it was a result of blood clots called chronic thromboembolic pulmonary hypertension, and that they were inoperable because what was left of them had become too fibrous in both lobes.

I transferred to a PH specialist closer to me, and right heart catheterization determined that my mean pulmonary arterial pressure was 55—when the normal should be no higher than 25—and that I fall under Class IV New York Heart Association functional capacity. My doctor started me on Tracleer (bosentan), and it took some time, but I did notice that I could do more.

That first year after diagnosis was what I call “The year of frustration.” Thankfully, I discovered the Pulmonary Hypertension Association, received their book *Pulmonary Hypertension: A Patient’s Survival Guide*, and met other PH patients through a support group. I now participate in awareness and advocacy.

At first, the words “rare and fatal” were constantly flashing in front of me like a neon sign. But now they do not control me.

Noël Holly was a patient speaker at the ATS 2014 International Conference in San Diego.

Noël Holly



Katie Lessard

PULMONARY HYPERTENSION

“I was given much hope and I still have hope, even in rejection, that I will live many more years and will continue to thrive.”

I was a Navy helicopter pilot for nine years. In 2005, I ran in a San Diego marathon but started to notice that my running wasn't very good. I took a trip to Ireland and noticed how exhausting it was to climb stairs. I went skiing, and it was torture getting to the lift.

I mentioned my struggles to my flight surgeon and she began ordering tests. Everything came back normal. But she knew something was not right so she just kept ordering more tests. I eventually had an echocardiogram. The hushed whispers and the gathering of doctors and nurses around the echo screen told me they had found the problem. After a right heart catheterization, I was told that I had idiopathic pulmonary arterial hypertension.

The diagnosis was absolutely shocking. I didn't understand. I was healthy, I was a helicopter pilot, and I ran marathons. I was sent to University of California, San Diego and saw Dr. Nick Kim. I started oral therapies to battle the damage being done to my lungs. I told my then-boyfriend, Jeff, to run fast and far—this disease was going to get interesting! But he stayed by my side, and we married in September 2008.

A month later I was out for a walk and I started to feel weird. I stepped into the street and passed out. I woke up covered in blood and was surrounded by people. I had split my lip, ripped open my knee, and bruised up my face. I took a taxi to the hospital. After being stabilized, I was told that I was beyond their capacity and needed to be seen at UCSD exclusively. Dr. Kim wanted to start me on Flolan immediately. I agreed to the central IV. But I found the PEER mentor network and asked another PH patient about Remodulin. After talking with Dr. Kim quite a bit, I convinced him to let me try the IV Remodulin first.

Katie Lessard

PULMONARY HYPERTENSION

The cause of PAH is often difficult to determine. PAH can be from some known causes, such as inherited (called familial) PAH, or be caused by reasons that are never known (called idiopathic PAH). Other known causes of PAH (called associated PAH) are:

- Connective tissue diseases such as scleroderma or lupus
- Use of prescription amphetamines or diet pills
- Use of illicit drugs such as cocaine and methamphetamines
- Congenital heart defects
- Liver disease/cirrhosis
- HIV

PAH is a serious disease that at this time has no cure but there are treatments available and new medications are in clinical trials. Early diagnosis and treatment are important to try to limit the progression.

*Learn more: ATS Patient Information Series.
"Pulmonary Arterial Hypertension" New York, NY:
American Thoracic Society 2013. patients.thoracic.org*

Unfortunately, my husband had orders to Pensacola, Florida, and we had to move away. None of my doctors here were happy about it. There is a military program called Exceptional Family Member that is designed for people with exceptional medical cases, but it was too late. I was there for three years and got worse during that time.

When I was classified a Category 5 Exceptional Family Member we were able to get back to San Diego. I knew I wasn't doing so great, so I went to see Dr. Kim for right heart catheterization in April 2012, and he told me that I needed a lung transplant. I received a double lung transplant on June 23, 2012. I barely survived the surgery but was out of the hospital in 21 days. I went into chronic rejection due to acid reflux nine months post transplant. I had the Linx surgery and have been doing photopheresis. My lung capacity dropped by half from the rejection but I have gained a bit and am now at 40 percent lung capacity and am doing really well.

My experience with military health care has been a relatively good one. I absolutely would not be here today without the Pulmonary Hypertension Association and the doctors at UCSD. I was given much hope and I still have hope, even in rejection, that I will live many more years and will continue to thrive. I know there is still a long road ahead for better treatments, cures, and better prognoses, but I'm confident that there will be a better quality of life for all lung disease patients.

Katie Lessard was a patient speaker at the ATS 2014 International Conference in San Diego.



Cheryl Misak, DPhil

ICU DELIRIUM AND
COGNITIVE IMPAIRMENT

“Taking critically ill patients as autonomous agents—part of the ‘decision-making team’—fails to respect the precarious position such patients find themselves in.”

In 1998, I spent almost a month in an ICU with invasive group A streptococcus infection and multiple organ failure, including severe acute respiratory distress syndrome (ARDS).

When I awoke in the middle of the night with screaming pain in all my joints, my general practitioner surmised that I might have some kind of arthritis. An appointment was made with a rheumatologist in a teaching hospital. As I waited, I was in rapid decline. The rheumatologist took one look at me, put a blood pressure cuff around my arm, declared that I had very little blood pressure, and called an ambulance to take me to the emergency room.

They had a very difficult time getting a line in; my lungs failed; and I descended quickly into multiple organ failure. Twice my husband was told that I wouldn't make it through the night. I owe everything to those critical care physicians who worked night and day to secure my survival.

The critically ill undergo much distress. Some of that distress is physical. But some of that distress is psychological and emotional. We know, for instance, that ICU delirium is rampant in the critically ill patient population.

I recently had an interesting view on one of my worst delusions. I was able to read volume one of my thick file that included a detailed account of my ICU stay and one of the most interesting sections concerned my trying to effect what is quite gloriously understated in the literature as an “unplanned extubation.”

The log makes it clear that things are going very badly: I'm in a psychotic state; my vital signs are plunging; my husband, who has for the first time in weeks been able to go home, is called; my brother is 'agitated'; and the writing is getting progressively more urgent and panicky. An anti-psychotic is dumped into me, which only makes things

ICU DELIRIUM

ICU Delirium can be one of the effects of Acute Respiratory Distress Syndrome (ARDS), in which the lungs are severely injured. Inflammation (swelling) occurs throughout the lungs. In the lung tissue tiny blood vessels leak fluid and the air sacs (alveoli) collapse or fill with fluid. This fluid buildup keeps the lungs from working well, and a patient will have trouble getting enough oxygen in and removing carbon dioxide from the body, and become short of breath. Therapies include:

- Breathing support from a mechanical ventilator (respirator) combined with oxygen therapy
- Medicines to keep the person calm while on the ventilator; antibiotics (to treat bacterial infections); vasopressors (to maintain blood pressure); diuretics (to treat excess fluid); or blood thinners (to prevent blood clots).

Learn more: ATS Patient Information Series. "What is Acute Respiratory Distress Syndrome?" New York, NY: American Thoracic Society 2013. patients.thoracic.org

worse. Then in capital letters, the following sentence appears: “PATIENT IS TOLD THAT IF SHE RIPS THE TUBE OUT OF HER THROAT, SHE WILL DIE. PATIENT CEASES ATTEMPT TO SELF-EXTUBATE.”

Once things had become less precarious, I was very anxious to get off the ventilator and out of the ICU. I was trying to cope with roving bouts of mental distress and psychotic episodes. It was still not clear to me whether the nurses and physician were trying help or kill me. I lobbied my way off the ventilator and out of the ICU before my physicians thought it desirable. Hence, my belief that taking critically ill patients as autonomous agents—part of the “decision-making team”—fails to respect the precarious position such patients find themselves in.

Indeed, once I was moved to the ward, I found myself wishing I were back in the oasis of the ICU. I also lobbied to get myself out of the ward after just a couple of days, despite this too being thought to be not a wise idea.

I was allowed to leave. No doubt I was a difficult and strong-willed patient. But perhaps I should not have been taken to be an autonomous, competent decision-maker, and my strong wishes should have been over-ridden. Nonetheless, I was on the way home, with all that entailed. In the absence of post-ICU clinics, it entails being set adrift, with countless issues to cope with, some serious and frightening.

The fact that even ICU delirium and cognitive impairment are so difficult to assess and have such a tremendous impact on outcomes is enough to give one pause about how well we are able to tell how the most vulnerable of patients are doing.

Dr. Cheryl Misak was a patient speaker at the ATS 2014 International Conference in San Diego.



Sandra Rock

IPF AND SEPSIS

“My sepsis attacks go something like this: I wake up to a normal day and the attacks can come anywhere, anytime. All of a sudden I start to shake all over, my heart rate goes crazy, my blood pressure plunges, then the fever and dry heaves set in.”

In 2001, I was diagnosed with idiopathic pulmonary fibrosis. I could not walk across the room without being short of breath. Back then, information on IPF was hard to find except these dreadful facts: three to five years to live, no FDA-approved treatment, and no cure, except through lung transplantation.

With the help of the Coalition for Pulmonary Fibrosis, I started the Central Valley Pulmonary Fibrosis Support group with another IPF patient, Kathy. We had five members at our first meeting. Over 10 years, the group had grown to more than 60 members and I calculated that I lost 103 members and friends, including Kathy.

Health-wise I was doing well until 2009, when I had toxic myelitis, a reaction to a medication, which inflamed my spine and left me paralyzed. I was in the hospital for eight days, and they also found a brain tumor on my spine, but I was too weak to have surgery.

I was diagnosed with a Merkel cell carcinoma, a rare aggressive skin cancer. Seven days after surgery, I had my first case of sepsis, a potentially fatal whole-body inflammation caused by severe infection. My sepsis attacks go something like this: I wake up to a normal day and the attacks can come anywhere, anytime. All of a sudden I start to shake all over, my heart rate goes crazy, my blood pressure plunges, then the fever and dry heaves set in.

In January 2010, I was finally strong enough to have surgery to remove the brain tumor on my cervical spine. I made it through 2012 without a sepsis attack, and regained about 75 percent of my motor skills. But in 2013, I had a staph infection on my skin in different areas of my body, and then had my worst sepsis attack, a very high fever over 104, and several more attacks that became more severe.

Sandra Rock

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

I will never forget that during all this chaos my husband, Jim, gently stroked my hair, and whispered to me that I was going to be OK as he placed cold compresses on my forehead and lips; or my daughter Julia at my beside, reading and saying, “It’s OK, mom. I’m here go back to sleep.” Gradually my strength returned.

It’s been 10 months since my last sepsis attack. The daily dose of Keflex is keeping it at bay for now. My husband is my guardian angel and caregiver—his goal is to provide me with “maximum happiness” every day. Without his love and care, I wouldn’t be here today. Jim and I went on a cruise that we have been trying to arrange for the past six years, and we will soon celebrate 44 years of marriage.

The fear of another attack is always there but we have to try and lead a normal life. It is hardest on my family because they watch what’s happening to me: all the IVs, the swelling from the fluids, sweating from the fever, exhaustion from the heaving, knowing that sepsis is very serious and often deadly so they wait and wait.

How I’ve managed to live through six attacks astounds everyone, especially my doctors. My friends call me the “Energizer Bunny” or “Wonder Woman” and are amazed that I lug my oxygen all around—traveling, shopping, to festivals, fairs, concerts everywhere, and dancing in the park on Sundays with Jim. All I know is somebody up there is watching over me and there must be something else I’m meant to do in this lifetime—I wish I knew what it was!

Sandra Rock was a patient speaker at the ATS 2014 International Conference in San Diego.



Emily Schaller

CYSTIC FIBROSIS

“My future is the brightest it has ever been, and I am beyond excited to see where we are heading.”

I was born in 1982 to a very athletic mom, a dad with a mustache, and two older brothers with bowl cuts. The only thing that made me different from them (besides being a short girl without a mustache and bowl cut) was that I was born with cystic fibrosis. My diagnosis came after 18 months of struggles to gain weight and a constant series of runny nose and ear infections. My CF diagnosis in 1983 was grim, but my parents made a decision to not let CF stop me from living.

During the first decade of my life, I was very active and healthy. The only medications I took were vitamins and digestive enzymes. My parents performed my chest physical therapy to help loosen up any mucus in my lungs. As I got older, more medications were developed, and patients were living longer. In my teens, however, I started to develop more lung infections and lose lung function, even though I was on all of the possible CF medications.

In 2007, I decided that there was more that I could do to get my life back. I changed my diet to a whole-foods-based vegetarian diet and started to run and cycle. It took four months to be able to run two miles, but once I could, I kept running. My pulmonary function went up and my hospital stays went down. Since 2007, I have completed 11 half marathons, and I have a goal of participating in one in every state. I also cycle 1,000 miles each year and have completed a half iron distance triathlon. Exercise has definitely given me life.

While I was busy getting healthy, there was some incredible research and development being done for the first drug of its kind being developed to treat cystic fibrosis at its core.

CYSTIC FIBROSIS

Cystic fibrosis (CF) is a life-threatening genetic disease that primarily affects the lungs and digestive system. An estimated 30,000 children and adults in the United States have CF. In people with CF, a defective gene and its protein product cause the body to produce unusually thick, sticky mucus that clogs the lungs and leads to life-threatening lung infections, and/or obstructs the pancreas and stops natural enzymes from helping the body break down food and absorb vital nutrients. Symptoms include:

- Very salty-tasting skin
- Persistent coughing, at times with phlegm
- Frequent lung infections
- Wheezing or shortness of breath
- Poor growth and slow weight gain, in spite of a good appetite
- Frequent greasy, bulky stools or difficulty in bowel movements

Source: Cystic Fibrosis Foundation. "About CF." cff.org

Born and raised in “The Motor City,” I am very familiar with rock ’n’ roll, sports, Motown, and, of course, cars. Everyone knows that Henry Ford’s Model T was where it all started. The more I think about it, my newest medication, Kalydeco, is the Model T of cystic fibrosis designer drugs. It is paving the way for more drugs that will help treat the underlying cause of CF.

I was lucky enough to be in the Phase 2 and Phase 3 clinical trials for Kalydeco. While I know I was on placebo for the Phase 2 trial, it was four days into the Phase 3 trial that I realized that I was on the real thing. Almost instantly I noticed that I could take a full, deep breath without coughing, or run 10 miles and still be full of energy. I feel better today than any time in my life. Soon enough, future generations may be using only a pill or two that is designed just for their gene mutation to control their CF. My future is the brightest it has ever been, and I am beyond excited to see where we are heading.

Emily Schaller was a patient speaker at the ATS 2014 International Conference in San Diego.



Heather Snyder

PULMONARY FIBROSIS

*“To this day, I always tell doctors:
‘A patient’s will to live is stronger
than a doctor’s opinion. Don’t give
up on someone who has not given
up on herself.’”*

I am a double-lung-transplant survivor of pulmonary fibrosis, which has affected my whole life, starting with my father. He was diagnosed with IPF and was given oxygen for palliative care. With three children, he was not about to give up. So, he participated in drug studies and even posed as a doctor to research his disease at the library of the National Institutes of Health. He would have a hard time breathing and would stop a lot to catch his breath. His back was covered with severe acne as a result of the high doses of prednisone and the drug studies.

He pushed himself to the limit, and although he prepared us as much as possible, nobody understood how this disease would continue to haunt our family. I was nine when he died. The divorce rate was not like it is now, so I didn't have too many friends that I could identify with. My mother worked 10-hour days, six days a week, which left my two sisters and me home alone fairly often.

I loved track-and-field in middle school, but that came to a halt in high school because of my breathing issues. In college, I would work out and run short distances, but I stopped often to cough and catch my breath. I switched to kickboxing, but the more I worked out, the more out of shape I felt.

In 1998, an allergist diagnosed me with asthma, loaded me up with meds and inhalers, and sent me on my way. After years of struggling to breathe with routine tasks, I finally said enough, my father died of lung disease—I need a chest X-ray. HCRT shortly followed.

My pulmonologist took one look at my “clubbed” fingernails and referred me to a cardio-thoracic surgeon for a thoracotomy. The lung biopsy indicated fibrosis.

Heather Snyder

PULMONARY FIBROSIS

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

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

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

In 2011, I went into full respiratory failure during an ice snowstorm in Hershey, Pennsylvania. When the storm finally let up, I was flown to University of Pittsburgh Medical Center with only a couple of hours to live. I was placed on a Venous-Arterial ECMO and waited for a donor. Five days later I received a double lung transplant. A week after transplant, I had an emergency partial colectomy and contracted the C-Diff virus. My chance of survival became extremely low.

Just like my father, I do not give up easily. I actually told my surgeons when I planned on going home. They were surprised, but I had a goal to reach. Not only was I discharged the following week, I was out one day earlier than I even planned. To this day, I always tell doctors: “A patient’s will to live is stronger than a doctor’s opinion. Don’t give up on someone who has not given up on herself.”

The day I was diagnosed in 2009 with PF was like a bad dream. How do we not have a treatment for something that killed my father 28 years before? It was a hard pill to swallow. My father had no hope, but I had some hope for a lung transplant. Looking at the future generations of my family I know there will be more options. I am now very blessed to witness a treatment on the horizon and the progress that has been made in genetic testing.

As a patient, I have a personal interest in the future of medicine. I have become a strong advocate for PF research and know that our hard work will pay off in the long run. As doctors and researchers move forward, I hope they remember my story. I am one of the many faces of pulmonary fibrosis.

I am not just a number, I am a life.

Heather Snyder was a patient speaker at the ATS 2014 International Conference in San Diego.

Previous Voices

Patients and speakers featured in *ATS Patient Voices 2*.

“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.”

–*Larry Hoffman*
Alpha-1 Antitrypsin Deficiency

“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!”

–*Vijai Sharma*
Chronic Obstructive Pulmonary Disorder (COPD)

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

–*Mary Kelly, Addison’s mother*
Childhood ILD

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

–*Patsy Menig*
Chronic Obstructive Pulmonary Disorder (COPD)

“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!”

–*Lynn Markwell*
Hypersensitivity Pneumonitis

Previous Voices

Patients and speakers featured in *ATS Patient Voices 2*.

“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.”

–*Connell Rodden*
Idiopathic Pulmonary Fibrosis

“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.”

–*Elayne Klein*
Lung Cancer

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

–*Tina Silks*
Pulmonary Arterial Hypertension

“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.”

–*Sara Whitlock*
Lung Cancer

Previous Voices

Patients and speakers featured in *ATS Patient Voices*.

“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

“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

“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
(COPD)

“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

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

–Beth Mittelstadt
Pulmonary
Fibrosis

“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

Previous Voices

Patients and speakers featured ATS Patient Voices.

“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

“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

“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

Previous Voices

Patients and speakers featured in *ATS Patient Voices*.

“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 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

“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

“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

Rally: Lung Disease Week at the ATS

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
<http://patients.thoracic.org/lung-disease-week/index.php>.



Learn: ATS Patient Information Series

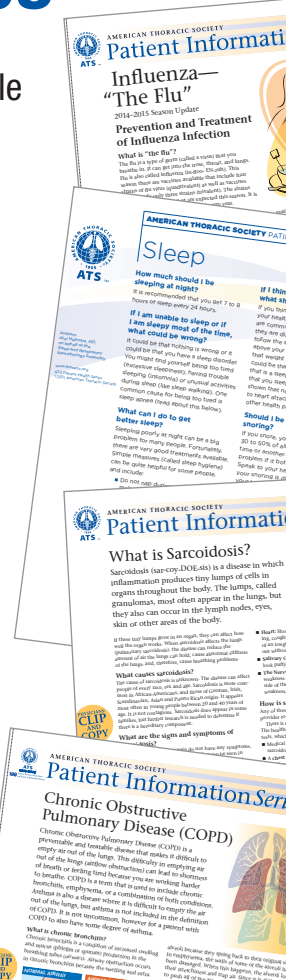
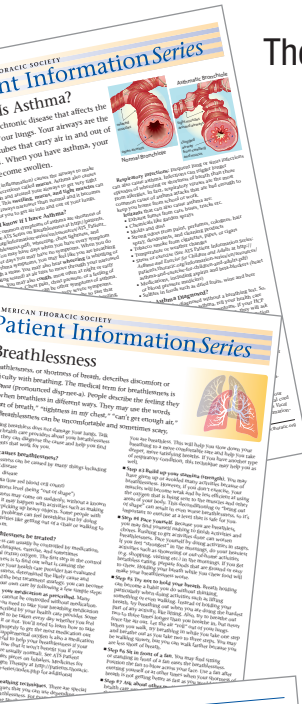
The American Thoracic Society's Patient Information Series features **free** downloadable fliers that describe lung diseases, treatments, and tests in patient-friendly terms.

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

<http://patients.thoracic.org>.



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ATS Patient Voices 3



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