




the new normal

LIFE AFTER DIAGNOSIS WITH PH





A decorative graphic on the left side of the page consists of several overlapping arrows pointing diagonally upwards and to the right. The largest arrow is teal. Overlapping its top-left corner is a smaller, light purple arrow. Overlapping its bottom-left corner is a dark blue arrow. Within the dark blue arrow, there is a light blue arrow pointing in the same direction. The arrows are layered to create a sense of movement and progression.

We have compiled the following stories to show how, while life may be different after a diagnosis of pulmonary hypertension, patients discover a way to live with meaning and find a “new normal.” Being newly diagnosed with PH can be challenging, frightening and overwhelming but we want you to know that you are not alone. There are many others waiting to welcome you and help you along the way.

This book is designed to be both a resource and a source of reflection, with stories representing a cross section of patients across gender, race, age and type of PH. We have also included journal prompts to encourage you to express what your diagnosis has meant in your life, and to find your voice and your strength.

While it won't always be easy, remember that you have an entire community of people living with PH on your side as you embark on this journey.



doug
TAYLOR

Diagnosed in 2003 at age 43, idiopathic

Each of us has our story of the journey we took to be diagnosed with pulmonary hypertension. I am no exception. Most of us have some striking similarities, but then there are differences, too.

Here are some of the things you and I probably have in common. About two years before being diagnosed, I started noticing some unusual symptoms. Unusual for me, anyway. My symptoms included shortness of breath with just moderate exertion, light-headedness, chest pains, fatigue, and fluid retention or “cankles.” You know what “cankles” are, don’t you? When your legs are so swollen that you don’t have ankles, and your calves just kind of merge into your feet.

I also had trouble sleeping due to sleep apnea and sometimes even insomnia. I was often afraid to fall asleep because I would frequently wake up gasping for air. I reached the point where walking up just three or four steps, or any kind of incline, was enough to leave me with severe chest pains and gasping for air.

Here is something else I bet we have in common; I was misdiagnosed with several other conditions before being diagnosed with PH. For me, these included:

- stress
- depression (I saw a great psychologist for the depression, but she didn’t help me get over the shortness of breath, light-headedness, or the fatigue.)
- altitude sickness, while driving in the Sierra Nevada Mountains (my “favorite” misdiagnosis).

- the last misdiagnosis, before being diagnosed with pulmonary hypertension, asthma.

At the time that I was being treated for asthma, my PCP was at a clinic in a teaching hospital, so I typically saw both a resident and the PCP. Sometimes they would ask patients to volunteer for various “tests” for medical students to conduct. My doctor and his resident ambushed me during a “routine” follow-up appointment, and I ended up agreeing to have an EKG.

During the EKG, the nurse told me that there was a problem and she needed to check with a physician to see if she should do the test over again. She thought the problem might be a loose electrode. So, she left the room and was gone for quite a while. When she came back all she said was it was “all good,” and the doctor would talk to me at my next appointment in about two weeks.

For the next appointment, my PCP and the resident came into the exam room together, which was very unusual. Usually the resident would come in and do an exam and then a little later the PCP and the resident would come in together. For this exam, they stood just inside the door like they were afraid to get close to me, both practically wringing their hands.

“Mr. Taylor, your EKG showed that you have significant tricuspid valve regurgitation.”

My first thought was, why can't doctors speak English? What did "tricuspid valve regurgitation" mean? Then, I thought it's a problem with a heart valve. They are going to tell me I need to have valve replacement surgery, and my heart sank at the prospect of heart surgery.

They went on. "The tricuspid valve is on the right side of the heart and right side heart issues almost always mean pulmonary hypertension." What a relief! I had never heard of pulmonary hypertension before, but at one time I had taken medication for hypertension, so I thought I was going to get off with just a prescription for a medication that I would have to take regularly.

But they weren't done talking. "Pulmonary hypertension is a rare disease. It is usually fatal and there is nothing that can be done for it. You are probably going to die within three to five years. You need a right heart catheterization right away."

I was stunned. Absolutely stunned. I had gone from asthmatic to terminal in just five minutes. I was only 45, and in just three sentences my doctors had taken every hope I had for a future and made it vanish—abracadabra and it was gone. Without hope, I didn't care if I had the right heart catheterization or not. I didn't see how having it would matter if the disease was fatal and there was nothing to be done. It was terrifying.

Six months later, I finally had an appointment with a pulmonary hypertension specialist, Dr. Kristin Highland, at the Medical University of South Carolina in Charleston. I can't describe what those six months were like, but I probably don't need to for those of you with pulmonary hypertension. Do I?

Dr. Highland confirmed that I had idiopathic pulmonary arterial hypertension (IPAH). And

although I knew the prognosis was grim, after one appointment with Dr. Highland and two days of testing, I saw that there was perhaps some hope.

At the time, there were three medications that had been FDA approved to treat pulmonary arterial hypertension. One was taken orally and the other two were therapies delivered by a medical pump. One of these required a central IV line and the other could be delivered either by line, or subcutaneously.

My choice was to try the oral medication first. Frankly, at that time, I was scared to try the other two medications. After just a few weeks I felt so much better. Dr. Highland showed me that there *was* hope and options for treatment. Eleven years later, there are several more PH medications available, and more still being developed.

I have been fortunate, at least as fortunate as someone with pulmonary hypertension can be. Although I had symptoms for more than two years and I had reached Functional Class IV (the worst class) before being diagnosed, my diagnosis was relatively early. I also responded well to the first PH medication that I was prescribed, and to a second medication added a couple years later.

Looking back, the prognosis of three to five years was more than 11 years ago now, and I am still fighting. My tolerance for exercise and physical activity has increased tremendously thanks to the two PH medications and my participation in pulmonary rehabilitation at a local hospital. When I began rehabilitation, a little more than seven years ago, I could only do about 18 minutes of exercise two days a week. Now I try to go three days a week and can exercise up to 60 minutes at a time, on top of other physical activities like shopping, light chores and traveling.

The two things I would like others to learn from my experience are the importance of seeing a well-qualified pulmonary hypertension specialist and the importance of holding onto hope. Hope is what keeps us going. Hope is that fire inside us that tells us maybe things can get better. Don't let anyone take away your hope, especially a doctor who may not know better.

A well-qualified pulmonary hypertension specialist can help lead the way to hope by showing us our options. Just because a doctor treats pulmonary hypertension doesn't mean that they fully understand all of the possible therapies and treatments. It is important for patients to find doctors who are familiar with all the options available. It is well worth the effort to have at least one exam with a doctor who specializes in treating pulmonary hypertension, even if it means the patient has to travel a great distance to see the doctor.

"Hope is what keeps us going. Hope is that fire inside us that tells us maybe things can get better."



Kia

THOMPSON

Diagnosed in 2007 at age 33, idiopathic



My story began after my son was born in 2006. My husband and I were on top of the world because we had tried for 10 years to have a child and never thought it would be our reality. But God blessed us with our son!

Soon after having him, I began to experience extreme fatigue, which I thought was from being a new mom. Not long after, I became very short of breath while walking up the stairs in my home, while cleaning, doing laundry and going to the grocery store. I spoke with my husband and told him I thought something was wrong.

So I went to the doctor. He did a few tests, and then called me back to tell me that my heart was mildly enlarged and that he thought I should see a cardiologist. From my symptoms, he thought maybe I could have something called pulmonary hypertension, but he didn't want me to mention that to the other doctor, because he wasn't sure. I went to the cardiologist, and he looked at what the other doctor sent. The cardiologist informed me that he didn't think I had PH. He said, "You're too young—look at all the old people here." He then sent me on my way.

I was happy, of course, because I had read about PH, and I didn't want it. But as time went on, my symptoms got worse, I was more out of breath, it became hard to care for my son and daily tasks were nearly impossible. One day while carrying my son up the stairs, I felt lightheaded and my heart was beating fast. I only remember getting into the first bedroom at the top, then waking up sweating, my son was crawling around on the floor tearing things out of his diaper bag. I remember thinking that he could've fallen down the stairs. I knew I had to get answers.

Finally, I met another doctor and cried to her that no one was listening. She vowed to find out what was wrong with me. After many emergency room visits, I finally was admitted to the hospital. By then, I had new symptoms—my belly, feet and ankles were swollen.

While in the hospital, I went to use the bathroom, and I passed out and woke up with a huge egg on my head, which left me with two black eyes. Finally, a doctor came in and told me that they thought I had pulmonary hypertension. My liver and kidneys were starting to fail: I was very sick. I was told I could have two to five years to live. I was devastated. I didn't want to leave my son and husband. Family was everything to me. But with prayer, treatment and support from friends and family, I got better.

Initially, there were things that I loved to do, before my PAH diagnosis, that I could no longer do. One of the main activities was exercising. I couldn't do that at all anymore, but now, with treatment, I have a gym membership and exercise at least three times a week! I even have a treadmill at home when I am unable to go to the gym, which makes me feel good about myself.

When I was diagnosed, I was told that my working days were over because I could barely even walk to bathe myself without help. But, since treatment, I have done some part-time phone work from home, and I've done some volunteering at a friend's business in my spare time. These activities are rewarding and help me feel a sense of independence, while still primarily being an active mommy and wife!

Life doesn't have to end with a PAH diagnosis. I have now been living with the disease for seven years. I find that keeping active has made some of my PAH symptoms even better than they were when I wasn't so active. So if you are able to get out and enjoy life, by all means, do so! I travel, shop, go out on lunch dates with friends and I'm very active at my son's school. These were all things I wasn't able to do, but I am thankfully able to do again with treatment! My advice is to never give up, to pray if you pray, to not let PH take over your life because there is hope—there are more treatments available today! I believe there will be a cure, and I believe we must think positively.

“Family was everything to me. But with prayer, treatment and support from friends and family, I got better.”

*“Ignore the
internet articles,
listen to your
body, heed the
doctors’ advice,
and rest when
you need it.”*



ICE

I am the Conqueror

Donna Ice

Nine years I've travelled this road.

In these nine years, I've cheered on one child as he earned three black belts in a martial arts discipline.

I've had another child earn her high school diploma with honors.

I've made countless trips to the hospital emergency room in the wee hours of the morning and endured an extended in-patient stay during a Christmas holiday break.

I've driven thousands of school carpools, chaperoned seemingly endless trips with the band, hauled musical instruments across football fields across the state, cheered at dozens of high school football games in the searing desert heat, attended parent-teacher conferences, band parent meetings, martial arts practices, and choir concerts.

I've trudged across soft sandy beaches, carrying bulky beach chairs and umbrellas in order to enjoy hours of relaxation seaside.

I've sung at weddings, funerals, weekly church services, and anniversary celebrations.

I've made small hikes along canyons and streams, stood at the top of a 9,000 vista, washed mountains of laundry, carried years of groceries, vacuumed floors, moved furniture, and hosted parties.

I cook, I clean, I drive, I shop, I sing, I volunteer. I am mom, wife, friend, and sister.

I am the face of Pulmonary Arterial Hypertension nine years later.

I am the Conqueror.

My name is Donna Ice. I am 52, and in July 2005 I was diagnosed with idiopathic or primary pulmonary hypertension. I started to notice symptoms in the fall of 2004—I began experiencing extreme fatigue and shortness of breath while doing mundane activities.

I currently take oral bosentan, use inhaled treprostinil, and have used a C-PAP since 2004 when I was diagnosed with sleep apnea. Treprostinil was the final key to the puzzle that allowed me to live my life as normally as I used to, prior to diagnosis.

I enjoy volunteering three days a week at two different schools. I help out in numerous capacities. In addition, I am currently looking for a part-time job that I can complete from home.

If I could go back in time and give myself some advice right after being diagnosed with PH, I would tell myself to ignore the Internet articles, to listen to my body, to heed the doctors' advice, and to rest when I need it.

There are a few positive things that PH has brought to my life. I've learned how to say "no," so as not to get overwhelmed; I learned to appreciate the ordinary and the mundane; I've learned to live in the moment; I've learned that laughter is the key to happiness and well-being. Also, I have learned that I can still exceed expectations, I recently visited Disneyland and was able to enjoy several rides that were previously "off limits" such as Space Mountain and Big Thunder Mountain, which was great, since I love roller coasters. I have even exceeded expectations by travelling to elevations above 5,500 ft. without oxygen, and didn't suffer any ill effects until reaching 9,000 ft.

To get a glimpse of my life as a mom of two high-achieving teens, please visit my blog at: www.dondidaze.blogspot.com



URRUTIA

My name is Reinee Urrutia, and I'm 32 years old. I was diagnosed with pulmonary hypertension in May 2003, just a few weeks shy of my 21st birthday. Since I was born with a heart defect and had open-heart surgery at the age of five, I was seeing my pediatrician, who knew my medical history, until I was around the age of 19. I never thought anything was really wrong with me because I associated all of my "shortness of breath" experiences with my open-heart surgery.

In high school I played basketball and volleyball and was very active in gym class, but always had a little hint of shortness of breath. Looking back, I think it was the beginnings of my PH.

After my pediatrician retired, he referred me to another doctor who was a cardiologist. Around one of my first appointments, I had a routine echocardiogram. My new doctor also took a look at my other organs just to make sure things were

working well. At that time, he noticed "stiffness" in my lungs, but I didn't think anything of it. I just thought to myself, "I'm totally out of shape—I haven't worked out since high school!" Turns out, it was PH.

Around the time of my diagnosis, I didn't really feel any symptoms. I just felt the way I always felt during activity—a little out of breath. I never felt dizzy, no chest pain, no fatigue, no REAL symptoms. So, naturally, I thought taking medication was useless, and I continued to live my life. I partied with my friends, I went to school full-time, I worked full-time, and I never let the thought of this disease bring me down because, honestly, I didn't care.

Then one night I woke up with a horrible cough and blood came up—a lot of blood. I felt it gather in my chest. My mom rushed me to the ER, and although it wasn't quite related to my PH (I just coughed way too hard and popped a vessel), it scared me. My doctor ran a bunch of tests on me and sent me

home with supplemental oxygen and increased my medication. I think at that time, I was on bosentan, sildenafil, and warfarin.

My health has been on quite a roller coaster. I've been in and out of the hospital for low O2 saturation, respiratory infections, and influenza. And once I finally started taking care of myself, I hit rock bottom. In May 2009, I lost my medical insurance and didn't have any way to get my medication.

I felt my body deteriorating, and I just wanted to die. I was hospitalized for 18 days while doctors and social workers got me on my feet. I was able to get on a bridge program that waived all of my medication costs, and I also went on disability. Being in the hospital for 18 days was my turning point. I was depressed, frustrated, and angry at the world. Every second I thought, "Why me?" I never wanted to feel that way again.

“Every second I thought ‘Why me?’ I never wanted to feel that way again.”

Diagnosed in 2003 at age 21, Congenital Heart Disease



It took me some time, but I finally turned my life around completely. I'm currently taking bosentan, sildenafil, cartizem (beta blocker), and inhaled treprostinil. I feel better than I have ever felt, even before my diagnosis! I'm able to work out at the gym, ride my bike, walk long distances, climb up to three flights of stairs, swim, and just enjoy life with my new husband.

We were married on Sept. 28, 2013, and I honestly never thought I'd see the day. My husband has been my main support during the ups and downs, and he never gave up on me. He always taught me to have something to look forward to, that way, I can focus on being healthy for that one thing.

Treprostinil is one of my newer medications, and I think it's one of the main reasons why I'm able to be so active. I started treprostinil in spring 2011. I didn't like the idea of having to carry around a huge nebulizer, especially since I tried iloprost before and that only lasted a week, but I didn't really have a choice at that point. It took a lot of getting used to. I

coughed a lot, had a sore throat, and felt nauseous, but I got over the side effects within a month. I noticed I was able to do a lot more things without having to stop or rest—like walking through the mall, fixing my bed, showering, carrying groceries, etc. Life started to get more and more easy!

I was on disability from 2008-2012, and I now work part-time as a registered dental assistant at an orthodontics office close to my home. I'm also a freelance makeup artist, and I'm quite busy during wedding season. There are still those days where I'm tired, and all I want to do is stay home in bed. That's okay because having pulmonary hypertension has made me embrace life and not take anything for granted. The things that “normal” or “healthy” people take for granted, such as fixing a bed, are actually moments when I'm proud of myself. Even while I'm doing 30 minutes on the elliptical at the gym, I always think back to the times when I could barely walk five steps without fighting for air.

It is truly an amazing feeling—I feel like a brand new me! Although I still have pulmonary hypertension, I am not living with PH—PH lives with me. I know my body and its limits, and I never do anything that I can't handle. I enjoy working out at the gym with my husband, biking, swimming, hanging out with friends and family, traveling to Hawaii, and paddle-boarding out into the ocean. I love being in and around water—that's what I enjoyed a lot as a kid—and I'm happy that I'm still able to enjoy it as an adult with PH.

Recently, thanks to technology, I started to have more contact with PH patients through my Instagram account. I get a lot of questions on how I'm able to workout with PH, and what medications I'm on. Instagram has brought me a lot closer to the PH community, and now I don't feel alone in this PHigh. I'm inspired every day by my PHigher PHriends. We are a STRONG group of people. #PHliveswithme

Have faith and take care of your body! You only get one life, so don't let yourself down.



serena

LAWRENCE

My name is Serena, and I was diagnosed with idiopathic pulmonary hypertension in December 2013. At diagnosis, I was between stages three and four.

My symptoms had come on very quickly. In October, I noticed that my asthma seemed to have flared up, which wasn't unusual because of the humidity. By November, I was struggling to go up the stairs, and I could no longer run to my bus stop after work. I had a sense that something was wrong. I went to the ER and told the doctor that this wasn't asthma, it felt different. She didn't believe me, and I was sent home with more inhalers.

"Adjusting to such uncertainty is difficult, but I am hoping that one day I can be a source of hope and inspiration for others."

Diagnosed in 2013 at age 25, idiopathic

By December, things had taken a turn for the worse. I could barely leave my desk at work to walk to the bathroom without feeling short of breath. I felt like I was going to blackout every time I tried to walk up the hill leading to my bus stop. I had woken up several times gasping for air during the middle of the night. I went to the ER again. They didn't know what was wrong, but they agreed that something wasn't right and this wasn't asthma. I eventually saw a specialist at an airway clinic who referred me to a PH specialist.

On Dec. 16, 2013, it was official—I had pulmonary hypertension. Here I was, a 25-year-old who, until the end of October, worked out for two hours every day. I did not drink or smoke. I was a vegetarian and overall health nut. I did everything to take care of my body, but for some reason I had developed a lung and heart disease.

When I was told it was fatal, I fell into a pit of depression. I felt like I had started my life, just to find out that it was ending. That June I had just started my first real job that I loved, I had been with my boyfriend for nearly four years and I felt like things were getting serious now that we were both finished school and had good jobs. I thought I would be getting married and having children soon. It felt like someone pulled the rug out from under me.

The months after diagnosis were hell. I felt awful. I would fall asleep some nights not knowing if I would wake up the next morning. I would cry all the time, and some days I would barely speak. I needed my father to carry me up the stairs, and had to use a stool to sit on in the shower.

But, it was also a few months after my diagnosis that I started to connect online with more people with PH. I learned that more of "us" are doing much better, post diagnosis. I decided I needed to do something in order to make this positive

information more available to the newly diagnosed. As such, I started the PHight or Flight Project. The mission of the PHight or Flight Project is to help share the incredible stories of those living with pulmonary hypertension, and to provide hope for the newly diagnosed.

The “fight or flight response” is a physiological response to a perceived threat or danger. It is our body’s primitive, automatic, inborn response that prepares the body to “fight” or “flee” from a perceived danger, attack or threat to our survival. I found the “fight or flight response” fitting because we all have different ways of responding to our diagnosis and PHighting.

As far as my post diagnosis story goes, I am now happy to say that I am around a stage 2, and I believe that I will continue to improve. I can now workout while using supplementary oxygen. I was in very bad shape when I was first diagnosed, so it is going to take some time to get “better.” My body needs time to recover, and I am starting to see this experience more as a second chance. I was in such bad shape at diagnosis that I know I could have gone at any moment. I am starting a second (just approved) oral medication next week, and I am hoping that this will be the last little push I need to be able to go back to work. I plan to start working part-time, and then return to full-time.

I have been on supplementary oxygen since diagnosis, but I have been able to turn down the amount of oxygen needed per minute. I also do not need to wear oxygen for sitting. I am on oxygen because my body decided to open a safety valve in my heart because the pressures were so high. My specialist said I will be off of oxygen completely in a few more months; we’re just waiting for the hole to close back up naturally. Being on oxygen is rough, it can take a toll on your self-esteem. However, the oxygen is saving my life and preventing damage so it is important that I use it.

Many of us are given certain limitations, and even timelines, at diagnosis. Medication has improved within the last five to ten years, and I don’t think the statistics have been able to consider that—our future isn’t written yet. We are a new generation that will be rewriting history and statistics about this mysterious disease, and providing more hope for those affected by PH. And, since I was diagnosed seven months ago, a few more medications have been released, along with the start of new studies and new scientific findings. More people with PH are living even longer while maintaining a better quality of life. Yes, this disease is devastating, but hope is on the way.

And, interestingly enough, I have found that the most useful and inspiring information out there has come from fellow PHers. Many of them have outlived

the dreaded life expectancy they were given and have gone on to do things that many PHers are told they will no longer be able to do, such as return to work, start a business, bike, participate in marathons, start a family through adoption, volunteer within the PH community and so on.

For every Friday that I have an inspiring PH journey to share, I post it on the *PHight or Flight Project* for “PHighter Friday.” The first PHighter Friday I posted was about a young woman who has had PH for more than 11 years. She is doing much better since diagnosis and has improved within the last couple of years. She works a physically demanding job and even goes to the gym a few times a week.

I also chronicle my life as it changes through my new diagnosis of PH, so that we can go through this together. Adjusting to such uncertainty is difficult, but I am hoping that one day I can be a source of hope and inspiration for others. I sincerely wish that others are able to find strength, hope and inspiration through the *PHight or Flight Project*.

If you, or someone you know, would like to submit their PH journey to the *PHight or Flight Project*, please email me at phightagainstp@gmail.com or visit my site at phightorflight.blogspot.com.





Kathy

MORTON

"To me I thought this was crazy. I felt great. I would just get winded when I worked out."

Diagnosed in 2005 at age 34, mild PH

In August of 2004, at the age of 34, I decided to take a taekwondo class with my children. I was a bit out of shape, but nothing too bad. As it progressed into spring, I began to notice that I was getting more out of breath and occasionally dizzy during a workout. It seemed odd that I was more out of shape after several months of exercise than I was when I started. So, I scheduled a physical with my general practitioner in March.

My general practitioner didn't think it was anything serious, but he was willing to look deeper. He decided to run an EKG just to err on the side of caution. He discovered a right bundle branch blockage. He said that it was probably unrelated, but just in case, he ordered several additional tests and referred me to a cardiologist.

When I saw the cardiologist, I was essentially told that I was middle-aged, overweight and a hypochondriac. However, he did say he wanted me to follow up with him in three months, in June, just to be sure. I nearly skipped that appointment because, during those three months, my symptoms didn't get any worse so I was not concerned. I didn't have any symptoms except when I worked out. At the follow up appointment, the doctor did a brief check-up EKG and was about to send me on my way when he went back and reviewed my initial test results. He said, "Let's do one more test while you are here." He took me to the back and they did another cardiac echo. He stood there and said, "I know exactly what you have. It's called pulmonary hypertension."

The new echo showed that the right side of my heart was enlarged. This had not shown up on the echo done three months prior. What he saw on the original test results were the pressures from the echo with the statement "mild pulmonary hypertension." At this point he told me to return in two weeks. In the meantime, I was not to exercise and to spend most

of my time in bed, resting. To me I thought this was crazy—I felt great and would just get winded when I would work out.

When I returned to the doctor after the two weeks, he told my husband and me that I needed to get my affairs in order since I had a life expectancy of 2-2 1/2 years. He ordered more tests and told me to return in another two weeks. During this time, I did my own research and decided that I needed to see a specialist. When I returned I requested a referral to see a doctor at the University of Chicago. The doctor seemed relieved that I was going to see a specialist.

I was able to get an appointment quickly and was seen in early August. At the University of Chicago, they confirmed my diagnosis and put me on sildenafil, which had just become available. At this point, however, I was still not experiencing daily symptoms. Therefore, I was encouraged to resume exercising as long as I didn't get short of breath. I chose to swim laps and work out on the treadmill.

After one year of sildenafil, my pressures went up, so I was placed on treprostinil. I was still not having symptoms, but my PH doctor was very proactive in treating me. I continued with the treprostinil until January 2012 when I was fortunate to be able to participate in a clinical trial.

In January 2013, I started taking kickboxing classes three days a week. Recently, I participated in Registers Annual Great Bike Across Iowa (RAGBRAI), and rode my bike across the state of Iowa in late July.

Due to the early diagnosis and my doctor's proactive approach to treatment, now age 44, I work full-time as a high school teacher, exercise regularly, and experience a virtually symptom-free life. An early diagnosis has allowed me, rather than PH, to call the shots in my life.

expressing your “*new normal*”

Write one characteristic or habit about yourself that you like and describe it...

This image shows a blank sheet of white paper with horizontal ruling lines. The lines are evenly spaced and run across the width of the page. There are no margins, text, or other markings on the paper.



susie
ALVAREZ

Diagnosed in 1992 at age 30, familial

The story of my journey begins in 1983. I moved to Los Angeles, met and married my late husband. I then began working full-time for the state of California in 1984. My life was full with family events, work projects, vacations, and much fun and promise.

While attending college and working full-time, I began to notice I was short of breath during my busy day. I continued to keep up with my daily work routine, but had to drop my college track and dance classes as I could not keep up with the physical activity the courses required. I continued to experience shortness of breath, fatigue, and generally felt out of shape. In June 1992, I made a mental note to visit my doctor after the upcoming Fourth of July holiday.

However, on June 30, 1992, my life was forever changed. I cashed my paycheck after work and began the drive home. Without warning, I had a crushing pain in my chest as though someone was sitting on it. I pulled over on a very busy city street during heavy afternoon traffic and parked right under the “no parking anytime” sign, which caught the attention of a parking officer. She approached my car, and I suddenly grabbed her by the chest and gasped, “I can’t breathe!” I saw the urgent look on her face as she called paramedics, who arrived and took me to Cedars Sinai Hospital in Los Angeles.

During the week I was there, I had several tests each day to find out why I was so sick. I remember coming in and out of consciousness and not knowing what day or time it was. Then on the morning of Saturday, July 4, 1992, the pulmonologist visited my room. He quickly sat down and said, “We found out what’s wrong with you. You have pulmonary hypertension, which is a fatal lung condition for which there is no cure. Go home, get your affairs in order, and the best we can tell you is that you have five years to live. We will send you home with a prescription to help you

breathe, but your illness will progressively get worse, and eventually you will need a lung transplant.” Then he left the room.

My mind went into a fog, I felt so confused and very scared. I was hyperventilating through my tears. My family lived only miles away, but I did not have the strength to pick up the phone and call them. I was alone in a cold hospital room and had just received the most devastating news someone could ever hear. I remember grabbing the nurse by the arm, crying all night—WHAT IS PULMONARY HYPERTENSION AND HOW DID I GET IT?

At the time of my diagnosis, the Internet and cell phones were not widely available to search for resources. I had no written information and no source of support to find out more information about this disease that was to end my life. My doctors informed me that there was not much information available about pulmonary hypertension and advised me to continue taking the medication I was given. For three years, I didn’t know anyone who had PH or who knew what it was. I continued to work, but the mystery of the disease always stayed in my mind.

My symptoms progressively worsened. I was short of breath, fatigued, dizzy most of the day. Eventually, I was referred to a PH center for further treatment. My life was forever changed again when I visited the Liu Center for Pulmonary Hypertension in Torrance, Calif. I met a cardiologist, Dr. Ronald Oudiz, and his mentor and PH center director, Dr. Bruce Brundage, who knew what PH was and how to treat it. He explained that I immediately needed an IV line placed in my chest with medicine attached to a pump (epoprostenol) that would open up the arteries in my lungs to help me breathe. I would have to wear this pump and medication 24 hours a day, which also required care of the IV site and mixing medication that would be attached to the pump. I refused and delayed his

recommendations for three months until one spring day when Dr. Brundage told me, “You are going to die unless you get this medication!”

I received the pump and medication in 1996 and would wear it for 11 and a half years. When I returned to work, where I was a supervisor, I made the effort to explain PH to my crew. I did not explain it very well because, at the time, I didn’t understand the disease very well. My crew and other co-workers were unsure what I was going through, but they understood I was sick and that they would see me walking around with the pump in a backpack. With this medication, I experienced four IV line infections and many hospital visits for treatment.

I was also referred to UCLA Medical Center, evaluated and placed on the transplant list for a double lung transplant. I remained on the transplant list for 13 months. During that time, I moved to the top of the donor list, and one evening in the middle of the night, I was called in for a transplant. I arrived quickly, was prepped for surgery, witnessed the lungs being flown in and delivered to the surgery room. After some time waiting, the surgeon told me that the donated lungs were not viable, and I was sent home. Shortly thereafter, I was removed from the transplant list by my PH doctor who informed me the medicine was improving my symptoms and quality of life. What I did not know at that time was that this IV medication (epoprostenol) and medical pump was the ONLY FDA approved medication available at that time to treat PH.

Finally, on Oct. 31, 2007, I was taken off the epoprostenol pump therapy and walked out of the hospital – PUMP FREE; two new medications had been approved. Finally! I was only on oral medications ambrisentan and tadalafil, and it felt so good to be free of the pump.

I continued to attend local support groups and even PH-related events nationwide. In 2011, I became the co-leader of the Greater L.A. PH support group at UCLA Medical Center/VA Hospital.

I’m also a proud founding member of a new group, SuperHeros: Long Term Living with PH, a group for those who have been living with pulmonary hypertension for eight years or more.

In 2012, I attended PHA’s Congressional Luncheon at the Capitol in Washington, D.C., to ask my senators to co-sponsor the Pulmonary Hypertension Research and Diagnosis Act (H.R.2073). We also taught them the importance of early diagnosis.

I’m often asked why I do so much for PHA. My answer is that I was once a newly diagnosed patient, living alone, and so afraid of dying without understanding PH. I’ve survived beyond the five-year timeline that I was given 22 years ago. My journey was not easy by any means. As a long-term survivor, every day is a blessing for me.

After my diagnosis, I discovered that others in my family have PH, too. The power of knowledge learned at PHA’s International Conference brought us together to raise awareness and hope for a cure!





james GREY

My name is James Grea. I write stories for children with a magical fountain pen named Solomon J. Inkwell. I spend a great deal of my time writing about Boogey Men and things that go bump in the night. I love spooky tales. They are my favorite. My monsters are harmless and even quite fun because they are imaginary. But there are times when monsters can be real, and sometimes they can even live inside of you. Unfortunately, I discovered this almost three years ago.

On Thursday, March 29, 2012, I arrived at outpatient registration at the hospital at 5:30 a.m. with my sister and my partner. I was surprisingly calm. I swear to you, I don't know why. Maybe I had convinced myself that I was going to receive good news. The mind has an odd way of masking the unimaginable, especially when it is standing right in front of you. We sat in the lobby waiting to be called. My sister patted my leg and smiled at me. My uneasiness wasn't apparent to those who didn't know me, but she knew I was shivering on the inside.

Soon they called my name and my chest tightened at the sound. I took a deep breath and followed the nurse, who showed me to my curtain-walled preparation area. Two older gentlemen were on either side of me, hidden behind their own curtain walls. They were apparently having the same procedure as I was that morning. I tried my best not to listen to the questions the nurses were asking them. "When did you have your first heart attack?" "Do you have a living will?" I stared at the ceiling tiles trying to think of other things, things I would do after I received the good news—movies I would see, food I would eat.

Right heart catheterization — it is a fairly simple and relatively painless procedure where a measuring device is inserted into the right femoral artery of the leg. It travels to the heart where pressures are measured and dyes are injected. It's a common test that can locate blockages in the arteries. It's also the only exact method of diagnosing pulmonary arterial hypertension (PAH).

There are instances where this disease is inherited. I come from a fairly large family consisting of eight children in total, of which I am the youngest. By the time I was 21 years old, I had lost my oldest brother (diagnosed age 24, passed age 27), my nephew, who was 10 months younger than me (diagnosed age 11, passed age 12), and my mother (diagnosed age 60, passed age 61) — all of them victims of PAH. You

could say that the disease has stalked my family and me all of our lives. We all spend a great deal of time praying it doesn't catch us.

My new adventure had begun with a routine EKG that had shown an abnormality, which led to further testing. They eventually found mild enlargement on the right side of my heart. I heard the word "right" and immediately knew what was happening to me.

The orderly wheeled me into the catheterization laboratory and began to prep me for the procedure. I welcomed the sedative and tried to focus on its feeling. Through my wooziness, I could see the large monitor above me. On the screen was my beating heart, and I watched as the thin catheterization line traveled into its chambers. I took a deep breath and tried to relax myself. Moments later it was over.

The rather abrupt cardiologist I had been working with had no experience with PAH. So, he thought nothing when he rounded the table and said nonchalantly, "Well, it looks like pulmonary arterial hypertension." Stunned, I sank into tears. And when I did, the doctor disappeared. He was so stunned by my reaction; he apparently had to get away. I, too, wanted to get away, but there was nowhere to go.

Yes, the Boogey Man under my bed had found me at last. I continued to cry for three solid days. To me,

“For today, I am alive — we are alive. And what could be better than that?”

Diagnosed in 2012 at age 38, familial



my life was over. I wondered how much time I had left. How long would it be before I couldn't walk to the bathroom without passing out? I was no longer James Grea. I no longer wrote scary stories. I was now a zombie who wandered around my house sobbing. Finally, I decided to reach out to PHA. I began reading everything I could on PHA's website, trying to find any sign of hope.

I obtained the *Envelope of Hope*, which contained a wealth of information. I went one step further and called PHA's Patient-to-Patient Support Line where I spoke with some great people who were living with PAH. I learned that in today's world I had great chances, far better chances than others in my family had. In the past, there was nothing at all that could treat PAH. But I learned that times had changed.

Then I began to do things I hadn't done in a while. I began to talk more with God. I began to see the value in little things in my life that I had taken for granted. I began to realize that not one of us is guaranteed to see tomorrow. At any moment, we could receive that test result that could drastically alter our lives forever—or even get hit by a moose.

While I have been living with a PAH diagnosis for more than two years, I've really been living with PAH all my life. By March 2015, I will be the longest living PAH survivor post-diagnosis in my family.

I am fortunate enough to still be able to get up every morning and go to work. I pay my bills and enjoy time with my partner, friends and family. I try to live life as normally as I possibly can, with one little exception: I now have an infusion pump that administers treprostinil therapy connected to me at all times. I have found very creative ways to deal with the medication—changing my site, taking a shower, wearing the pump, and so on. Sometimes, it's like it's not there at all.

Recently, I have also started tadalafil, so I am extremely fortunate to have my PH specialist and PHA. I am also fortunate to have the Caring Voice Coalition, for without their assistance I could never, in a million years, afford my medication.

I often wonder what the future holds for me. Will my symptoms continue to worsen over time regardless of the medication? Will I eventually have to stop working? If so, how will I survive on disability? Will they find a cure soon? Yes, those questions sometimes haunt me. I shake it off, and carry on. I am certain there is a part of my mind that denies what is happening to me. It is a coping mechanism, you see. Regardless, I have a heart catheterization each year, and the pressures aren't exactly perfect, but they are getting better. Anything lower is a step in the right direction, right?

I've never said, “Why me?” Why not me? What makes me better than anyone else? Absolutely nothing. So, I choose to have hope. For the only alternative is to lose hope, and that is unacceptable. Why am I telling you this? I'm really not sure. Maybe I want you to come along with me. Maybe I want us to be like old friends, sitting in a room, drinking coffee, and talking about the frailty of life. Maybe I just want you to listen. I'm not certain. I only know that I am moved to tell you these things because a writer must write. It could be possible that you, too, are afraid — for yourself or someone you love— and maybe it will make you feel just a tad less lonely to know that somewhere out there I am here, being afraid along with you, living life as best as I can with this disease that no one understands. Who knows what tomorrow holds. Miracles happen every day, and my miracle could be right around the corner...our miracle. Ironically, being diagnosed with this disease could very well prove to be my miracle.

So, I will look toward today and value what I have in my present world, my present health, and my present life. For today, I am alive — we are alive. And what could be better than that? And yes, I am finding my identity again. My name is James Grea. I write stories for children with a magical fountain pen named Solomon J. Inkwell. And I just happen to have pulmonary arterial hypertension.



kimberlee FORD

Diagnosed in 2009 at age 30, idiopathic

I was diagnosed with idiopathic pulmonary hypertension in July 2009 at the age of 30. I believe I started experiencing symptoms six months to a year prior: extreme shortness of breath, syncope, severe chest pain, dizziness, fatigue, cough and headaches. I worked as a nurse, and on a daily basis, I would walk and stop several times to rest on floors, sidewalks, steps—wherever I could find a place to sit. I was short of breath, and my limbs felt like they were bricks.

Later, I would learn that adequate oxygen was not being supplied to my body. In addition, I was incapable of doing chores, and I struggled to bathe and groom myself; therefore I missed days of

work and school. I knew something was medically wrong with me. So, I scheduled an appointment with my primary care physician who then referred me to a pulmonologist. The physician did electrocardiograms and pulmonary function tests, and concluded that I had exercise-induced asthma.

I knew it had to be something else because my mother is a severe asthmatic and told me, “I do not experience what you do on my worst days.” I continued to seek medical attention from hospitals and physicians, but was always told it was asthma, so use inhalers and lose weight—later it would be noted the weight gain was from fluid retention. I was also told that I was fine, and what I was experiencing was all in my mind. I was told to exercise more, but at the time, I was going to Curves at least three times a week. And, finally, I was told that I must be using illegal drugs because of the symptoms (I never used illegal drugs or smoked). I thought I was going crazy because I knew I was sick, but the medical community was of no help to me—I was dumbfounded. Even though I struggled daily to get out of the bed, I continued to pray and hope whatever was wrong would be revealed soon.

Each day I went to work, I had a rough time with my patients at the hospital because I could not perform my duties, due to my limited mobility and declining health. However, I took several breaks throughout my 12-hour shift, and I needed assistance with minor physical tasks. On one particular day I was very symptomatic and fainted, so my best friend, who was also working, and another nurse took my vital signs, which were normal with the exception of my oxygen saturation and heart rate. I determined I had to go home, so we immediately notified the Charge Nurse, and my friend took me to my car in a wheelchair; she offered to take me home, but I insisted I could do it.

I drove about 40 miles at night on the Capital Beltway around Washington, D.C. to my parents’ house

because I knew I could not make it up the stairs to my apartment. As soon as I arrived I immediately told my mom what happened, so she called 911. While she called, I vomited and fainted; my other family members assisted me to the couch. I was taken to the emergency room by ambulance and tests were periodically done, but doctors did not believe I had a serious condition, and the hospital wanted to release me. My mother immediately spoke on my behalf for them to transfer me to the hospital of her choosing, and they complied with my mother’s demand. Upon arrival I became unresponsive and tests were urgently performed, including various x-rays, echocardiogram and right heart catheterization, which lead to the diagnosis of pulmonary hypertension by two physicians who were also pulmonary hypertension specialists in separate facilities. It was a joint effort by the hospital’s emergency medicine, cardiology and pulmonary departments.

My family was told that if the first hospital had released me, my chances of survival were very low because I was not responding to treatment. Phenomenally, I survived, and once I became responsive, I was told the news that ultimately changed my life forever. “You have pulmonary hypertension.” I was clueless on what this meant for my future.

I am now 35 and living a “new normal.” The PH specialists were aggressive with treatment, so I am currently on combination therapy: treprostinil, ambrisentan, tadalafil and a calcium channel blocker. I also use several other medications to help alleviate the side effects of the PH medications. I used supplemental oxygen 24/7 at the beginning; then after doing pulmonary rehabilitation for three months, I was able to switch to during exertion, or as needed. After two more separate admissions to the pulmonary rehabilitation program, I am fortunate that I now only need supplemental oxygen during air travel.

Through the grace of God and the medications, I am able to successfully continue my educational studies; I am working on my third degree. Most importantly, I am determined not to allow PH to stop me from pursuing my goal. If I give PH the power, then I lose respect for myself.

Also, I happily lead four support groups because I believe no one should face this journey alone. In addition, I actively participate in PHA events and disability awareness events within my community: I have spoken at PHA's Congressional Luncheon and two of PHA's International PH Conferences; I volunteer by consulting with disability departments at universities to improve facilities and programs for those living with seen and unseen medical disabilities so that they may have an enjoyable college experience despite their challenges. I also speak at community events at local centers, and at my church which has awareness services biannually for PH (it helps when your father is the Pastor).

Additionally, I am honored to be a member of the Social Media Advisory Board for PHA, a member of PHA's Generation Hope, a blog for young adults with PH, and a peer mentor for United Therapeutics. I am also the co-founder of a very active and successful patient-led PH group on Facebook, PH Family, a growing global forum since 2010 for patients and their families to voice questions and concerns about this life-threatening disease and to support each other. I am excited to be an active participant in the PH community.

Pulmonary hypertension has taught me to live life; I am more appreciative of my quality of life rather than the quantity. I am blessed to have loving parents because without them and my faith, I would not have been able to overcome the challenges I have with my diagnosis. My parents are always encouraging me to be a better person than I was

yesterday, by making my dreams a reality and helping others; this is my journey and I must be a testimony, so that others will see that there is life after being diagnosed with a chronic illness. It is what you do after the diagnosis that defines who you really are.

As a PH community we must stand together and be "empowered by hope." Every day I want there to be a medical cure for this disease, but until there is, I must cherish my moments with my family, friends, and church family because they are a wonderful support system that wants to see me blossom. Pulmonary hypertension has changed my life drastically, but I am determined to ensure it does not negatively impact my life entirely. I will continue to do the impossible and aspire to be a phenomenal woman so I can lead by example. I know what is important to me; therefore, I will not allow PH to have power over my destiny. People often say, "I wonder what she will do next, because each day is an adventure and obstacle for her to overcome." Although it took time, I have accomplished a lot since the diagnosis, and I will continue to do so with guidance from God and my parents.

My next goal is to return to the working world and have the most important titles: "Mrs." and "Mommy." My journey is not yet complete — to be continued!



"Pulmonary hypertension has taught me to live life; I am more appreciative of my quality of life rather than the quantity."



noël

HOLLY

*“Now, in all honesty,
I still have what we
call our ‘PH days.’
But, I am moving
forward in life and
not letting PH get
the better of me.”*

Diagnosed in 2008 at age 53, CTEPH

My name is Noël Holly, and I am a PH patient. I am 59. Prior to being diagnosed with PH, I was someone who was relatively energetic. My last job was as an office manager, so I multitasked and was always “on the go.” Outside of work I stayed busy with gardening, teaching Sunday school to active children, and even dancing. I lived by myself, so there was, and still is, the daily upkeep and running of a household. All in all, a rather normal life!

In 2007, I was diagnosed with Type 2 diabetes. Now, of course that has nothing to do with PH, but one of the ways to help manage one’s blood sugar is to exercise. So I started walking. Not miles — just around a small lake in a local park. It helped my blood sugar management, but I also found that I began to cough a lot. At first I chalked it up to being out of shape for this kind of exercise and left it at that.

Then came a morning when I got up to go to work as usual, and I found myself feeling very weak. My thought was, “I can hardly breathe. I think I need to go to the ER and have them give me some oxygen, and then I will go to work.” I drove to the local hospital, marched into the ER and said, “Hi, I am having a hard time breathing. I need some oxygen, please.” I was given a bed, and they tested my O2 saturation which was in the low 80s. I was put on oxygen and they took me for several tests including an MRI, CAT scan and blood work. Then, I waited. What I anticipated they would say was that all was well with the test results, and I was good to go.

At last, the doctor on duty came and informed me that I had massive blood clots in both lungs. Initially, I was in disbelief—I figured that blood clots would be something that someone who smoked, or had smoked, got, and I did not smoke. I started to get rather indignant and said that I needed to go to work and that the test results were ridiculous. But, they were insistent that this was real and that I needed to be admitted to the hospital as soon as a room could be prepared for me.

Finally, I said to the staff that I would be admitted but asked if I could go home first and get my jammies and some other things to make my stay, which I expected to

be a short one, a little more comfortable. Their response was, “No—we are very concerned that if you leave, you will not make it back.” I was incredulous.

I was admitted, and other tests were done, including a VQ scan and an ultrasound scan of my legs. The doctor said that I had initially had a blood clot (DVT) in my leg that had traveled up my leg and through my heart and into my lungs. It was a silent clot because I never had the symptoms that one might associate with a DVT, such as redness, bruising, swelling or pain.

After 10 days in the hospital and being hooked up to an IV with heparin, I was released and started back to work. I continued to cough, but it seemed to be less than before, so I was happy that my condition appeared to be improving. I went to see the doctor for follow-up visits, and the doctor seemed pleased with my progress. However, when he saw me, it was usually after I had sat in his office for 20 minutes or so, and he didn’t witness the coughing and shortness of breath.

As 2008 continued, the coughing had once again worsened, and all other normal life functions were almost impossible. I had no clue that this wasn’t normal for what I had been through. I was ignorant and continued to just trudge along in life—through quicksand.

Finally, one day I went to see the doctor, and he said, “You have a disease called pulmonary hypertension. It is rare, incurable and life expectancy is usually three years.” He told me that the echo showed that both the left and right sides of my heart were enlarged. I remember saying, “Well, a large heart was good for the Grinch, isn’t it good for me?” Sighing deeply, he told me it wasn’t and then gave me some literature and the name of a specialist at UCLA to see, and that was that.

Needless to say, I was a bit numb. Once home, I got online and looked up what I could find about pulmonary hypertension. Yep. There were those words: rare and fatal. I hadn’t yet seen the words “unless treated.” More tests were done, and I went to see the doctor at UCLA. He showed me the x-rays of my lungs and

explained to me that my PH was a result of the blood clots, also known as chronic thromboembolic pulmonary hypertension (CTEPH). He told me that my clots were inoperable because what was left of them had become too fibrous in both lobes. He also mentioned the test called the right heart catheterization.

My doctor also prescribed a sleep study, and I learned that I also had sleep apnea. It was so severe that in the span of an hour and a half, I had stopped breathing 70 times. I did the second half of the study with a CPAP on, and in the same amount of time, I stopped breathing seven times. Thus began my love/hate relationship with the CPAP. It took me forever to be able to sleep with it, but I knew the numbers and, so, I stuck it out.

Eventually, my doctor started me on bosentan. He was very thoughtful, treating both the disease and my fears. It took some time, but I did notice that I could do more without feeling as if I could hardly breathe. I was able to increase my activity.

I began to learn about what many of us call “The new normal,” or “life, but different from how we knew it.” My doctor also put me in touch with one of his other patients, and it was such a relief to talk to someone who “got it!” This other patient and our doctor began a local support group, and I met even more patients.

In June 2008, the Pulmonary Hypertension Association had one of its *PHA on the Road* events near where I live. This one-day, educational conference was my first time meeting many PH patients and hearing of different therapies, ideas, opportunities, etc. I was quite shy and did not interact as much as I would have nowadays, but the event opened my eyes to a lot of new information.

In 2010, I attended PHA’s International PH Conference and Scientific Sessions in Anaheim, Calif., and was so amazed at how many people had come from all over, even from other countries, once again affirming that I was not alone. This was very important too because,

since the disease can be very socially alienating, often those closest to us do not understand it. So, to meet others who “get it” is a valuable thing.

Over the years, other PH support groups in my area came into being, and I have attended several of them. I also connect with other PH patients on social media such as Facebook. This is something that is a positive—connecting with people that I probably would not have ever come in contact with had circumstances been different. These are special and wonderful people who understand and support one another.

As new PH therapies emerge, my management strategy only gets better—recently, my doctor has added riociguat to the management of my PH. It has once again taken some time to notice any improvement, but it is happening gradually. For example, one day, I had to drop my car off for some work to be done, and I ended up deciding not to wait for three hours, but to walk home, four blocks away. And then, I walked back. That’s eight blocks, which for me, is an improvement indeed! And, yes, I still huffed and puffed, but I did not feel as if I was trudging through quicksand!

Now, (cue the angels singing), I no longer have that chronic coughing! When first diagnosed, I had to stop gardening, but I have been able to get back into it. I just garden differently than before—no more power gardening. It is actually better than therapy for me—and I get tomatoes. I also decided to learn how to preserve the garden bounty through canning, and I have found that to be therapeutic as well. Also, I have always sung at church, and though it took a while to get to where I could sing without having coughing fits, I can still do so! In fact, I think singing is great therapy for your lungs.

I feel empowered and encouraged when I can give back to my own community. The PH community is special to me, and I found that I wanted to do more to raise awareness of this disease, both in the community and in the medical field. When I learned of one event held in my area, Taylor’s Wish 5K Walk/

Run, held in honor of a young PH patient who had passed away at 4 and a half, I asked some friends to participate on my behalf and they did. “Team Noël” was born, since I, myself, could not walk or run. The first year there were six members of the team, the second year there were about 25, and the third year, there were more than 40 proud Team Noël participants! We were the largest of all the teams.

I have also volunteered at other events, such as the Wojo Swing 4 A Cure Golf Tournament, and have participated in the Team PHenomenal Hope Unity Walk. And outside of PH-related events, I am connected with a local organization to help homeless children and their families.

In May 2014, I had the honor and pleasure to speak to a large room of doctors and medical professionals at the American Thoracic Society’s Annual Convention as a patient speaker. I have also met with my Congressman to help educate him about PH awareness and to ask that he support the Pulmonary Hypertension Research and Diagnosis Act (H.R.2073). I am not someone who normally would do this, but with the tools available on PHA’s website, I was able to step out of my comfort zone. I was so elated after the meeting even though he did not agree to sponsor the bill at that time. I have followed up with his office and will continue to do so.

Being diagnosed with PH was scary, and it took time for me to come out of the “gloom of diagnosis.” Now, in all honesty, I still have what we call our “PH days,” but I am moving forward in life and am not letting PH get the better of me. I have learned many lessons of faith, growth, strength, and love along this journey, and by the grace of God, I am not through with learning!

If one person can be encouraged by my story, then I am encouraged as well. And as the saying goes, “Never give up! Never surrender!”



melanie KOZAK

"I always tell people that I wouldn't change my life or my diagnosis. I feel like it's taught me so much."

Diagnosed in 1998 at age 22, idiopathic

In 1997, at 21 years old, I had shortness of breath, palpitations, chest pain, and I was having seizures—my awesome body decided fainting just wasn't cool enough. On May 1, 1998, I was diagnosed with primary, or as they say now, idiopathic pulmonary hypertension. Currently, I am on bosentan, sildenafil and treprostinil. But I have been on almost all of the therapies approved for PH.

It's funny because I have had PH for 17 years now, and I really can't remember life before it. I always tell people that I wouldn't change my life or my diagnosis. I feel like it's taught me so much. I've learned to slow down, be more patient, be more tolerant, and have compassion for others. I have also become active in the PH community, volunteering for the Patient-to-Patient Support Line, running a support group, fundraising and advocating.

I feel like PH has truly given my life purpose! I've made so many friends and have helped others. I have also been blessed because others have reached out to help me. Having PH has made me a better and stronger person.

Even more so, PHA's biennial International PH Conference is another positive in the life of a PHer! It's like a family reunion where you aren't "the sick one." I also speak with friends living with PH on a daily basis through Facebook, I speak with some local PH friends weekly (sometimes daily, too), and I even meet up with them for lunch or a shopping date!

If I could go back and give myself advice when I was first diagnosed, I would advise

myself to remember that everything happens for a reason—just go with it and follow the doctor's orders. You will find your new rhythm. When I was diagnosed in 1998, the only medications available were epoprostenol and calcium channel blockers, so things were very different then. I was told I had to take things easy and never elevate my heart rate, but I am happy to say that this is no longer the case. I take yoga classes, I am going back to pulmonary rehab and I enjoy playing soccer with my nephews—all things I was told I could never do with PH.

Also, my biggest accomplishment with PH is traveling. Since being diagnosed, I can't even count how many places I've gone, from the west coast to east coast, down to Houston and up to Canada, and even venturing out of the country to Bermuda, the Bahamas and Mexico. The list goes on because I've really made it a point to live life to the fullest and travel as much as possible. Flying with oxygen and traveling with my pump and all my pills is not going to stop me! And I hope it won't stop *you* either.

expressing your “*new normal*”

What is your favorite hobby? Why do you enjoy it?

[illegible]



Diagnosed in 2011 at age 29, CTEPH

It was dusk, and the men at the Afghan National Police headquarters were turning on the generator that powered the electric lights in the station. We sat in a haphazard circle that stretched across the furnitureless room, our bellies full of kabob and naan bread. My interpreter and I had spent the week working with the Afghan men who formed the police force in this village, and had earned ourselves an invitation to join them as they unwound from a day's work. Now relaxed, we sipped green tea and played a traditional board game, similar to the air hockey I had grown up playing with my brother in Virginia. Late into the evening, the pleasant sound of overlapping conversations mixed with laughter and the thrum of Afghan pop music. It was a perfect evening.

Two days later, I lay in a hospital bed on Bagram Airfield. I had collapsed the morning after returning from two weeks in the field, falling in and out of consciousness for more than four hours before I was able to belly crawl to a phone and call for help. I was physically unable to stand. The doctors were initially confused as to why a seemingly fit and healthy young woman would present with a blood oxygen level of only 58; frankly, I was confused, too.

At 29 years of age, I was in the best shape of my life. I had been in Afghanistan for seven months, participating in all-day patrols while carrying an excess of 90 pounds of gear at least six days a week. On my days "off," I hit the gym hard, working to develop the balancing muscles needed to support my heavy kit; and

running six miles a day. Prior to my deployment I'd played rugby, competing at the national level, and was an avid runner. Then, in April, I was walking to the chow hall and noticed I was breathing hard. I chalked it up to the poor air quality on the base and made a mental note to start wearing a dust mask during my runs. Two weeks later, I began feeling strange while on patrol. With my heavy breathing drawing looks and teasing remarks from my buddies, I volunteered to go back to the vehicle, where I blacked out. I woke up shortly before we returned to post where a visit to the "med shed" yielded an asthma inhaler.

A month later, I was patrolling another village on top of a mountain. At 17,000 feet, in the most sensitive area I'd worked in, I suddenly found myself unable to breathe. To the distress of my patrol sergeant, I took off my protective body armor, shameful with the knowledge that I was putting the entire patrol at risk, but no matter how much air I took into my lungs, it felt like I wasn't breathing. We aborted the mission and I was taken back to the post hospital. This time, I was given a bag of fluid and two asthma inhalers.

This last collapse occurred on June 17, two months from the start of my symptoms. I was scared, but it now seemed that the CT scan had identified the problem: I had developed five pulmonary emboli (PE). The prognosis was not good. Within 48 hours I was in a second hospital, this time in Germany. The doctors in Germany stabilized me, releasing me after a week into the hands of a young Staff Sergeant who had been assigned to see that I returned safely to the U.S. Unbeknownst to me, the doctors in Germany had expressed to my superiors their concern that I might not survive the trip.

In October, just four months after my evacuation, I was cleared to resume my exercise regime. Returning to my daily runs, I found that I struggled to complete a single mile. I also noticed chest pain from the exertion, but decided that these were things I needed to work through, and that exercise could only improve my condition. After just three weeks of exercising regularly I became convinced that my body had beaten the PE, and decided to see some non-military doctors for a second opinion.

Thinking back, Dr. Svetlecic must have thought I was a crazy person. Here I was, less than four months after a major PE event, hopping up on her table as I announced that the purpose of my visit was to have her confirm my clean bill of health, and sign the paperwork clearing me to return to Afghanistan. Instead, she hospitalized me. And there I sat, two days before Halloween, looking sullen as she explained something I had never even heard of: pulmonary hypertension. She brought in a cardiologist and a hematologist. The three of them showed me images of my chest and explained that, while my lungs were inflating, very little of the oxygen was reaching my bloodstream. I had effectively lost use of 75 percent of one lung and 25 percent of the other. My heart was working overtime

to try and push through the blockages, and I was at immediate risk for heart failure. My parents were called and flew in for the conversation that no parent ever wants to have. I would not be returning to Afghanistan. I was a ticking time bomb. My heart could go at any moment. At best, my condition was only sustainable for another five months. I was going to die. And then, I saw my father cry.

Two days later I was released from the hospital. Quite simply, we were told, there was nothing else they could do. My parents and I returned to my apartment in a fog. That afternoon, as we sat in my kitchen not knowing what to say to one another, Dr. Svetlecic called. She had done some research, and had learned of a center at the University of California, San Diego. UCSD had pioneered a procedure called a pulmonary thromboendarterectomy (PTE). As an otherwise healthy CTEPH patient, I was a good candidate, but it was a long shot. Their waiting lists were long. I gave her permission to refer my case.

On Jan. 20, 2012, I underwent a pulmonary thromboendarterectomy at the Sulpizio Medical Center at UCSD, once again embarking on a long, hard road. It was painful; more painful than I ever could have imagined. Dealing with the pain from the fresh surgical site and the loss of my former life, launched me into a period of deep depression. Bit by bit over those first three months, my outlook improved. I joined a church in my neighborhood, I made some friends, and then, an absolutely

wonderful thing happened: I met someone special. Really special. Someone who could accept me, scars and all, and who, after I opened up to him about my illness, asked only one question: "If something ever happens, what symptoms do I look for, and what can I do to take the best care of you?"

Last September we were married, and I began my married life with the profound sense of being extremely blessed. Despite stretching across three countries and five U.S. states, my diagnosis had come relatively quickly, within six months, as opposed to the average of 2.8 years that most pulmonary hypertension patients go without diagnosis. I was fortunate to have been in the care of a pulmonologist who was unwilling to give up on me, even when her hospital's administrators said there was nothing to be done. I was also fortunate to be connected to UCSD's PTE program, which only began in 1990 – 22 years before it saved my life, and to have access to the Sulpizio Cardiovascular Center, which opened less than one year before my surgery. I was blessed by all the researchers and all the patients who had come before me, who had done the hard work that led to my happy ending. I had a second chance at life, and at a good life, and I was grateful to have it.

Then, on Nov. 6, 2013, we got the news that my 29-year-old cousin, Christina, had died in her sleep of right heart failure. A member of my own family had died from undiagnosed CTEPH! My reaction was a myriad of emotions: I felt profound guilt for

not knowing that Christina had been sick and for not making sure that everyone that I knew was aware of the symptoms. I felt overwhelmed by the extreme disparities in our outcomes. I felt grateful to be alive, and then instantly ashamed of that emotion.

Christina's death changed my life, but not in the way you might imagine. In my grappling to understand how this could have happened, I finally began to study the disease in earnest. I learned that not only do most pulmonary hypertension patients go an average of 2.8 years before being successfully diagnosed, but that the average survivability without treatment is also 2.8 years. I learned that, in addition to a curative surgery for CTEPH, there were 9, now 12, approved therapies for pulmonary hypertension. That week I made a decision to dedicate my life to changing the patient experience for patients like Christina, and for patients with PAH and IPAH, and PH with associated conditions. Earlier this year, I honored that pledge by joining PHA's staff and leading it's *Early Diagnosis Campaign* – www.SometimesItsPH.org, a dedicated effort to reduce the time between symptom onset and accurate PH diagnosis. Every day I work to reduce the knowledge gaps and the barriers contributing to the high rate of missed and delayed diagnoses. I do it for Christina, I do it for myself, and each day, I am humbled by the incredible people who stand beside me.

"I had a second chance at life, and at a good life, and I was grateful to have it."





"Looking back, I remember how angry I was after my diagnosis. Today, all I feel is gratitude for everything I've received."

Diagnosed in 2009 at age 48, idiopathic

perry

MAMIGONIAN

My name is Perry Mamigonian. I am 53 years old and live in Fresno, Calif. I first started to experience shortness of breath in 2001. We had tickets to our local college football games for years, but that season I noticed that I could no longer walk long distances or climb the stadium stairs without stopping to rest. I just blamed it on my weight, age and lack of exercise. For seven years this was my only symptom, and whenever I mentioned it to my physician he told me that I needed to exercise and lose weight.

But after eight years of just shortness of breath, I started to experience more severe symptoms. My legs and abdomen were so swollen that my clothes and shoes would not fit. My shortness of breath became so severe that I could not even go back and forth to the mailbox without stopping to rest. I developed a chronic hacking cough and felt as if I had the flu and couldn't shake it. But when I started having dizzy spells I called my doctor immediately. At the time he was treating me for systemic hypertension, and he thought it was due to the dose of my medication. He recommended that I cut the dose in half and see if the dizziness subsided, which it did.

March 11, 2009, began as a typical day for me as I drove off for work. Shortly, my life would change forever. I started coughing badly and felt very dizzy. I had just exited the freeway and was headed toward a major intersection as I blacked out. My last conscious thought was that I was not going to survive. When I woke up, I was completely surrounded by white light. I thought, "It's true—when you die you do see a white light." But when I stuck my hand out, it was just the airbag that had covered my head. Fortunately, my car drifted off the road and struck a parked, unoccupied truck. I was relieved that no one was hurt, except me. A passerby called an ambulance and the paramedics took me to the nearest hospital.

At the hospital they began treating my injuries, but when I explained that I had passed out they started running tests to determine the cause. One test after another showed nothing until they finally performed an echo of my heart. They immediately saw the enlarged right ventricle and called in a cardiologist. Fortunately, he was familiar with the signs of pulmonary hypertension and called in a pulmonologist. Subsequently, a right-heart

catheterization was performed, and my diagnosis of Class 3, severe idiopathic pulmonary hypertension was confirmed on March 16, 2009.

All I understood about my diagnosis was "hypertension." I expected them to give me a pill and send me home. But, the pulmonologist, Dr. Vijay Balasubramanian, was also the head of a new PH program in Fresno. He was very sincere and honest as he explained to me that my condition was very serious and that, unless I began treatment immediately, my condition would worsen. Before I made the decision to start therapy I wanted to learn everything about PH. I studied every brochure and DVD I was given. After spending 16 days in the hospital, I was released and began my life on subcutaneous treprostinil therapy.

My life had been turned upside-down by my diagnosis. I was single, living alone and unable to return to work. Because I had blacked out while driving, my license was suspended. I felt helpless and completely dependent on the care of relatives and friends. I was angry for what this disease had done

to my life. There was not a support group in my city, and although there were other resources available to me, I was too stubborn to use them.

Soon after I started therapy, I received a phone call that would change my outlook. My specialty pharmacy had a patient advocate who contacted me to check on my needs and concerns. Her name was Kelli Danner, and she convinced me to attend a patient education conference in Northern California that summer. She felt that I would benefit from the education and opportunity to meet other patients. I reluctantly agreed to go, but I'm happy to say that the experience changed my life. Everyone was so welcoming and friendly. I made friendships with other patients that continue to this day. I learned so much about everyday life with this disease that my outlook about the future became positive.

After I returned home, I soon began to feel the positive effects of my therapy. I was walking more and feeling less symptomatic. Soon after, I was prescribed a second therapy, tadalafil. I was also diagnosed with sleep apnea and started CPAP therapy. All my therapies, including the water pills, were having a positive effect. I was losing weight and gaining energy.

In the fall of 2009, a PH support group was started in Fresno by two nurses in the UCSF/Fresno PH program. It was my first opportunity to experience the support group process. At first I was reluctant to share my feelings because I didn't want to burden others with my problems. But soon I realized that many people were expressing the same thoughts as me. I came to understand that support groups are safe environments where attendees could express any feelings or thoughts without fear of judgment. As our support group grew, I became more interested in helping out at the meetings. And, in July 2010, I was asked to become the patient co-leader of the group.

It was 16 months since my diagnosis, and I never thought my path would lead me this far. I developed an interest in advocacy and raising awareness and joined the grassroots 435 Campaign organized by the Pulmonary Hypertension Association. With the help of our support group members, we visited with representatives in eight districts and convinced six of them to co-sponsor PH legislation. We also met with local leaders during awareness month and worked to spread awareness in the community. Additionally, since 2012, I have chaired our fun walk committee. Last year our Fresno "Six-Minute Marathon" fun walk and BBQ hosted 180 people and raised more than \$5,000. This year we anticipate more than 200 people will attend.

I have been fortunate to attend three of PHA's International PH Conference and Scientific Sessions. At the 2012 Conference in Florida, I enrolled in United Therapeutics' PEER Mentor Network which taught me to be a mentor for other patients who were on, or considering, my therapy. It has been very rewarding to connect with other patients and share my personal experiences of living with PH on my therapy. I hope that I am able to give back for all that I have received and help others take advantage of the support services that I was once too stubborn to use. The program has given me the opportunity to travel and share my story with others and experience people and places I never thought possible. I am very grateful that I have the opportunity to make a small contribution in fighting this disease.

My family instilled a sense of community service in me at a young age. After diagnosis, I had to give up those activities, but eventually I regained the energy and desire to get involved again. Today, I chair a committee for a community service organization that awards college scholarships to deserving students. I also volunteer my time for a university organization that raises funds for student-athlete scholarships.

Personally though, there was one part of life I assumed would fall victim to having PH. At diagnosis I was single and not dating anyone, so I assumed that the complications of living with a chronic disease made starting a relationship impossible. Eventually, I learned that I was wrong. At the 2012 PHA Conference in Florida, I met another patient who was also from California. We had known of each other for a while because we are both support group leaders. Susie and I soon developed a friendship based on our similar experiences and mutual admiration. Before long, it bloomed into something more as I developed feelings for her beyond friendship. I was reluctant to admit my feelings because I still believed my PH was an obstacle. Susie suspected it, and coaxed me into admitting my feelings. She explained to me that I was wrong and that having PH is not a reason to deny ourselves the things that make life worth living. If anything, we need to embrace life even more. I am very grateful to have her in my life.

Looking back, I remember how angry I was after my diagnosis. Today, all I feel is gratitude for everything I've received. I'm grateful for my supportive friends and family, my healthcare providers and the "team" we've become, our support group, and PHA for all the opportunities and hope that they provide. But most of all, I'm grateful for finding love when I thought it was not possible anymore.

If I could go back and give myself advice when I was diagnosed, I would say: "It's perfectly normal to feel angry or depressed. That's part of the process. But you can empower yourself through knowledge and gain control of your life again. Having PH does not have to mean the end of your life; it can be a new beginning."



"Who knew I would be around to see the number 44? Not me and not the pulmonologist who first diagnosed me almost exactly six years ago to the day, but my family did."

Diagnosed in 2008 at age 37,
idiopathic

August 2008 was a great month. I was running nearly 40 miles a week. Next marathon on the notch was to be Gasparilla in Tampa, Fla., my home city, for the second time. Over the next month of training, my running got much slower, and I had to stop for frequent walk breaks. I could tell there was something wrong with my breathing but didn't think anything of it.

In October, I couldn't even run to the end of my street without stopping. I finally called the doctor. After a brief visit with my primary care provider, I asked for a referral to a pulmonologist. I picked a name out of a hat. I had no idea who to see; all I knew was that I wanted to be seen fast so that my running could get back on track. After calling around, I found one who could see me the very next week. He said that my symptoms sounded like exercise-induced asthma, so he prescribed two inhalers, and we made a follow-up appointment in two weeks.

After the first week I felt no relief, so I called the doctor, and he recommended I inhale more often while running. That did not help, which I told him at the second visit. He suggested I get a chest x-ray and referred me to his cardiologist friend down the street. I had an echocardiogram performed by the cardiologist, and my follow-up appointment with the pulmonologist was the day before Thanksgiving. We needed to get out of the appointment quickly because my entire family would be arriving in just 12 short hours. I brought my husband and 6-year-old daughter to the appointment. Why did I bring them? Thinking back, I have no idea what initially made me want to have them there. Remember, I thought I only had exercise-induced asthma.

The doctor came into the room and told me I had pulmonary hypertension. I had no idea what that was and just sat there. I may have even started laughing at that point. To tell you the truth, everything went numb as soon as he said, "Marla, this is serious business. Your life expectancy is three to five years, and you need a lung transplant."

All I heard was that was it for me. I was a 37-year-old woman with a husband and 6-year-old daughter. Who would take care of my daughter? Who would drive her to school? Who would buy her clothes and watch her get married? We left. What else was there to do at that point? Of course, we called our families and told them the news. Everybody was shocked and in deep sorrow. Imagine how our Thanksgiving was.

On Thanksgiving Day, my father told me about a pulmonologist who specialized in PH. My cousin Scott recommended him to us. Without that recommendation, who knows what kind of care I would have received. That pulmonologist was Dr. James Gossage at the Medical College of Georgia in the Georgia Health Sciences University.

Three weeks later, in the middle of December, we were in Augusta, Ga. The initial diagnosis was confirmed, but there was no life expectancy given this time. I was started on sildenafil at 20mg, three times per day.

After a couple months, I was passing out while walking at work, so I needed something stronger. I had also gained 25 pounds. My cardiologist in Tampa never said a word about it and told me to stop eating so much. When I got back to Georgia in February 2009, I was in right-heart failure and in critical need. My pulmonary pressures were in the 100s, and Dr. Gossage immediately admitted me into the hospital and started epoprostenol within 24 hours. At that time, I also started bosentan. After two weeks in the hospital, I was finally ready to go home and be a mommy again. Oh yeah, and did I mention I work full time as a speech language pathologist? The truth was, it was tough. I was crying every day because I was afraid I was going to die. It's just hard to get past that.

I started seeing a therapist and walking for exercise. I started attending support groups through PHA. I attended two groups, one in Sarasota and one in Lakeland, Fla. I went back to work. My mood started to get better and was becoming somewhat normal

again. In September 2009 after seven months on epoprostenol, I had a right-heart catheterization. My pulmonary pressures had gone back to normal. Me? Normal? "Maybe only from the neck down," Dr. Gossage would say.

In October, my doctor and I began weaning me off the pump. In December, I started on treprostinil, an inhaled medication. Since February 2010, I have been on the inhaled medication ambrisentan and sildenafil. I feel great. I exercise five times a week; I take my daughter to amusement parks, the beach, roller skating and bowling, and I work full time.

I also attended my first PHA International PH Conference and Scientific Sessions in California in 2010, where I got the nerve to start running again. It is a very slow process, but I feel awesome. I attribute my mental and medical success to my very supportive family, the support of the PH

support groups, and my awesome pulmonologist Dr. Gossage. I couldn't have gotten to where I am today without their help.

Now, four years later as I write this, it's Nov. 10, 2014 – my birthday. Who knew I would be around to see the number 44? Not me and not the pulmonologist who first diagnosed me almost exactly six years ago to the day, but my family did. They never stopped supporting me or giving me positive encouragement. This is a disease you cannot deal with alone, and it's unbelievable that I get to continue my story.

So, after attending the PHA Conference in California, I was motivated to start a local PHA support group here in Tampa and haven't missed an International PH Conference since. PHA is a great network and offers many ways to support people with PH. The support group has been going strong for five years now, and we have close to 30 members.

As for me, I am still working full time as a speech language pathologist in a skilled nursing facility where I meet and see people everyday who battle with their own ailments and sicknesses. I feel grateful to be where I am now.

I am happy to say my daughter is 12, and I feel lucky that one day I will be around to see her get married, and I will be a grandma! (Not any time soon, of course.) We still love to travel, take cruises about once a year, and I exercise on a regular basis. Of course, I won't be running a marathon anytime soon, but I exercise the best I can without passing my limits.

Again, I owe everything to my family. Thank you.



expressing your “new normal”

Dear Past Me... Dear Future Me...

[illegible]



maddy SHANAHAN

“My new normal was fantastic and remains so to this day.”

Diagnosed in 2007 at age 36, CREST, PH

In 1996, I was 25 years old, living in Las Vegas with my military husband of three years. Life was great. Then what I affectionately called my “dead white finger” appeared. My right index finger began turning white for extended periods of time. I saw a doctor who recognized it immediately as Raynaud’s, and I was eventually diagnosed with CREST Syndrome, a form of scleroderma.

My husband and I adjusted to the news and life went on. I finished my teaching degree and taught school. We had two boys, and the Air Force moved us every couple of years. CREST was always there in the background, but it didn’t stop me from living a very full life.

After the birth of my second child, I never seemed to recover completely. I was out breath after carrying laundry upstairs or pushing the stroller up our street. I tried to work out more, but nothing seemed to help. Doctors treated me for asthma, but for 18 months my symptoms progressed and I became very ill.

In October 2007, at the age of 36, I was diagnosed with pulmonary hypertension secondary to CREST. It hit my husband and me like a ton of bricks. We were told I had two to three years to live. I remember going home to my two sons, ages 6 years and 20 months at the time. I lay on my couch and watched them play, heartbroken at my new future.

I immediately began treatment. Oral meds, inhaled meds, enough diuretics to dose a small town, and a lot of faith and prayer set me on the right path. By April 2008, I had my life back. I began to live like a normal military wife with two children. I carpooled, went to church, drove to soccer practice, volunteered at school, and participated in all the usual events as a commander’s spouse. My new normal was fantastic and remains so to this day. We have since adopted a little girl from Taiwan, moved several more times, and I even completed a 5K. I currently homeschool all three of my children. I am immensely thankful for my crazy, busy life. This whole “new normal” thing turned out better than I could have ever imagined.

I do still remember how terrifying it was to hear the diagnosis. I remember scouring the Internet for stories of hope, and they were few and far between.

I constantly browsed the PHA website to arm myself with information. But there are some things I wish a fellow PHER could have told me.

First, no doctor can predict your expiration date, so don’t let them. The treatments and medications available now present a very different outlook to people with PH. Second, it is OKAY to have a bad day sometimes. They happen to everyone so just accept it and know that tomorrow is a fresh start. Third, be proactive. Research, ask questions, and be your own best advocate. Lastly, don’t let PH and scleroderma or any other disease define who you are or what you do. You might have to adapt activities you enjoy, or find ones that fit your lifestyle better, but it’s worth the effort to figure it out. I’m a happy mom and wife who just happens to have PH and scleroderma. The diagnosis did change my life, but I am so much more than a diagnosis, and so are you.

Follow Maddy’s blog: www.shanahan5.blogspot.com



Kevin



MAYHOOD-PASKAWYCH

“All the world’s a stage, and all the men and women merely players. They have their exits and their entrances, and one man in his time plays many parts.”
- William Shakespeare, As You Like It

Truer words could never have been spoken. As long as we are here, we have a part to play, and over time that part changes. Despite the years of training in the performing arts, despite the time I spent as a director and actor at community, collegiate and professional levels, I never really knew the meaning of these words until pulmonary hypertension invaded my life.

I have always enjoyed entertaining people. A part of me has always, even from my early childhood, thought that if you are making someone laugh, cry or gasp, then you are keeping them from the worries of the world. For that instant that they are watching, nothing else matters. While I have had many other jobs and another career, theatre has always been in my life in some way, shape or form.

When my family and I realized something was wrong with my health, my first concern was if I would be able to continue working for our local community theatre and working on the projects that I had started. As I started down the rabbit hole that we have all traveled, that leads to a PH diagnosis, I quickly realized, as I am sure many of you have, that I had to do something to keep sane, to keep my mind off of what was happening.

My wife, Karen, had to keep me motivated some days to just get me off the couch, and at times, I think she and my father both saw the life slipping away from me as much as I felt it was. While my family encouraged me as best they could, I struggled to find a meaning for why I was stricken with this disease. I never did find a reason, and I finally learned that getting tied up in the search for “why” was fruitless.

I struggled to cope, until one of my friends came up with an idea. After some gentle encouragement from Karen and my family, I agreed to hear my friend out about his idea and discovered that not only was it something I had wanted to do for years, but something that we had even gotten to the planning stage of, at one point, in my earlier days. I can’t say this project saved me, but it did open up doors, both mentally and physically.

The project my friend proposed was to start our own Shakespeare in the Park Festival in Marietta, Ohio. I jumped at the opportunity, figuratively, and we began meeting about the project in May 2012, right in the middle of my journey between “something is wrong” and “you have pulmonary hypertension.” Our initial group was four men—two theatre professors, the director of a theatre restoration project and myself.

In the beginning, a major part of me thought of how it would be a legacy for me to leave my community, and that it would be helpful to get my work off the ground, but I secretly doubted I would make it to an opening night for the festival. Remember, I was still going down that rabbit hole, and it was getting darker and darker, and no matter how bright this speck of light was to me, that light felt as though it was getting further and further away.

I worked on the project. I gave my two cents worth, with Karen dutifully taking me to the early meetings. All the while I hoped that just one of my ideas would make it to the final festival. That way, some little part of me would survive in this piece of art we were creating for our community.

On August 26, 2012, I found the bottom of the rabbit hole. Once there, I discovered that there was light at the bottom. That light came in the form of Dr. Curt

“You can choose to make PH this horrible dark cloud that covers your sky, or you can choose to see it as a traveling companion, a whiny, draining, annoying travel companion, but something that YOU can have some control over.”

Daniels and The Ohio State Medical Center. Dr. Daniels gave me my diagnosis, but then he also gave me hope. I returned to Marietta, and very quickly was on the first of my three medications. Tadalafil came first, in September, followed by treprostnil in October and bosentan in December. Slowly, I was realizing that this was a whole new world, and quickly, I learned that I can make this disease part of my life, not just let it rule my life. I continued to work on the festival, but with a renewed sense that I would make it to opening night. At least with only some reservation, I saw it as a real possibility.

In December 2012, I took on a last-minute directing project in my local theatre. Still on the road to recovery, I was incredibly unsure if I could do it, but three friends in the cast, including Karen, gave me the encouragement I needed. The rehearsal process was a rough road—the original director had dropped out due to medical problems and left no notes. It was only three weeks to opening night and the actresses in the show, “A Piece of My Heart,” did not have their movements down, had not rehearsed in weeks, and we needed to get the show under way. It was rough. At times I thought, “How can I do this?” but I never said, “I can’t do this.” The show made it to opening night, and we were subsequently selected to represent our theatre in a regional festival. All the while, Shakespeare and our “future” festival continued in the background.

Then, on closing weekend of our show, Teresa, a friend and fellow PH patient, encouraged me to run for local office. She had become part of my support network and her encouragement was, “If I think I can do it, then I think you could do it.”

On Feb. 1, 2013, I filed to run for City Council President, in Marietta. Around the same time, I had started writing for the Generation Hope Blog, a blog for young adults with PH, and was becoming involved in the online PH community, even being asked to sit on PHA’s newly diagnosed patient

advisory board. Then as the winter broke into spring 2013, I realized how much work running for office was, and I admit that I was more than a little apprehensive about what I had just gotten myself into. Physically, though, I was improving. My six-minute walk distances had gone from 290 meters before my diagnosis to almost 600 meters in about nine months, and I felt better than I had in the last few years. But still, I was worried about whether or not I could keep up the pace required for even a run for local office.

And, as spring changed into summer 2013, I learned that I could. Karen pointed out to me one day, “Look at all you are doing—we can do the festival.” I hadn’t even realized it, but the conversation and preparation work with the Shakespeare festival was continuing. I was still working at the theatre, and was assigned two directing slots for the forthcoming season. I was also still working with my actresses, who had won our regional theatre competition and who were now heading to the state festival, as well as campaigning for office. In short, I was probably more involved than most healthy folks I know.

The more I worked, the more I realized I wasn’t letting PH and fear rule me like it had in the beginning. I was learning to live with it. If I was tired, I was learning to back off and let someone else take the reins. I was learning to work with PH.

Pulmonary hypertension is not fun, but I have seen PH as a challenge and I have learned that there are different ways you can live with it. You can choose to make PH this horrible dark cloud that covers your sky, or you can choose to see it as a traveling companion, a whiny, draining, annoying travel companion, but something that you can have some control over. I know it is not a great analogy, but that is how I realized it. PH was going to be there, and I had to deal with it, but I had to do it on my terms. I just had to learn how to do it.

As summer turned into the fall of 2013, I was learning how to do just that, with some help from my friends. Karen was learning my moods and my signs of getting overworked. She became diligent toward my medical schedule and was generally the voice of reason when I let my mind get ahead of my body. My father, David, managed to step in and learn as well. He had been there from the beginning and was trying to learn to adapt as well as the rest of us, and it was his “work smarter, not harder” mantra that really started helping us.

During the fall door-to-door campaign route for my office run, I applied the rules. Karen helped, and did so much as to start at the top of hills so that we could work our way down. We also planned out our schedules according to the weather as, I am sure many of you know, a humid day could mean a reduced ability for me to get door to door. With every campaign day, we learned something new, and all the while, all my other projects moved right along, right up to Election Day.

Tuesday, Nov. 5, 2013, is a day I will remember, and not for the reasons you might think. My Shakespeare group, as we had started calling ourselves, learned we were tentatively a “go” for our summer 2014 date. And, of course, there was an election. I lost, but not by much, and in our community of 14,000, it is not uncommon to see election results where winners and losers are separated by 500-800 votes, particularly when a well-known candidate is running. I was unknown, running against a known name. I was the underfunded underdog of a candidate, having been outspent by slightly more than 2 to 1, and in the end I did lose but by just 3 percentage points, or about 200 votes.

I couldn’t have been more proud. I looked at what I had accomplished, and it was amazing that I was standing there with the small success that I had. How can you be sad about something like that? Even a loss can be a personal victory when the conditions are right. And, all the while, I still had

the Shakespeare festival to work on, and I still had things to accomplish with our community. So, it was time to buckle down, look at what I had learned, and apply it to every other aspect of life as well.

Through the winter, we continued to work and meet on the Shakespeare festival. I directed my first musical show, and I started lobbying my local and state politicians regarding PH. I made phone calls, sent emails, whatever I could do to convince my representatives that they should throw their support behind us.

In March, we learned the final piece of our puzzle was in place for Shakespeare. In April, I was part of an organized effort to get PHA public service announcements on-air and to conduct a “media blitz” of Canadian and American news outlets to get them to recognize PH. In May, I directed my third play within 12 months and had the honor of speaking to an FDA panel with other patients to get FDA direct their efforts to finding medications that best benefit patients. Finally in June, that small spark of light in the darkness years before, had exploded.

The first of June is probably one of my new favorite days—it is the day that actors report for the “Shakespeare by the River” Festival. Nine student actors from all over the country, as well as six actors from our local community, banded together to begin rehearsals for our first ever show, a production of Shakespeare’s comedy *Love’s Labour’s Lost*. During rehearsals, we had intense sessions, and as the date drew closer to our opening, the city threw a wrench in our plans, and other issues threatened our production. But, in an odd way, PH came to the rescue.

Living with pulmonary hypertension, I have learned one very important lesson; nothing is worth getting stressed about. You sit back, you breathe deep, and you go to work. For every problem, there is a solution, and you won’t find said solution if you are freaking out about it. PH taught me that, and I applied that lesson and did what I needed to do. I lobbied our director to make some changes, rallied the actors to help, and when a local actor dropped out, I took up his role because it was the quickest solution to the problem. As a cast, we overcame many issues, and I have PH to thank for helping. Bet you weren’t expecting that, were you?

On July 11, 2014, we opened our inaugural “Shakespeare by the River” production in Marietta. Personally, I couldn’t believe it. Two years prior, I had started this project thinking it would be nothing more than a distraction as I waited for the worst news of my life. I hadn’t actually prepared myself to make it to opening night, but luckily everything I had done in the last two years, everything I had accomplished while I was working on this project, prepared me for it.

I think it is ironic that the character I played has the last line in the play. Spoken to the audience, the Don Armado character informs the audience that, “The words of Mercury are harsh after the songs of Apollo. You that way: we this way.” He is informing the audience that the play is over, the audience to one direction, and the players to the other. You can make many interpretations of the line, at least within the context of the play, about what he is saying. For me, though, it is much more in a very symbolic way. Closing night of the festival, with that line uttered, I closed the book on Act I of my life with PH.

Many patients choose to view their life before the disease with this rose-colored haze that is sometimes true and sometimes not. The words of our doctors change that. The words that create our diagnosis are harsh after the “disease free” life many of us had lead before. This is where we part ways, a normal life that way, we must go this way. It is just another act in the play. We do get to choose our characters though, sometimes consciously, sometimes not so much.

Since my diagnosis, I have chosen to be an activist for my community, an activist for the patient community, a director, an actor, a husband, a father, a business owner. I am a patient, but that doesn’t mean I have to play just that one role for the rest of my time on the stage known as life. There are so many characters to choose from that fit, and we all can be more than just a patient. I challenge you to find the role for you. We all have multiple parts to play on this grand stage. Who do you want to be for Act II?



expressing your “*new normal*”

What do you consider your greatest accomplishment to date and why?

[illegible]



stacey GAUSLING

Diagnosed in 2006 at age 39, Lupus, Mixed Connective Tissue Disease

How has PH changed my life? That is a loaded question for all of us. But, with any PH story, it all starts with a diagnosis. It was the summer of 2006, six weeks before my 39th birthday. It was a Friday afternoon, and my husband and I had planned to leave for a weekend trip to the Gulf Coast. I got home from my busy day of being a rehab medical sales representative, and started up the stairs to my bedroom. About half way up, I felt like my lungs were caving in and that I had just run a marathon. I rested for a couple of minutes and finished the rest of the way. There it is...in those few seconds, my life changed forever. My background is in occupational therapy, and I knew deep down that something wasn't right. But, I decided to still go on our trip and that if the shortness of breath remained, then I would go to the ER.

The drive was about four hours. The plan was to scope out beach properties and just have a relaxing weekend. The drive was fine, but our hotel room was upstairs. Self-check – yes, still short of breath. We had a nice dinner, which I am sure was loaded with salt. My abdomen started to swell. The next day, my walk from the flat parking lot to the flat beach resulted in more shortness of breath I made it halfway and told my husband that I couldn't walk the rest of the way.

I got back in the car, and we decided to head home and I would call the triage nurse. But first, we needed to gas up. I wanted to throw out some trash and walked from one side of the car to the other and again became short of breath. I went from “being

concerned” to “scared.” Another four hours back home and then to the ER. They ran some basic tests, but never did an echo so they sent me home saying, “We can't find anything wrong, but you might want to follow up with a cardiologist for a mild EKG abnormality.”

Most people celebrate the words “we can't find anything wrong,” but those are the most frustrating words to come out of a doctor's mouth when you KNOW that something is wrong. I went home and opened up my “Living with Lupus” book and looked up pulmonary conditions and also researched “right axis deviation.” Both searches came up with pulmonary arterial hypertension, amongst other conditions, but that one seemed to stand out. I made my appointment with a cardiologist and said, “I want to be ruled out for pulmonary arterial hypertension.”

“Do you know how rare that is?” He asked. “If you have this condition, you will likely be the only patient I will ever see during my career with it.” I was thinking...”Just humor me.” But I hoped that he would go his entire career without diagnosing anyone, including myself.

The echo was supposed to be followed with a stress test. After the echo, I lay there with my half paper gown and sticky ultrasound gel for what seemed like a long time. The ultrasound tech finally came back and said, “You can get dressed. The doctor wants to talk to you.” It was a very short conversation: “You have PH. Your pressures are close to 100, which is severe.” I asked about resources, and he said he didn't know of any, but that I should put my affairs in order. At this point, I had already read all the scary stuff that everyone else reads on the Internet before discovering the PHA website. I started crying and said, “Okay, thank you,” and that was that.

Life went in slow motion for the next month. I have always been a planner, so planning for death made sense to me. I wrote a living will and discussed end-of-life plans with my husband. That's where some newly diagnosed PH patients think their story will end, but there's more.

After telling my rheumatologist about my diagnosis, he researched and found a PH specialist in San Antonio. My life went from slow motion to a flurry of tests, prescriptions and a glimmer of hope. I didn't have time anymore to plan on dying. I had to adjust to oxygen 24/7, large medication boxes, a tube coming out of my chest, managing my job juggling oxygen tanks, the CADD pump and IV line, and running to the bathroom several times a day.

I trudged forward each day with my job, in order to keep my health insurance, but looking back, it was a blessing in disguise. I would wake up some mornings and think to myself, "Why am I doing this?" and indulge myself in self-pity. But, after a day of working with clients who had severe head injuries, strokes and spinal cord injuries, my self-talk would change to, "But, I can still walk...and think...and do some things for myself."

Despite having PH, I knew the key to sanity was turning self-pity for what I didn't have any more, into gratitude for all that I still had. My husband did everything that was humanly possible to help me get through each day, from packing up my car, ordering my meds, cooking meals and doing household tasks. I "therapized" myself and applied treatment to myself that I would normally give to others. That meant frequent pursed lipped breathing, pacing activities throughout the day and week, conserving energy for important things, and adapting my environment.

It wasn't just my efforts but also those of my husband's that got me through those first few years. After three years, and I was able to ditch the daytime oxygen. It's funny how resistant we can be to wearing something that is essential for health because of the outward signs of what it means. But, I waited for the green light from my doc, and when my body was ready, I began to only use the oxygen at night time.



Once I was no longer "planning on dying," the topic of adoption had come up again. But, we decided to forego that route after my husband told me, "If I had to choose between having kids and having you here longer, I choose you." We were in our mid 40s and there was a future to consider.

Now that my health had reached a "new normal" point, we were able to start making some definitive plans. "What is important at this stage?" we asked ourselves, and the answer pointed to moving back to California from Texas, where the weather would be better for my health. The two-year plan was to transition our jobs to allow us to live anywhere, to simplify our lives as much as possible, to reduce stress and to adopt as many healthy lifestyle practices as we could.

We arrived back in California Christmas Eve of 2012, and with the move came more exercise, healthier eating, fresher ocean air, and finally having the time and desire to become involved in the PH community. After six and a half years, I was finally attending support group meetings where I made new phriends and found others, more like myself.

I had a new perspective on living with PH as an almost long-term survivor, pump wearer and working PHer. I started a private Facebook group called "Pulmonary Hypertension: Maximizing Independence and Living." I thought that, using my therapy background, there had to be something that I could contribute to the community. It didn't take long for my PH activities to escalate to participating in PH advocacy, fundraising and PHA events.

Now, it is the summer of 2014, and it's been eight years of living with PH. We sometimes tell newly diagnosed PHers that they will learn to live their new normal. That is true, but I can also share that there is hope to live a "Near Normal." This is where I am today.

I am at a crossroads of transitioning from Living with PH to Living Near Normal. I work, travel, spend time with friends and phriends, and try to find a purpose in all that has been given to me. No more planning for death. My only plans these days are for the future, and it's looking pretty bright.

"That's where some newly diagnosed PH patients think their story ends but there's more..."

expressing your “new normal”

Dear PH...

[illegible]

This image shows a single sheet of white paper with horizontal ruling lines. The lines are evenly spaced and run across the width of the page. There are no margins, text, or other markings on the paper.



PHA'S PATIENT RESOURCES

FIND A PH DOCTOR

Look here for a list of PH-treating physicians from around the world. While you're on this page, watch the short video and read the FAQs on how to talk to your doctor.

www.PHAssociation.org/FindADoctor

NEWLY DIAGNOSED? NEW TO PH?

Find information specifically for you as you are getting started on your PH journey.

Find information about PH, coping and day-to-day living created by experts in the field.

www.PHAssociation.org/Patients/NewlyDiagnosed

TREATMENT ACCESS PROGRAM

This PHA program is here to help you. Find answers to questions about Social Security Disability, Medicare and more with our online insurance guide.

www.PHAssociation.org/Patients/Insurance

PULMONARY HYPERTENSION: A PATIENT'S SURVIVAL GUIDE

Our comprehensive guide to understanding and coping with PH.

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Join one of our 245+ support groups nationwide to connect with people who understand what it's like to live with PH. To find support groups in your area: www.PHAssociation.org/SupportGroups

We also offer telephone support. Join one of our monthly telephone support groups for patients, parents and caregivers. From the comfort of your own home, connect, learn and share strategies and experiences with others who get it. www.PHAssociation.org/TelephoneSupport

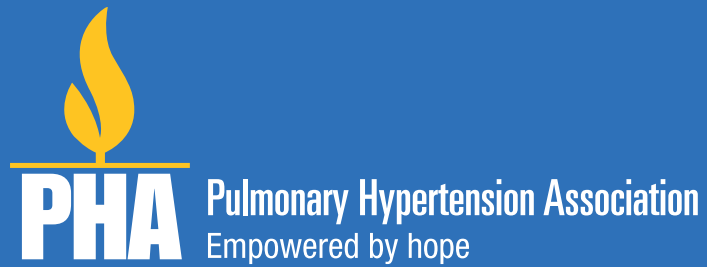
ONLINE OPPORTUNITIES

Our discussion boards, email groups and chats help you instantly connect with other patients and family members. www.PHAssociation.org/Community

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