SUMMER 2018

emphasis

The magazine for supporters of PHA UK www.phauk.org

Travelling with pulmonary hypertension

Your holiday questions answered

Help for anxiety

The difference it can make

Studying with PH

The financial help available

My fight to become a mother

Sonia's adoption story

Summer reads

Our book review feature is back!

PLUS...

Scotland says 'yes' to Selexipag, your fundraising news and much more!

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pha UK | Influence. Hope. Integrity.

Latest News...

Living with PH...

Regular Features...

This magazine is printed on paper from sustainably managed sources

Welcome

Welcome to the summer 2018 issue of Emphasis. We're still smiling following our first ever PHact Finding and Fun Weekend, which saw us collaborate with Great Ormond Street Children's Hospital (GOSH) to host a special event in Oxfordshire for young people under their care.

"PH shouldn't stop you going away and making precious memories"

We couldn't have asked for better weather, and a novel fundraising idea helped some of us cool off - you can see photographic evidence of myself and Dr Shahin Moledina of GOSH being pelted with wet sponges on page 26. Thanks to everyone who made the weekend such a success; it was great to see so many families enjoying a happy break away together.

On the subject of holidays, at this time of year we get lots of calls to the office for advice on various aspects of travelling abroad with PH, so we've put together a special feature on page 12 that answers some of the most common questions we get asked. Ultimately, it just takes a bit more planning – but having PH shouldn't stop you going away and making precious memories. We all deserve that.

Thank you, as always, to everyone who has shared their thoughts and experiences with us this issue. Adopting as a single mother, getting help for anxiety, life after surgery, and bringing up a child who has PH are all covered in special features that we hope readers will find both informative and inspirational.

Finally, we're delighted to bring back our book review feature on page 15 and we'd love you to share your favourite health-related reads with us too. Enjoy the summer.

Iain Armstrong
Chair of PHA UK
editor@phauk.org

Emphasis *exchange*

If you have any news and views to share via the Emphasis exchange please e-mail **editor@phauk.org** and don't forget to keep in touch via **Facebook** and **Twitter** too.

Drive to end 'PYJAMA PARALYSIS' *in hospitals*

A campaign to get people up, dressed and moving while in hospital will run until 26th June.

The drive to end 'pyjama paralysis' aims to give patients back one million days of their precious time that would otherwise be wasted in bed.

Professor Jane Cummings, Chief Nursing Officer for England, launched the 70-day **#EndPJparalysis** challenge, which started on 17th April.

A recent pilot gave patients back 91,728 days' - or 250 years' - worth of time across nine trusts in the East of England as a result of getting patients up and dressed.

The **#EndPJparalysis** challenge aims to build on its success, with an ambition to have a million patient days captured in just 70 days.

Studies show that three-in-five immobile, older patients in hospital had no medical reason that required bed rest and doubling the amount of walking while in hospital reduces the length of stay.

Professor Cummings said: *"For many, wearing pyjamas reinforces feeling unwell and can prevent a speedy recovery. One of the most valuable resources is a patients' time and getting people up and dressed is a vital step in ensuring that they do not spend any longer than is clinically necessary in hospital."*

Professor Brian Dolan, Visiting Professor of Nursing, Oxford Institute of Nursing, Midwifery & Allied Health Research (OxINMAHR), added: *"End PJ paralysis has galvanised nurses, therapists, doctors and managers in a way I've not witnessed in a 30 plus year career, and so many are passionate about doing the right thing."*

Financial impact survey update

Thank you to everyone who shared their voices in our survey into the financial impact of PH. The analysis of the results is being finalised and we'll be sharing the findings soon. *Watch this space!*

Social Media Round-up

Top tweets and popular posts from PHA UK's Facebook and Twitter pages

@alicetg
Thanks @PHA_UK for a great w'end. Was good to meet other families of kids with same heart & lung condition that Molly has. Plate spinning & disco were a real hit!

@JessicaMRussell
Disappointing day brightened by seeing @GrangerKate with @PointonChris #hellomynameis article in my @PHA_UK magazine. Much missed on Twitter but her important message lives on

Capital B Media
Charlotte enjoying some magic at the PHA UK PHact Finding and Fun Weekend! We had a great time filming and chatting to families and meeting so many inspirational young people.

@JuliaPHNurse
I work in a highly specialised area. I am an expert in my field #IAmANurse Our patients travel from across the region to @NewcastleHosps. They trust our knowledge and expertise in this rare disease.

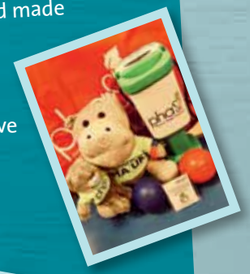
Donna Welch
My daughter has PH and her school is doing a 6-mile sand walk for charity on July 6th. Hannah (and her wheelchair) plus seven friends are taking part and they have decided to donate their sponsor money to PHA UK.

Tess Jewson
I am feeling very humbled that due to my raising awareness of PH every day, people have been contacting me to chat about their PH. I have been introduced to many PH'ers... and some have never spoken to anyone about it before... You are not alone!

@yorke_janelle
Symptom burden in PH is significant. We need more research in this group of patients to support symptom management

@thisisgerty
Thank you Mr Postie... cookies, a brew and the spring edition of @PHA_UK emPHAsis magazine. #findmypic

Janine Bennett
The PHact Finding and Fun Weekend was really great. Thank you to everyone who made it happen and thanks too to all the lovely people who were there and made it so much fun.



@kazawazamate
@PHA_UK some early guests have arrived for the SPVU patient day

@SEGrai32
Thank you @PHA_UK for sharing my story with others. Means so much to me to be able to tell my side of the story. I love the layout of what you've done. #PH #CTEPH

@DrGillianSmith
Scleroderma Family Day at Royal Free Hospital with @PHA_UK. Dr Dan Knight reminded us that 8-12% of scleroderma patients are affected by pulmonary hypertension. #SclerodermaFreeWorld #RareDisease @RoyalFreeNHS @SclerodermaRF @NIHRCRN_nthames

Thank you to everyone who took part in May's 'Twitter chat' to mark World PH Day.
It was great to see patients, carers, professionals and supporters come together online to put pulmonary hypertension under the spotlight.

Support day in Scotland

In March, PH patients in Scotland gathered for another successful Scottish Pulmonary Vascular Unit (SPVU) support day.

Organised by the PH team from the Golden Jubilee National Hospital near Glasgow, delegates were able to meet fellow patients and benefit from expert talks on topics including PH therapies, breathlessness, genetics and research.

The event area at the Golden Jubilee Conference Hotel also included stands to promote the help available from charities including the PHA UK and Breathing Space.

Golden Jubilee National Hospital
Patients at the heart of progress

Comments from patients who attended the day included:

"I had a great time catching up with PHriends and of course our fantastic team! Thanks for organising everything, looking forward to the next one."

"A lot of organising required for this and it all went splendidly. Thank you to all."

Crafting clever

Earlier this year, a craft day to raise money for the PHA UK generated a fantastic £1737.50. Organiser Sarah Colledge sent us this email to tell us all about it...

"The idea for a craft event to raise money for the PHA UK charity came last year when chatting to my mum, Suzanne Calder. She had started to crochet again and I discovered a love of sewing when I tried a beginner's sewing class. We had never made items to sell but we thought we could make small gifts and things for the home.

We started to make things last year and finally over a period of ten months we felt we had enough items to sell. I promoted the event to friends and family and very quickly the day arrived. It was held in my home and mum and dad came up to support and help out on the day.

A lovely friend had made us some knitted toys that all sold. My sewing teacher had donated fabric and had made some cushions and doorstops to sell as well. A huge thank you to my friend Sue and her little helper Daisy. Sue baked all the lovely cakes and we offered all the guests tea, coffee or Prosecco for a small donation.

We raised £960 on the day - far more than I thought we would! I had also created a Just Giving Page which raised even more.

The response and support we had was amazing. We were so busy and people loved the items we had made. All our efforts had paid off and we had such lovely comments."

Sip in style this summer

Now you can raise awareness of PH every day, just by enjoying your morning coffee.

Our new PHA UK branded thermal insulated mugs are dishwasher and microwave safe, BPA free, made in the UK and come in lime green, red or blue. With a handy screw-on lid and rubber-grip collar, these stylish mugs keep tea and coffee warm on-the-go, and they are also perfect for keeping cold drinks cool this summer.

Order yours from **www.phauk.org** *now for £4.99 plus delivery.*

PLAN AHEAD TO PROTECT

We might be in the swing of summer, but it's not too early to start thinking about protecting yourself from winter illnesses. Play it safe by planning ahead for your flu and pneumonia vaccinations, which are available for free on the NHS for those with conditions like PH. Visit www.nhs.uk for details.

Behind the HEADLINES

Earlier this year, the BBC reported the discovery of the genes that cause heritable and idiopathic pulmonary hypertension. We spoke to *Professor Nick Morrell*, who led the research on behalf of the British Heart Foundation, to find out what it really means for patients today.

What made this research unique?

"This research is the largest systematic search for genetic causes of PAH ever conducted. Previous studies have used small numbers of patients in single families to try to identify new genetic causes of PAH. This study recruited over 1000 patients and set a very high threshold (a high degree of certainty) for identifying new genes.
Our findings call into question some previously reported genetic causes of PAH, whilst providing definitive proof for others. *In addition, we identified four new genes that cause PAH and which can now be included in routing clinical genetic testing for PAH patients in the NHS."*


Professor Nick Morrell

What do the results mean for people living with PAH?

"For patients with a family history of PAH, these results mean that we should be able to provide patients with a genetic diagnosis (the cause of their PAH) in over 90 per cent of cases. For patients with a diagnosis of idiopathic PAH without a family history we will be able to identify the genetic cause of the condition in around 20-25 per cent of cases. For many people living with PAH this means we can provide an answer to the question "why me?". In addition, the finding of a genetic cause for the disease in a patient has immediate implications for family members who are at risk of carrying the mutation and developing PAH. If a mutation is found in a PAH gene in a patient with the disease, family members can also be offered genetic screening. It should be noted that the results apply to patients with heritable and idiopathic PAH. We have not yet looked to see whether these genetic findings are applicable to people with other forms of PAH."

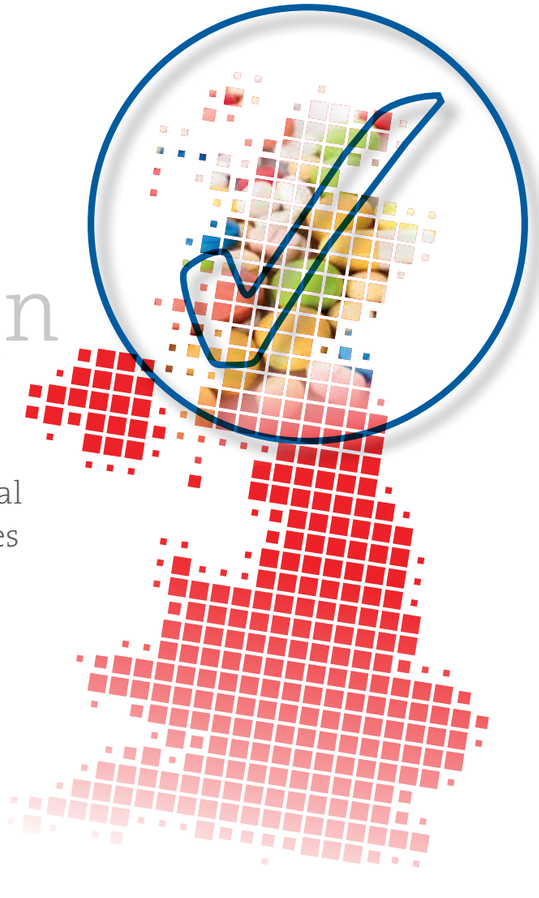
When do you think people living with PAH will benefit from the findings?

"Some benefits will be immediate. Our findings will be fed back to patients who consented to receiving the genetic results. There are many families with PAH, and patients with idiopathic PAH, who do not have an explanation for their disease or any means of predicting who in their family is at risk of developing the disease. Our results have extended the number of genes that can now be tested for in these individuals. Providing an answer for many patients. ***Our findings also shed new light on the causes of PAH,*** *which will stimulate much new research on the pathways identified, potentially leading to new ways to treat the disease."*

What's next? What do the findings mean for the future in terms of drug development?

"The reason why a lot of drug development fails is because we do not know exactly what causes a disease, but we make educated guesses about the biology and try drugs that work on particular pathways. When we find a genetic cause for a disease it means that gene or pathway is 'hard-wired' into the biology of that disease, so we can be sure it is fundamentally involved. This gives much greater confidence that if we 'cold design' a drug that affects the genetic pathway, it would have a much higher chance of success. ***Our findings will stimulate a lot of new research around the new PAH genes we have identified.*** *One in particular is already the subject of a new drug development attempt. Our study revealed that some patients with PAH have mutations in a gene called GDF2 (also called BMP9), suggesting that a lack of BMP9 in the blood can cause PAH. BMP9 is already being developed as a potential therapy for PAH."*

Victory in the campaign for selexipag



Your voices have helped secure the approval of a new drug designed to improve the lives of people affected by PH in Scotland.

Following months of campaigning and high-level talks with policy makers, it has been confirmed that selexipag has been approved for use by the NHS in Scotland.

The Scottish Medicines Consortium (SMC) made the announcement in May, after rejecting the drug in July last year because evidence of the clinical and economic benefits was 'not robust enough'.

Selexipag, also known as uptravi, helps to relax and widen the pulmonary arteries, relieving symptoms of pulmonary hypertension and slowing down progression of the disease. It is taken as a tablet, providing an alternative for the first time to drugs usually only available intravenously or by inhalation.

The PHA UK campaigned hard and in collaboration for the decision to be overturned, meeting with agencies and stakeholders and submitting key evidence in the form of the Living with PH survey.

The open letter we wrote expressing our dismay at the decision to reject the drug last July secured a national audience when it was printed in the influential Scottish newspaper the Sunday Herald.

Iain Armstrong, chair of the PHA UK, said: ***"This is a victory for everyone who shared their experiences in the Living with PH survey****, as your voices provided undisputable proof that the symptom burden of PH has a major impact on quality of life.*

"The depth and insight that this evidence brought to the decision-making process was crucial in securing this turnaround. We've been told by decision-makers that it was enlightening and refreshing to see compelling, relevant evidence that harnessed the patient voice.

"This is a great example of how, when you engage with our surveys, you are doing more than simply sharing your experiences. *You are making a significant difference and contributing to tangible outcomes – and it's vital that this continues."*

England and Wales have already reviewed selexipag once and said no, so the PHA UK is also working in collaboration with the National Institute of Clinical Excellence (NICE) and All Wales Medicines Strategy Group (AWMSG) to help them understand why funding the drug is so vital.

"We won't give up until we have the same equality and access to treatment as other disease areas like cancer."

Iain added: ***"The approval of selexipag in Scotland is a step in the right direction but we want to see equality across the UK.*** *Any decision to deny PH patients a drug that will improve their quality of life is grossly unfair and we won't give up until we have the same equality and access to treatment as other disease areas like cancer."*

For updates on the fight for access to selexipag in England and Wales, keep an eye on our website, Facebook and Twitter, and the next issue of Emphasis.

The PHA UK campaigned for access to selexipag via its advocacy group **PHocus2021**, a consortium of health professionals and patients pushing for public policy changes to improve lives for people affected by PH. Find out more about its aims at **www.PHocus2021.org**

Travelling abroad with PH

Having pulmonary hypertension shouldn't stop you enjoying a summer holiday overseas - it just requires a bit more planning. Here, we answer the travel questions we hear most often in the PHA UK office.

Visit the PHA UK Facebook page for additional travel advice in a special video with our co-founder Kay Yeowart MBE

What do I need to think about in terms of oxygen?

Travelling by air with oxygen will require a fair amount of additional planning and unfortunately, due to tightening regulations and security, it has become more difficult to organise your own oxygen for flights.

Well before you begin planning your holiday, on your next clinic visit, ask your specialist if you need oxygen to fly. Not everyone with PH needs to, but it's your specialist, not your GP, who should make that decision.

As a rule of thumb, the further the flight is, the more likely it is that you will need oxygen.

The majority of airlines will charge extra for oxygen. As an organisation we think that's wrong, but unfortunately that's the way it is at the moment.

There are UK-based companies that can help by supplying PH patients with portable concentrators for flights that don't provide oxygen themselves, and for use during the holiday. One of them is Pure O2 (see p15).

A HOOF (Home Oxygen Order Form) may be required; please check this with whoever prescribes your oxygen.

Although more expensive, a cruising holiday could be an alternative if flying presents too many obstacles. If you do decide to fly, then our ultimate advice is to check requirements directly with the airline before booking a flight to avoid wasting money.

When should I ask about a fit to fly letter?

It used to be that fit to fly letters lasted six months or a year, but things have changed. *Now, we advise phoning your specialist centre at least six weeks before you fly, as you'll need a letter dated as close to your departure date as possible. Airlines are becoming much more exacting and will vary in terms of what they require, so again, be sure to check with them in good time - regardless of what they said last time you flew.*

Fit to fly letters must be obtained from your specialist centre, not from your GP.

What happens if I get ill abroad?

If you're travelling within Europe, a European Health Insurance Card (EHIC) entitles you to get state healthcare in other European Economic Area (EEA) countries and Switzerland at a reduced cost, or sometimes for free.

It does not replace insurance, so you will still need to organise this before you travel. It is not yet known whether leaving the EU will affect the use of EHIC cards. At present, they can be used as normal. If you're travelling outside of Europe, having adequate insurance in place is even more vital.

It's common to get a stomach upset when abroad, so we advise visiting your local pharmacy before you travel to stock up on over-the-counter medication and rehydration sachets, just in case. It's also a good idea to take a first aid kit away with you.

If you fall ill and can't find anyone who speaks English, having a smartphone allows you to use 'Google translate' to communicate – simply type in the words in English and select the language you need to convert it to. Always keep a list of your medications on you too.

Our top Travel Tips

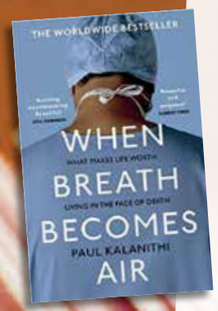
- Consider your destination carefully – will it be too hot at certain times of the year? How easy will it be to get to the hotel room?
- Split your medication between hand luggage and hold luggage, and always take more than you think you will need
- If you need to take medical equipment or drugs through airport security, ask your doctor for a letter to explain why you need them
- Always keep a list of your medication and important telephone numbers on you, in case of emergency, and make sure the details of your specialist centre are included
- Visit your local pharmacy before you travel if you have any questions about medication – they are experts and you don't need an appointment
- Keep your medication in a safe in your hotel room
- Consider booking through a travel agent if you want one point of contact for questions about your holiday

Wherever and however you travel this summer, we wish you a ***very happy holiday!***

BookReview

Would you like to review a book which may be of interest to Emphasis readers? Please contact **editor@phauk.org**

Looking for something to read in the garden or on the beach this summer? Check out these two very different titles...



When Breath Becomes Air by Paul Kalanithi

"This is the true story, in his own words, of a neurosurgeon whose life changes with his own cancer diagnosis. The doctor becomes the patient and comes to terms with his imminent death. This book is a testimonial to love. It also highlights both sides of healthcare; what it is like to be a doctor helping critically ill people, and the other side. Paul recognises how it feels to be the patient and it changes him and the doctors around him. There is nothing about this book that disappointed me, or that could have been improved. It's wonderful. After I had finished reading it, this book made me feel sad, yet uplifted. Paul lived his life well and faced his death with strength. I hope I can have such dignity when my time comes. I want every doctor to read it as it articulates the fear of being a patient; afraid, and out of control of your own body. All doctors should have this book as required reading. ***I would absolutely recommend this book to others as it is a subject that touches everyone."***

Reviewed by Karen Mosely, PHA UK member

MARKS OUT OF 5? **5.5!**

The Selfish Pig's Guide to Caring by Hugh Marriott

"Hugh writes with humour and breath-taking honesty about what it means to care for a loved one, addressing issues that few people openly discuss.
With chapters on sex, 'toilet mess', and even a section on thoughts of murder, the author doesn't just push boundaries with this book. He shoves them, hard, and smashes taboos with a sledgehammer. Ultimately, The Selfish Pig's Guide to Caring normalises the secret thoughts that carers may have and offers an important insight into what it really means to look after someone who depends on you for everything.
Covering emotions like guilt, isolation, anger and fear, it also offers practical advice on a wide range of issues such as finances, looking after your own body and accessing information and support. Refreshing and at times entertaining, the book can be read front to back like a novel (which is how I chose to read it), but you can also refer straight to chapters that chime with you.
This book isn't for everyone; if you're sensitive about your role as a carer, you could easily be offended by some of its content. But I imagine that for many people, its contents could be something of a lifeline."

Reviewed by Mary Ferguson, Emphasis editor

MARKS OUT OF 5? **4**

MY FIGHT TO BECOME **A MOTHER**

When Sonia was diagnosed with pulmonary hypertension at the age of 30, it left her shell-shocked and bewildered. She was terrified her quality of life would be non-existent and she would be left housebound and constantly attached to an oxygen machine.

"I really was very anxious," recalls Sonia, from Stockton-On-Tees, Cleveland. *"I'd suffered with medical issues most of my life. I'd been diagnosed with scoliosis when I was three as well as asthma a few years later, both of which I'd managed to keep under control.*
"However, this diagnosis seemed far more daunting. I envisaged being housebound and my life as I knew it being over." But despite Sonia initially being wired up to an oxygen machine overnight to help her breathing, she soon discovered that her diagnosis wasn't as restrictive as it first seemed.
She recalls: *"Obviously it took some adjustment and took some getting used to but I still managed to go about my day to day life. I could still meet up with my Mum, go shopping and socialise. I also managed to take my dogs out for a walk every day to try and keep myself as fit and healthy as possible."*
And even when Sonia was told she needed oxygen 24 hours a day, she refused to let it faze her. Instead she carried the portable pack around with her everywhere she went.
"I had two options," recalls Sonia. *"I could let the condition rule me or I could get on with life. I knew which one I was going to opt for."*

> "Every time I look at Anthony, I count my blessings."

But there was one overriding worry that did concern Sonia – she was desperate to become a mum.
"As I turned 38, I really felt like I needed to start looking at my options. My consultant had explained having a baby naturally was too risky and not a viable option.
"But even if it was something I could consider, I was single."
Desperate to fulfil her dream of becoming a parent, Sonia tackled the issue head on.

"To me it felt like the right time. I was feeling healthy, studying childcare at college and working part time.
"And adoption seemed the natural route for me to take."
But Sonia explains it wasn't all plain sailing. Despite Middlesbrough Social Services initially being extremely encouraging, Sonia was told, based on her diagnosis of pulmonary hypertension, she wasn't medically fit enough to be considered as a potential adoptive parent.
Sonia recalls: *"I was absolutely devastated. I felt like my only chance to become a Mum was being taken away from me. I hadn't even been offered a medical assessment."*
But undeterred, Sonia refused to be beaten and spoke to a solicitor who agreed to take her case on.
"Thankfully, they apologised and the process then began. I agreed to every medical, questionnaire and interview. I would be lying if I said it wasn't draining but I kept my mind on the end result and remained focussed.
"Finally, after several assessments from my own GP, a lung specialist and a cardiologist, it was stated I was medically well enough to look after children.
"I also had to prove I had support which wasn't a problem – I had a very close knit family who couldn't wait to welcome a new child into the fold."
Four years after stating her journey to become a parent, Sonia had one final intense interview with social services and in May 2015 got the news she had been longing for.
She recalls: *"After I had been quizzed by the panel of ten professionals, I was asked to wait outside. Those few minutes seemed to last forever but finally a social worker came out and said I'd been approved.*
"I was absolutely elated. All my hard work and determination had paid off."
Sonia was warned it may take a while to pair her with a suitable child but within a week, she received a call to say there was a 16-month old little boy that needed a loving home.

"I just cried and cried with happiness," said Sonia.
"All my dreams were beginning to come true."
In October 2015, Sonia's first visit with Anthony was arranged. *"As I saw him for the first time, I thought my heart would actually melt,"* she recalls.
"Anthony was sat in his highchair and as I knelt down next to him, he grabbed my finger, squeezed it tightly and smiled. We stayed like that for several minutes and instantly our bond was cemented.

> "I couldn't recall ever feeling so happy."

"I was totally besotted with his beautiful brown eyes, curly hair and infectious giggles. I couldn't recall ever feeling so happy.
"Even the social worker was mesmerised – she said she had never seen a child gel with a new adult so quickly."
Over the next three weeks, Sonia spent hours a day with Anthony, getting used to his routine and personality.
Anthony did suffer with a slight developmental delay. He hadn't started walking and his speech had been affected after being born tongue-tied but Sonia felt well equipped to cope with any problems her new little boy faced.
"I think having always had medical issues of my own gave me the skills and resilience to accept and understand that with help and support you could come overcome anything," said Sonia.
"Anthony hadn't progressed as quickly as other children his age but during one of my visits he took his very first steps. I already felt so proud of him and clapped and cheered. In that moment I knew together we would tackle anything."
Finally came the day for Sonia to take her son home.
"It was just a surreal moment driving Anthony home and showing him the nursery I had decorated for him. I finally had the child I'd always dreamt of."
Sonia and Anthony soon settled into their little family unit.
"He was such a happy little boy. Anthony was always smiling and made becoming a Mum incredibly easy.
After three weeks, I was giving Anthony a cuddle when he looked up at me and said Mum. I couldn't control the tears – it was just an incredible moment and I knew then, Anthony and I were always meant to be. I know my own limitations and never overdo it. On the days I'm tired, instead of chasing Anthony around a park, we go to an indoor soft play where he can run and climb until his heart's content.
"And my parents are always on-hand to help out, delighted to be able to spoil their grandson.
"Anthony is now coming up to his fourth birthday. I gave up my job to be a full-time Mum. We have to be careful with money but it's a small price to pay. I was given the chance to be a parent and didn't want to miss a single minute.
"Anthony and I are a great team. I spend hours helping him with his speech and he has got used to my oxygen tubes. They don't faze him and he knows not to pull at them.
"Every time I look at Anthony, I count my blessings. He was worth every minute of the time I spent fighting to become his mum."





If you would like to share your family's PH story, please email editor@phauk.org

Refusing to give up.

When *Kaylee Mynot* was born with PH, doctors said there was no cure, and she would die before she was five. For her mum, *Kaye*, that just wasn't good enough. Last year the pair spoke to online magazine Folks, who have kindly allowed us to reproduce their article here. >>>

Refusing to give up.

A pair of foil balloons – a one, and a seven – bob at the end of the table in Kaylee Mynot's South London home. It's her birthday weekend, and Kaylee has just turned seventeen.

Small and pale, with delicate features and long brown hair tied back in a ponytail, Kaylee doesn't look her age, but despite her childlike appearance, Kaylee is poised and articulate. She flashes a grin at me from across the table, revealing silver braces. I've just asked her if she thinks living with a condition gives her any advantages.

"Um, yes!", she exclaims, raising a small white hand in a flourish. ***"I jump all the queues, especially if I'm in my wheelchair. I got to make a wish once, I went to central London in a mo and went to Rainforest Cafe. I met [pop group] Girls Aloud, and went back home in a limo."***

Kaylee's mum, Kaye, is sitting beside her, drinking a cup of tea. She's a stylish, older version of Kaylee, with a smile that's a little more guarded than her daughter's. She reminds her daughter to eat. *"She's talking too much,"* she explains to me. *"She forgets to eat."* Kaylee obediently winds noodles around a fork. Noodles are one of the few foods she can stomach, along with chicken, mashed potato and carrots. Sometimes she'll have green beans. Meals are repetitive in Kaylee's house.

"If you can't find the good in something then what are you doing in this world?"

The pair don't argue much, but they agree food is a contentious subject. Food has been a problem that dates back to Kaylee's babyhood, when she couldn't keep anything down. For Kaye, it was a flashing neon sign telling her something was seriously wrong. The health professionals she spoke to brushed off her concerns, and by the time her daughter was eleven months old, Kaye was tired of Kaylee's problems being dismissed as "failure to thrive".

"I went to the doctor one day and said, 'I'm not leaving until you find out what's wrong'."

The doctor sighed, and to humour Kaye, did an oxygen test. The sixty-eight percent reading was so critically low the doctor assumed the machine was broken. When a second machine gave the same reading, Kaylee was blue lighted to hospital, where tests flagged up a congenital heart defect, intestinal malrotation and pulmonary hypertension, high blood pressure in the lungs. Kaye was told her daughter might live to be five.

"I came home and thought, this isn't it. It can't be. I started Googling." Kaye found a pulmonary hypertension specialist at Great Ormond Street Hospital and managed to get her daughter transferred into her care. Baby Kaylee became one of the youngest candidate for Bosentan, a drug which was being trialled on adults to slow progression of the disease. It worked, and sixteen years later Kaylee is alive and kicking – and still using the drug.

When I ask Kaylee what she thinks of that initial diagnosis, the confident teen disappears and she stumbles a little.

"Um. I don't know. I think it's amazing. I don't know how mum did it."

Kaye is team leader, coach, and head organiser. She has a plan, and if that doesn't work, she'll come up with a back-up. *"We have options. If you can't find the good in something then what are you doing in this world? That goes for everybody. You need to have nice things to look forward to. Yes, plans change, but you just find another way."*

The first thing Kaye decided was that her daughter would have as normal an upbringing as possible. So, like many other London children, Kaylee danced, rode, swam and trampolined her way through childhood. The difference was, her mum would cart an oxygen tank along. *"She did all the normal stuff kids do, we'd just do it slower or find a different way. We'd go to soft play [an indoor play centre] and I'd have to go climbing up into the play area with oxygen when she got stuck at the top and felt unwell."*

These days, Kaylee is careful to not push herself too hard. If she doesn't get enough oxygen she suffers debilitating headaches, and experiences numbness in her hands and legs. She walks – slowly – to the shops with her friends, watches fashion YouTubers and takes photos. She's studying photography at school and loves to take macro shots, like close ups of a leaf, detailing its veins. At school, she stands in the quiet dark room and develops prints, the old-school way.

Kaylee's had a bumpy ride through high school. Her peers quickly noticed the smaller-than-average girl couldn't carry her own bag, and left lessons five minutes early to avoid the crush of students hurrying between classes. Kaylee also spent her free time in "base", a common room for students with special educational needs, and had to leave school frequently for her regular medical appointments. She might as well have walked around with a sign over her head saying "I'm different", she says.

"Once [other students] know you're from base, and they saw I left early, got my bag carried, they formed their own opinions. I was just a different animal, and they never wanted to talk to me."

When they did talk to her, it was to call her "slow", or to make fun of her small stature. In the last couple of years students in the years below Kaylee at school have begun to comment, which stands out to the senior as particularly unfair.

She's armed herself with a few tactics to shut down questions and comments. ***"I'm very outspoken, I'm not afraid to say something. When people ask me why I had my bag carried I would say, 'because I'm special, unlike you'. Or I'd just tell them to go away."***

"I look scary on paper, but I'm fine in real life."

"I look scary on paper, but I'm fine in real life," she says, explaining her frustration at being left out of peer groups.

Now, she has a small but close group of friends. If she ever moves out of home, it'll be with her best friend, Chloe, she says. Chloe isn't from base, but like Kaylee she's on the periphery of school life, and the two became friends after Kaylee noticed her sitting alone on the edge of the schoolyard each day, watching the other students.

Kaylee and Chloe dream of living in a mansion with a white living room. Fantasy mansions don't come cheap, and Kaylee will need to find a job. As long as she can remember, she's wanted to be a nurse. But as she's grown older and her condition has come into sharper focus, the dream is starting to look unlikely.

"Nurses work long hours, they're on their feet all day," Kaye says. *"We've had to change things as we go along."*

Now, she's thinking about working in childcare. Teaching life skills to children with disabilities, or maybe caring for preschoolers. Her older sister has two small children, and Kaylee loves spending time with them.

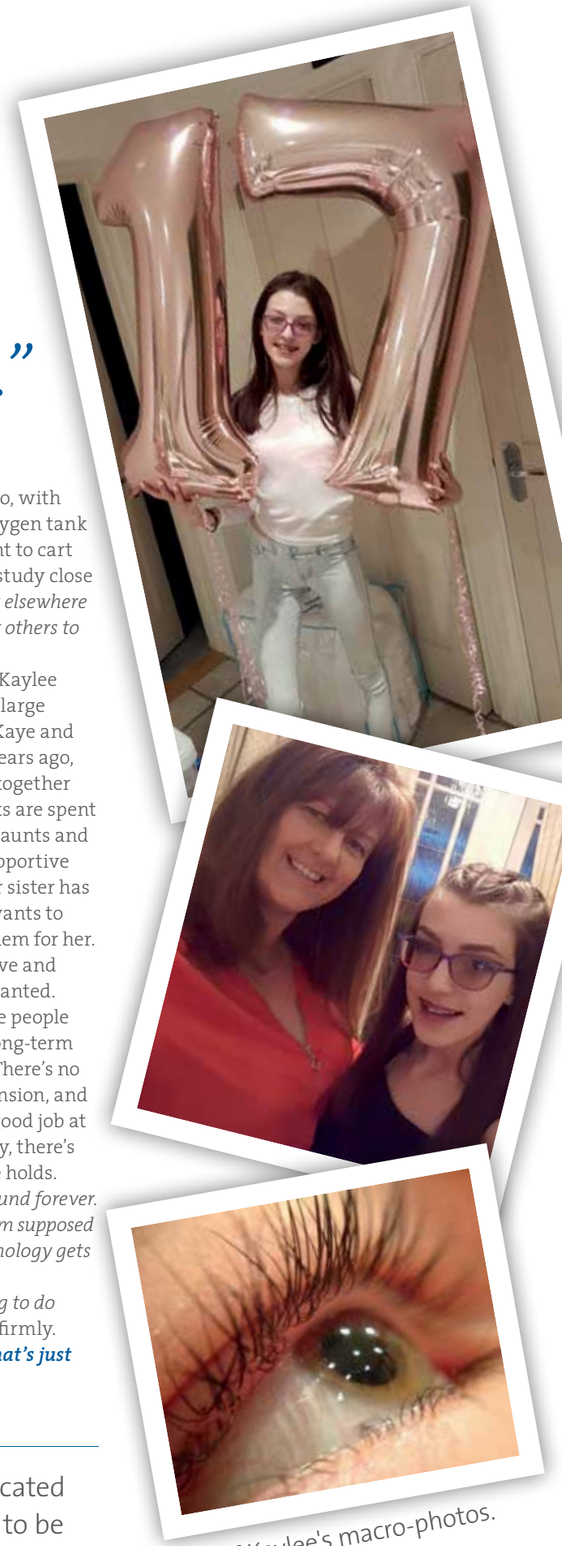
Whatever she chooses to do, with an array of medicines, an oxygen tank and other medical equipment to cart around, Kaylee will need to study close to home. *"It's too hard to stay elsewhere away overnight, too much for others to deal with,"* Kaylee says.

As well as her older sister, Kaylee has a younger brother and a large extended family. Although Kaye and her husband split up a few years ago, they're still friends, and eat together each week. Other weeknights are spent with cousins, grandparents, aunts and uncles. The family sound supportive and protective of Kaylee. Her sister has told her that if Kaylee ever wants to have children, she'll carry them for her.

Kaylee doesn't take this love and unconditional support for granted. Her greatest fear is losing the people she loves, she tells me. Her long-term prognosis is also a concern. There's no cure for pulmonary hypertension, and while medication's doing a good job at keeping her symptoms at bay, there's no knowing what the future holds.

"Mum's not going to be around forever. And, there's the thought that I'm supposed to outlive her. I hope the technology gets better and I can carry on."

"You will if I've got anything to do with it, babe," her mum says firmly. ***"We'll always have a plan, that's just what mums do."***



One of Kaylee's macro-photos.

Originally published by **Folks**, an online magazine dedicated to telling the stories of remarkable people who refuse to be defined by their health issues. Visit **www.folks.pillpack.com**

PH WEEK IS COMING!

PH AWARENESS WEEK 2018

Monday 22nd – Sunday 28th October
Join us for our third annual week dedicated to encouraging conversations about pulmonary hypertension.

Keep an eye out for ways to get involved!

www.phauk.org
@PULHAUK
@PHA_UK

To talk to us about PH Week, please email **office@phauk.org** or call **01709 761450**

Going to great lengths

For teenager *Paige Wilson*, a fun run or bake sale just wasn't going to 'cut' it when it came to raising money for the PHA UK. Instead, she decided to shave off her hair.

In February this year, at the age of 18, Paige raised £650 by being sponsored to have her locks lopped off. She donated the hair – all 12 inches of it – to the Little Princess Trust, a charity that provides wigs for children who have suffered hair loss through illness.

Paige, who lives in Kent, was diagnosed with idiopathic pulmonary hypertension at the age of ten and has spent her teenage years learning to live with the condition.

She now volunteers part-time at a charity shop and dedicates her time to helping others, which was her motivation for the shave.

> "It was a drastic change...It's been a lot to get used to!"

"There were about three months between making the decision and actually having it done, and there were times I started to doubt what I was doing," she said. *"But I just kept thinking of all the good that would have come of it and those thoughts got me through. It was a drastic change, as the shortest I'd ever gone before this was a long bob. It's been a lot to get used to!"*

Although she admits things got a bit cold over the winter months, Paige has decided to stick with the shave over the summer.

"Everyone was so supportive about what I was doing. My best friend told me he couldn't afford to sponsor me much, but he was there to help me through it - and that's all that mattered. It was amazing though to raise so much and it was a lot more than I expected."

Growing up with PH, Paige has tried various hobbies – including photography and music – but after a chance encounter with a homeless man in London, she now finds that helping others is what motivates her most in life.

"I was coming out of Great Ormond Street Hospital with my family and we had some leftover pizza we had all enjoyed for tea. Instead of taking it home, we gave it to a man lying on the pavement in the cold, and his gratitude brought something out in me that I hadn't felt before. It was then I decided I wanted to do more to help other people."

"Most of my life has revolved around PH, and it's been tough. But in some ways, I'm grateful for it because it's made me realise how lucky I am to have so much support. I could be in far worse situations, and I'm just really happy that I'm not. I've met a lot of people along the way, and I think a lot about a friend I lost to PH, Liam. He was a fighter, a soldier, and I want to be just like him." ●

Paige before and after her cut.

I feel like I've come such a long way

Ten years after her pulmonary endarterectomy operation, *Chitra Tripathi* won two awards in a Mrs India UK competition – providing a confidence boost and a platform to raise awareness of pulmonary hypertension. Here, Chitra tells Emphasis about living with CTEPH in both India and England, and how her care has changed her life.

Chitra is taking part in a 10km run in London in July to raise money for the PHA UK. You can support her at www.justgiving.com/fundraising/chitra-tripathi or follow her on Instagram at chitra.tripathi

My aim was to raise awareness of PH through the platform of Mrs India UK

Soon after my 23rd birthday and just three months before my first marriage anniversary, I found myself in an intensive care ward in India, being diagnosed with pulmonary hypertension. I was told it was a life-threatening disease which affects just one to three people in a million. 'Guess my chances', I thought.

What was more devastating was to be told its lifelong complications for a woman of my age, who had hardly experienced the essence of married life, or for that matter even dreamt of having kids. In one word, I was completely devastated.

It all started following a sudden episode of DVT (Deep Vein Thrombosis, clots in the big vessels) in my leg, which travelled to both of my lungs (a condition known as a pulmonary embolism).

At the time, I was working in my dream job at a multinational bank in India. The clots in my lungs changed me from a healthy, sporty, academically-excelling girl to a bed-bound, breathless person who struggled to speak a short sentence or even walk ten steps without taking frequent breaks.

After a series of tests and scans over the next three months, doctors finally concluded that I had developed pulmonary hypertension secondary to the clots in my lungs. Apparently, it was the delay in the diagnosis and treatment of the pulmonary embolism that led to treatment-resistant blood clots, which otherwise would have dissolved with blood thinning medications.

I was further devastated to know that the only treatment available in India was a double lung transplant, which had a five-year survival rate of just 40 to 50 per cent. I was also told that I would require blood thinning medication for the rest of my life.

Fortunately, my fate took a positive turn in 2007 when I moved to the UK with my husband, who had secured a job as a trainee doctor in the NHS. As my condition was deteriorating, my GP made an urgent referral to the local cardiology department, who then referred me to my nearest specialist centre.

I still remember sitting in Dr. Howard's outpatient clinic and praying to hear that there was a treatment for my condition

I was fortunate to see Dr Simon Gibbs and Dr Luke Howard, who are both pioneers of the field. Needless to say, they cared for me like my family, promptly carried out my tests and showed me a ray of hope. I still remember sitting in Dr. Howard's outpatient clinic and praying to hear that there was a treatment for my condition.

At last my prayers were heard, and I had a second chance when I was offered life saving surgery, a pulmonary endarterectomy, at Papworth Hospital.

Little did I know at that time that the girl who was sitting in the pulmonary hypertension outpatient unit, suffering with pain and depression, would be pleasantly introducing herself as a finalist in Mrs India UK 2018.

Yet there I was in March this year, Chitra Tripathi, a PHighter, fitness freak, HR professional and Mrs India UK 2018 finalist. It felt like I was in a dream world and none of it would have been possible without the care and support I received from my PH team. I feel like I've come such a long way.

The final of Mrs India UK was held in April and I was honoured and humbled to be awarded 'Mrs Congeniality' and 'Mrs Glowing Skin'.

My aim was to raise awareness of PH through the platform of Mrs India UK, so that more people could join hands in spreading awareness with the aim of investing in early identification and better treatments.

I think the words I used in my Instagram post to announce my awards sum up how I feel about the last few years:

Life is a journey and not a destination. I'm so thrilled and grateful for every learning, experience and lesson that I can take forward in life.

NHS
Royal Papworth Hospital
NHS Foundation Trust

New CTEPH procedure **now available on NHS**

Patients with Chronic Thromboembolic Pulmonary Hypertension (CTEPH) are now able to benefit from a pioneering procedure that uses tiny balloons to inflate and compress blockages in the blood vessels.

The procedure, called balloon pulmonary angioplasty (BPA), is being funded by the NHS and will be carried out at Cambridge's Royal Papworth Hospital.

The procedure sees cardiologists insert a very fine wire into blood vessels in the lungs and a tiny balloon – approximately 4mm in length – is then guided into position over the wire. The balloon is inflated for a few seconds, to around the size of a pea, which pushes the blockage aside and restores blood flow to the lung tissue. The balloon is then deflated and removed. This can be repeated several times in different parts of the lung during a single treatment session.

"It's a life-changing technique; I can do so much more than I could"

Dr Joanna Pepke-Zaba, Consultant Respiratory Physician at Royal Papworth Hospital, was part of an international delegation which travelled to Japan to research the procedure in 2014.

She said: ***"Because of the success of the pilot study launched at Royal Papworth Hospital three years ago we can provide balloon pulmonary angioplasty in the UK. It allows us to get to those hard-to-reach areas of the lung and offer CTEPH patients a better chance of survival and a much-improved quality of life."***

One patient to benefit from the treatment is Elizabeth Irons, 69, a retired teacher from Nottingham who was rushed to hospital gasping for air after blood clots formed in her lungs.

After being diagnosed with CTEPH, Elizabeth was told the location of the clots were too hard to reach using the only surgical treatment at the time. However, she was given fresh hope when doctors at Cambridge's Royal Papworth Hospital offered her the new treatment.

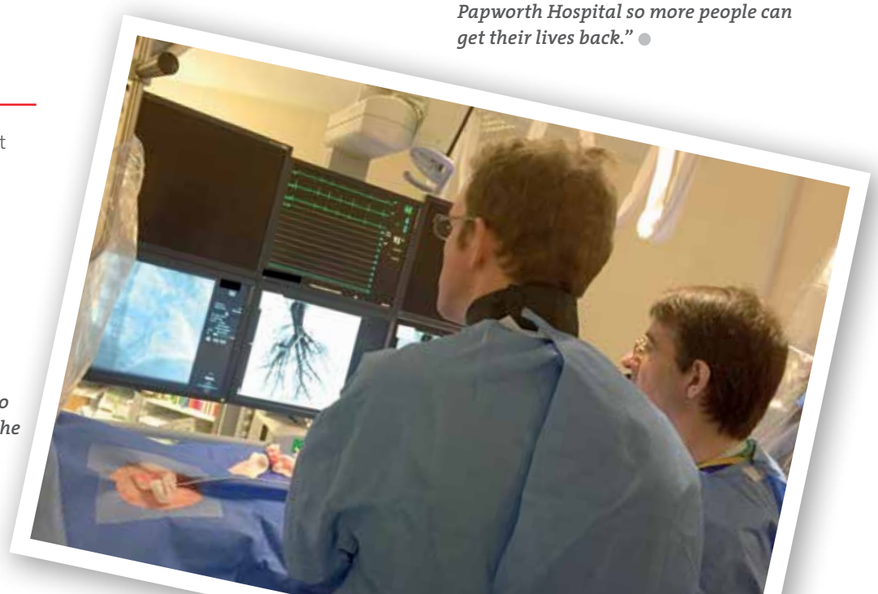
Now Elizabeth is looking forward to plenty more days playing with her young granddaughter.

"It's been an amazing treatment," she said. ***"I was awake through the whole thing – I needed to hold my breath for the clinicians at certain points – but I never felt worried because I had such trust in the medical team. "It's a life-changing technique; I can do so much more than I could – I've been able to go on holidays and fly around the world to see all my sisters. My quality of life has improved enormously.***

"I'm also able to be a lot more active with my toddler granddaughter. She was due to be born just as I fell ill – my biggest fear was that I wouldn't be around to see her grow up.

"It's wonderful news that NHS England is commissioning BPA at Royal Papworth Hospital so more people can get their lives back."

"It allows us to get to those hard-to-reach areas of the lung"



Heythrop Park Hotel
5-7th May 2018

Sun, sponges and a 'superb weekend'

The weather was firmly on our side for the first-ever PHact Finding and Fun Weekend for children with PH under the care of Great Ormond Street Children's Hospital (GOSH).

Held at the beautiful Heythrop Park Resort in Oxfordshire over the sunny early May Bank Holiday weekend, the event - which was funded by the PHA UK - saw families relax, learn more about PH, make friends and enjoy precious time away together.

Activities for children and teenagers included arts and crafts, street dance, African drumming, circus skills and mini golf. A programme of educational talks by experts from GOSH gave adults insight into research and advice on talking to siblings, and a Q&A panel session addressed a range of questions submitted by parents.

Before the Sunday night dinner and disco, children cooled off in the heat by throwing soaking sponges at Dr Shahin Moledina, consultant at GOSH, and Iain Armstrong, chair of the PHA UK. Parents donated £1 per sponge to the PHA UK and the pair took the battering in style.

Away from the activity programme, families were able to explore and enjoy the leisure facilities and beautiful grounds of Heythrop Park. And, of course, there were plenty of opportunities to snap selfies with cut-outs of The Green Leaf Crew.

Speaking at the event, Lorna Gravenstede, whose seven-year-old daughter Emily has PH, said: *"We really looked forward to this weekend. It's been so useful to meet other families, and other parents of children with PH. In the last six to 12 months Emily has become more aware that she has this condition, and has been really keen to meet other children with PH. She's met six or seven so far in the last day, and I'm sure we'll meet more."*

Kevin, whose ten-year-old daughter Holly has PH, added: *"It's been a superb weekend. It's given all the kids chance to meet and play with each other so they know they are not the only ones out there (with PH). As well as the food and the place itself, the highlight for me has definitely been meeting different people."*

"It's been so useful to meet other families, and other parents of children with PH."

Green Leaf Crew!

GREEN LEAF CREW Q&A
LEWIS & ETHAN HOLMES

This issue, brothers Lewis and Ethan Holmes from Newcastle answer our Q&A. Lewis is four years old and lives with his mum, dad and brother Ethan, who is eight. Lewis was diagnosed with PH when he was born and is cared for by Great Ormond Street Children's Hospital. Here, the boys share some of their favourite things.

Q. If you could be any super hero, who would it be?
A. *Lewis: Superman because he can fly.*
A. *Ethan: The Black Panther because claws can slice through metal.*

Q. What are your favourite things to do at weekends?
A. *Lewis: Soft play. Going to birthday parties. I like getting party bags.*
A. *Ethan: Trampolining. Going swimming at the wave pool. Going to Cub camp. Playing with Lewis.*

Q. What's your favourite food?
A. *Lewis: Cheese sandwiches and spaghetti Bolognese.*
A. *Ethan: Curry and cheese and onion crisps.*

Q. What has been the best holiday you've been on?
A. *Lewis: I like the seaside and building sandcastles.*
A. *Ethan: Hemsby because I liked the beach and the arcades.*

Q. Where would you like to go next?
A. *Ethan: Devon because there are loads of interesting things to see like the bee farm, the aquarium and lots of sandy beaches.*

Q. Do you have a favourite film?
A. *Lewis: Monsters University because it's funny.*
A. *Ethan: Jurassic Park/World because I love dinosaurs.*

Q. What's the best thing about the summer holidays?
A. *Ethan: Going on holiday and doing lots of activities.*

Q. If you could be anyone in the world for a day, who would it be and why?
A. *Lewis: Batman because he's got a black car that is fast.*
A. *Ethan: A paleontologist because I love learning about fossils and dinosaurs.*

Q. What would you like to be when you grow up?
A. *Lewis: A Bus Driver.*
A. *Ethan: A paleontologist.*

Patch

by David Banks

The GREEN LEAF CREW

BY DAVID BANKS

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.**

£337 RAISED

Dress Down Day for PHA UK

Richard Annett organised a Dress Down Day within his team at Lloyd's Banking Group to raise money for PHA UK. Richard said: "My wife has suffered with PH since 2009. It was discovered whilst pregnant with our daughter. The hardest thing for me personally with supporting her is the feeling of not being able to solve the problem for her, so raising funds is the next best thing I can do. I try to raise funds each year and will continue to do so through this year and many more to come."

Run to the sun

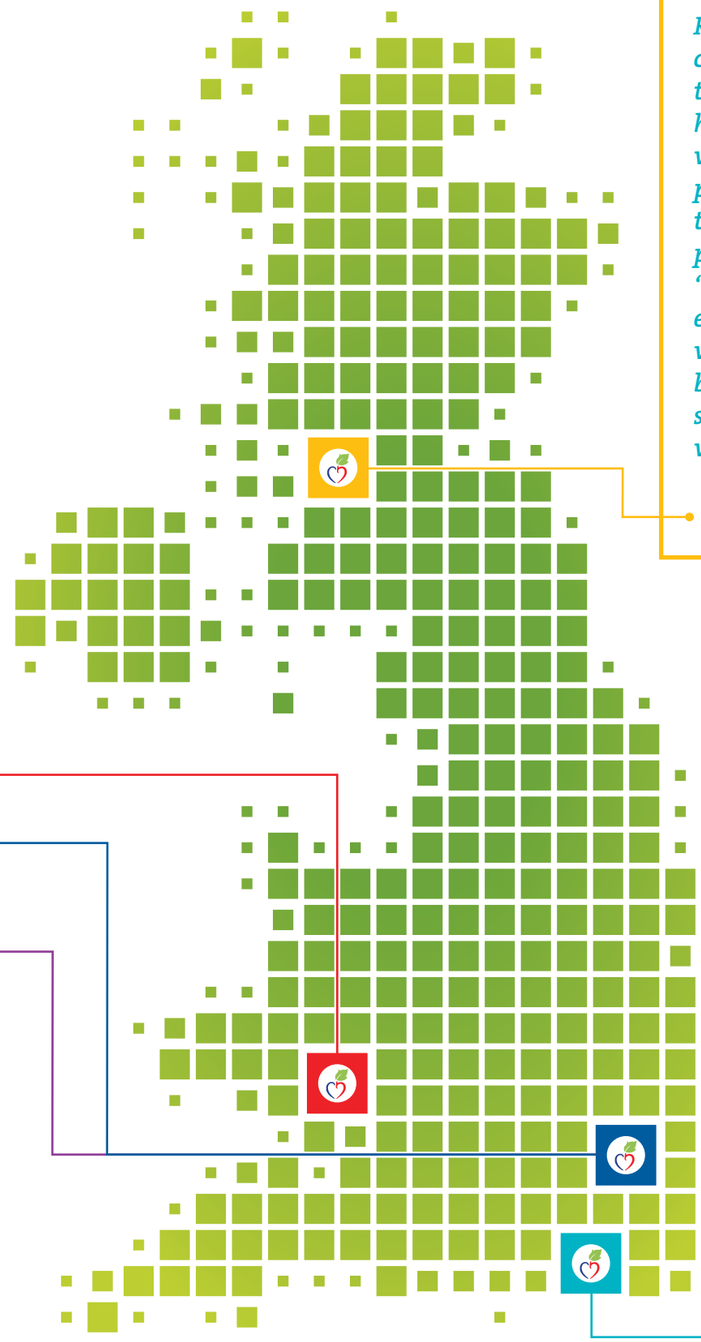
AssuredPartners London participated in the Chase the Sun run at the Queen Elizabeth Olympic Park to raise funds for the PHA UK. Lisa Wemborne of AssuredPartners said: "PHA UK is a charity which is personal to one of our colleagues and with the help of family, friends and colleagues, we were proud to be able to raise so much. We believe that fundraising to raise awareness and investment in research is very important and a personal aim for us."

£2,950 RAISED

Conquering the capital

Ellie Price completed the London Marathon for PHA UK in memory of Alexandra Beeston, who passed away in 2012. Ellie said: "The experience was tougher than I could have ever imagined. I trained really hard for months but the hot weather made conditions extremely tough for everyone on the day. The bigger achievement has to be my fundraising for PHA UK which both the family of Alex and I are so grateful for."

£3,490 RAISED



Sugar free fundraising

Robbie Dalziel, seven, gave up sweets, chocolate and fizzy drinks for a month to raise money for the PHA UK after his grandma Sandra was diagnosed with PH in 2015. He also made a presentation which he showed at school to explain what PH is and how it affects patients. Robbie's mum LeeAnn said: "I am immensely proud of Robbie. The excitement on his face as his donations were rising not only made me smile, but made me emotional to think I have such a loving, caring and wonderful little boy."

£375 RAISED

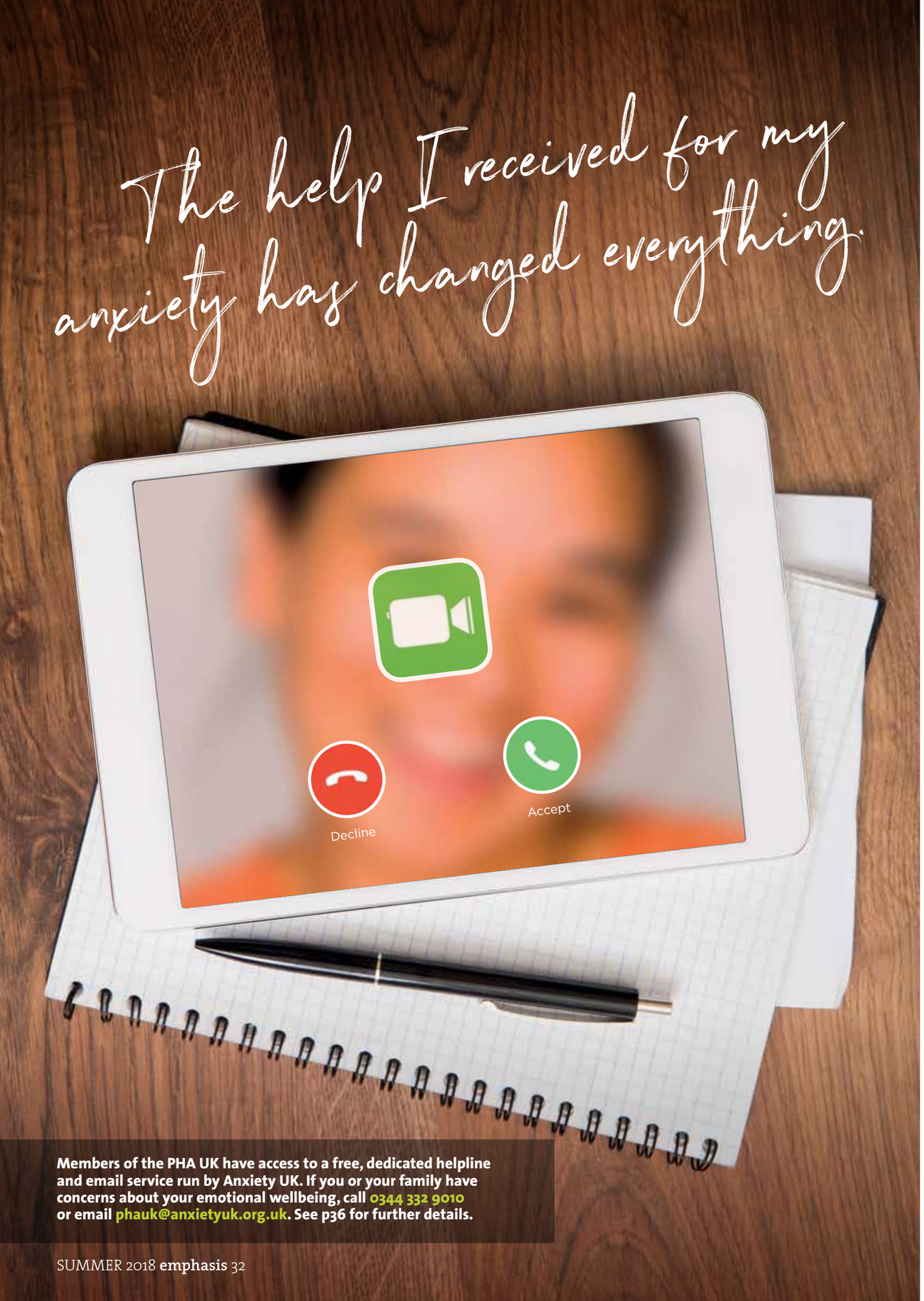
Taking the Lead for PH

David Leader from Worthing completed the Brighton Marathon to raise money for PHA UK in memory of his mother, who was diagnosed with PH in 2012 and died in 2016. David said: "There were so many people but it was a great experience at the same time. It had been so cold in training and snowing only days before so 16 degrees on the day was a huge shock to the system!"

£512 RAISED

If you have any ideas for raising money for PHA UK visit www.phauk.org. 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

The help I received for my anxiety has changed everything



Members of the PHA UK have access to a free, dedicated helpline and email service run by Anxiety UK. If you or your family have concerns about your emotional wellbeing, call 0344 332 9010 or email phauk@anxietyuk.org.uk. See p36 for further details.

Wendy Clemmet struggled to cope when she was diagnosed with PH, but after accessing help through Anxiety UK, the former childminder has learned to manage her mental health and live a happy life. She spoke to Mary Ferguson at her home in Barnsley, South Yorkshire.

Like many people with pulmonary hypertension, 47-year-old Wendy's journey to diagnosis involved increasing breathlessness and multiple tests.

Initially thinking she simply needed to increase her fitness, Wendy delayed visiting her GP – but when she started to experience palpitations, chest pains and dizziness too, she was referred to her local hospital. Here, an echo revealed the right side of her heart was enlarged, and she was sent for an MRI.

A long wait for results ensued, and when Wendy's chest sensations became increasingly alarming she went back to her GP, who referred her to her nearest specialist centre.

Here, a right heart catheter in 2017 confirmed Wendy had idiopathic pulmonary hypertension – three years after she first started experiencing symptoms.

As soon as PH was mentioned as a possibility, Wendy started googling.

"I found a lot of really harsh stuff online", she said, "and although I tried to only read reliable websites, all the information about quality of life and life expectancy really upset me and sent me into a bit of a decline. The bottom just fell out of my world."

As she waited for her diagnosis, Wendy's worries were affecting her physically as well as mentally.

"When I went for my first visit to the specialist centre it coincided with experiencing debilitating Irritable Bowel Syndrome (IBS) symptoms, which I think was down to the stress of it all. My whole body felt like it was fizzing. There was so much adrenaline. I found it really difficult to function."

One of the things making Wendy so anxious was the thought of having a right heart catheter and she wasn't the only one worrying about it. Her son Daniel overheard her talking on the phone about what the procedure involved and Wendy believes the stress of thinking about it, at only 11 years old, caused him to develop shingles.

"My husband couldn't get his head around it either, he can't even watch Casualty, so it was a very stressful time for all of us."

It was at that point that Wendy decided to travel with her family to the PHA UK Resource Centre in Sheffield and find out more about the condition. It was during this visit that Wendy found out about the charity's partnership with Anxiety UK, but she was initially reticent to get in touch.

"Because I was responding well to my PH medication, physically I felt like one of the lucky ones, and that made me feel guilty about using resources that other people more in need could access. But I decided to contact Anxiety UK anyway, and in hindsight I can now see how much I needed to."

After getting in touch with Anxiety UK through the dedicated PHA UK contact channels, Wendy was offered a choice of finding a therapist local to her that she could see face-to-face, or accessing therapy via Skype or Face Time. She chose Face Time sessions, and once a week for eight weeks, retired somewhere quiet with her iPad to talk to Tina, a mental health professional based in Bournemouth.

Wendy said: ***"I didn't feel like I was missing out by not seeing someone face-to-face. It took the stress out of having to drive somewhere and find a parking space, plus the time it would have taken to get there.***

"Tina taught me about relaxation techniques, breathing exercises and de-catastrophising. She introduced me to mindfulness, and a whole range of other techniques, without ever over-facing me. They were all introduced gradually, over the course of the sessions."

The therapy helped Wendy cope with her second right heart catheter, and she now feels prepared for the time when she will need another - whenever that may be.

"I kept reliving the experience of having the catheter, over and over again. But Tina taught me to imagine locking the memory in a leather-bound book, sealing it with a good memory, and shutting it all away in a room in my mind. It doesn't eat me up all the time now."

Wendy made notes after every therapy session, and now goes back to them when she feels she needs to. ***"It makes me feel a lot better knowing they are there, and it also helps me see how far I've come."***

Wendy, who had never accessed psychological therapy before, didn't expect it to help as much as it did. ***"I always thought I had a strong mind, so never needed that sort of thing. But you'd have to be a pretty tough nut to have all this happen to you and not be affected by it. The help I received from Anxiety UK has changed everything. I feel so much better about my situation now. It's made a massive, huge difference to me."***

Wendy is now using physical activity as a way of maintaining her mental health, and credits going for a walk every day with helping keep her on track. She's also started Pilates and is in the process of persuading her husband they should get a dog.

"I do think that learning to manage my anxiety has helped my PH too", she added. ***"I remember being told by a specialist nurse that one of the most important things you can do is to keep stress under control. For so many reasons, I'd encourage anyone struggling with anxiety to access the support that's there."*** ●

IN THE NEWS

RED LION QUEST FOR PH **RAISES £££s...**

Kayleigh and Paul Tyack from Oxfordshire have completed their quest to visit all 632 Red Lion pubs in Britain, raising thousands for the PHA UK. *Charlotte Goldthorpe* reports.

The pair began the pub crawl to end all pub crawls in September 2015 and two and a half years later they ticked the last one off their list in Boldre near Lymington, Hampshire, in February.

During the challenge they clocked up 20,500 miles and gained the attention of the national press, featuring in The Metro, The Daily Mail, The Sun and even Dutch paper De Telegraaf.

Having originally hoped to raise £650, the pair smashed their target, raising over £2,200.

Kayleigh said: *"The challenge was amazing and everyone we met was so lovely. Paul's seven-year-old niece was diagnosed with the disease at the age of just three. When we reached 200 Red Lions, someone said to us 'why didn't we do it for charity?', so we decided to raise money for PHA UK which is a charity close to our hearts.*

"The locals would give us a fiver and ask us about PH. It gave us the chance to help raise awareness of this rare disease which is little known."

On their visit to the Red Lion in Preston, a lady whose daughter lives with PH heard about what they were doing and got in touch to arrange to meet them at the pub. Kayleigh said: *"It was really heartwarming to talk to others affected by the disease and to feel part of the PH community."*

The couple travelled by plane as far as Inverness in Scotland, 500 miles away from their home and spent endless hours planning rail and road trips to reach the destinations. Not fazed by all the travelling, Kayleigh said: *"We enjoyed visiting different parts of the country, especially the Peak District where neither of us had been before. We'll definitely be going back."*

When some of the pubs learned about their mission they were extremely welcoming, including at The Red Lion in Merthyr Tydfil where they were treated to food and free drinks. But their favourite of the hundreds of pubs was in their home county of Oxfordshire.

The couple celebrated again when they got married in May, but this time they swapped pints for champagne. ●

Sun's out, **everyone's out!**

Some medicines can make the skin more sensitive to UV rays, meaning choosing the right sun protection this summer is even more important. Here, consultant pharmacist *Neil Hamilton* offer his advice.

If you are anything like me, you'll be relieved to see the back of our wet and snowy start to 2018. After heavy snow as late as April this year, I hope we can now enjoy a prolonged warm spring and summer. I love it when the sun comes out and cheers everyone up. It is great to have the longer evenings back; to enjoy in your garden, go walking with your dog or socialise with friends and family.

As much as we enjoy the sunshine and even getting a healthy tan, we all know sun burn is no fun at all. Now is a great time to think about sun protection. Not least because your skin will be extra sensitive when it is exposed for the first time to this year's warm sun. I'm sure we can all recall that time when we forgot to use sunscreen and regretted it! Adequate protection from harmful Ultraviolet (UV) rays is easy to come by with a seemingly endless range. We can choose from an array of sunscreen lotions, oils, sprays and gels, with some now proven to protect after just a once-daily application for extra convenience.

Protection from the sun is crucial for us all to ensure we can enjoy our summer. However, it is even more vital to people prescribed a certain group of medicines. These medicines may be either oral or topical but have the effect of making the skin even more sensitive to UV. The precise incidence of medicine-induced photo-sensitivity is unknown, but men are more likely to have reactions than women.

In most cases, there are no ongoing effects once the causing agent – either the medicine or the sunshine - is removed, but complete resolution may take weeks to months depending on the severity. Voriconazole (anti-fungal, listed right) is directly associated with a risk of skin cancer. However, recurrent over-exposure to UV would increase anyone's risk of developing skin cancer in any event.

The list in the box on the right shows the most commonly prescribed photo-sensitising medicines. This list is not exhaustive, because rather like any side effect, there are one-off reports from individuals who have experienced problems with other medicines.

As a result, if you are prescribed one of these medicines, I advise you to take extra precautions – solar-protective clothing and eyewear are strongly advised. If avoidance of direct sunlight altogether is not an option, it is worth investing in very high factor sunscreen (SPF 50 or even a total sunblock). SPF stands for Sun Protection Factor. This is an indication of the level of protection against sunburn, primarily caused by UV-B. However, most medicine-induced reactions are caused by UV-A. Hence purchasing sunscreen that protects against UV-A should be used if you are prescribed any of the above list.

In the event that adequate solar protection is not enough, and you unfortunately suffer some sunburn, liberal application of emollients and cooling lotions may soothe and moisturise the affected areas. Anti-histamine tablets may reduce some of the redness, itch and irritation of the burnt areas. In severe cases, topical steroids such as hydrocortisone cream may help, but they should be confined to small areas only unless under medical advice.

Your pharmacist will be happy to advise if you have specific queries about your particular medicines, whether I have mentioned them here or not. Being prepared and taking care is, as always, part of effective holiday and summer planning. More than anything I hope that you all have a great time this summer wherever you go – whether it's a 'staycation' or a holiday abroad.

Antibiotics
Tetracyclines
(e.g. doxycycline, tetracycline)
Fluoroquinolones
(e.g. ciprofloxacin, ofloxacin, levofloxacin)
Azithromycin

Anti-fungals
Itraconazole, Voraconazole, Griseofulvin

Non-Steroidal Anti-Inflammatory (NSAIDs)
Ibuprofen, Naproxen, Ketoprofen, Celecoxib

Diuretics
Furosemide, Hydrochlorthiazide

Retinoids
Isotretinoin, Acetretin

Statins
Atorvastatin, Fluvastatin, Lovastatin, pravastatin, simvastatin

Tricyclic antidepressants
amitryptiline, doxepin, nortriptyline

SSRI Anti-depressants
duloxetine, fluoxetine, fluvoxamine, paroxetine

Chemotherapy
Bortezomib, dasatinib, hydroxycarbamide, imatinib, pentostatin, methotrexate

Other medicines
(various classes)
Amiodarone, diltiazem, Quinidine, Hydroxochloroquine, coal tar, promethazine, prochlorperazine, tacrolimus, valaciclovir.

the*interview*

Clinical Lead for PH at the Royal Brompton Hospital, London

DR JOHN WORT

in conversation with

Deborah Wain

John, who hails from Wiltshire, first studied for a chemistry degree at the University of Oxford, where he decided he wanted to be a doctor and not a chemist. After graduating he moved to London and studied medicine at University College. He took time out from clinical medicine to study for a PhD at Imperial College London; this was the starting point for his career in PH. The work involved understanding why pulmonary blood vessels became thicker in PH and, in particular, found an important role for endothelin-1 – a molecule that causes blood vessels to narrow.

After qualifying in general, respiratory and intensive care medicines, John took up a consultant post at the Royal Brompton in 2006 and became Clinical Lead for PH three years later. John's particular academic interests include: the role of 'inflammation' in the development of PH; the reasons why patients with lung disease develop PH; and, more recently, the importance of skeletal muscle weakness in PH. He relaxes by spending time with his family and dogs, keeping fit, and painting.

Q. Growing up, what did you envisage your career would be?

A. *My mum was convinced that I should be a postman. I had a holiday job as a Christmas postman, which gave me my first pay packet. She liked the idea of that; no more pocket money! I can remember wanting to be a marine biologist and astrophysicist although I am sure that I didn't know what they entailed. I went to university to study chemistry because I was good at it and it got me into Oxford. The school liked that.*

Q. What was it that prompted the realisation you wanted to pursue medicine?

A. *Whilst studying chemistry I think I just grew up and started thinking about what I wanted to do. I had gone to university very young and didn't really have an idea of what I wanted to do in the future. I think it was a combination of becoming more interested in biology and a desire to help people that enticed me to medicine. My girlfriend at the time (now my wife) was studying medicine and it looked a lot more interesting than chemistry. I volunteered with Nightline, a Samaritans-based charity, which provided support for fellow students in need. I enjoyed the empathy in that and the feeling that I could help, even just by listening.*

Q. How did you find your way to specialising in PH?

A. *Well, I fell into it really. I had decided to do a PhD with Professor Tim Evans at the intensive care unit at the Royal Brompton. He was integral in setting up the designated PH services in the UK. Initially I thought the PhD would be on a condition called acute respiratory distress syndrome but he said "the future is in the pulmonary blood vessels" and he was right. The rest is history.*

Q. How does your research inform your clinical medicine work?

A. *That's a good question! I think the research we are involved in is very broad, from basic research on specific molecules to large, international studies testing new drugs or old drugs in different ways. So, I am hoping that, one day, the work on new molecules will lead to a new treatment that will make a real difference to patients with PH. The clinical studies help us decide how to use drugs in the best way; for instance we have learned that it is best to treat earlier and to use drugs in combination. And, of course, whether new drugs will be effective...*

Q. Please tell us more about one of those important molecules - endothelin-1; the focus of your earlier work. What's the significance of research around it?

A. *It is a very small molecule that causes blood vessels to narrow, both by constriction and by increasing the number of cells in the vessel wall. It is related to a protein found in snake venom. I showed that the smooth muscle cells, forming the majority of the blood vessel wall, produce endothelin and that this causes these cells to increase in number. This explains to some extent why PH develops. Obviously, other people were working on this and soon after drugs that block the effect of...* >>>

the*interview*

Dr **JOHN WORT**

"People have inspired me all through my life: from my parents, my teachers, my tutors, the team I work with, to the patients we treat."

endothelin (endothelin receptor antagonists, such as bosentan) were shown to be effective in patients with PH.

Q. What major developments have you seen in the department since 2009?

A. *The team started very small with Carl Harries, clinical nurse specialist, Professor Michael Gatzoulis and myself. We now have five consultants, three clinical nurse specialists, three research coordinators, a service manager, secretary, data entry manager, as well as a host of other staff! So growth has been the most obvious thing that I have noticed. But also the number of patients we look after and the complexity of the treatments we use. I think the National Audit for PH, led by Simon Gibbs, has been a big success and the constant desire to keep up with the nationally-agreed standards has improved the quality of care that we deliver.*

Q. What might a day look like for you now?

A. *What I like about my job is the variety. Each day and week is different. I don't think I have a normal day. But as everything is so busy nowadays, I need to juggle all the things I do. I suppose most of time is either direct care of patients or management of the ever-growing service. However, I try and fit in as much research as possible. Days are generally long but rewarding. And they always start and finish with a cycle ride, which keeps me sane!*

Q. What kind of a team do you have?

A. *The best! They are an utter pleasure to work with and keep me going when things seem too tough. They work incredibly hard, are generous and funny. When you look for a job, people always say the most important thing is the people you work with. That is so true. I am very lucky.*

Q. How is your current research breaking new ground?

A. *We are fortunate to be involved in the national cohort study for idiopathic and heritable PAH. By collecting blood samples from all patients with this condition and extracting DNA, the team, led by Professor Nick Morrell at Cambridge, have discovered several new genes associated with PAH. This is important, as understanding how the abnormal genes produce abnormal proteins, and how these abnormal proteins act, will allow us to create new drugs. In fact, it opens up the door for personalised medicine. I really think this will come in the not too distant future. Closer to home I would like to think that we have produced a lot of research that has helped in the diagnosis and management of patients with lung disease and PH and also in the management of PH associated with congenital heart disease. We have some exciting lab-based work going on at the moment looking at how inflammation and abnormalities in iron handling can cause thickening of pulmonary blood vessels. I really hope that this will lead to trials of novel drugs in patients soon.*

Q. How much optimism should there be about the potential for advances in treatment for PH?

A. *Well, I have mentioned it before; I think the advances in understanding exact genetic defects underlying PAH and the specific proteins circulating in the blood will allow us to personalise medicine in quite a few patients. This is very exciting. Most of the treatments so far have been 'pulmonary vasodilators' but they only treat a part of the narrowing we see in the tiny pulmonary blood vessels. We need drugs to reverse the thickening caused by proliferation of cells in the vessel wall. Imatanib was a drug that acted like this but had too many side effects. Similar drugs are being developed which will be safer to take hopefully. In fact, the UK has just finished a novel trial - led by Dr Mark Toshner; Transform UK - that looked at antibody treatment (tocilizumab) to treat PAH. The results should be published soon. So I would say there are grounds for a lot of optimism.*

Q. And finally, who or what inspires you?

A. *There is no single person or thing. People have inspired me all through my life: from my parents, my teachers, my tutors, the team I work with, to the patients we treat. If I had to pick a few individuals I would have to mention Professor Tim Evans, who introduced me to PH and taught me how not to give in until you have the correct diagnosis; Dr Hazel Rossotti, who saw the potential in me when I first went to university and taught me to always question and never rest until you understand; and finally, my lovely family, of course!* ●

The family of a young man who passed away earlier this year have honoured his memory with a unique online challenge designed to raise awareness of pulmonary hypertension.

Will Acres died in February at the age of just 22, after being diagnosed with PH in the summer of 2016.

Inspired by a video of her brother using his forehead to crack an egg taped to his wardrobe door, his sister Annabelle, 21, encouraged people to film their own 'egg smashes' and post the video on Facebook, along with information about PH.

Participants were also encouraged to nominate someone else to take on the challenge and make a text donation to the PHA UK when they uploaded their video, along with the hashtag #PHAEggHeadChallenge.

Annabelle said: ***"I've been quite overwhelmed by the response to the Facebook challenge and I think Will would have found it all hilarious. He was fun, loving, and kind-hearted, with an amazing determination to make the most out of life, especially after his diagnosis.***

"Will's courage became very apparent as he became more ill and Mum, Dad and I did all that we could to make him realise how proud we were of him. We were also blown away by the respect that so many of Will's friends had for him, as the response to the Facebook challenge shows."

At the time of going to print, hundreds of videos have been shared and over £1500 raised through text donations.

The challenge also caught the attention of the media, and Annabelle appeared in newspapers and on various radio stations, giving her a valuable platform to raise awareness of PH and remember her brother.

PHA UK chair, Iain Armstrong, also joined Annabelle in a special ITV news programme which was aired in the south of England.

The 'egghead challenge' is just one of many fundraising initiatives organised by friends and family in Will's memory. A t-shirt sale, summer ball, half marathons, an event at his university - and more - are all helping people understand PH and encourage earlier diagnosis. Will was a successful musician and DJ and a group of friends are also set to release a single that he wrote before he passed away.

Annabelle added: ***"I think my family are still in disbelief that Will has passed, but we find comfort in knowing that in his short life, he became such an inspiration to so many people.*** *PH is such a complex condition and there are many causes and symptoms, which are often mistaken for more common conditions like asthma.*

"Will spent a lot of time thinking that some of his breathing problems were asthma-related but if he had known about his PH earlier, his heart would perhaps not have gotten to the point of irreversible damage as quickly as it did."

Search #PHAEggHeadChallenge on Facebook to see some of the videos made in Will's memory. ●

Studying with PH

What help is out there?

For anyone, applying to study a university degree is a big decision. Moving away from home for the first time, having to live independently, and keeping control of your finances are all challenges. Having a long-term health condition like PH is another huge factor to consider, but there is help available, as *Chris Coates* reports.

Government funding

If you live in the UK, you can apply for the Disabled Student Allowance (DSA). This is for students who have a health condition or disability which affects their ability to study. DSAs are allocated as grants, so they do not need to be repaid, and can be claimed in addition to any other student finance, such as a maintenance loan. The amount you receive depends on your individual needs and income, so if you want to get a part-time job or keep your current one as you study, this may affect how much you're entitled to. Undergraduates can claim up to £25,677 per year, while postgraduates are entitled to £10,993. These figures are the maximum amount - most students will receive less.

To get the funding, you have to submit proof of your health condition, and once approved you will be asked to attend a local centre for a needs assessment. This is your opportunity to inform your funding provider about how your condition affects your studying. After your assessment, you'll be sent a report which shows the equipment and support you need, and the amount of funding you'll be granted.

The Open University

The Open University (OU) provides the opportunity for students to study from home, full-time or part-time, with moral support throughout the course and extra financial help when necessary.

Lynda Culley, advisor for the Disability Support Team at the OU, told Emphasis: "We encourage any student with a long-term health condition such as PH to make an application for a Disabled Students Allowance as soon as they are reserved on a module. However, we also have a Study Support Fund which can help to meet some of a student's study-related costs. This is means tested and based on household income but covers costs like books, childcare and consumables such as printer ink and paper.

"We offer to write a personalised 'Disability Profile' which outlines the main impact of a student's health condition on their study and makes anticipatory adjustments. This confidential profile would be used by tutors and others involved in study support.

"We can also fund non-medical helpers where they are not covered by the Disabled Student's Allowance. For someone with PH, this might be a practical support assistant to help at home, in the library, or in exams and tutorials. They might act as a companion, note-taker or even carry equipment."

Union support

The National Union of Students(NUS) has been running a campaign for the last four years to ensure that disabled students are represented and supported across all universities. In 2014, when the Government announced proposed cuts to the Disabled Students Allowance, the NUS campaigned fiercely and the idea was eventually scrapped.

Rachel O'Brien, Disability Support Officer at the NUS, told Emphasis: "People are shocked when I tell them that 92 per cent of disabled people don't use a wheelchair. Far too often, disabled people are seen to be inaccessible or irrelevant and we want to make sure that universities do not restrict their chance to study. It's also vital that decisions about support for disabled people are led by disabled people.

"The Disabled Students' Allowance is an incredibly important means of support for so many students, but it is only a small part of the help they need. At the NUS, we have done great work in the past four years to ensure that disabled student support is not reduced and we have lobbied universities to review and improve their welfare support services for disabled people."

Non-financial support

Every university in the UK is required to have a specialist team for students with health conditions and disabilities. When you enrol on a course, you will be contacted by the Disability Liaison Officer (DLA) from your academic department to discuss how they can support you throughout your studies.

Lynda Culley from the Open University told us: "Most qualifications can be studied over several years and therefore 'part-time'. If unexpected circumstances occur, students can return to a module at a later date, or apply to postpone an examination. We also offer a variety of formats for module materials to suit study at home, such as PDFs, e-books to download onto portable devices, audio files, transcripts and comb-bound books for ease of handling."

PHA UK member Shani Fernando has spent the last six years studying aerospace engineering at a university in London. ***"The financial support I received was brilliant,"*** she said. ***"Before I could drive, I had to get a taxi to lectures because using public transport was too exhausting for me. That was all paid for by student finance without any issues."*** Although she was pleased with the financial support she received, Shani thinks that universities could do more to help students with long-term health conditions like PH. She added: ***"Aside from financial support, my university wasn't great. There were delays with mitigation when I asked for extra time with assessments. It was my lecturers that were really understanding of my condition. It would be great if universities could improve their support management because the system is not that good for people who are going through the same thing as me."***

Meet the new PH professionals

Nikki, Pam, Jennifer, Mia, Nadine and Sian tell us about their new roles within the specialist centres across the UK.

Before becoming a nurse, I was an outdoor education instructor. And despite living in the UK I still play Australian football (AFL) on the weekends for fun. Also, in my eyes there is nothing better than ice cream.

"I was working in the Cardiac Intensive Care unit at GOSH before taking on this role and prior to this I trained and worked in Sydney, Australia. As a nurse specialist I've become part of the expert team in assessing and supporting families with their new or existing diagnosis.

I'm really enjoying having the opportunity to get to know all of the wonderful children and their families under GOSH. I am also looking forward to attending outreach clinics at our eight locations around the UK and Ireland and supporting families within their own community. I am really enjoying learning more about PH from our specialist team at GOSH."

I believe laughter is a good tonic of life and not only inflates one's lungs but neutralises the atmosphere of all horrible issues around our world today. I am always smiling!

"I have worked here in Sheffield for just over 12 years as a (general) PH nurse, but my new role provides more of a link between the patients and the doctors.

Our team is instrumental in supporting our patient group by organising fit to fly letters, oxygen therapy and generally assessing patients remotely with the aid of their local GPs, so they don't have to travel many miles to come and see us.

This role has opened up even more opportunities to work closely with families. I am enjoying the fact that I am able to be that direct link for them, in times of both happiness and distress, and I'm looking forward to being that pillar of strength they need."

I went to Bali in 2016 and climbed the Mount Batur volcano. I really wanted to watch the sun rise there and although it was a hard climb, it was worth it.

"I've been in nursing for nine years and was previously a senior nurse on an intensive care unit. My new role includes monitoring and assessing patients, taking them through tests, initiating treatment plans and educating them about their therapy. I'm really enjoying the patient contact and building up a rapport, speaking to patients on a regular basis, and being with them along their journey. What's really struck me since I've been in this role is just how rare PH is, and how much it impacts on people's lives."

I have been playing drums in a heavy metal band for over ten years. It's a great outlet for stress and lets me be creative (in a 'hitting things' sort of way!)

"Prior to accepting this secondment role, I worked in the aseptic preparation services department of pharmacy for a year after qualifying as a pharmacy technician. Overall, my new role is to provide support to the pulmonary vascular disease team, tackling mostly medication-related issues. Currently, I most enjoy interacting with patients, both face-to-face and over the phone, to help them solve medication queries and provide support. I am looking forward to being able to answer more complex queries when I am more experienced and have more confidence – although this role has massively helped me improve my professional confidence and my patient skills."

I grew up on a remote (and beautiful!) island in the Outer Hebrides called the Isle of Berneray

"I have about ten years' experience working in various hospitals in Glasgow and my most recent role was in the respiratory unit in the Queen Elizabeth University Hospital.

My new role involves prescribing the pulmonary hypertension medicines and ensuring their supply to all our patients across the whole of Scotland. I ensure their safe and effective use and advise on the best choice of medications and doses. I also educate patients so that they have greater knowledge about why they are taking these drugs and the best way to take them. I am trying to have a greater presence outside of the hospital so that I can meet all our patients and am looking forward to fully establishing a pharmacy service to the clinics."

My family are Welsh. I'm incredibly proud of my heritage and can speak a little of the language!

"For the last 23 years I have been working in palliative care in the local hospice and most recently at the Royal Hallamshire and Weston Park Hospital. My new role involves communicating daily with patients, offering support, education and advice, which includes training for new equipment and therapies.

I also liaise with families and all the members of the pulmonary hypertension team in Sheffield and further afield. I'm definitely enjoying the continuity of patients and having been in post for eight months I am slowly getting to meet people face to face that I have spoken to on the helpline."

Full steam *ahead*

If having pulmonary hypertension makes travelling by train difficult, you could be entitled to discounted tickets. *Deborah Wain* finds out more about the ***Disabled Persons Railcard***.

The rail network in Britain is among the oldest in the world and stretches more than 10,000 miles. In 2016 there were 1.7 billion passenger journeys, but travelling by train in Britain is more expensive than in most of Europe. In January fares increased on average by 3.4 per cent – the biggest increase in five years.

Government figures show that the Disabled Persons Railcard was issued to more than 207,000 people in 2016-17, a rise of 7.7 per cent, giving passengers a third off their fares.

Among those who qualify are people who:

- *Receive Personal Independence Payments (PIP) at any level for either the mobility or daily living components*
- *Receive Disability Living Allowance (DLA) at either the higher or lower rate for the mobility component, or the higher or middle rate for the care component*
- *Receive Attendance Allowance or Severe Disablement Allowance*
- *Buy or lease a vehicle through the Motability scheme*

A one-year Disabled Persons Railcard costs £20, or you can get a card that lasts for three years for £54.

What are the benefits?

You get a third off adult rail fares for travel on the National Rail network in Great Britain. There are no time restrictions on the Disabled Persons Railcard. You can use it to get a discount on Anytime, Off-Peak and Advance fares, both standard and first-class. The average annual saving has been calculated to be more than £123.

If you're travelling with another adult friend or family member they will also get a third off their rail fare.

The discount also applies to other services in London, cross country sleeper services and some rail/sea journeys. See the website **disabledpersons-railcard.co.uk** for full details as there are some restrictions.

How do you apply?

You can buy a railcard online at **disabledpersons-railcard.co.uk** or by downloading a form and then submitting it by post. You can't buy a card at a station. You will need to provide proof of eligibility, like a copy of your award letter.

How long does it take for the card to arrive?

The scheme recommends allowing five working days for an online application and 10 working days for a postal application. You must remember to take your railcard with you when you travel.

Are there other railcards?

Those who don't qualify for a Disabled Persons Railcard may qualify for another kind of Railcard. These are:

- ***Senior Railcard for anyone aged 60 or over***
- ***16-25 Railcard for anyone aged 16 to 25 or in full-time study***
- ***Family & Friends Railcard for anyone travelling with one child or more***
- ***Two Together Railcard for two named adults travelling together***

People who stay in their own wheelchair for a journey but don't have a Disabled Persons Railcard will qualify for a concessionary fare.

Is there other help available while travelling by train?

The free Passenger Assist service provides support such as help getting on and off the train or with luggage. It is available by booking ahead. It can also be accessed as you go, where reasonably practicable.

For more information about the Disabled Persons Railcard email disability@raildeliverygroup.com, call 0345 605 0525 (minicom/ textphone 0345 601 0132) or write to Disabled Persons Railcard Office, PO Box 6613, Arbroath, DD11 9AN.

IMPHACT study shows *true symptom burden of PH*

The results of a major study into how PH affects quality of life show that patients experience 'high levels of symptom severity' that don't diminish over time. *Mary Ferguson* reports.

The IMPHACT study, the first ever in-depth research into the long-term impact of living with PH, was conducted by the University of Manchester in partnership with the PHA UK.

A total of 126 patients from across the UK's specialist centres completed the 18-month study, which concludes that although many people with PH are living for longer, they are doing so **'under the weight of a considerable burden of symptoms that negatively affect their quality of life'.**

Participants completed questionnaire packs at the start of the study, and after six, 12 and 18 months. Questions were based around breathing, fatigue, anxiety, and depression, and the emPHasis-10 quality of life tool, developed in association with the PHA UK, was also used as a measure during clinical appointments.

The results showed that patients had a 'significant' impairment of quality of life at the start of the study, which didn't change over the course of the 18 months. The use of oxygen therapy, in particular, was associated with a significant impact on quality of life.

The results of the study were published through the British Medical Journal's Open Respiratory Research resource in March this year.

Professor Janelle Yorke, who headed up the study at the University of Manchester, said: *"Despite improvements in survival of patients with PH, this study has illustrated that symptom severity is high, and although there were improvements in anxiety,* ***the negative impact on health-related quality of life was unchanged over time.*** *This highlights the need for further study of factors influencing, and additional interventions to manage, the impact of this chronic condition on health-related quality of life.*

"Recognising the importance of these symptoms and the need for their regular assessment may help us to offer specific interventions to patients to help improve or maintain health-related quality of life in the long-term."

Iain Armstrong, Chair of the PHA UK, added: *"This is a really significant piece of research that shows just how much PH impacts upon quality of life.* ***The results provide another vital piece of evidence to help influence important decisions about funding, treatments, and service provision.*** *We're very grateful to everyone who took part in the study."*

The results of the IMPHACT study have been published in the British Medical Journal (BMJ)'s Open Journal, to enable them to be accessed by all. ●

As well as being shared widely via medical journals and global healthcare conferences, the results of the IMPHACT study will be used to further the work of PHocus2021, the advocacy group set up by the PHA UK to campaign for changes to public policy. To find out more about its aims, which include equity of access to treatment, reducing time to diagnosis, improving quality of life and reducing financial hardship, visit **www.phocus2021.org**

ME & MY JOB

Welcome to our regular column where PH professionals tell us more about themselves and their work. This issue, meet **Sheila Forshaw**, assistant service manager at the Sheffield Pulmonary Vascular Disease Unit.

HOW LONG HAVE YOU WORKED WITH PH PATIENTS? *I have worked in the Sheffield PH service for the last ten years. I initially applied to work here on the UK Audit for Pulmonary Hypertension* but the role then gained momentum and has been extended to include managing the administration and secretarial team. This is the team that ensures appointments are made and sent out, as well as the letters and reports from clinics and investigations to GPs and other appropriate medical teams.*

WHAT DOES YOUR JOB INVOLVE? *I work closely with the wider PH team to make sure we see patients in a timely manner and ensure their treatment pathway is as seamless as possible. This includes scheduling clinical activity, lots of administration, and liaison with other medical departments. My job also involves inputting and analysing our data to ensure we meet all the national standards required of a PH centre. This also includes working closely with our PH shared care centres.*

WHAT IS THE BEST THING ABOUT YOUR JOB? *Working as part of a small, very friendly and dedicated team. I really enjoy co-ordinating and bringing together all the elements of the service.*

WHAT INSPIRES YOU IN YOUR WORK? *Making sure we offer the best service with the resources that we have. Every day brings a new challenge and I enjoy solving these challenges.*

WHAT'S ON YOUR DESK? *At the moment, lots of data sheets as we near the deadline for submission for the National Audit of PH.*

TEA OR COFFEE? *Definitely coffee, and lots of it!*

WHAT DO YOU LIKE TO DO WHEN YOU'RE NOT AT WORK? *I've got lots to fit in outside of work! I enjoy the company of my family, especially spoiling my grandchildren, dog walking, gardening, and catching up with close friends.*

Our report into the latest National Audit of Pulmonary Hypertension can be accessed via* ***www.phauk.org*

If you work with people with PH and would like to answer our Q&A please e-mail **editor@phauk.org**

We know it's still summer...

...but if you want to get organised early for Christmas, look out for details of our new festive greetings cards in the next issue of Emphasis.

In your Autumn issue of Emphasis...

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

My journey with PH
Maria Fernandes shares her story

Keeping active
a closer look at vibration plates and yoga

The financial impact of PH
the results of our latest research

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

You can get involved in Emphasis too:

Emphasis Reviews - read a book, used an app or seen a film you think other readers may be interested in? Tell us about it!
Family Matters - let us know if you'd like to tell us about your family's PH experience in our regular feature.
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 .

We always love to hear from you – contact editor@phauk.org with any feedback or ideas.

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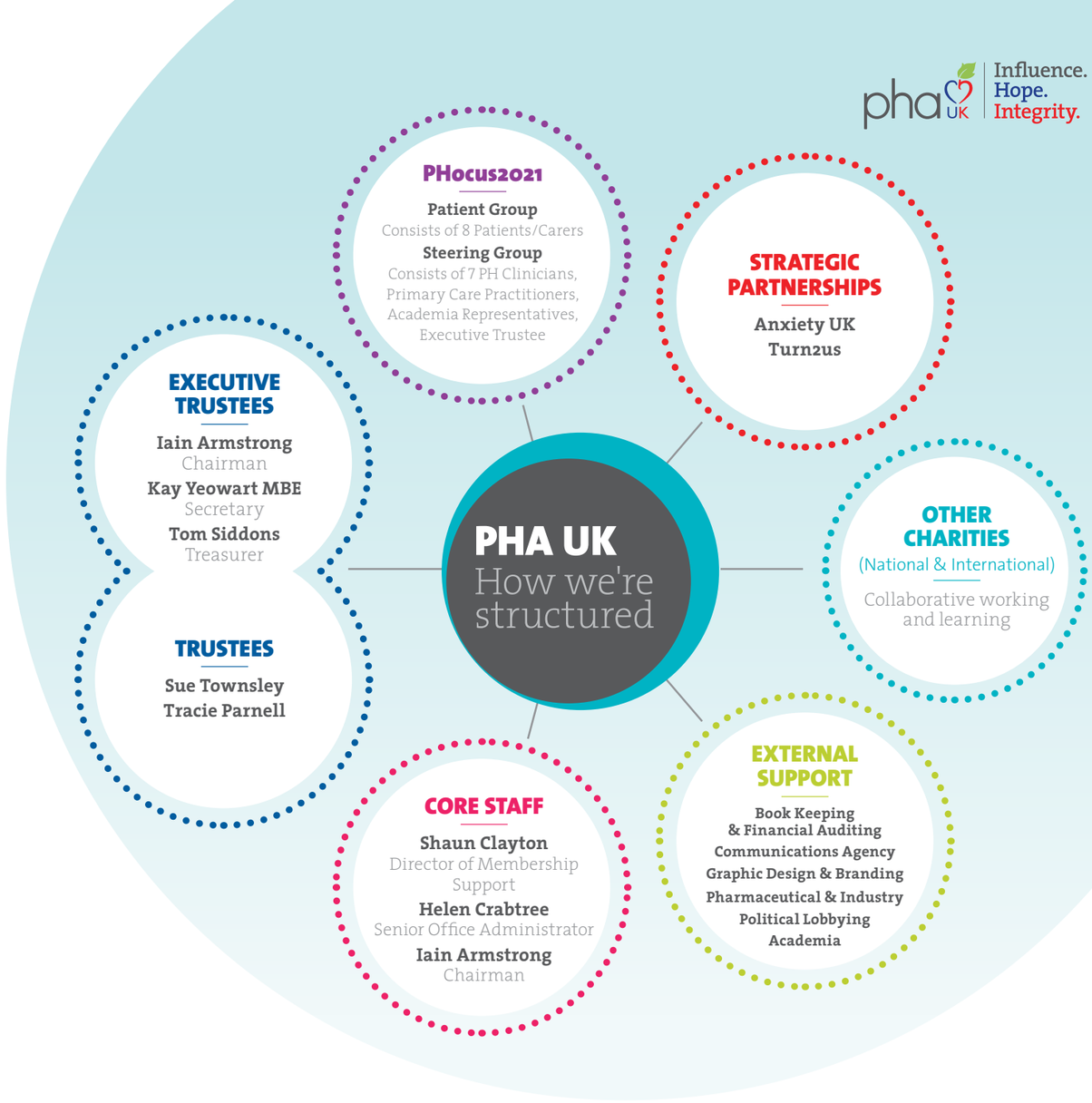

PHA UK Contact Details

Office hours: 9am to 3pm, Mon to Fri for general enquiries
Tel: 01709 761450
Web: www.phauk.org
Email: office@phauk.org
Address: PHA UK Resource Centre,
Unit 1, Newton Business Centre, Newton Chambers Road,
Thorncliffe Park, Chapeltown, Sheffield, S35 2PH
Registered Charity Number: 1120756

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** or email: **phauk@anxietyuk.org.uk**

Turn2us

PHA UK has joined forces with Turn2us, a national charity that helps people in financial hardship in the UK. The charity aims to help people in need to access support; and provides a range of information and resources on welfare benefits, charitable grants and other services via its website: **www.turn2us.org.uk**. Through our partnership with Turn2us, PHA UK members can also use the Turn2us Benefits Calculator and Grants Search on our website at **www.phauk.org**

Do we have your correct details?

Please email us on **office@phauk.org** if any of the following apply to you:

- Are the details incorrect on the mailing you've just received?
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