



Pulmonary Hypertension Association - Ireland

# PHA NEWS

Volume 1 : Issue 4 : December 2016



## CONTENTS:

- Changing landscape of PH
- Bio Banking
- World PH Day
- Fundraising Activities
- Patients' Stories

## ***Message from the Editor***

It has been a long time since we produced the last PHA News. As the cost of professionally designed & printed booklets is very expensive – we are producing this in house version which can be produced quarterly. The opinion of our readers is very important to us – so please let us know what you think.

### **New name for our Newsletter**

A new name for our Newsletter is also an idea. PHA Europe name their newsletter **Mariposa Journal** and PHA UK booklet is **EMPHASIS**. We are inviting our members to make a suggestion for our Newsletter. A voucher for €50 will be given to the person who comes up with the best name. An independent adjudication panel will be set up to decide on the winning suggestion.

### **Support Group Meetings**

I am very happy to report that our PH Support Group meetings have gone from strength to strength in recent times. During 2016 we held 7 meetings in Dublin, one in Mullingar and one in Cork. A number of venues were tested throughout Dublin including the Red Cow Moran Hotel, St Alphonsus Centre, Drumcondra, Crofton Airport Hotel and the Crowne Plaza Hotel Blanchardstown. We think the Crowne Plaza is the most suitable and have had recent meetings there. Having tried evening meetings – we found that attendance was better when held in the morning – however we are always open to suggestions and will be happy to discuss. Minutes of meetings are circulated to those on email and we encourage our members to submit their email address if they have not already done so. The strength of our group is only as strong as its members and we would encourage you to attend group meetings. Those who have been attending on a regular basis say they find the meetings very beneficial and many have formed new friendships. Together we can create new ideas to increase awareness of PH. If you would like to set a group up in your area – we will be very happy to help you in any way possible.

### **Christmas Lunch**

The inaugural PH Christmas lunch was held on 7<sup>th</sup> December in the Crowne Plaza Hotel. It was a most enjoyable afternoon and great to see everybody in good form and getting ready for the forthcoming festivities.

### **Annual meeting for patients & families – Date for your diary**

The 2016 meeting was a great success with 131 people in attendance. Our annual patient & family meeting will be held in the Crowne Plaza Hotel Blanchardstown on Saturday, 22<sup>nd</sup> April 2017. Please mark the following dates in your diary:  
Dublin Support Group meeting: **23rd Jan 2017** at 11 am in Crowne Plaza. Cork Support Meeting **20<sup>th</sup> Feb 2017** at 12.30pm in the Clarion Hotel, Cork & **22 April 2017** Annual Patient & Family meeting will be held in the Crowne Plaza Blanchardstown at 10.30 am (programme will follow in the New Year).

**Best Wishes:** Finally I would like to wish all our members a very happy and peaceful Christmas. I hope 2017 will bring much happiness and great health to you and your families. I look forward to seeing you all soon.

***Regina***

# Pulmonary Hypertension: A changing landscape

Dr Brian McCullagh, Consultant Respiratory Physician, National Pulmonary Hypertension Unit

The landscape of pulmonary hypertension (PH) medicine has changed greatly over the past quarter of a century. Twenty five years ago our understanding of this disease was poor and we lacked therapies that could make an impact on outcomes. Nevertheless there has been a lot of progress in the intervening period. During the 1990s the first major advance was the approval of prostacyclin therapy in the form of epoprostanol which was given through a continuous intravenous (IV) drip. While this drug was no doubt very effective it was also cumbersome to administer requiring a permanent indwelling venous catheter to be placed. Since then advances have included the discovery of many genetic pathways responsible for causing PH and a number of new drugs have become available that directly improve quality and duration of life.

In Ireland we now have 9 therapies that we frequently prescribe for the treatment of PH. These therapies all target one of three distinct molecular pathways. Broadly speaking people with PH have too little 'nitric oxide', too little 'prostacyclin' and too much 'endothelin' in their circulation and so we target each of these with the therapies at our disposal. Many of these treatments are in the form of oral medication with the obvious advantage of ease of administration. Sildenafil (Revatio), Tadalafil (Adcirca) and Riociguat (Adempas) are three such tablets that target the nitric oxide pathway. We have a further three oral therapies, Bosentan (Tracleer), Ambrisentan (Volibris) and Macitentan (Opsumit) that block the deleterious effects of endothelin.

Remarkably the latest and most exciting advances have occurred with the 'prostacyclin' pathway, the first pathway to have been targeted back in the 1990s. Traditionally we in the Mater PH Unit have utilized prostacyclin therapies in the form of inhaled Iloprost (Ventavis) and subcutaneous (under the skin) Treprostinil (Remodulin). These therapies require a significant amount of education and training and have a predictable side effect profile that include headache, light-headedness, flushing, and jaw pain. Inhaled iloprost can be quite cumbersome with its frequency of administration (6 times daily) and subcutaneous treprostinil has the added burden of being painful at the site of administration. Therefore the addition of a new synthetic epoprostanol (Veletri) given as a continuous infusion through a permanent catheter is an eagerly anticipated addition to the prostacyclin therapies currently available. We expect to have access to Veletri in 2017 and for the select few we expect it will make a difference to both quality and quantity of life.

Finally there is a new oral prostacyclin treatment called Selexipag (Uptravi) that has been approved in the United States in 2016 and currently going through the approval process here in Ireland. Some of you have had access to this drug through a clinical trial that took place in the Mater over the past number of years. The results of that trial were published in an international journal last year and demonstrated overall efficacy of this drug. The availability of an approved effective oral prostacyclin therapy is a new departure in the world of PH and one that we are very excited about in the Mater PH unit. The ease of administration would be the principle advantage and the potential to treat advanced PH with three different oral therapies in select patients is appealing.

In conclusion, while there remains much to be discovered and progress to be made we are nevertheless fortunate to have witnessed some great advances in the management of PH over the past quarter of a century and with continued scientific endeavour, on-going clinical research trials and support from the pharmaceutical industry the future looks bright.

# BIO BANKING

**Clare Hughes M.Sc. B.A**

Hi, my name is Clare Hughes. I am a PhD student working with Dr. Christine Costello in the Conway Institute, UCD. Over the past few months I have been visiting the Pulmonary Hypertension Clinic in the Mater Hospital and requesting that patients donate an extra blood sample at their appointment.

Our research group focuses on investigating and understanding the mechanisms that lead to the development of pulmonary hypertension. We have identified a number of proteins that we believe may be involved in the structural changes that occur in the blood vessels of the lung in this disease. Currently we are most interested in assessing levels of a protein, known as CXCL11, in patients' blood with the hope that this may be used by doctors as a marker of disease, so that patients may be diagnosed earlier, or it may represent a new target for the next generation of therapeutics that may help combat this condition.

Blood samples, kindly donated by patients visiting the pulmonary hypertension clinic, are stored as part of a 'biobank'. This biobank will allow researchers, like me, to investigate this debilitating disease and ultimately develop new therapeutics. All donated samples are coded in order to protect patients' privacy and confidentiality.

We would like to extend our sincere gratitude to all patients who have donated blood for our study so far. We firmly believe that your participation will assist in improving outcome for patients in the future and contribute to the worldwide fight against pulmonary hypertension.

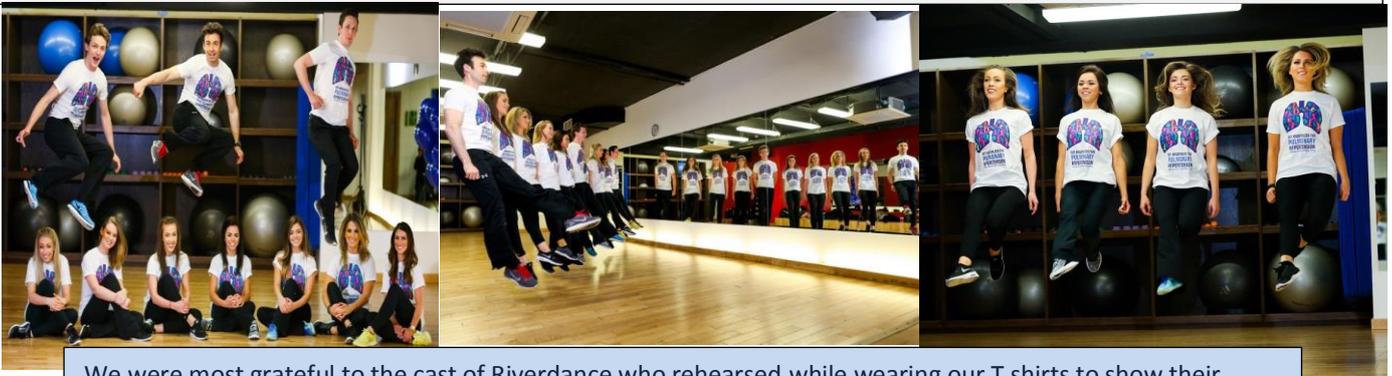
## **Some relevant literature from our group:**

1. Costello CM, Howell K, Cahill E, McBryan J, Konigshoff M, Eickelberg O, et al. Lung-selective gene responses to alveolar hypoxia: potential role for the bone morphogenetic antagonist gremlin in pulmonary hypertension. *American journal of physiology Lung cellular and molecular physiology*. 2008;295(2):L272-84.
2. McCullagh BN, Costello CM, Li L, O'Connell C, Codd M, Lawrie A, et al. Elevated plasma CXCL12a is associated with a poorer prognosis in pulmonary arterial hypertension. *PloS one*. 2015;10(4):e0123709.
3. Rowan SC, Keane MP, Gaine S, McLoughlin P. Hypoxic pulmonary hypertension in chronic lung diseases: novel vasoconstrictor pathways. *The Lancet Respiratory Medicine*. 2016;4(3):225-36.

## World PH Day

Pulmonary Hypertension Awareness day is held throughout the world on 5<sup>th</sup> May. It's not always possible to organise the event on the actual day but we try to organise it somewhere around that date. The theme of the awareness campaign is 'Get Breathless for PH'. Sports personnel, dancers etc are encouraged to wear our T shirts while training / performing to raise awareness of the fact that people with PH suffer breathlessness on a daily basis and not only when exerting themselves. Other countries across Europe have been very successful with their annual campaigns – receiving attention from National TV & Radio stations as well as print media publications. Let's make it our 2017 New Year Resolution that we will strive to raise awareness through the organisation of enjoyable events. We have an exciting idea involving Line Dancing for our 2017 World PH Awareness Day. This is still at the planning stage and if it works – it will be wonderful!! The plan is that we have number of heats throughout the country with the grand finale taking place around World PH Day. If we get media attention everywhere a heat is held – PH will be a household name by the time the Grand Finale is held in late April / early May (date to be confirmed)

**Awareness Month:** For some years the USA have nominated the month of November as awareness month for PH. I note the UK marked it this year. We will work together towards organising some interesting events in November 2017.



We were most grateful to the cast of Riverdance who rehearsed while wearing our T shirts to show their support for World PH Day 2015. Thanks to members of the Dublin Senior Football Team for their support of World PH Day 2016. A fun day was had by all on the Viking Splash Bus.



## FUNDRAISING EVENTS HELD DURING 2016



The Pulmonary Hypertension Association is affiliated to the Mater Foundation (CHY 9768)

### Colin Darker's Dread Chop for Pulmonary Hypertension

By: Avril Burbridge – organiser of event

Kathleen Darker was diagnosed with Pulmonary Hypertension about four years ago. PH is not her primary illness; it developed as a result of having an auto-immune disease. It was a relief in a way to finally have a prognosis to explain her extreme breathlessness, but it also came with the reality that she was living with a life-long illness with no cure. Unfortunately for Kathleen her diagnosis was not detected straight away and for many her PH remained untreated. However when a diagnosis was confirmed the treatment received was a life changing moment for Kathleen.

Kathleen's son Colin Darker had a very distinguished hair style - dread locks. After more than a decade long affair with the dreadlocks, Colin had decided he had enough and was looking for the opportune moment to shave his head. It had been on his mind that such an iconic moment should be recognised by an event such as a charity event. He felt that it would be a shame to shave his head in quiet and not try raise some awareness and money for a charity while doing so.



Suzie from  
Abner Browns  
Barber Shop  
prepares for  
the chop !!

Last year his Mother, Kathleen approached him and asked if he would shave his head for Pulmonary Hypertension. At this stage Kathleen had become a regular attender at the monthly PH Association meetings - Colin didn't think twice and was very happy to do this for Kathleen. So we started organising Colly's dread chop for charity which was held on February the 6th 2016. He had the support of his family and friends, not only through donating money but with providing help in organising the event and providing music. Everyone who knew Colly associated him with his beloved dreadlocks. People were certainly intrigued at the thought of Colly shaving his hair. What would he look like without his mane? Countless friends and family offered support and were willing to donate. Many knew of Kathleen's illness and many more were interested to know more about PH.

We set up an online charity page as well as promoting the event on Facebook - both of which we would recommend to anyone hoping to run a similar event. Through the whole process the Pulmonary Hypertension association were extremely helpful in providing support. They provided charity sponsor cards, t-shirts, pencils, teddy-bears and more. The event itself took place in Slattery's bar in Rathmines, who were very kind to let us run the event for free in their upstairs bar. The night was extremely well attended by family and friends who were all there to lend support, donate and have a good night. Kathleen was also there to witness the chop along with all her family. We were also especially delighted to see so many friends from the Pulmonary Hypertension group there to support us.

On the night we had a raffle organised in the hope of raising some extra donations. We were completely overwhelmed by the generosity of friends and local businesses who donated raffle prizes. In the end we were able to make up eight quality prizes and hampers including perfumes, bottles of wine and whiskey, vouchers, chocolates, a round of golf and even extremely sought after concert tickets donated by Aiken promotions. We were so very grateful to everybody. Such excellent prizes led to furious donations from attendees on the night. Indeed there was a great buzz about Slattery's throughout the evening. Everyone wanted the winning ticket and to guess the weight of the chopped dreadlocks which turned out to be over 13 ounces.



**Colly with his Mum  
Kathleen Darker having  
sheld 13oz of his  
beloved Dread Locks**

Music was provided on the night by Cold Blooded Moon, (of which Colly is a member) along with internationally acclaimed Dirt Blue Jean and the wonderful Tony Collins' band. The M.C. for the entire night was Gavin Collins who did a fantastic job, especially when it came to announcing the raffle prizes and commentating the live hair cut. All in all the performers were superb and we thank them hugely for their time and effort given to the event. It really lent to the atmosphere of the night and it would not have been a success without them.

At 11 O'clock the music was briefly interrupted for the main event, the chopping of the dreadlocks. A local hairdresser Suzie from Abner Browns barber shop, Rathmines, was kind enough to donate her time and services to the big task at hand, getting rid of the dreads. She didn't waste any time getting her scissors out, a hairdresser's dream or maybe nightmare. Nonetheless within a matter of minutes the lion's mane was shed and Colly rose from the ashes of his dreads. A little light headed at first, he slowly got used to his new do.

In all the night was a huge success. So many people came out to support us and donated money, Slattery's was jam packed. Between the night, online donations and sponsor cards we managed to raise, the by no means modest sum of €4,800. We couldn't have been more thrilled with the result. We are so thankful to everyone that donated and turned out that evening to support the Pulmonary Hypertension event. We were also grateful to our local newspaper *The Echo, Tallaght* who printed an article about Colly's dread chop and about Pulmonary Hypertension in general.

As it is a charity very close to our hearts we were extremely thankful to all the help we received. We hope that the Pulmonary Hypertension foundation continues to garner support and donations, so they can carry on providing such a great service to people living with PH and also research into new possible medications. We firmly believe that the whole event not only raised money but went some way to educating people about an illness they may not have known much about or even heard of before. We understand that not only money can help but rather it is awareness that is vital. It is so important in providing early diagnosis so that patients will benefit from treatment before the illness strengthens. As it stands there is no cure for PH but early diagnosis can provide longer life expectancy and better quality of life for an individual.



Macy Darker Batten has the first chop of Colly's mane!

Colin Darker with his Mum Kathleen with Euan Darker Atkinson & Macy

Kathleen Darker is an extremely special woman, a friend to all and an inspiration to everyone that knows her. Kathleen's positive attitude is a tonic to everyone and definitely goes a long way to bettering her health. However it can be said that she depends to some extent on the support given to her by the Pulmonary Hypertension Association Ireland of which she attends their meetings throughout the year. These meetings provide a great support network and also share important current information regarding the illness. They also provide much needed services for those living with PH, which is why it is such a worthy charity.

Needless to say it was our absolute pleasure to run this event for Pulmonary Hypertension in honour of Kathleen. Colly had no regrets chopping his dreads off for his Mother, and I'm sure he would do more if he could to help Kathleen and others find a cure for PH. One thing he might miss is being mistaken for Bob Marley. We never imagined that we could raise so much money for a long overdue haircut. We are over the moon with the outcome, not only by the money raised but also, we hope, the awareness it raised too. It would not have been possible without the extremely generous support from friends and family and the Pulmonary Hypertension Association.

Many thanks from the Darker family. *Avril Burbridge*

**Note: Monies raised on behalf of the Pulmonary Hypertension Association are used for the following:**

- Continuous awareness events of pulmonary hypertension
- Monthly support group meetings giving patients the opportunity to meet and learn
- Provision of a counselling service for patients & carers
- Guest speakers at support group meetings
- Annual conference for patients and their families
- Newsletter
- Website, containing information, news etc
- Merchandise

# Race night in Templeogue

By: Sally-Anne Breen

Thursday Night the 10<sup>th</sup> of March 2016 was a cold wet night and we were seriously worried – the tables were set out, we had all our helpers lined up, the balloons inflated, the banners were flying and then we thought ..... What if people don't turn up? We had organised a race night to both fund raise and also to raise awareness of Pulmonary Hypertension. The venue was St Mary's College RFC in Templeogue.

We needn't have worried..... Our friends, neighbours, colleagues and supporters started to arrive and by 8 pm, the Club was packed to capacity and the atmosphere in the room was electric. The races started and while some were luckier than others – we had a great night.

Then we held the raffle – initially we thought we would have a couple of bottles of wine, chocolates etc. The generosity of people – (some who didn't even know us) was overwhelming. We even got a voucher from Trump International Golf Course.

Our profits for the night were €10,000. For a single event, this was a tremendous success and we were humbled by the support we received. I was asked to say a few words about Pulmonary Hypertension. Initially I felt apprehensive but then I thought – we're all in this together and we must support each other by raising awareness. Before the night, many of our friends didn't really understand what PH was, but thankfully I think my presentation explained in lay man's terms what we go through every day. I was delighted with the attention given to the presentation and many people asked me questions about PH afterwards.

We were very touched by the generosity of not only those who were there but also those who couldn't come to support us.

It was great to see support from our PH community with financial support and expressions of good wishes from many while on the night Karen Doherty, Maria Flynn & Regina Prenderville flew the flag for PH.

It took a lot of hard work and we could not have delivered so much without the support of family and friends.



**Sal giving a presentation of how PH has affected her life and how she copes with it**

**Hardworking tellers who diligently collected bets and calculated winnings**



Marie-Therese Walnutt, Aoibhinn & Clare Garrihy



Sarah-Kate Breen & Eugene Garrihy

Thank you everyone.

**Sal**

# OUR GROWING ASSOCIATION

By: Karen Doherty

Today I feel very proud to be a member of our developing PH Association. Thankfully our members are increasing. In the past year, we also set up support group meetings in Mullingar & Cork. In the meantime, it's business as usual in Dublin.

From a personal note, I feel very lucky to be surrounded by lovely people in our support group. It's a very positive experience, listening to other patients sharing their stories. We can give each other words of advice, for making life and living with PH, a little more bearable.

We all discuss how we've been feeling since our previous meeting. It feels like having therapy among people who understand what you're going through. We have a good few laughs together, as we have got to know each other more. As a patient, who can feel isolated at times, I really look forward to our meetings.

On 17<sup>th</sup> October, Sally Ann Breen and I attended the first support group meeting in Cork. It was very successful. The patients living locally were very happy, as travelling to our Dublin meetings can be inconvenient. We met some very interesting, vociferous and entertaining people and we look forward to our next meeting in Cork on 20<sup>th</sup> February 2017.

Every day, it takes courage and grit to live with the challenges we face by having PH, so having support group meetings is extremely beneficial. In 2017, we will continue expanding and getting stronger. Helping each other, one step at a time.

Seven months ago I was placed on the lung transplant list. Initially I was overwhelmed, as it's a massive mind-set adjustment - and with the biggest transition ahead, I'm very lucky to have great love and support from family and friends. In 2017, I hope to raise more awareness for organ donation as well as Pulmonary Hypertension.

I would like to wish you all a wonderful Christmas and here's to 2017.



**New additions to  
our growing  
Association:**

*Merchandise  
available from the PH  
Association*

**Karen**

# PHA International Conference: 17-19 June 2016

## 25 years of progress: changing the history of PH

By: Regina Prenderville

*The Pulmonary Hypertension Association, USA (PHA) held its 12<sup>th</sup> bi annual conference in the Omni Hotel, Dallas, Texas in June 2016. Over 1,500 patients, families and healthcare professionals attended for the largest gathering of the PH community in the world. I was very privileged to be invited by PHA USA to attend this amazing meeting.*

*I met two wonderful people—Susie & Perry, who spoke about their experiences with PH. Their presentations were so stimulating that I asked them for a copy for publication in our Newsletter. They very kindly agreed and I have copied their presentations below. I think you will like their stories.*

## PHA STORY

By: Susie Alvarez, Los Angeles, California

Hello PHamily!!

Let me begin by giving you a positive Message of Hope. We are all ONE PHamily looking towards the day a cure for Pulmonary Hypertension is found. The global PH community is counting on each one of you to join the global cause to help find a cure for PH by getting involved and motivated to raise awareness of this chronic, life-changing, often misdiagnosed, complex, and rare disease. Invite your family, friends, and community members to join you in educating the community by sharing your PH story. Each story is unique and only you as a patient, caregiver, family member or friend who's life has been touched by PH can tell it!!

The story of MY journey begins in 1983. At the young age of 21, I moved to Los Angeles to accept a full time job. My life was full with family events, work projects, vacations, and much fun and promise. While attending evening college classes for working adults, 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 exercise classes as I could not keep up with the physical activity the courses required.

However, on June 30, 1992, my life was forever changed. Without warning, I had a crushing pain in my chest as though someone was sitting on it. The paramedics arrived and took me to Cedars Sinai Hospital in Los Angeles. During the week I was there, several tests were performed 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 entered my room and gave me news that would forever change my life. He says "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 an oral medication (Vasotec) to delay the progression 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 miles away and 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, asking —**WHAT IS PULMONARY HYPERTENSION AND HOW DID I GET IT?**

Over the next four years, my symptoms progressively worsened. I was short of breath, fatigued, and dizzy most of the day. In 1996, 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, California. 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. I was also referred to UCLA Medical Center, evaluated and placed on the transplant list for a double lung transplant. I did not know at that time, but the therapy I was placed on, an IV medication (epoprostenol-Flolan) delivered by a medical pump, was the ONLY FDA approved medication available at that time to treat PH. I remained on the transplant list for 13 months. The IV Flolan was slowly improving my quality of life and I was removed from the transplant list.

In 1996 I began attending local support groups and PH-related events nationwide. In 2011, I volunteered to become co-leader of the Greater L.A. PH support group at UCLA Medical Center/VA Hospital, Los Angeles. I reflected on my journey at this stage of my diagnosis and believed that helping others' in their time of need is a confident and selfless act that all of us can achieve every day. I had personally benefited by the generous donations of time, research, and funds gifted by many generous donors to PHA, people like yourself here today who have saved my life in so many ways, and felt the call to "give back" by volunteering my time to support other patients who were diagnosed with PAH.

I attended the 2012 PHA International Conference in Orlando FL. While volunteering at the registration desk, I met Perry Mamigonian, a support group leader and PH patient who had emailed me the year before seeking support of other California support group leaders to secure a proclamation for November's PH Awareness month. Diagnosed only 3 years, Perry was inspired to meet a long term survivor of 20 years. I clearly remember Perry reaching out and giving me a hug, expressing his message of inspiration to me that he had never met a long term PH survivor!! We chatted and exchanged contact information and continued to communicate after Conference.

In 2012, I attended PHA's Congressional Luncheon/Lobby Day at the Capitol in Washington, D.C., to seek co-sponsorship of the Pulmonary Hypertension Research and Diagnosis Act (H.R.2073) and the importance of early diagnosis through PHA's Early Diagnosis Campaign. Perry was also invited to join the group for the luncheon. The night before the luncheon, Perry and I enjoyed dinner and took a walk around downtown Silver Spring, MD sharing our journey stories together. We talked about my experience living alone with a rare disease and the amazing amount of support the PHA Conference provides to patients and families and the opportunity Lobby Day provided to meet with members of Congress and advocate for PH causes that would help many patients.

After Conference, we continued to support and attend PHA events in California. We traveled 210 miles to visit one another in our home towns of Los Angeles and Fresno, attended CA PH support group meetings, special events, and PHA California Chapter events. We discovered we shared a love to volunteer as support group leaders and were inspired to fundraise and accept speaking invitations asking us to share our stories of hope. The Fresno CA and Greater L.A. PH support groups of Southern California each founded and host an annual fundraising Walk. We are so inspired to see the PH community gather and watch the attendance grow larger each year.

I felt in my heart I had found a special person who loved me for who I was, who accepted the PH journey I had experienced, and looked forward with excitement for what we planned to accomplish together, (pause) as co-patients and caregivers in love. Our PH doctors and nurses expressed AMAZEMENT at our union and gave their support to get our story out to other patients who need to hear of patients, like us, who are living life and surviving their 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 24 years ago. My journey was not easy by any means. As a long-term survivor, every day is a blessing for me and keeps me motivated to work towards awareness, support, and hope for a cure.

Thanks so much for allowing us share our journey stories with you today. Please continue to support the efforts of the Pulmonary Hypertension Association and the PHA Chapters. Your efforts WILL help so many lives in unbelievable ways.



Suzie & Perry have been together 3 years and became engaged last year

# PHA STORY

By: Perry Mamigonian, Fresno, California

Good morning everyone! I don't know about you but I've really been enjoying this Texas hospitality! We've experienced some of the best PH education here at conference, and one thing I've learned is that all of our journeys are unique but our paths have brought us all together by PHA. I'm humbled by amazing devotion and work the PHA staff does for all of us.

When I was diagnosed with pulmonary hypertension at age 48, I thought my life was over. The truth is, it was just beginning, and I credit the power of this community for teaching me that I was wrong.

Like many of us, for several years prior to my diagnosis I experienced shortness of breath during normal activity. My primary care doctor thought it was due to age, lack of exercise and excess weight, and I had no reason to doubt him. But in early 2009, I began to feel flu-like symptoms which would not improve. Then one morning while driving to work, I began to cough uncontrollably – another pre-diagnosis symptom I developed. I had just exited the freeway and was headed toward a major intersection as my head started to spin and slip into unconsciousness. My last thought before blacking out was, "I'm going to die." But when I woke up, I was covered in white light and for a brief moment I thought, "It's true. When you die you really see a white light". But when I lifted my hand, it was just the airbag that was covering my head. Somehow, my car had drifted to the side of the street and struck a parked car. No one was hurt but me. But my injuries were severe and a passerby called an ambulance.

Once I was in the hospital and explained that I had passed out, the doctors began running tests to figure out why. One test after another showed nothing, until an ultrasound of my heart revealed an enlarged right-ventricle. The cardiologist recognized it as a sign of pulmonary hypertension and called in a PH specialist, Dr. Vijay Balasubramanian, Community Regional Medical Center, Fresno, CA. and he confirmed my diagnosis.

At first, all I understood was "hypertension", and I expected them to give me pills and send me home. But Dr. Bala (as we call him) calmly explained that it was very serious and that unless I began treatment immediately I would get much worse. I appreciated his concern and honesty, and after 16 days in the hospital I was released on infused prostacyclin therapy.

But I was just focused on the physical, not the mental effects of this disease. My life had been turned upside-down. I was unable to return to work, and because I blacked out in the car accident my driver's license was suspended. I was single, alone and completely dependent on my family and friends for help. It all made me very angry, and I'm sure many of the people in this room know the exact kind of anger I'm talking about. We didn't have a support group in Central California at that time, and although I was aware of other resources for help, I was just too stubborn to use them.

Several months into my diagnosis, I received a phone call that would ultimately change my life. My specialty pharmacy had a patient advocate who was part of a team hosting a PH conference in San Francisco, and she encouraged me to go. She felt that the education and opportunity to meet other patients would benefit me. But I was reluctant at first. I didn't even have a driver's license. But she was persistent, and pushed me to go. So I earned back my suspended license and attended the conference.

It was my first experience with the PH Community – which I would later come to see as a PHamily. I learned so much about living with this disease, but more importantly, the patients and caregivers I met were so friendly and inspiring that I learned from them that there can be life after diagnosis. For the first time in months, my outlook was positive.

Shortly after I returned home, two nurses in Fresno started a support group and we began to form our own PH family. Over the past seven years, I've learned that we are more than just patients coming together to share similar problems – we are all unique people: husbands and wives; mothers and fathers; brothers and sisters; sons and daughters – all

coming together to help to us cope and support each other, and sometimes even laugh and cry together. In short, we are a PHamily.

I'm grateful for all this community has given me, so I feel a desire to give back. In 2010, I was asked to be a co-leader of our support group, and I soon became active in advocacy. With the help of our members, we've convinced several local congressmen to co-sponsor each of the last three PH bills in Congress. In 2011, I had an idea to ask our state capitol to issue a proclamation for Awareness Month. But I wanted our representatives to be aware of the power and scope of this PH community, so I contacted all the California leaders asking permission to include their names in the request. I began friendships with many, but one became special – Susie Alvarez from Los Angeles.

Susie and I, both from California, finally met in person in Florida (of all places) at the 2012 PHA Conference. We began a friendship and over the next 18 months would see each other more frequently at various PH-related events. Soon I began to develop feelings for her beyond friendship, but I was reluctant to admit it. Being middle-aged with PH gave me confidence issues, but Susie sensed my feelings and coaxed me to admit them. When I explained my reluctance to her, she told me that I was wrong – that having PH is no reason to deny ourselves the things that make life worth living. If anything, it's a reason to embrace life even more. We've been together for almost three years now, and became engaged last summer.

Looking back, I remember how angry I was after my diagnosis. Today, all I feel is gratitude for all I've received since then. I'm grateful for my friends and relatives, my healthcare team, the devoted staff at PHA, and to all of you – this amazing PHAMILY. But most of all, I'm grateful for Susie, who showed me that hope and love is still possible with pulmonary hypertension.

## Christmas Lunch 2016

Date: Wednesday 7<sup>th</sup> December 2016    Venue: Crowne Plaza Hotel, Blanchardstown, Dublin 15



Karen Doherty & Addis Doyle



Kathleen Darker & SallyAnne Breen



Kathryn Rooney & Maria Flynn



Ann Duignan, SallyAnn Breen & Rose O'Neill



John Doyle & Susan Sheehan



Joan Doyle, Kate Keating & John Bolger

# **pulmonary Hypertension Association - Ireland**

**Wishing you and your families a very happy & peaceful  
Christmas. Look forward to working together in 2017!!**

**Happy New Year to one and all!!!**



Pulmonary Hypertension Association - Ireland

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