

WINTER 2015

# emphasis

The magazine for supporters of PHA UK [www.phassociation.uk.com](http://www.phassociation.uk.com)

## Brothers and sisters

Vicky Kelly on supporting the siblings of young people with PH

## National Audit of PH

Simon Gibb talks facts and figures

## My heart and lungs

Stacie Priddon shares her PH transplant story

## 15 years of treatment for pulmonary hypertension

A look back at key developments in PH drugs

## The Interview

Professor Paul Corris talks about his career

## On the up

Lois Parker tackles a snowy mountain

## A patient's guide to surgery

Launching a new publication about PEA surgery

## A PHantastic time

Photos from the PHA UK 15th anniversary conference





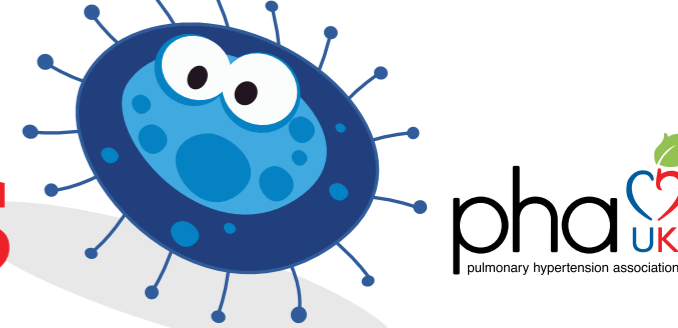
# Free support & advice...



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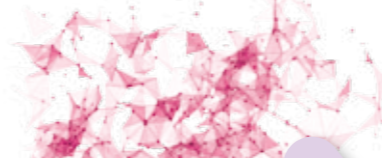


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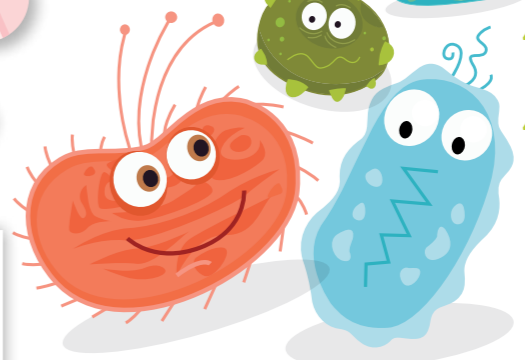
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# MAKE YOUR VOICE HEARD

Hundreds of patients will soon be invited to take part in a **national survey** about living with pulmonary hypertension.

The PHA UK survey, which will be launched early in 2016, aims to gather evidence about what it means to have PH in the UK today. It will ask people about their experiences of diagnosis, treatment and care and also the wider impact of the condition on things like their family life, finances and mental health.

Around 500 people participated in a similar survey carried out in 2010 and their responses provided vital information about matters such as the effectiveness of drugs; experiences of NHS services and other important aspects of living with PH.

Chair of PHA UK Iain Armstrong said: "As an association we are committed to representing the interests of people with PH, their families and carers and, therefore, it is essential that we gain a good understanding of people's real life

experiences of living with the condition. "What we learn through our survey will help us to make informed decisions about the charity's future spending priorities; what issues we need to campaign on; what research we need to invest in and what support services we need to provide to best meet members' needs.

.....  
**“As an association we are committed to representing the interests of people with PH, their families and carers.”**  
.....

"The survey's findings also greatly strengthen our voice with NHS clinical commissioners and so on as it provides frontline qualitative evidence direct from

patients about, say, the impact of certain drugs and the value of certain services.

"We would love to hear from as many people as possible in our 2016 survey – the larger the statistical group, the louder the voice."

Details will be announced via the PHA UK website, Facebook and Twitter, as well as via the Homecare PH delivery service early next year.

People interested in taking part will be asked to get in touch and then emailed a login password to allow them to complete the online survey. Patients who would prefer to complete the survey on paper can also phone 01709 761450 and a questionnaire will be posted to them.

The survey will be quick, multiple choice and with a maximum of 30 questions. It is open to everyone with PH, not just PHA UK members. ●



# Welcome

.....

Welcome to the winter issue of **Emphasis** magazine. We've certainly got a packed issue for you, with lots of news, views, information and advice from right across the PH community.

As our 15th anniversary year comes to a close, we celebrate our phenomenally 'phantastic' conference with photos and feedback on **page 28**. And you can also turn to the centre-spread for a look back at the many drug developments we've seen in PH treatment over the decade and half the charity has been in existence. Plus, there is a look forward to the possible advances we may hope to see through stem cell research in future, on **page 16**.

The life-changing significance of heart and lungs transplantation as an option for many with pulmonary hypertension is also highlighted. Patient Stacie Priddon shares her personal experience of undergoing a double transplant earlier this year on **page 18** and the UK's world-renowned transplant specialist Professor Paul Corris, based at the Freeman Hospital in Newcastle, is interviewed on **page 42**.

We've news of a new patients guide to PEA surgery at the Papworth Hospital; a poem from PH blogger Tess Dunn; and a focus on the incredibly important National Audit of PH, to which staff at our PH centres constantly contribute information which supports the ongoing development of PH services.

Finally, clinical psychologist Vicky Kelly of Great Ormond Street

Hospital gives us an expert view on an important subject we haven't covered before in these pages; how best to support the brothers and sisters of children with PH. As we all know the impact of a chronic health condition doesn't just affect the one person and PHA UK is committed to supporting whole families affected by the disease.

There are lots of opportunities to get involved in future magazines, so please keep in touch. And in the meantime, everyone in the Emphasis team here at PHA UK would like to share the compliments of the season with all our readers and send all good wishes for the new year.



Iain Armstrong  
Chair of PHA UK  
editor@phassociation.uk.com

.....  
**“PHA UK would like to share the compliments of the season with all our readers and send all good wishes for the new year.”**  
.....





# Emphasis *exchange*

If you have any news and views to share via the Emphasis exchange please e-mail [editor@phassociation.uk.com](mailto:editor@phassociation.uk.com) and don't forget to keep in touch via Facebook and Twitter too.

## Happy Couple

Guests at a Brighton wedding were all given PHA UK badges in lieu of favours to raise awareness of pulmonary hypertension on the happy couple's special day.

Bride and groom Sereena and Richard Adams were keen to spread awareness of PH as the condition plays such a part in their lives together.

Sereena, who was diagnosed with PH in 2011 at the age of 25, said: "We knew there would be a lot of friends and family at the wedding who wouldn't really have heard of the condition and were not aware of what it means for me - and what Richard is getting in to! PH did come up in the speeches and it was really nice to have people understanding so much more about it, asking questions and all wearing the little badges by the end of the night. It was a happy day and we were very pleased to share our story with our friends and family."

Richard and Sereena, who met in the same year she was diagnosed, both work as teachers. The newlyweds made a donation to PHA UK for the pin badge table decorations.



## Great heights

A Newcastle nurse has gone above and beyond the call of duty to raise awareness of pulmonary arterial hypertension.

Rachel Crackett, a PH nurse specialist at the northern PH unit at Freeman Hospital, jumped out of an aeroplane at 10,000 feet to draw attention to the disease and raise funds.

Her sponsored skydiving feat raised more than £2,000 for PHA UK and also raised the profile of the condition.

Rachel, who has worked with PH patients for 14 years, said: "As a team it frustrates us all that PH does not get the attention that other serious and potentially fatal conditions do, and so we wanted to do something that would help make more people aware of it."

"I'd like to thank all colleagues, patients and the PHA UK for their support and donations. And give special thanks to the patients who were able to come and support on the day. This was a once in a life time experience for me - in other words never again! But it seemed to work."

Rachel's jump featured in the Newcastle Chronicle, attracting thousands of views on its website and driving more visitors to PHA UK's website too.



## TALKING TO TEENAGERS

Teenagers and young adults with PH are invited to get involved in a new review of how services are delivered to meet their needs.

The transition between children's and adult services has long been acknowledged as a sensitive period which needs careful planning and support.

Children up to the age of 16 are treated by the dedicated PH paediatric team at Great Ormond Street Hospital London; then their care is transferred to one of the other PH centres in the UK, most often the one closest to their home.

This new PHA UK-funded research project aims to take a closer look at this transition period and gather frontline views to inform future service developments. It will be led by former lead clinical nurse specialist at GOSH, Yvette Flynn.

Yvette said: "We would like to talk to teenagers about how the transition period currently affects them; what is positive and what is difficult during this time and what they may like to see happening differently."

"In parallel with juggling long term health issues, teenagers with PH face all the usual demands of growing up, thinking about their futures, taking on more responsibility for their own decisions and so on. We'd like to talk to them about living with PH in relation to all the usual concerns for young people such as education, careers, relationships and developing their own independence and self-confidence.

"Our aim is to make the transition from childhood to adulthood with the PH services a more comprehensive and easier process for all. The involvement of young people at this stage will ensure their needs and ideas will be integral to current and future development of the process."

Yvette wants to hear from young people aged 12 to 24, interested in getting involved. She would particularly like to hear from those who have already experienced moving from the paediatric to adult PH services.

To find out more, please contact Yvette via the PHA UK office on 01709 761450 or [office@phassociation.uk.com](mailto:office@phassociation.uk.com)



### Giving in memory

Thank you so much to all our supporters who have collected donations in lieu of flowers at a loved one's funeral.

Retired librarian Glennis Walmsley of Chorley in Lancashire died a year ago on November 30th at the Royal Hallamshire Hospital in Sheffield. Her husband John would like to thank everyone involved in her care - and thank friends and family who raised £900 through donations at her funeral. Glennis, aged 65, also leaves a daughter Jill and son Neil, who lives in Dubai and previously took part in a 10k run there to raise money for PHA UK. Donations in honour of someone's memory are very much appreciated by everyone at PHA UK. These personal collections help us to continue campaigning for better diagnosis and treatment for people with PH and raise awareness of the condition.

If you would like to support the charity's work by giving in memory of a loved one, please post your donation to Helen Crabtree, PHA UK, Unit 2, Concept Court, Manvers, Rotherham, South Yorkshire, S63 5BD. You will receive a personal letter of thanks to confirm receipt of your gift.



### In case of emergency

Did you know there's a new feature on iPhones that can store vital health information, in case of an emergency?

News of this handy little feature is being spread widely via social media and more and more emergency personnel are reaching for people's smartphones in case they hold information that could save a life.

The 'Medical ID' function allows you to enter data such as who to contact in an emergency, blood type, any medical conditions, allergies and medications you are taking.

The feature is built in to the 'Health' application on any iPhone running iOS8 and it is easy to set up. When launching the app, click on Medical ID

in the bottom right-hand corner and set up a new profile. Finally, to make sure emergency staff do not have to know the passcode to unlock your phone - enable the function 'Show When Locked'.

Once set up, you just have to tap the word Emergency in the bottom left-hand corner of your screen, and the Medical ID information tag will appear.



## Meet the Doctors

PHA UK has raised awareness of pulmonary hypertension at two high-profile medical exhibitions this autumn.

Representatives of the charity attended the Acute and General Medicine exhibition in London and also took a stand at the European Respiratory Society (ERS) exhibition in Amsterdam.

Sheffield PH centre ward manager Paul Sephton, who attended representing PHA UK, said: "These events are a great opportunity to meet a range of healthcare professionals, who may not already be familiar with PH. "We aim to raise awareness across the board so that many more clinicians, not directly working with the condition, may recognise the symptoms earlier when they see it in their patients."

"The events are a great way to interact with professionals we wouldn't otherwise meet such as GPs, specialist registrars who are training to becoming tomorrow's consultants, nurses working outside our PH specialist centres and so on.

"Ultimately we want to talk to them all about PH and pass on our materials so that if they have a patient who is breathless, for example, and the more common medicines are not working, they think about the possibility of PH. As we all know, an earlier referral can lead to an earlier diagnosis which can often lead to a better prognosis."

Both exhibitions were also a chance for PHA UK to network with pharmaceutical companies and, at ERS, to catch up with other PH associations in Europe and promote awareness of PH globally - visitors to the PHA UK stand included doctors from as far afield as the United Arab Emirates, Uzbekistan and India.



## Social Media Round-up

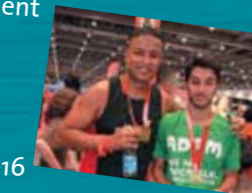
Here are some top tweets and popular posts from PHA UK Facebook and Twitter



@wginsing Great presentation from Dr Simon Gibbs on the PH National Audit @PHA\_UK



@Arafmo Great moment @TheLondonTri being able to complete the #LondonTri with my Son #ProudDad in aid of @PHA\_UK roll on 2016



@DJChurch94 Delighted to be in this summer's edition of the @PHA\_UK's emphasis magazine - a feature on a remarkable couple. #PHA



Katie Somerfield Saw this and thought of everyone



Mum-of-two saved by double lung and heart transplant says: My life rested on a cliff edge



@PHA\_UK A positive outlook can make a huge difference! "I'm a very determined lady and I won't go easy"



Catherine Makin Thank you PHA, was a fab weekend. My first conference and I really enjoyed it. See you all next year xx



Pulmonary Hypertension Association UK When times get hard, please don't forget, there is always someone to talk to.... We have a free dedicated helpline set up with Anxiety UK (official) just waiting for you call. Support is only a phone call away - 0844 332 9010



@bfc\_official Barnsley FC captains past and present join forces to support local charity event!



Follow us on Twitter @PHA\_UK and like our page on Facebook at www.facebook.com/PULHAUK



## Fresh look for fundraising

A brand new range of posters, stickers, collection boxes and balloons has been launched to help people raise funds for pulmonary hypertension.

PHA UK has revamped its free fundraising pack to highlight that this is the only charity dedicated to supporting people with PH in the UK and that it supports a 3,000-strong network of people affected by the condition.

Anyone interested in receiving a copy of the bright new pack to use at their next fundraising event, please contact the PHA UK office via office@phassociation.uk.com or call 01709 761450.



## SHARE WHAT YOU THINK OF YOUR CARE

Would you like to share your experience of the care you've received at the doctor's, dentist's, in a hospital, care home or any other setting?

The Care Quality Commission (CQC) is giving patients and families the opportunity to give feedback on any poor – and good – experiences they have had, through their website. CQC is the body that makes sure health and care providers in England provide safe, effective and high-quality services.

To give people more of a voice, they have now introduced an online feedback form called 'Share Your Experience.' The form is NOT a substitute for complaints which must be made through the official channels for each provider, but is a chance to share a little positive or negative feedback with the CQC. You can opt to share your experience anonymously and it is also possible to complete the form on someone else's behalf. The CQC may then use this information to help continue their work in assessing the quality and safety of services. For more details go to [www.cqc.org.uk/share-your-experience](http://www.cqc.org.uk/share-your-experience)

# Latest research is subject of discussion

Health professionals from all PH specialist centres in the UK gathered to discuss latest research and developments last month.

The annual Pulmonary Hypertension Physicians Research Forum, which is supported by PHA UK, was held at the historic Chelsea Physic Garden in London.

This informal two-day meeting has become a key date on the calendar for doctors, nurses and other professionals involved in developing ever better treatment and care for people with PH.

Forum co-ordinator Dr John Wort, who is clinical lead for PH at the Royal Brompton Hospital, said: "The forum is a chance for us to get together and share news of on-going research and studies into many different aspects of PH.

"Every PH centre is invited to make up to three presentations about projects of their choice and that may be an in-depth piece of basic research or a simple patient audit.

"It has become a fantastic way to gain feedback and comment, facilitate the cross-fertilisation of ideas and promote collaboration."

The PH Physicians Forum is unique in the way that it brings together all the designated PH centres and therefore specialists from the fields of cardiology,

respiratory medicine, rheumatology and paediatrics. It involves scientists, doctors, nurses, pharmacists, technicians and physiotherapists.

This year's forum focused amongst other things, on matters within the fields of genetics, physical exercise, narrative writing as an insight into disease and haematology.

PHA UK has supported this forum with generous financial grants to cover some of its costs over the past few years.

Dr Wort said: "We really do appreciate the support of the PHA UK to help make the Physicians Forum happen. The annual event provides a relaxed yet stimulating environment where everyone can speak freely

*"It has become a fantastic way to gain feedback and comment, facilitate the cross-fertilisation of ideas and promote collaboration."*

The setting for the forum is also an inspiring one as the Chelsea Physic Garden was established by the Society of Apothecaries in 1673 and is home to one of the most important collections of herbs and medicinal plants in the world.

and raise matters of interest.

"Our shared aim is to improve our understanding of PH and the forum really does help to build relationships and advance the research and development work of professionals across the national PH community."



# PH POET TESS DUNN SHARES HER STORY



Maritess Dunn, known to her family and friends as Tess, was diagnosed with PH in June 2008. She was born with a hole in the heart, and an operation at the age of six months left her with just one lung.

**T**ess, now 25, has always taken an interest in poetry and music and, despite only having three fingers on her left hand, she has played the piano from a very young age. She took up the trumpet but this had a negative impact on her heart so she had to give it up. Oh and she also plays cello!

"Music is my life. I live and breathe music, which is where the name of my blog, 'Music helps me to Breathe' comes from," said Tess.

"When I got diagnosed with PH, I felt my world had fallen apart, but my music was there for me. My piano playing allows me to express my emotions without having to explain to people how I feel about having PH."

"I wrote 'From the heart' last year because I think people tend to judge

me because I don't look ill. I've always written poems for family occasions. It's something I find easy to do and being a musician, I tend to think of them as lyrics."

Her love of music inspired Tess to take a music degree at Southampton University and then start her own music business, Treble Tess Music. She then decided to go to Qatar working as a music teacher, but returned home in July 2014 and reopened her music business.

"I really enjoy teaching people of all ages and abilities, and sharing my love of music," said Tess.

She now plays with the London Shostakovich Orchestra and aims to complete a Masters degree in music therapy so she can use her experience

to help others.

"Music has got me to where I am today, along with support from my family and my boyfriend Terry who has been a rock since my dad passed away last Christmas."

"Music has helped me through difficult times, so I'm hoping I can use this to give hope to other people with PH and similar conditions," she added.

In 2015 Tess became one of the faces of the British Heart Foundation's 'Wear It. Beat It' campaign and her picture was used in a poster campaign - helping to raise £1.5 million for life-saving research.

*Tess writes a blog about living with PH - why not check it out at [www.musichelpsmebreathe.blogspot.co.uk](http://www.musichelpsmebreathe.blogspot.co.uk)*



## FROM THE HEART BY TESS DUNN

So, 2008 HERE'S THE DEAL

I GOT DIAGNOSED WITH A HEART CONDITION,  
HOW DO I FEEL?

TO TELL MY STORY WHERE DO I BEGIN,  
EXCEPT TO SAY I'M FEELING RATHER GRIM.

I'M LYING IN A HOSPITAL BED,  
I'VE HAD LOADS OF SCANS AND TESTS, ALL THE  
THINGS WE DREAD.

I THEN GOT TOLD I MIGHT NOT LIVE 'TIL I'M 18,  
I'M HOPING ALL THIS IS JUST A DREAM!

ALL THIS IS DUE TO A HOLE IN THE HEART,  
THIS MAKES ME TERRIFIED FOR A START.

THIS HAD LED TO AN ILLNESS CALLED PH,  
NOT KNOWING WHAT THIS IS, I WAS IN A STATE.

I THEN GET TOLD THAT THERE IS NO CURE,

SO I'M ALWAYS HOPING FOR ONE, THAT'S FOR SURE.

PH IS A LUNG AND HEART CONDITION UNKNOWN TO ALL,  
I TRY TO KEEP STRONG, BUT SOMETIMES I FALL.

THE QUESTIONS I ASK MYSELF, LIKE, AM I DYING?

THEN PULL MYSELF TOGETHER, LOOK AT THE  
OBSTACLES I'M SO FAR DEFYING!

IT'S HARD TO LIVE WHEN IT TRIES TO HOLD ME BACK,  
BUT I'M DETERMINED TO KEEP GOING ON THE  
RIGHT TRACK.

CHEST PAIN AND BREATHLESSNESS IS WHAT I DEAL WITH,  
BUT I'M THANKFUL FOR EACH DAY THAT I GET TO LIVE.

I HAVE TO TAKE LOTS OF TABLETS EACH DAY,  
AND KEEP INFECTIONS, COUGHS AND GERMS AT BAY.

THE PAINS IN MY BODY ARE SOMETIMES SO BAD,  
THAT YES I DO CRY AND GET SO SAD.

YES LIFE CAN BE TOUGH, BUT I GET BY,  
A DOUBLE TRANSPLANT ONE DAY I'LL HAVE TO TRY.

BUT 'TIL THAT DAY WHEN MY HEALTH GETS WORSE,  
I GET TO LEAD THIS WONDERFUL LIFE FIRST.

TRADE KNOWS ME AS A MUSICIAN,

PLAYING PIANO, CELLO, TRUMPET, DESPITE MY CONDITION.  
MY MUSIC HAS GOT ME THROUGH SUCH TOUGH TIMES,  
JUST BEING ABLE TO PLAY, READING LINE AFTER LINE.

WITH HOPE AND DETERMINATION, I WENT TO  
SOUTHAMPTON UNI,

AND AFTER GETTING MY DEGREE I KNEW NOTHING  
COULD STOP ME!

I CAME HOME AND OPENED MY OWN MUSIC BUSINESS,  
IT WORKED WELL WITH ALL MY HOSPITAL VISITS.

I THEN GOT A JOB OFFER IN THE MIDDLE EAST,  
THE DOCS ALLOWED ME TO TRAVEL, WHICH WAS  
A FEAT!

I'M NOW OFF TO CAMBRIDGE TO STUDY A  
MASTERS DEGREE,

TO GET A QUALIFICATION IN MUSIC THERAPY.

THIS IS WHERE I USE MUSIC TO HELP THOSE  
WHO ARE ILL,

IT'S WORKED FOR ME, SO I'M HOPING FOR THE IT WILL.

AS FOR FITNESS, I CAN'T GO FOR A JOG OR A RUN,  
SO I POLE DANCE INSTEAD, WHICH IS EVER SUCH FUN!

MY PARENTS AND CLOSE FRIENDS WILL ALWAYS  
BE THERE,

WITH LOVE AND SUPPORT BECAUSE I KNOW THEY CARE.

STRONG AND POSITIVE I WILL ALWAYS WILL BE,  
FIGHTING A BATTLE YOU WILL NEVER SEE.

FINALLY I WANT TO THANK THOSE AT ROYAL  
BROMPTON HOSPITAL,

AS WITHOUT THEIR TEAM I WOULDN'T HAVE GOT  
THROUGH IT ALL!

SO DON'T JUDGE ME, TRY AND WALK IN MY SHOES,  
THERE REALLY IS NOTHING TO LOSE.

JUST SEE HOW FAR YOU CAN REALLY GET,

'CAUSE YOU'LL REALISE HOW STRONG THIS  
GIRL IS THAT YOU'VE JUST MET!





# CHRISTMAS GIFT LIST

HAVE YOU DONE YOUR CHRISTMAS SHOPPING YET? HERE ARE A FEW FESTIVE IDEAS FOR YOUR LIST FROM PHA UK MEMBERS.

## 1. COLOUR BEAUTIFUL

A colouring book for grown-ups with 50 beautiful garden designs to lose yourself in. "What an excellent idea", says Kay Yeowart of Sussex 'Garden: 50 designs to help you de-stress (Colouring for mindfulness)' - **Amazon, price £4.67.**

## 2. VRRROOOOM

For little ones who are going places... **K's Kids Jumbo Go Go Go, for six months plus, available from Smyths Toys for £39.**

## 3. FITNESS TRACKER

Wearing a fitness tracker, such as the Jawbone UP3, the Nike+ FuelBand SE, and the Fitbit Charge, is the smart way to keep track of your exercise routine, whatever goals you set yourself. These interactive wrist bands monitor your movements and can connect and compare you to friends' data too! "The best present I've ever had," says Toby Pettifor who likes to take exercise to help manage his PH. **The Fitbit Charge, in different sizes, is available from £79.99 from Currys, Argos and John Lewis.**



## 4. SOUNDS OF MUSIC

Intrepid Toby also tells us what he'd like for Christmas this year... a set of good quality headphones to wear on his world travels! Well, he did spend last Christmas in Canada! **The DN-12 Trident noise-isolating earphones are available from Amazon, priced £20.**

## 5. COMFORT AND JOY

Every festive gift from Lush is a joy to look at – and then you get to open it up and enjoy the luxurious bathtime products. "What's not to like," says Helen Crabtree of the PHA UK HQ **Comfort and Joy luxurious shower gift pack from Lush, £20.**



# Ain't no mountain high enough



Travelling with oxygen can be a concern for people with PH, and needs careful planning. But it doesn't have to stop you. Here, seventy-year-old *Lois Parker* of Leicestershire, tells us about her determination to go up a mountain.

**"Back in 2008 my husband Alan and I climbed a mountain! Well, we didn't exactly climb it with ropes and crampons, we sat in a train. We were on a touring holiday in our campervan in Switzerland and, when we were in Interlaken, we decided to take an excursion up to the Jungfrauoch, a 11333 feet high tourist attraction and research station.**

We spent a day up there, but unfortunately it was cloudy so we determined to come again, in better weather.

Then in 2012 at the age of 67 I was diagnosed with pulmonary hypertension. This was obviously quite a shock, even more so when I was told that an endarterectomy was out of the question. Luckily for me the medication (Sildenafil, Warfarin, and oxygen overnight) helped to overcome the breathlessness and within a few months I was starting to feel a lot better. I was looked after very well by the PH Unit at Sheffield and it seemed that my problem had been caught early.

So when we planned our holiday for this year we wondered if it would be

possible to go up the mountain again. I asked my consultant what he thought, and was told "Go for it, but take your Oxygen with you".

So on a bright sunny day in July this year we set off from our campsite in Interlaken to take the Bernese Oberland train to Kleine Scheidegg, where we boarded the train to take us up the mountain.

The railway was built over 100 years ago and travels through tunnels in the Eiger. It takes about an hour, stopping twice to allow people to view the snowy slopes, but more importantly to allow everyone to acclimatise gradually to the increased pressure and lessening of oxygen.

We alighted from the train at the summit with my Inogen One portable oxygen concentrator on its small trolley. It was the first time I had used the trolley as normally I only use the Inogen overnight. I managed without

the oxygen for about ten minutes, but then started to feel peculiar and Alan commented that I was a bit blue! So on with the oxygen, and we started to re-explore. We stayed up there for about four hours, going out on the slopes to look at the glacier in bright sunshine, watching the snow-boarding, sliding through the ice palace and enjoying the exhibitions, shopping and lunch!

At first I felt a little self-conscious about having my oxygen on but I very soon realised that it was helping me to enjoy my day up there. In fact I believe that quite a few people would have liked to share it with me! Altitude sickness is not very pleasant.

*So I decided that I would like to share my experience with PH friends and show that it is possible to do most things with a bit of planning. I would recommend this adventure of ours to everyone whether you need oxygen to keep you going or not."* ●

Have you got a story to tell about travelling with PH? Let us know via [editor@phassociation.uk.com](mailto:editor@phassociation.uk.com)





# CELLSEARCHING

*Dr Dan Reed, a research associate at the National Heart and Lung Institute, Imperial College London, was recently commissioned by PHA UK to publish a guide to stem cell research, aimed at patients. Sally Brown spoke to Dan about his interest in stem cell research and how to communicate the implications to patients.*



**A**s an Imperial College scientist and an Honorary Research Fellow at the Royal Brompton Hospital, Dr Dan Reed currently works on a range of projects that study stem cells and their possible benefits in the study and treatment of pulmonary hypertension (PH).

His latest book for PHA, called 'Stem cells and scientific research in pulmonary hypertension', attempts to explain stem cell therapy in layman's terms and looks at how ongoing research into the use of adult stem cells could help conditions like PH by stimulating repair to blood vessels in the lungs.

The book also highlights how stem cells can be used, not just as therapies, but as tools to help scientists learn more about the disease in the laboratory and develop new treatments this way.

These days it is possible to take blood samples from anyone, and grow stem cells from their blood which can then be used to produce new blood vessels or a piece of muscle. This enables scientists and doctors to learn about diseases in the lab without invasive procedures.

Dan is particularly interested in 'personalised medicine' where medical decisions, practices, and products are tailored to the needs of the individual patient. His guide explains the biology

of stem cells, stem cell therapy and how stem cells are used in laboratory research into PH.

Scientists and doctors fully understand that having a life-threatening disease like PH makes people feel desperate, and sufferers often look abroad for cures and treatments. However leading doctors, and charities such as the PHA, recommend that patients don't go down the stem therapy route at this time.

"Stem cell therapy is an exciting treatment because of the potential to repair damaged tissue, but our knowledge of how stem cells work on the lung or the heart isn't advanced

enough for this treatment to be ready," said Dan.

"It is a very different stem cell system to the one we use in bone marrow transplants in cancer treatment for example."

**"Because of the extensive media attention around stem cell research, several institutions have started offering stem cell therapy for a range of diseases, including PH.**

"But there are still a lot of questions to be answered and many scientists are rightly wary of making false claims."

Some companies charge thousands of pounds for so-called stem cell therapies which don't necessarily work. There are many different types of stem cells which all do different jobs and until scientists and doctors know what they all do, there are a lot of unanswered questions.

Dan is currently working with Professor Jane Mitchell using stem cells from patients' blood to make discoveries about PH. The main challenge is the funding to support the research, as well as the scientific expertise. Funding is very competitive and scientists often don't know in advance whether their work will make a small difference or whether it will be a quantum leap forward.

Keen to demystify the subject, Dan attends numerous discussion forums to help patients understand the current

research. One of his presentations was at the PHA annual conference in April 2014, and again in October 2015, where he presented the findings of the field's latest research and took questions from the audience.

"This was my first real opportunity to speak with patients about research in PH, and this isn't something scientists normally have the chance to do" said Dan.

"In meeting patients and their families the importance of good communication between scientists and the patient community became clear to me, and this was something I wanted to get involved in"

Dan's work with the PHA and their patients has really changed his perspective on the importance of the research that he's involved in and how that research is communicated to patients.

**"It can really impact on patients' quality of life and their healthcare. We need to guide them through stem cell and other PH research and be there to give the right support and advice.**

"It's a minefield for people to navigate and it can add to their anxiety or confusion. Scientists need to be at the heart of this to explain and reassure."

Dan firmly believes that people's quality of life can be influenced not just by what happens in the lab but

by what is said in the media, and by scientists, about their condition.

"Having worked with the PHA for the past few years I really hope that this will be the start of more opportunities for scientists to interact with patients. This is, of course, good for patients, but also encourages us, as scientists, to really think about our research from the patient perspective, and how we communicate what we find."

*Free copies of Dan's book, and accompanying DVD, are available from the PHA, telephone: 01709 761450 or email: [office@phassociation.uk.com](mailto:office@phassociation.uk.com)*

## SOME FACTS ABOUT STEM CELLS

- Stem cells are a special type of cell – they can change themselves into other types of cells in the body, such as heart cells or lung cells.
- There are broadly two types of stem cell – embryonic stem cells and adult stem cells (sometimes called progenitor cells).
- Some types of adult stem cells, taken from bone marrow, are currently being used to treat cancer and some types of immune disease.
- In the future it might be possible to replace damaged lung blood vessels with cells made from stem cells – however, there is still a lot of research to be carried out.
- Scientists are, however, already able to use blood vessels made from adult stem cells in the laboratory, and use them to make new discoveries about PH, develop new drugs, or determine the best drugs for individual patients based on their cells.



Photo courtesy of National Heart and Lung Institute, Imperial College London



THREE YEAR WAIT FOR TRANSPLANT  
**DEALING WITH IT TOGETHER**

*Pulmonary Hypertension had been part of Stacie Priddon's life for 13 years before she had her heart and double lung transplant in May this year. Here Stacie, aged 24, who lives in Swindon, shares her family's story...*

**“I** was diagnosed with PH at 11 years old after living with congenital heart disease my entire life, so I've never known what normal is. Normal to me and my family has always been 'Stacie the ill child with regular hospital appointments, blood tests, surgeries, medications, oxygen...' That is our normal when it comes to me..

The diagnosis of Pulmonary Hypertension was a shock, but it was just another hurdle that me and my family would overcome really, because that's who we are. When presented with a challenge you don't simply give in you attack it head on and that's what we did.

Growing up I had an older sister, Candice, and a twin Megan. I suppose it's easy for parents and siblings to become over cautious when one of them is diagnosed with a terminal illness. At the time the prognosis was still only a few years so presumably you would think my family would want to wrap me in cotton wool and never let me leave the house. I got lucky! I got a normal teenage experience with the odd hospital appointment thrown in. I was treated like the normal teenager I wanted to be. My sisters never let my PH be an excuse. I was just Stacie to them. I'm sure my parents worried, especially as I deteriorated over time and oxygen got added to the mix, and more medication with the worsening of my PH, but they let me get on with my life the way I wanted. The only difference I would say I really had was that I couldn't do P.E. and quite simply who would be bothered by that? Who wants to do hockey, outside, in the middle of winter, in the rain? I certainly didn't.

I was in my second or third year at college when things started to get worse so this was around 2009/2010. As anyone with PH knows, stairs are really not our friends even when we are

at our best, but with me I could kind of do them so when a few steps started to become hard for me, that's when we knew I was getting worse. A few feet seemed like 10 miles. I was missing lessons because I was just too tired to get out of bed. I was reducing my hours at work because it was just too much. My family stepped up the support though and drove me around as much as they could. I couldn't go to university like I wanted, but it was a small price to pay and I figured out a way to do my degree from home. By 2011, I was much, much worse. I had exhausted all possible medication options available to me at the time and the only way to get any more was to be on the transplant list.

Transplant had always been the very last option for me. My doctors knew this and so I knew they would only bring it up if they believed it was the only way forward. Transplant isn't simply 'be put on the list, wait a little bit, get your transplant, go have a normal life'. We all knew it would take a very long time to get me organs. The process involved 11 months of going to different transplant centres and being assessed and the addition of a nebuliser and then a IV line with a 24/7 infusion of Flolan. Firstly, I went to Harefield Hospital where they frankly told me no. At the time Harefield were only able to perform heart transplants or double lung transplants but not both together. They wouldn't perform a double lung transplant on me as they believed my heart was too damaged and would be unable to cope with a new set of lungs. This therefore led me to another assessment at Papworth Hospital, where I was eventually told they would put me on the list for a heart and double lung transplant, on the condition that I kept a fit as I could and lost 10 kilos to be at the optimum weight for transplant. It took 11 months to get on the list but I eventually



got there on April 20th 2012. What we didn't know was I was in for a very long wait. On average the wait is about two years and they only put you on the list if they think you have less than three years to live. I had the best support system behind me my family could not have been any better! I'm a positive person generally, for those of you who have met me you will know this, but when you're on the waiting list little things tend to get to you. Stupid things really; a comment meant in

a joking manner, someone being late for something, not being able to go somewhere, just things that happen every day that normally would just go over your head... Everything becomes very intense and affects you in the stupidest of ways. My family would let me be down because you do need it sometimes, but then they would be there to lift me back up again. I'm not sure I would have made it through my wait if it hadn't have been for them. **Continued overleaf** ▶



Stacie's appearance on ITV's Good Morning Britain.



## Family Matters

..... Stacie Priddon .....



*I did eventually get my transplant after an excruciating wait of three years, one month and 10 days, with a few near death experiences added into the mix. In those three years I had three false alarms, which were devastating at the time, but they prepared me for what I needed to do the fourth time I was called. I was the most prepared I could possibly be for my transplant. You'll never know exactly the kind of strength you need to get through that experience because if you did people would probably say no, but all I can say is my family and friends all provided me with an ample supply of it when I was lacking. Them and the life I had been waiting for is what pushed me through. After my transplant I was told that my lungs were in a horrific state. They couldn't even remove them in a block, they had to take them out in bits they were that bad. My surgeon even wondered how I was even alive. I'm a strong person, I defy the odds when presented with a challenge and I don't ever give in and, even with my doubts towards the end*

*of my wait, transplant was not going to be any different. I can't say that having PH is fun. It's really not and I have to watch friends suffer with it like I did, but the thing is PH was a part of me. It made me who I was, it brought me to this point in time. Without it, yes maybe I would be living the life I dreamed of as a ten-year-old, yes maybe I would be off having a family of my own and a dream job somewhere. But, the thing is I wouldn't have some of the friends I have, I wouldn't have an amazing community of people that I've been a part of for 14 years, I simply would not be me. So yes I now have a new heart and new lungs and I'm on a new journey but PH will always be a part of me and my family. ••*

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 If you would like to share your family's PH story, please email [editor@phassociation.uk.com](mailto:editor@phassociation.uk.com)



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 Read Stacie's blog at [www.stacie-lifeisworththefight.blogspot.co.uk](http://www.stacie-lifeisworththefight.blogspot.co.uk)  
 .....

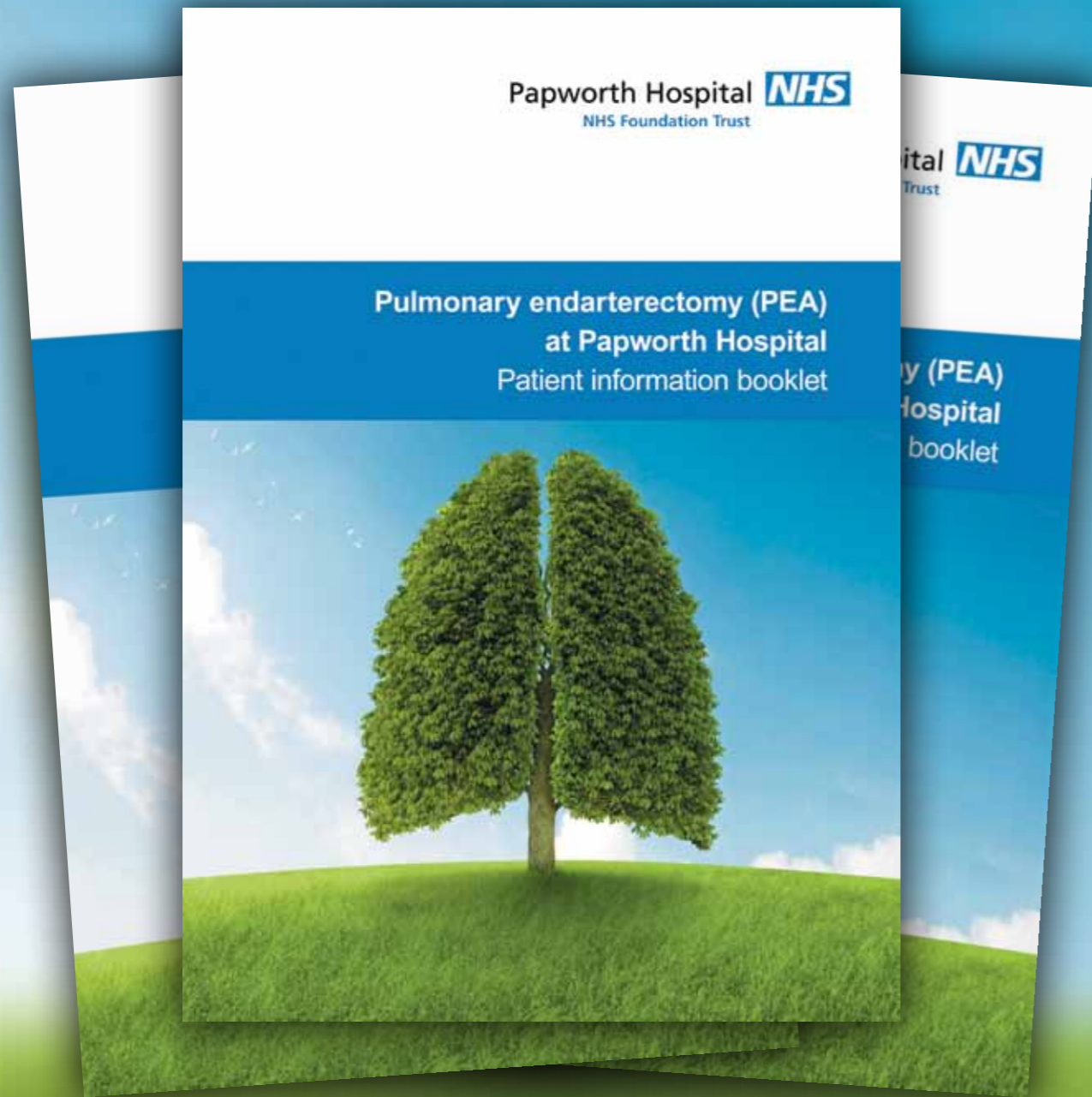
# Are you struggling with stress or anxiety?

.....  
 Help is available to members of PHA UK if you or your family have concerns about your emotional wellbeing

- Dedicated helpline service 0844 332 9010
- Assessment and therapy provision for those who need emotional support
- Training and resources for PHA UK professionals and members

To find out more about how Anxiety UK can support the emotional aspect of having a diagnosis of PH contact us today on **0844 332 9010** or visit **[www.anxietyuk.org.uk](http://www.anxietyuk.org.uk)**





# NEW PATIENT GUIDE TO PEA SURGERY PUBLISHED

*Surgery is an option for a growing number of patients with a certain type of PH called Chronic Thromboembolic Pulmonary Hypertension (CTEPH). Staff at Papworth Hospital, where this operation called a Pulmonary Endarterectomy (PEA), is performed are helping patients to keep informed each step of the way through a new booklet.*

**The booklet has been carefully written to include all the information and guidance needed by patients and their families. A patient group reading panel from PHA UK and from Papworth has received the new material and given it a very favourable response.**

Anie Ponnaberanam, lead nurse for PEA at Papworth, led the project. Anie says: "The aim was to update and expand upon a similar booklet published in 2000 using what had been learnt from the expectations and experiences of patients over the years. Input was gained from members of the team, including the retired PEA specialist nurse Maureen Rootes, consultants, surgeons and feedback from patient groups.

"The booklet explains to patients what CTEPH is and what PEA surgery involves. It covers what is likely to happen from the point of the first clinic appointment at Papworth right through to the procedure itself, recovery, and going home. Alongside detailed medical information is plenty of practical advice such as what to pack for the stay in hospital. There is also guidance for families and visitors."

Explaining the process, Anie says: "Patients with a diagnosis of CTEPH are referred to Papworth from designated PH centres. All data and images are assessed at weekly multi-disciplinary team meeting for consideration of surgery. Patients who may potentially benefit are invited to attend an

out-patient clinic at Papworth for a face-to-face information session with a specialist PEA nurse and surgeon.

"The information booklet is handed to patients at their first meeting with a PEA specialist nurse at the group information session involving patients and relatives. Following this, patients will go on to have a one-to-one meeting with the nurse and then have an individual consultation with a surgeon, who will make the final decision on the operation and explain the risks and benefits of the surgery. When possible, patients may also get the opportunity to hear the first-hand experiences from an ex-patient who underwent surgery a few years ago.

*"The booklet compliments the face-to-face education with patients. It helps them to understand the complexity of their disease and management with surgery and guides them to give informed consent. It is a very good source of information for patients and their families to refer to at any time as they go through their journey with us. The feedback received from patients and their families so far is very positive. Patients have praised the fact it's so detailed."* ●



## **What is CTEPH?**

Chronic Thromboembolic Pulmonary Hypertension is a form of PH that is caused by blockages in the blood vessels to the lungs due to scar tissue. The scars are the result of blood clots that the body has not properly cleared. These scars cause a complete or partial obstruction leading to difficulties in blood flow through the lungs. This causes high blood pressure in the lung arteries, which in turn affects the right side of the heart, which pumps blood through the obstructed lung blood vessels. The heart is forced to work harder than normal and gets bigger and weaker as it tries to cope. If left untreated, the heart will eventually start to fail. Initially people get short of breath on exertion and, with time, this could progress to shortness of breath even at rest. Patients may struggle to do any type of physical activity. Swelling of the legs and abdomen may be noticed by some patients. Treatment with blood thinning agents such as Warfarin stops more clots from forming, but has no effect on the

scarring left by previous clots and has a poor prognosis without treatment. Management for CTEPH used to be a heart and lung or lung transplant, now it can be treated and potentially cured with pulmonary endarterectomy.

## **What are the benefits of PEA surgery?**

Pulmonary Endarterectomy (PEA) is potentially curative surgery to remove the scar tissue which is causing the obstruction in the lung blood vessels and can result in immediate reduction in pressure in the pulmonary artery. Most people who have operable obstructions in the big blood vessels are benefited by this surgery with improved symptoms, less breathlessness, improved quality of life and live longer after their operation. However, some people have obstructions not only in the big blood vessels that are removed by surgery, but also in smaller blood vessels that are more difficult to reach. Following surgery for these people with 'mixed disease' the pressure in the

pulmonary artery will not completely return to normal, but their quality of life should still get better. Unfortunately, a very small number of people may not notice any benefit after surgery. If this happens, they will be closely monitored by their PH centre.



## **PEA at Papworth Hospital**

The world-famous Papworth Hospital, in Cambridgeshire, is the only hospital in the UK that carries out this specialist procedure. The first PEA surgery at Papworth Hospital was carried out by Mr John Dunning in 1997. The service was then commissioned in 2000 as Papworth Hospital NHS Foundation Trust to be the national PEA centre. Since then the service has expanded considerably in response to increasing referrals over the last 10 years. Now, there are four surgeons performing the operation. Mr David Jenkins is the lead PEA surgeon with Mr John Dunning, Mr Steven Tsui, and Mr Choo Ng. The number of patients being considered for possible surgery increases every year.



# Fundraising **roundup**

Catching up on some of your fantastic fundraising antics from all corners of the UK.

To see more of the fundraising that's taking place for PHA UK, or to publicise your event, join us on Facebook & Twitter  

## Run-away success

Congratulations to Jennifer McConnell, Gillian Mulraney, Donna Hunter and Jill Gallacher who completed the Great Women's 10k Glasgow Run for PHA UK. "Not only did we achieve the unimaginable, by completing the 10k" said Jen, whose mum has PH and scleroderma, "But we set ourselves a target we never thought we would reach, and beat it." Jen was also very pleased to wear PHA UK running vests. "A lot of people had never heard of PH so it was good to be able to spread the word."



£2,132  
RAISED

## Birthday present

When Anne Mountain's mum lost her battle with PH in 2011, her daughter decided to raise awareness of the condition. So when Anne came to celebrate her 60th birthday at Childwall Golf Club in Liverpool she asked friends and relatives to make a donation to the PHA instead of giving her presents. About 40 people turned up and with the help of a raffle, including a PHA goody bag, she raised £525. "It was a really lovely night and people were so generous. I'm just so glad we did it," said Anne.



£525  
RAISED

## Community action in full colour

Regulars at the Stag and Hounds pub in the heart of Farnham Common, Buckinghamshire, held a PHA UK community fundraising day to support their friend Clare Quinton. Clare, who was diagnosed with the condition 15 years ago, said; "Their generosity exceeded my expectations and I would particularly like to thank Jo, Kat and Leah who completed an obstacle course whilst being pelted with coloured dye."



£610  
RAISED

## Taking a dip for PH

Andy Ross's girlfriend Gemma Cousins was diagnosed with PH one year ago, and watching her manage her condition with 20 tablets a day inspired him to tackle a gruelling four hour, 10km swim in the chilly waters of the Manchester ship canal in Salford recently. Andy said: "I know it would have been much worse for Gemma if she had been diagnosed with PH just a few years earlier. Treatments have come a long way. But there is still no cure and much more research to be done."



£1,640  
RAISED

## Helen is a friend indeed

Helen Weller is one determined lady. She was going to run the Berlin marathon for PHA UK last year, in support of her best friend Camilla Keyte who has PH. But then her own diagnosis of breast cancer got in the way. Undeterred, as soon as Helen's treatment finished and she was given the all clear, she signed up for this year's event. Helen, who lives in London, said: "It was my first thought to enter again, I said I'd do it for Camilla and I didn't want to just let it go."



£300  
RAISED

If you have any ideas for raising money for PHA UK visit [www.phassociation.uk.com](http://www.phassociation.uk.com). Fundraising mini-packs are also available. And, don't forget to share news of your achievements via our Facebook page or tweet about them tagging @PHA\_UK



# Medical advances over 15 years...



PHA UK celebrated its 15th anniversary this year. During the decade and half that the charity has existed, we have seen amazing advancements in drugs, treatments and therapies for those affected by pulmonary hypertension. There is plenty more we hope to see improved and achieved through further research and development, but here are some of the key steps taken to a better prognosis for PH patients over the past 15 years.

**2000**

PHA UK formed to support all patients with PH, their families and carers and advance PH treatment and care in the UK.

**2001**

**Epoprostenol** is approved for use in the UK after evidence shows it is effective for those with advanced or rapidly progressing PH symptoms. This is a prostaglandin drug continuously infused intravenously through a Hickman or Groshong line.

**2002**

**Bosentan** (brand-name Tracleer) becomes the first tablet specifically developed for PH to be licensed in the UK. To this day, it remains the most commonly prescribed Endothelin Receptor Antagonist (ERA).

**SIGNIFICANT OTHERS**

Consultant Pharmacist Neil Hamilton says: "As with lots of rare diseases, not all of the treatments we use are licensed like the ones on our timeline, but this doesn't mean they are not helpful or commonly prescribed. Medicines such as treprostinil and iloprost, which for one reason or another have never been licensed in the UK, have been used to treat patients with significant PH since the late 1990s."

"Over the last decade we have witnessed major advances in drug and surgical treatment that have significantly improved the outcome for PH patients. The UK has been at the forefront of research and development in pulmonary hypertension, but perhaps most importantly, recognised over ten years ago that integrated clinical pathways are key in allowing patients to access high quality diagnostics, treatments and support networks. The provision of a network of national specialist centres in 2001 was the first step in this journey... The UK now has a model of care which is highly respected throughout the world."

David Kiely, Chair of UK and Ireland Pulmonary Hypertension Physicians Committee

**2003**

The Department of Health publishes the first standards of patient care, quality and safety requirements for specialist PH centres. Our network of expert centres providing high quality care remains the envy of the world.

**2003**

**Iloprost** (brand-name Ventavis) is another prostaglandin approved for use in the UK. This drug needs to be inhaled using a nebuliser every three hours.

**2007**

NICE's decision to not recommend funding for PH drugs because of cost is overturned following PHA UK campaign.

**2005**

**Sildenafil**, a PDE-5i, is approved for use with PH patients and this oral treatment is now the first choice treatment, according to UK prescribing guidelines. Liquid and injectable forms of sildenafil are also now available.

**2014**

**Riociguat** (brand name Adempus) is the first medical treatment proven to be effective in treating chronic thromboembolic pulmonary hypertension that is either not operable or persisting after surgery.

**2013**

**Macitentan** (brand name Opsumit) is licensed. This is the first PH treatment to be shown to improve 'morbidity' ie it delays worsening of symptoms and/or 'mortality', ie it may improve life expectancy.

**2015**

Whilst **warfarin** has been around for decades, Novel Oral AntiCoagulants (NOACs) have now arrived. **Rivaroxaban**, **apixaban** and **dabigatran** have been shown in other conditions to be just as safe and effective but without the need for regular INR blood tests. In future more patients with PH and CTEPH may be given NOACs.

**KEY DRUG TYPE DEFINITIONS:**

A **Prostaglandin** drug boosts the level of prostaglandin in the blood. Prostaglandin is a substance produced by our bodies which causes the blood vessels in the lungs to dilate, or become wider, allowing blood to flow more easily. When the body does not produce enough prostaglandin, it can narrow the arteries and put pressure on the heart; and cause scarring and cell growth in the vessels and lungs too. These drugs introduce synthetic prostaglandin to help boost natural levels.  
An **Endothelin Receptor Antagonist (ERA)** drug reduces the amount of endothelin in the blood. Endothelin is a substance made in the cells of heart and blood vessels linings. Excessive amounts can cause the blood vessels to thicken, constrict and become narrower increasing pressure in the pulmonary arteries. ERAs work by reducing the amount of endothelin the body produces.  
A **Phosphodiesterase type 5 inhibitor (PDE-5i)** acts by opening up blood vessels, in certain organs around the body. Having originally been developed as a treatment for angina, the 'side-effect' affecting men with erectile dysfunction was discovered. More recently it was proven to be safe and effective for PH.

Please talk to your PH team if you have any queries about your own medication.

**2008**

**Ambrisentan** (brand name Volibris) is licensed in the UK. This ERA tablet's lack of interactions with other medicines is a major step forward as patients are taking more and more different treatments.

**2010**

**Sitaxentan** is withdrawn worldwide after it is linked with causing liver damage. It was available for nearly three years in the UK. Although the other ERAs are safer, regular monitoring of liver function through blood tests remains vital.

**2011**

The first report from the National Audit of Pulmonary Hypertension is published. The audit collects vital information regarding treatment of every patient at PH centres.

**2017...**

The year after next should hopefully see the next tablet to be launched. **Selexipag** works in the body like a prostaglandin. We await the published results of its clinical trial.



# A PHantastic time was had by all!

More than 270 people of all ages attended PHA UK's 15th Anniversary Conference at Heythrop Park in Oxfordshire from Friday 30th October to Sunday 1st November 2015. A packed programme included presentations by professionals about the latest developments in PH treatment and care, as well as insights from people living with pulmonary hypertension. There were also social and creative activities for children and adults and plenty of opportunities to get to know others affected by PH. A fabulous gala dinner and fireworks rounded off the weekend to celebrate our association's anniversary – and look forward to the future.



*"An excellent event and trusted speakers to update us all on exciting times ahead for PH sufferers! It was a privilege to be amongst so many amazing and strong people. PH sufferers are always optimistic and the most valuable gift from the conference still remains the contacts made between people from all over the country."*  
**Julie Royle, Manchester**

*"Thank you PHA UK, it was fantastic and Mark and Lucy said the fireworks were amazing. We look forward to next year's event."*  
**Anna Bowen, Llandysul, Wales**

*"We had a great time including our two kids who got to understand a bit more about PH and enjoyed themselves so much. It was great for them to see PH isn't all about doom and gloom and hospital appointments."*  
**Laura and James Ling, Dover**

*"Congratulations on a wonderful conference. It was inspirational to speak with so many patients and carers and parents during the gala dinner - and I realised why I do what I do. Utterly wonderful experience and very moving."*  
**Professor Paul Corris, Newcastle**

*"The children provided a vibrant atmosphere not experienced at conference before. There was a greater sense of community. And parents had the opportunity to attend knowing they could take part in the sessions with their children safely looked after and enjoying themselves. The fireworks and gala dinner were the perfect finale."*  
**Kay Yeowart, Suffolk**

*"As well as meeting so many friendly people it was very informative and helped us get a better understanding of PH. Well done to all at the PHA who did us all proud."*  
**Mark Kingston, Bognor Regis, West Sussex**

*"The talks were very informative and the seminars were interesting too. I especially enjoyed learning CPR with St John's Ambulance. The promising and positive attitude given by each speaker was such a comfort. There are so many new treatments in the pipeline, that there is no way that any of us cannot have continuous hope!"*  
**Tess Dunn, Essex**

*"I found it extremely informative and so helpful to meet others with PH. The kids are already asking if they can come again next year!"*  
**Jayne Venables, Kent**

*"One of the best weekends ever and our daughter made new PHriends! One of her highlights was having her photo taken with the lovely Stacie Pridden."*  
**Kelly and Kevin Brand, Lancashire**





# The PH National Audit



The National Audit of PH has been running for five years now. But what's it all about? Here, *Simon Gibbs* gives an overview of this important statistical exercise to *Steph Pollard*.

**The NHS' specialised service for PH benefits from one of the most robust, routine monitoring exercises in the UK – if not the world.**

The NHS' specialised service for PH benefits from one of the most robust, routine monitoring exercises in the UK – if not the world.

The National Audit of PH has almost unprecedented levels of engagement, with every PH centre in the country recording key facts and figures about assessments, diagnoses, treatments, therapies and outcomes on a day-to-day basis.

This thorough data collection provides statistical evidence to inform and support our long-term understanding of PH as a disease as well as our knowledge of patients' experiences and how PH services are delivered.

It informs decisions about planning, funding and improving services for the future – and could help protect services against any potential cuts in a cash-strapped NHS too.

The National Audit's founder and lead clinician Simon Gibbs says: "We are very fortunate to have 100 per cent participation in the PH National Audit and we really appreciate everyone's efforts to input this data on a regular basis at all eight PH specialist centres.

"This input makes this one of the most effective audits of a specialised service for a rare condition carried out in the UK – if not the world.

"And it means the data is as robust as possible and provides an excellent rolling review of PH services and patient experiences which can be monitored, measured and reviewed year-on-year."

Simon put his proposal forward for a National Audit of PH more than five years ago and PHA UK played a vital role in setting up the project by funding the very first audit in 2009.

It is now funded via NHS Specialised Commissioning in England, and managed by the Health and Social Care Information Centre (HSCIC) in Leeds.

Simon said: "The charity's funding to get the audit off the ground was crucial. It laid the groundwork for future audits and showed what could be done. It undoubtedly helped to

secure long-term NHS funding for this important work with the HSCIC."

Simon explains here how the National Audit works:

Each specialist PH centre in the UK records data on patients for their own use such as visits, investigations, drugs prescribed and so on. This data is then also fed to the audit database at the HSCIC. All this information gathering is done in accordance with the relevant legal statutes for the country the centre is in (for example the Data Protection Act 1998 and the Health and Social Care act 2001 for England). If any patient wishes to not take part they should inform the hospital treating them; and all patients can be sure that being removed from the audit will not affect the quality of care they receive.

The data is then analysed, verified and published by the Health and Social Care Information Centre (HSCIC) in its annual 'National Audit of Pulmonary Hypertension' report; which is a series of tables of charts which make sure no individual is identified.

Information about this national audit can be found in the leaflet 'What is the National Audit of Pulmonary Hypertension?' which is available from the PHA UK or your hospital's PH service.

The report is aimed at health professionals and policy makers to work with, and is also published online

by HSCIC, available for all to see.

For example, this year's report showed that 8,431 patients were seen at PH centres in the previous year. During those 12 months, 4,126 patients received targeted drug therapy. And of the new PH patients starting therapies, 81 per cent were prescribed sildenafil as the first line drug.

The 2014 report also includes new analysis on diagnosis and therapy, analysis by specialist centre, a comparison of operated and not operated chronic thromboembolic pulmonary hypertension, and an analysis of distance from a specialist centre as an influence on referral patterns.

The audit's purpose is to help further improve the quality and safety of care and its clinical and cost effectiveness. NHS England relies on such statistical evidence to inform its decisions about planning the future of all services.

Simon, consultant cardiologist at Hammersmith Hospital in London, chairs the National Audit of PH project board and works closely with HSCIC on the audit, leading on some analysis of the data.

Simon said: "The analysed data we gather through audit helps us monitor all sorts of things from simply how many people are diagnosed with PH each year to which drugs and treatments are being prescribed for PH and outcomes for patients.

"It maps the delivery of the service across the UK and enables us to enquire into certain aspects of care or identify and monitor any changes. It provides accurate clinical data about the progression of the disease.

"All of this information provides evidence of PH services' activity and impact on patients which all helps to protect, review and develop services.

"As a PH community, we should be very proud that we work together to deliver this audit in support of the provision of high quality care in PH services."

*All five National Pulmonary Hypertension Audits are available to view online at [www.hscic.gov.uk](http://www.hscic.gov.uk)* ●

*"Advances in medical science have introduced important new therapies to the treatment of different forms of pulmonary hypertension... As with other uncommon diseases these developments have brought the need to establish a specialist service, where experience and expertise can grow, audit and research can flourish, and so provide the most up-to-date care of the highest quality. This national audit ... makes a detailed assessment of the clinical epidemiology of the condition; the service demand... the pathways of diagnosis and treatment; and the outcomes over time. It supports studies to better understand the natural history of these conditions, to guide diagnostic and treatment strategies and clinical research."*

**Professor Dame Carol Black,**  
writing in the first National Audit of PH (published 2010)

*"The NHS continues to strive to provide high quality equitable care in a very challenging environment. Nowhere is this challenge greater than in the provision of services for rare diseases. Audit is key in improving outcomes for patients and in helping to plan how we deliver our services. In an environment where resources are finite it is key we ensure that these are used carefully. Over the five years since its inception, the quality of data in the audit has continued to improve and we are now able to not only provide data on outcomes, but also understand more about the forms of pulmonary hypertension that we see in our specialist services. This information will be key in ensuring that the UK continues to provide high quality care for patients and their families over the coming years."*

**Professor David Kiely,**  
**Chair of UK and Ireland Pulmonary Hypertension Physicians Committee,**  
writing in the latest National Audit of PH (published 2015)

The National Audit of Pulmonary Hypertension involves an alliance of NHS professional bodies including PHA-UK; all the clinicians and audit database users at each PH centre; NHS England; the Pulmonary Hypertension Clinical Reference Group of the NHS Commissioning Board; the National Services Division, NHS Scotland; Welsh Health Specialised Services Committee, Northern Ireland Local Commissioning Group, and the Health and Social Care Information Centre.



Simon Gibbs



# At full Stretch



Vicki in yoga pose in Lake District.

Thirty one year old *Vicki MacDonald* was diagnosed with PH at the age of 13 after a year of being misdiagnosed with asthma. With a busy job in London how does she combat the stress?

**“Working as a finance manager in the West End of London can be quite demanding,”** said Vicki.

“And travelling on the underground really turns up my anger dial, so I was keen to do something which would reduce my stress levels.”

Joining a gym in London can be really expensive and, because Vicki has PH, there were lots of classes she couldn't attend because they involved high intensity cardio vascular exercises. So a couple of years ago she decided that joining a yoga class was more suitable to her needs.

“I can't do strenuous cardiovascular exercise but I kept seeing yoga flyers coming through my front door, so decided to head to a class and give it a try.”

Now Vicki attends a weekly class,

held at work, and practices at home at least twice a week.

She finds that yoga really helps with her breathing: “I always used to take short shallow breaths when walking uphill or upstairs and really struggled, but with yoga I've learnt about how to breathe properly and fill my lungs.”

**“The good thing about yoga is that it can be practised for its meditative and relaxing benefits as well as fitness.”**

And, after a busy day at work, yoga helps her to clear her mind of any worries: “The good thing about yoga is that it can be practiced for its meditative and relaxing benefits as well as fitness,” she said.

Her family have been really

supportive too: “When I first told my mum she was apprehensive, because of my PH. She knows that I like to push myself.

“She had concerns that I was overdoing it and not putting my condition first. However over time she has seen the benefits for me, even

though I haven't managed to get her on a yoga mat ... yet!”

Vicki's husband also understands her love of yoga, and helps out by making dinner so that she can practice at home after a busy day at work: “Having family who always puts my health and

Vicki in yoga pose in Koh Lanta in Thailand.



## VICKI'S TIPS...

IF YOU HAVE PH AND ARE THINKING OF TAKING UP YOGA TELL YOUR PH CONSULTANT AND ALSO TELL THE YOGA INSTRUCTOR.

FIND A CLASS THAT SUITS YOUR LEVEL, EG SOME CLASSES ARE SPECIFICALLY AIMED AT OLDER PEOPLE WHICH ARE AT A MUCH GENTLER PACE.

SOME POSES INVOLVE BENDING DOWN FOR PERIODS OF TIME SO BE SURE TO COME UP SLOWLY AND ALWAYS WORK WITHIN YOUR OWN LIMITS.

DON'T PUSH YOURSELF TOO HARD. IF YOU NEED A BREAK, MOVE INTO 'CHILD'S POSE' AND STEADY OUT YOUR BREATH BEFORE RESUMING.

PRACTISING YOGA GIVES YOU A MORE POSITIVE OUTLOOK ON LIFE AND CAN REDUCE ANXIETY. STARTING YOUR DAY WITH A SHORT 10-15 MINUTE GENTLE STRETCHING SESSION OR TAKING 10-15 MINUTES BEFORE BED CAN MAKE A BIG DIFFERENCE TO YOUR DAY AND HELP YOU SLEEP BETTER.

wellbeing first is a blessing,” said Vicki.

Vicki firmly believes that her fitness and flexibility has increased as a result of doing yoga: “When I told my PH consultant that I was starting yoga they were really supportive.

“I have definitely seen an improvement in my cardiovascular levels, and core upper body strength, as a result and I got my best result in my last bike test at Papworth, which is really encouraging.”

And Vicki has some advice for anyone thinking of taking up Yoga: “It's is not a competitive sport, so let your body and breath guide you, and listen to what your body is telling you.”

**“And don't be afraid to try different classes. Find a teacher who inspires and encourages you to achieve what you want in your practice.”** ●



# Supporting brothers & sisters



When a child or young person is diagnosed with PH it has an impact on the whole family. The child's brothers and sisters may be affected in all sorts of ways. Here paediatric clinical psychologist at Great Ormond Street Hospital *Vicky Kelly* talks to *Steph Pollard* about siblings and considers some ways to support them too.

**H**aving a brother or sister with a chronic condition like PH can affect a child in many ways and it is normal for a young person in this situation to experience a wide array of difficult and conflicting emotions.

They may feel worried and sad about their sibling's illness but also jealous of the 'extra attention' they perceive their sibling to be getting and angry about how disruptive the illness is to their family's life.

Research suggests that these children might be, therefore, more vulnerable to developing emotional difficulties such as anxiety or depression and also behavioural difficulties, as they try to elicit more attention from their parents and express their anger through tantrums.

However, it is also important to note that some siblings develop stronger resistance to emotional difficulties later on in life because they have had to develop coping strategies to manage difficulties earlier than they might have done otherwise. Thus they can become more emotionally resilient adults.

Vicky says: "One of the most important things for a parent to do is let their other child know that every

feeling they have is 'normal' and to provide a safe space for these emotions to be talked about in order to help make sense of them.

"Siblings often worry about burdening their parents further or feel ashamed about what they think or feel in relation to their brother or sister. It is perhaps even more important for parents to continue to regularly let them know how unconditionally they are loved no matter what they say or do and that any feeling is okay and can be talked about in an open and supportive way."

Lots of different things can have an impact on brothers and sisters of a child with PH. Regular trips to hospital with their family and witnessing their sibling waiting for and under-going tests and treatments can be just as traumatic for brothers and sisters as they are for the child with PH.

The, often unavoidable, time out of normal life can also disrupt the development of friendship groups for siblings, as well as the child with PH, which can lead to difficulties with their peer group.

On the other hand, if they don't go with their family on hospital visits,

they can feel left out, and might mistakenly think they are missing out on a 'fun' day out in the city. Not knowing what the visit actually involves, can also lead to further anxiety as, depending on their age, they might imagine something far worse happening to their brother and sister at hospital.

One of the most difficult things for parents to manage is dividing their time and attention between all their children when one is ill. Good intentions to spend more time with the other children can end up not happening, through no fault of the parent, as the demands of everyday life and managing PH, dominate the family's life.

Vicky works as part of the multidisciplinary PH team at GOSH and says that in assessing the emotional support needed by any young person with PH, she always tries to involve the whole family in initial meetings.

**"It is very important to assess all the potential needs for support. We work collaboratively with families and local services to try and coordinate support for the whole family where needed."**



## How to support siblings of children with PH

Vicky Kelly shares some strategies and ideas

**> Keep your child in the loop.** Explain what is happening, in a way that is appropriate to their age and level of understanding, and involve them in discussion. It may be appropriate to take siblings along to PH appointments as it can be good for brothers and sisters to see the situation is under control, people are caring and the hospital is a friendly place. But make sure you talk through any concerns they might have about doing this first and don't push them to do anything they do not want to do.

**> Plan for special one-to-one time with all your children as routine.** Even if it is just ten minutes a day after meals. Or it might be an extra bedtime story or another special activity you always do together. These dedicated moments when your focus is just on them can be extremely valuable for their self-esteem and a great opportunity to let them know how much you love them and enjoy spending time with them. They can also be key times to talk through feelings.

**> Encourage friends and family to remember all your children.** If they do bring gifts and treats for your child with PH, encourage them to include all of them. If they talk a lot about your child with PH, gently try and steer the conversation back to include all your children.

**> Use a 'worry box' in to which your child can post little notes about things they are worrying about.** This can then be opened at a set time each week and the 'worry notes' can be talked through. This can be useful to capture and acknowledge anxieties as they happen and then put them to one side to be dealt with at a safe, pre-arranged time.

Vicky would like to acknowledge colleagues, Jacqueline Blyth and Hilary Griggs for some of the ideas talked about in this article. Research referred to in this article: Alderfer, M. A., Long, K. A., Lown, A., Marsland, A. L., Ostrowski, N. L., Hock, J. M., & Ewing, L. J. (2009). Psychosocial adjustment of siblings of children with cancer. A systematic review. *Psycho-oncology*, 19, 789-805; And, Houtzager, B. A., Oort, F. J., Hoekstra-Weebers, J. E. H. M., Caron, H. N., Grootenhuis, M. A. & Last, B. F. (2004). Coping and family functioning predict longitudinal psychological adaptation of siblings of childhood cancer patients. *Journal of Paediatric Psychology*, 29, 591-605

**> Encourage your child to keep a diary or scrap book.** Younger children can collect bus tickets and pictures of hospital visits and look back on these things as they talk with their parents about the visit. Older children can use the diary as a safe place to express their feelings and reflect on their experiences.

**> Develop a culture of openness.** Talk about your feelings and encourage your child to talk about theirs. Give lots of reassurance that all their feelings and emotions are normal, okay to have and you can deal with them together. And let them know you love them no matter what.

**> Make sure school is aware that your child has a brother or sister with a chronic health condition.** And talk about how this may affect them. Parents often liaise with school about their child with PH but it is important to do the same for siblings too.

**> Set and maintain boundaries around acceptable behaviour.** There can be a tendency for some parents to make allowances for brothers' and sisters' disruptive behaviour because they feel they have so much to put up with already, but all children need clear boundaries. Be clear about what you expect of their behaviour – and deal with their emotional needs separately.

**> Use puppets and soft toys.** This can be an excellent way to encourage younger children to talk about their feelings in a non-threatening way. Play, listen and reassure through play.

**> Remember support is out there.** If these ideas do not help on their own, contact your child's medical team for further assistance. ●





# Green Leaf

## Crew!



### GREEN LEAF CREW Q&A RHIANNON LLEWELLYN-BISHOP

Seven-year-old Rhiannon Llewellyn-Bishop lives in Cardiff with her mum Emma who was diagnosed with PH in 2011. Rhiannon is mad about karate and has entered competitions all over the country and even trained with European medallist Emma Lucraft. Here, she answers our Green Leaf survey!

**Q.** What is your favourite biscuit?

**A.** Oreo.

**Q.** What's your favourite thing to do?

**A.** Karate. I train six days a week.

**Q.** What superpower would you most like to have?

**A.** I'd love the ability to fly.

**Q.** What is your favourite food?

**A.** Fish.

**Q.** Who is your favourite singer or band?

**A.** Ed Sheeran. And ALL his songs are my favourites.

**Q.** What is your favourite and least favourite thing to do at school?

**A.** I don't like the hard work. I like the karate lesson I have at school – and the nurture group I go to.



Would you like to answer the Green Leaf Q&A? Contact us at [editor@phassociation.uk.com](mailto:editor@phassociation.uk.com)

**Q.** What famous person would you most like to have a selfie with?

**A.** Ed Sheeran.

**Q.** Who is the most famous person you have met?

**A.** Gareth Bale came to our school.

**Q.** Do you have a big ambition?

**A.** To go up the grades and get a black belt in karate.

**Q.** What's your favourite film?

**A.** Willy Wonka and the Chocolate Factory.

**Q.** What do you love about Christmas?

**A.** Going to see Santa!

### Were you there?

It was fun for all at PHA UK's 'phantastic' 15th anniversary conference with plenty to do. Hope you had a cracker of time if you were there too!



## The Green Leaf Crew

by David Banks



## Patch

by David Banks

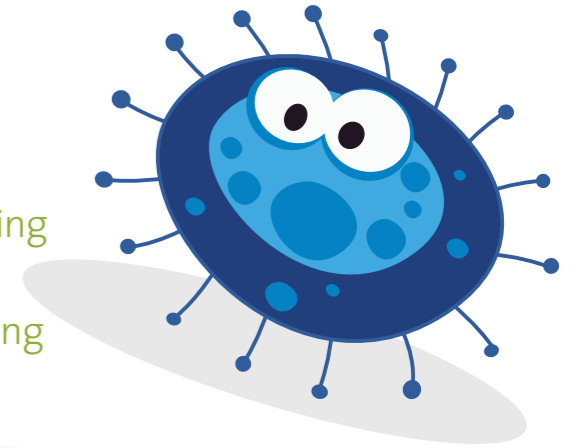




# Coping with winter vomiting bugs



A common query asked of the team at the PHA UK office at this time of year is about coping with 'winter' vomiting bugs. Here consultant pharmacist *Neil Hamilton* gives advice on dealing with this unpleasant little bug.



**V**omiting bugs can theoretically occur at any time of year but are more common in winter. There are many different strains, the most common of these is called Norovirus.

Norovirus is a highly contagious stomach bug causing a high temperature, stomach cramps, vomiting and diarrhoea. It is estimated that annually it affects up to a million people in the UK.

Whilst this is highly unpleasant for anyone, it is usually a very short-term problem lasting only a few days and virtually everyone will make a full recovery without needing to see a GP or visit A&E. In fact, I would advise patients to definitely not go to the surgery unless symptoms persist more than 72 hours. If this is the case, contact them by phone for advice. This is due to the highly contagious nature of the bug, meaning that contact with others could almost certainly spread the infection.

To reduce the chances of passing on stomach bugs to others, extra care with hand hygiene is essential. This means frequent hand washing, not sharing flannels or towels and maybe using alcohol hand gel if you want to make really sure. These measures are particularly important if you have either small children or older people in the house.

PH patients do not cope with any infection well. The body diverts all its attention to fighting off the infection so patients often notice an increase in breathlessness. This is likely to be made all the more significant if the stomach



bug means that usual medication doesn't 'stay down'.

In addition, patients who take diuretics (which are absorbed despite the bug) may notice more problems with thirst and dehydration. Careful management is needed because light-headedness or dizziness may be down to either dehydration or the PH. If these are occurring at rest, they are far more likely down to norovirus than PH.

If this is the case for you, there is no specific advice to cover all eventualities. However the most vital consideration through the (hopefully short) time of the bug is keeping well hydrated and keeping blood sugars as normal as possible. This is achieved by gentle sips of clear fluids. Rehydration sachets can be helpful to replace valuable salts.

Paracetamol can help with the symptoms of stomach cramps but there is little else that can be taken to treat norovirus. The bug will run its course in only a few days. If possible I would advise against the use of loperamide

or other agents for diarrhoea. These can prolong the duration so should be avoided.

In terms of PH medication, I would simply apply common sense. In general, tablets are absorbed from the stomach into the system within about an hour, but that depends on many factors and will vary hugely. However if you are sick after taking your medication we cannot be sure how much has been absorbed or not. I would not 'double up' by taking more just in case. I would concentrate on keeping up your hydration and blood sugars and try again when your next dose is due.

*As this is only likely to be a short-term problem there are more risks from taking too much of your medication than missing a few doses whilst poorly with a stomach bug. However, if you are unfortunate enough to catch norovirus and you have any specific concerns or questions, don't hesitate to give your PH centre a ring. ●*



# Keep Warm THIS WINTER

(WITH A LITTLE ADVICE FROM OUR FRIENDS)

The winter months are upon us so it's time to wrap up against the cold. When you've got a chronic health condition like PH which can affect your circulation and slow you down, it is particularly important to keep warm.

Our friends at the Raynaud's and Scleroderma Association and Scleroderma Society are also focused on helping people to defend themselves against winter chills, particularly in their extremities. Many PHA UK members are familiar with Raynaud's and scleroderma as there are strong links between the conditions and PH.

Raynaud's affects around five per cent of the population. The condition narrows the blood vessels and can severely reduce blood flow especially in the fingers and toes. A small proportion of people with Raynaud's also develop scleroderma, a rare autoimmune condition, through which individuals overproduce collagen which deposits within the body causing hardening and scarring. The condition can affect the skin, muscle tissue and internal organs. Some patients with scleroderma go on to develop PH as the condition stiffens and thickens the walls of the pulmonary artery, which, in turn, causes a build-up of pressure and puts strain on the heart. This type of PH is called connective-tissue disease-associated pulmonary hypertension. For more information please visit the Scleroderma Society website at [www.sclerodermauk.org](http://www.sclerodermauk.org)

And for a little expert advice to help you keep warm this season, visit the Raynaud's and Scleroderma Association (RSA) shop online at [www.rsa-shop.co.uk](http://www.rsa-shop.co.uk)

*The page opposite shows a few cosy suggestions from the RSA shop.*

**NEWS FLASH:** The Raynaud's and Scleroderma Association and the Scleroderma Society have announced plans to merge in early 2016 to form Scleroderma and Raynaud's UK (SRUK). We wish our partner charities every success going forward together!



## SILVER TOUCH

Did you know that pure silver is a great insulator and minimises radiative body heat loss by reflecting the body's energy back to the skin? Silver actually has one of the lowest radiant heat loss rates of any natural substance and so keeps warmer for longer. For cold hands on chilly days, fine knit gloves which are 8 per cent silver are sold via the RSA website priced £9.50. Long and shorts socks, with between 9 and 12 per cent silver content, are also available priced between £6 and £17.



## HEAT PACKS

A lot of people keep disposable Mycoal hand, body or foot warmer pads in the car or at home, to pick up and use whenever chills set in. These disposable packs can provide up to seven hours of heat once you've given them a little snap and squeeze. They can be popped in pockets, boots or even on your lap or behind your back when seated.



## THERMALS

There is nothing like a layer of thermal underwear to help keep us warm in winter. Fans of thermals include members of the Royal family wearing nice, smart dresses or dapper suits at formal outdoor engagements in windy spots; market traders who stand in some of the draughtiest workplaces in the land and thousands of uniformed professionals! Pull on the thermal underwear before you dress for a reassuring layer of warmth on a cold day.



## FEET HEAT

Another top seller on the RSA website is self-heating Astec insoles (just under £25 a pair) which use natural materials and patented nano technology to generate warmth by stimulating and invigorating the feet.



## BLANKET

The words 'comfort' and 'blanket' have always gone together. Have you got a good one? If you are snuggling up at home you may like to invest in a premium super-soft, lightweight, 1.4 tog fleece blanket (£25).

## KEEP WARM, KEEP WELL - TOP TIPS FROM [www.nhs.uk](http://www.nhs.uk)

- If you have reduced mobility, are 65 or over, or have a health condition such as heart or lung disease, you should heat your home to at least 18C.
- You can also use a hot water bottle or electric blanket (but not both at the same time) to keep warm while you're in bed.
- Food is a vital source of fuel, which helps keep your body warm. Try to make sure that you have regular hot meals and drinks throughout the day.
- Wrap up warm, inside and out. Layer your clothing to stay warm. If possible, stay inside during a cold period if you have heart or respiratory problems.
- Catch up on sleep on the long dark nights. The Sleep Council recommends seven to nine hours a night. It's okay to catch up in winter and hibernate a little!
- Eat fruits and vegetables and drink some warm milk to help boost your immune system and fend off colds.



.....  
**theinterview**  
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.....  
Professor  
**PAUL CORRIS**

in conversation with  
**Deborah Wain**

.....

Professor Paul Corris, based in Newcastle, is a leading physician in the field of heart and lung transplantation and holds many eminent positions. He is Professor of Thoracic Medicine for the Institute of Cellular Medicine at the University of Newcastle and the Regional Cardiothoracic Centre at Newcastle's Freeman Hospital.



In addition, he is Director of Cardiopulmonary Transplantation and the Northern Pulmonary Vascular Unit in Newcastle. Over the last three years, the Freeman has performed more cardiopulmonary transplants than anywhere else in the UK and is widely regarded as the premier lung transplant service in the country. Paul's other roles include academic lead and deputy chairman of the Institute of Transplantation. Paul is also a past president of the International Society for Heart and Lung Transplantation and vice president and president elect of the British Thoracic Society. He sits on the Councils of the British Thoracic Society and British Lung Foundation. Paul is a past president of the UK Pulmonary Hypertension Physicians Group and also sits on the Medical Advisory Council of PHA UK. Paul spoke on the subject of future therapies at the PHA UK conference.

.....  
**Q. Briefly, how did you get to where you are?**

**A.** Interesting! I would probably have been the person voted in my year at medical school least likely to have become a professor because I spent too much time playing rugby and partying. However, I always had confidence and once I qualified I concentrated on learning and was always inquisitive. I like thinking outside the box. I studied in London, Leicester, and Newcastle before my first consultant appointment in 1986. I was inspired by many. I spent a year on sabbatical as visiting Professor at McGill University Montreal Canada, in 1994-1995, and returned to Newcastle achieving a full professorship in 2000. I hold visiting professorships in Australia and Italy. I followed leadership advice from Sir Ernest Shackleton.

.....  
**Q. How many patients attend the Newcastle PH service?**

**A.** The Newcastle service is comprised of a hub at Freeman Hospital in Newcastle with spokes to satellite centres in Hull and Belfast. Clinics are held weekly in Newcastle and Belfast and monthly in Hull. On average the service will see four to five new patients each week and approximately 600 patients are currently followed.

.....  
**Q. How do the Newcastle PH service and transplant service interact?**

**A.** There are very close relationships because three of the physicians are appointed to work in both services. This means that patients with PH attending the Newcastle centre get seamless referral to the Transplant Service when necessary. I should point out that referrals for transplantation from other

centres are promptly dealt with and patients from Scotland and Northern Ireland may be seen in satellite transplant clinics in Edinburgh, Glasgow or Belfast to facilitate assessment because the patient will always be at the centre of what we do.

.....  
**Q. When was the first PH patient transplanted in Newcastle?**

**A.** That is an easy question because the patient and transplant are so memorable. The patient, a young boy came from Aberdeen and had complex congenital heart disease. It was 1989 and there were no targeted PH therapies available. He had a heart-lung transplant and is still alive and well today 26 years later! I just saw him recently in clinic. He was very ill during the first month post transplantation with a severe infection and I had little sleep for days when looking after him. It was worth the loss of sleep! We started our lung transplant program in Newcastle in 1987 and performed the first successful single lung transplant in Europe. I am probably the longest serving lung transplant physician in the world just now but still have much to learn.

.....  
**Q. Can you tell us a bit more about a patient's journey to transplantation?**

**A.** It can be long, arduous and a bit frightening but we try to make it smooth. Generally we would receive a referral from a colleague when the patient had reached a point when referral was advised from published international guideline, which I played a role in writing. We would review the letter and if there were pieces of information missing write back for this. With all >>>



theinterview

Professor  
PAUL CORRIS

.....  
“We started our lung transplant program in Newcastle in 1987 and performed the first successful single lung transplant in Europe. I am probably the longest serving lung transplant physician in the world just now but still have much to learn.”  
.....

the information either originally supplied or received we would generally either offer a patient an outpatient appointment to discuss further or, if everything looked as if transplant was possible and necessary, a direct four day assessment admission. Some patients may have contraindications clear from the referral letter and would be turned down at that point. Patients ultimately admitted for assessment might be accepted in principle but regarded as too well at that time to be listed. Others would be listed immediately. A few patients may have contraindications found at inpatient assessment and be turned down. A few patients decide that they do not want to undergo transplantation at this final assessment. Patients listed must then wait until donor lungs become available and this can be a very stressful period of time for obvious reasons. There has been much research and innovation leading to new ways of bridging highly selected patients with PH to transplantation who become very sick whilst waiting, though sadly some patients will not survive the wait. When suitable donor organs are available and transplantation becomes a reality for the majority, patients can expect to be in hospital for a month before return home cured of PH. Yippee!!

.....  
**Q. What do you consider your best achievement to date?**

**A.** No question. Successfully bringing up a family in a loving environment with my wife over 35 years of wedded bliss, despite the stresses and strains of my career is number one. I'm a very recent grandfather to Lilia Rose - 8 lb, 3 oz of sheer magic! Professionally, I'd say my efforts to improve the outcomes of patients following lung transplantation and patients with PH including a major role in developing national services for both will be my legacy, as will the numerous excellent fellows that it has been my privilege to train.  
.....

**Q. You talked about future therapies at the PHA UK conference. What developments are around the corner?**

**A.** The physician scientists and basic scientists continue to work hard to find new treatments and new ways of using existing drugs. The pharmaceutical industry also has many new potentially useful drugs in the pipeline. Early use of triple combination therapy has in a preliminary trial been shown to result in very good responses and a full trial is currently planned. For many patients treatment with prostaglandin therapy is not possible because of the method of delivery but a new oral prostacyclin agonist drug called Selexipag has just been shown to be effective and will allow many more patients to be treated. Drugs based on improving the functionality of a receptor called BMPR2 which is often defective in patients with PAH are available and trials ongoing. Inflammation may play an important role in disease progression in PAH and a new UK based trial of an anti-inflammatory drug will soon commence. Translational research, or research leading to direct patient benefit in PAH, is very active at present and greatly enhanced by the collaborative research spirit in the UK. Together we are striving for success. Finally stem cell therapy research continues and the PHA has recently produced a very useful patient guide written by Dan Reed.

.....  
**Q. And finally, how would you describe yourself in three words?**

**A.** That's impossible, but here goes... 'magnanimous, not mean', 'edgy but not moody', 'fun-loving and effective but not magnificent'. I'm failing the three words! ●

IN THE



## THE SKY'S THE LIMIT FOR GRANDDAUGHTER

Determined granddaughter *Rio Maddock* did a three-mile high plane jump to let people know about the impact of PH, and publicity around her efforts had just the desired effect.

**T**housands of newspaper readers in Essex know more about the effects of PH thanks to a nerve-jangling challenge by the granddaughter of patient *Jacky Cant*.

Rio Maddock, 25, leapt 17,000 feet out of a plane to raise awareness of Jacky's condition and money for PHA UK.

Her efforts were featured in the family's local newspapers, the Colchester Evening Gazette and Essex County Standard.

Jacky said many people had spoken to her after the story was published to tell her they hadn't previously heard of PH but now understood more about it. An old friend of Jacky's who she had lost touch with, even turned up on her doorstep and promptly presented a cheque for £500!

Mum-of-one Rio, who is a hairdresser, paid for the sky dive herself so every penny could go to PH UK. Her original target was £500, however the total donated so far has already topped £3,300. Her JustGiving page was in the top three per cent of fund-raisers out of over 83,000 during one particular month.

Proud Jacky said: "The sky dive was a real challenge for Rio and especially since high winds meant she couldn't jump the

first time she went up. She had to wait for an hour and a half on the ground before she could go up again!

"We watched her do the jump and it was nerve-wracking for us too. It's fantastic that the sky dive has brought in so much money for PHA UK but I'm just as pleased that the coverage has raised awareness."

Jacky, 67, believes she has been suffering from PH for several years. She was finally diagnosed at London's Royal Brompton Hospital in 2011. She is on oxygen 24 hours a day and can't walk far but uses a scooter.

She said the support of her large, close family, helps her to cope – especially husband David who "does so much for me". The couple are celebrating their 50th wedding anniversary next year.

Jacky said: "Before I became ill I was a very active person and I still try to get out every day, even if it's only for an hour. Although it's hard sometimes, I try to make the most of every day,"

Rio is considering taking on another fund-raiser next year and Jacky has given her some firm pointers: "I've told her I don't mind what she does as long as she keeps her feet firmly on the ground!" ●



Thank you to everyone who raises awareness of PH through their local media. Please do share your stories with us.



# Our fundraising PROMISE



There has been a lot in the news lately about the impact of fundraising activities by some of the UK's larger charities. We don't want to comment on concerns about the practices of others, but we would like to take this opportunity to reassure you of our commitment to very high standards.

PHA UK is extremely fortunate to have hundreds of dedicated supporters who put a tremendous amount of time and effort into sponsored events, activities and collections to support the charity's work. Every penny donated is appreciated and helps us to support the PH community and invest in research to better understand PH and develop new treatments and – hopefully one day – a cure.

As a charity, we do everything we can to support our fundraisers and fundraising. We have created a new fundraising pack this month (see page 10). And we are currently busy promoting our new PHA UK Lottery, which gives people the opportunity to make a regular donation to the charity - with the chance of winning a cash prize too.

We also support a well-used Just Giving page through which people can manage their sponsored events and easily attract gift aid if their supporters are tax payers. We absolutely love to celebrate our fundraisers' achievements through Facebook and Twitter and in the pages of this magazine.

We do not work with fundraising contractors; we do not make cold-calls; we don't invest in mass mail-outs or pay for door-knocking

exercises. (We do appreciate these activities are part of some charities' fundraising strategies and we support the view that such activities should be ethically managed and abide by stringent regulations, which may now be under review.)

PHA UK is a member of the Fundraising Standards Board (FRSB), a self-regulatory scheme designed to ensure all organisations raising money for charity do so honestly and properly. We are fully committed to the principles set out in the FRSB's code of practice.

For example, we can assure you we do not share our members' contact details with anyone. We keep your details safe and secure, according to data protection law.

We aim to be honest, open and clear about how we use the money donated to PHA UK. Our annual report summary is due to be published in the spring issue of *Emphasis* - and if anyone has any questions about spending, you are welcome to get in touch. We will certainly never put undue pressure on anyone to make a gift if they do not want to; and will respect anyone's wish to cease giving. We respect the rights, dignities and privacy of all our supporters and beneficiaries – and aim to be responsive to their needs and requests.

If you have any concerns at all about fundraising, you can contact us on **01709 761450** or **office@phassociation.uk.com**. And, thank you again to all our supporters.

**Iain Armstrong**  
Chair of PHA UK

To see the FRSB code of practice in full, please visit [www.phassociation.uk.com](http://www.phassociation.uk.com)

# Win! Win!

Make your New Year resolution a good one – sign up to play the **PHA UK Lottery**. Raise vital funds to improve the lives of people with PH and fund research and development. Enter the weekly draw for your chance to win a cash prize.

It's easy to sign up! Visit the **PHA UK** page at [www.unitylottery.co.uk](http://www.unitylottery.co.uk) to register and play. Or call the **PHA UK** office on **01709 761450** for an entry form.



## In your spring issue of *Emphasis*...

The next issue of *Emphasis* is due out in spring 2016 and we have planned features about:

- **Handle with care** – how to use the internet to safely research health matters.
- **Exercise training for people with PH** – a special report from the Scottish Pulmonary Vascular Unit in Glasgow.
- **Me and my job** – working with pulmonary hypertension. We talk to some people with PH about how they manage their condition at work.

Plus, lots more articles, interviews and news...

You can get involved in *Emphasis* too:

**Emphasis Exchange and Reviews** – don't forget we'd like to hear from you if you have news to share or if you'd like to review a book, app or film you think other readers may be interested in.

**Family Matters** – if you're interested in telling your family's PH experience in our regular feature, drop us a line.

**In the News** – let us know if you raise awareness of PH through the media.

**Take the biscuit!** – and please get in touch if you'd like to answer the Green Leaf Crew Q&A.

To get involved and to offer feedback and ideas, contact [editor@phassociation.uk.com](mailto:editor@phassociation.uk.com)





# Keep Calm at Christmas

Rebecca Rose talks about surviving seasonal stress.

Article reproduced with kind permission of Anxiety UK

**W**e are constantly told that this time of year is the season to be jolly. But I, for one, view it more as the season to be stressed. The build up to Christmas (that seems to begin earlier every year!) can seem like a pressure cooker; and with the combination of anxiety-inducing situations it can be the most traumatic time of year for some. The holiday season will probably never bring you the magic it did during childhood, but there are ways to make it much more bearable, and dare I say, enjoyable.

One of the biggest issues we face during this period is the burden of keeping other people happy. Whether it be worrying about who to buy presents for, cooking Christmas dinner, making sure we catch up with everyone we need to - it all amounts to putting other people first and forgetting ourselves. The key here is not to burn-out. If anything feels like too much then take it down a notch. It is easy at this time of year to forget that Christmas is not the most important thing in the world so figure out what needs to be done and prioritise accordingly. It may help to write it all down in to-do lists. Attend to the most urgent first, and keep your expectations realistic.

The cost of the holiday season can be horrific, but again do not have too high expectations of yourself and budget wisely. Everyone is going through this and it really is the thought that counts with presents; try craft sites, homemade bath salts, jams or biscuits go down a treat and show that you really care.

A side symptom of this season is the dark and cold nights, making it harder to get going in the mornings and it can have a severe effect on some people's moods. For this it is just a matter of coping until spring sadly, but there are a few things that can help such as making your house as cosy and comforting as possible, using a gradual lighting alarm or a lightbox, investing in a shawl (my favourite winter purchase!), and generally being aware that it is the season affecting you and that this will change.

Crowds are a huge issue for many people and they are hard to escape. My advice would be to plan your trips, avoiding late night or weekend shopping as this is when places will be busiest. Most shopping can be done online these days too. Sticking to pubs, bars and restaurants you know can be preferable if you do go out socially, but remember to book in advance! If you do find yourself in a panic-attack situation



If you would like to speak to someone at Anxiety UK, please call them in **0844 332 9010**. This is a free dedicated Anxiety UK helpline for PHA UK members if they are feeling depressed or anxious and need to talk. The helpline is open from 9.30am to 5.30pm Monday-Friday. You can also visit [www.anxietyuk.org.uk](http://www.anxietyuk.org.uk)

then find a quiet spot (toilet cubicles if nowhere else) and focus your attention on the moment and on soothing memories. Imagine that someone that loves you is there and picture how they would comfort you. Whenever I feel too panicked in crowds I find a book shop as they generally have quiet corners in which to re-coup.

Avoidance is never the best thing in the long-run though. If your anxiety or mood leads you to avoidance tactics then it really is worth seeking help. Simple CBT can be highly effective but longer-term therapies or medication may mean you can truly have the life you deserve and desire back.

The hardest thing over Christmas for most is feeling alone. The strong focus on family can trigger all kinds of deep emotions; sadness, loss, grief, anger and even jealousy. It can feel like a mirror is being held up and all you can see is what you did or should have. Loneliness is not about physical proximity but more a feeling of isolation, whether that be real or felt. To tackle this we must first remember that we are not the only ones feeling this way; there is always someone worse off than yourself that you could be helping! It is a good time to talk about it and reach out to others, and you never know, by admitting your own struggle you may be able to help yourself as well as someone else. Community events and volunteering roles are a great way to seek out people to share the season with, and forget the clichés because there will be a great variety of people at such occasions with real life experience and advice (and who are probably sharing your emotions.)

*This time of year does not have to feel hopeless. It can be a fresh start if we just change our perspectives slightly. It can be a time to truly address issues we may have, to reconnect with loved ones we have neglected, to open up about the battle we have with our emotions; and make new friends. So go ahead and run at Christmas head on because it's the season of togetherness and it may be the best time ever to reach out. ●*



# Join us free today and be part of a 3,000 strong national support network.



*The only charity in the UK dedicated  
to people with pulmonary hypertension.*

*Are you living with PH? Do you have family or friends who are? Are you interested in knowing more about the treatments available? Would you like to get involved with fundraising or stay up to date with the latest PH news & events? If so, we are here to support people like you.*

*At PHA UK we are committed to helping improve the lives of people with PH, and supporting the PH community across the country through funding research, raising awareness, and helping our members in their day-to-day lives. Our website provides useful advice, access to publications, audio and video presentations, plus ways to get involved with the organisation.*

*Our regular **Emphasis** magazine keeps people informed and engaged with all things PH. In addition our friendly, knowledgeable office staff are just a phone call away with one to one advice. But most importantly it's our 3,000 members who form a unique network of support and inspiration to each other. We think of it as one big family and there's always room for new people.*

**Join FREE today at [www.phassociation.uk.com](http://www.phassociation.uk.com), call us on 01709 761450, email us at [office@phassociation.uk.com](mailto:office@phassociation.uk.com)**



#### Support, advice and much more...

- Be part of a nationwide network
- Advice & support for patients, friends & family
- Fundraising ideas
- Latest news & events
- Regular supporters magazine
- FREE membership



## Inside PHA UK

#### PHA UK Contact Details

**Office hours:** 9am to 3pm, Mon to Fri for general enquiries  
**Tel:** 01709 761450  
**Web:** [www.phassociation.uk.com](http://www.phassociation.uk.com)  
**Email:** [office@phassociation.uk.com](mailto:office@phassociation.uk.com)  
**Address:** PHA UK, Unit 2 Concept Court, Manvers, Rotherham, S63 5BD

#### Anxiety UK

PHA UK works closely with our partners at Anxiety UK. To speak to someone about how you are feeling, call the dedicated PHA UK helpline on: 0844 332 9010

#### Do we have your correct details?

Please email us on [office@phassociation.uk.com](mailto:office@phassociation.uk.com) if any of the following apply to you:

- Are the details incorrect on the mailing you've just received?
- Have you moved house recently?
- Has your contact number changed?

#### Contact a friend

**Kathy Davis:** Kathy regularly meets with other PH-ers who live in the Portsmouth area of Hampshire. If you're interested in meeting up with this small and friendly group, just contact Kathy on 02392 365760 or email her at [davik7hmd@ntlworld.com](mailto:davik7hmd@ntlworld.com)

**Jocelyn Barker:** Jocelyn's friends support group is still going strong in London and meeting on the first Wednesday of every month between 10am and 1pm. Anyone is welcome, but it's best to call Jocelyn first in case the date has to change (Although this rarely happens) on 0207 738 7085 or email [jocelynab@gmail.com](mailto:jocelynab@gmail.com)

The PHA UK annual general meeting will be held at the Pullman St Pancras hotel, London, on Thursday 3rd December. Please contact [office@phassociation.uk.com](mailto:office@phassociation.uk.com) for further information.

#### PHA UK Trustees

**Dr Iain Armstrong**, *Chairman*  
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*Clinical Nurse Specialist – Hammersmith Hospital, London*  
**Yvette Flynn**  
*Clinical Nurse Specialist – GOSH, London*



# What is PH? How can it be identified? How can it be treated?



GPs, doctors, nurses and healthcare workers across all disciplines can learn more about pulmonary hypertension through PHA UK's medical education programme.



## *'Pulmonary Hypertension - diagnosis and management'*

This flexible e-learning resource incorporates text, video and interactive tools to develop knowledge and skills. It can be self-managed with online assessments.

The course can be taken as part of personal career development and gains a certificate on completion. Or, it can be studied as a module worth 20 points as part of a degree or masters course. PHA UK worked with Edgehill University for this accreditation.



To find out more contact [education@phassociation.uk.com](mailto:education@phassociation.uk.com) or visit [www.paheducation.co.uk](http://www.paheducation.co.uk)