

RESEARCH · REMISSION · TRIALS · HOPE

PEM Lives

ISSUE No: 4
JANUARY 2022



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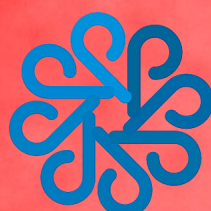
Patient Voice Publication

What PEM Friends means to me

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And more...

This magazine by PEM Friends is for people in the UK who suffer from Pemphigus or Pemphigoid or those who care for them.



PEM Friends
You are not alone

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JANUARY 2022

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PEM Friends

You are not alone

PEM Friends is for people in the UK who suffer from Pemphigus or Pemphigoid or those who care for them.

Welcome to another edition of the PEM Friends magazine

It's been another tough year, hasn't it? So we thought we would try to make the theme of this edition of PEM Life one of hope.

We have another picture from our little star Felicity who filled our hearts in the last magazine - we will have an update in the next PEM Lives. And we have a story from one PEM Friend who has found the holy grail of remission.

We also have the story of one of our members who is finding their way through the aftermath of the diagnosis of Bullous Pemphigoid.

And there is hope of new treatments on the horizon. We were asked to help a medical trials company called Richmond Pharmacology to find PV patients willing and able to participate in a new trial. They tell us something about how trials work. And one of our own PEM Friends will regale us in the next edition of PEM Life with his experience of going through the trial.

But that isn't all the work going on to find causes and treatments for our blistering diseases. PEM Friends is working with doctors and researchers to help them understand, prioritise the focus for future research and manage our autoimmune conditions. It's our strength as a community that enables us to be a part of these important potential breakthroughs.

We also heard about some of the work going on in Europe and the USA that we hope will also lead to developments in the treatment of PEM. We have a short update on the various studies and the conferences attended that talked about some of the work that is going on.

News on the Covid-19 vaccines,

which have dominated the conversations, as well as many of the advisories and papers on our diseases, is kept on our website, which is regularly updated by our own Ingrid. Do take a look from time to time as there is always something new to read.

I think you will agree that PEM Friends has been a good friend to many during these bleak months (and beyond) and it is so encouraging to read some of the messages of appreciation from our members. If you haven't joined the private Facebook group and are a PEM sufferer or carer and live in the UK, please do join us. Or sign up to make sure you are included in any e-mail updates. That includes those who are not patients or in the UK. We are at : <http://eepurl.com/hoye09>.

We also finished the year with a good Zoomie Christmas bash. Our weekly calls are always well attended, but this one was one to say farewell to 2021 and welcome a better year. If you joined, thank you for coming.

We welcome feedback and any comments, in addition to suggestions for useful articles for future magazines. Or just to tell us your own stories.

But, most of all, we hope that 2022 is going to be a very good year for you, your family and everyone.



PEM Friends Update

By Isobel Davies

Autumn is a time of conferences and webinars (all virtual) and we have been busy attending as many of these as we can. Not always easy, as some of them are on US time and concentration can wane a little.

But many of these events confirm the benefit of being associated with several key partners. In brief:

The European Association of Dermatology and Venereology Congress brings together Dermatologists from around the world, not just Europe, and invites many pharma and bio companies to exhibit too. We are invited as Patient representatives to occupy a “stand”, enabling medics to talk to us about our diseases. Our French Friends, the Association Pemphigus Pemphigoid France also attend. This year, we didn’t have much interaction with any delegates sadly, as I don’t think the patient area was very visible (even though well advertised). The true benefit was in the opportunity to attend any of the sessions about our skin diseases and relevant topics. The talks on itching, for example, were very educational and it seems that this is an area that hasn’t been well understood, but is the subject of much research. We also attended talks on treatments for PEM, as well as other presentations. I was particularly interested in the Keynote speech on the future of Dermatology, to kick off the event

Two or three of us also attended the International Pemphigus and Pemphigoid (IPPF) Conference. Here, we heard about the current understanding of PEM diseases (much of which can be heard in the Patient Education Series on the IPPF website). I can’t recommend these webinars enough. There was a lot about the work that the IPPF are doing in conjunction with Bio-Pharma companies into new treatments. PEM Friends were even featured towards the end of the conference by video.

GlobalSkin also had a conference which talked about developments in the understanding of and recognition of skin disease worldwide. Of particular note was the Patient Reported Impact of Dermatological Diseases programme to which many of you have contributed. This is a major international study to create an understanding in the dermatology community of the broader impacts of having a skin disease. You can still contribute if you would like to fill in the questionnaire.

The IPPF and GlobalSkin are also setting up European groups and we are intimately involved in the work of creating these.

Trina has been busy with the creation of a poster and video which featured at the Cambridge Rare Disease Forum, as well as the (still to be broadcast) appearance on 24 Hours in A&E. That makes it sound like it was easy, but it took many hours of being interviewed and filmed, etc. And Ingrid has featured in 2 magazines, having written detailed blogs. One for the British Skin Council and one for the British Dermatology Nursing Group magazine.



SCAN TO GO TO WEBSITE

If that isn’t enough zooming, videoing and writing to keep us going, we have had one of the bi-monthly meetings of the British association of Dermatologists Patient Support Group and the Dermatology Council of England.

The main focus of our PEM Friends meetings this half year, though, has been in trying to prioritise our work and plan for the future of PEM Friends. We hope to tell you more in our next edition, but do let us know if you’d like to get involved in one way or another.

To find out more about these events do ask. It’s difficult to know how much detail to include.

Hope for everyone..

By Peta

I was diagnosed in 2008 at the grand old age of 28. My diagnosis was relatively quick thanks to a fast acting GP and an incredibly knowledgeable ENT specialist at Frimley Park Hospital. After the biopsies and blood tests I was given a final diagnosis of Pemphigus Vulgaris, Pemphigus Foliaceus and Pemphigus Vegetans. Triple whammy!

Fast forward 5 years and after long periods of high doses of steroids (120mg at one point), Methotrexate, Azathioprine, Mycophenolate Mofetil and Mycophenolate acid, my Pemphigus was still not under control. The dermatologist I had been seeing then referred me to Dr Groves at Guys and St Thomas who was so kind and sympathetic and promised he could help me. I had my first two rounds of Rituxan in 2013, and that allowed me to reduce my steroids to 5mg and I remained on Mycophenolate Acid at a maintenance dose. I had 4 years of blister free life before I relapsed. This time I was given Rituxan almost immediately, but my steroids were also increased back up to 30mg. I then had to begin the slow taper back down to 5mg, alongside my maintenance dose of Mycophenolate acid. I did this with success, and the Rituxan once again did its job of ridding me of my blisters. 15 months ago I took my last Mycophenolate tablet!

Last month I had an appointment at Guys Hospital and they informed me that I have been rocking a negative pemphigus blood test for the last 15 months! I could have cried – well I did cry! Finally, some light at the end of this long and dark tunnel. The dermatologist explained that the next step would be to taper me off this final 5mg of steroids. I have now been referred for an appointment with an Endocrinologist who will be performing a cortisol stress test on me to see if my adrenal system is coping after all these years on steroids. In theory, your adrenal system becomes a bit lazy on steroids, so we need to know if mine will wake up on its own! If all goes well, we have a plan to reduce my steroids by 1mg a month (slower if necessary) with a view to being off them entirely in 6 months' time. I am currently on just 5mg but have been on this dose for nearly 4 years now.

I am a little nervous, but very excited at the prospect of coming off these little pills. As patients, we have a love/hate relationship with them. We have all

experienced the "road rage", the crying at nothing, the balmy mood swings and the sleepless nights with endless thoughts in your head. At the same time these little pills are a lifeline, and have changed the prognosis for us Pem patients, allowing us to get a step closer to remission. I shall not miss them, but I will always be grateful for them.



My Experience of Bullous Pemphigoid (BP)

By Lynette Hardy

An innocuous little blister on my hip - that was my introduction to Bullous Pemphigoid.

I covered the blister with a plaster and applied anti biotic cream, but boy was it

itchy and it grew big until it eventually pushed the plaster off! There followed extensive blistering over my body and limbs; I would dread mornings when new blisters had occurred overnight some of which covered the back of my thighs. The blisters joined up, so when a plaster was removed, it also removed the top layer of skin and I was left with large raw burning patches. One such night there was intense itching around my tummy, eventually I put the light on and found my navel to be full of blood.

I had 3 GPs prodding and poking me and I stood there with some shame (note to self,

wear best underwear next time!) An Irish nurse with a very lilty accent was also present who told me my skin looked 'awful dry' and gave

me a big tub of Epimax. It was horrible going to bed covered in thick goo and worse it didn't work, but at least the antibiotics stopped any infection. One of the GPs diagnosed Eczema Herpeticum and gave me.....more anti biotics....oh and continue the Epimax. This went on for 3 weeks during which the blistering got so much worse and so did the itching. My chest particularly was a raw area which made a comfortable laying position very difficult. At night I was in cold baths of water trying to stop the itching. I wasn't getting much sleep and it was a very lonely experience at 2am trying to deal with this. I cried for the state of my own body. I would return to bed trying to keep as cold as possible lest the warmth set off the itching again.

It created large quantities of laundry as I would usually be stuck to the sheets by morning due to seeping blisters or I would

bleed on them and the same for my own clothes. At one point, I floated around in a cotton lawn Victorian nightie like a demented ghost but at least air going through helped the sticking problem.

Finally I called my GP again to report on progress, or in my case lack of it! He asked me to take a photograph of it but I am the equivalent of a Luddite in the technological sense so he told me to come in as their Dermatologist was there today – Hallelujah! She thought it was Pemphigoid Bullae and took a biopsy and that confirmed it so at least I knew what it was. She made an urgent referral to Dermatology where she told me 'they have all kinds of good stuff' and indeed they did. When the Hospital called, their idea of urgent and mine did not coincide! It was a long long waiting list so I said 'don't bother I'll be dead by then' (I think part of me believed that) to which they replied, can you come in tomorrow then. Of course that was a 'yes'!

That's when things started to happen for the better, the Dermatologist I saw was fantastic, very sympathetic, but most of all knowledgeable;

She checked me out and then handed me over to one of the nurses, who plastered me in Dermovate (the laying on of hands somehow has such healing feelings, in the case of this Nurse I am not even sure it wasn't Biblical!) She covered raw places with special plasters which did not stick to the wound and then wrapped me from head to toe in some kind of meshy stuff which she told me to wear all night so the Dermovate could sink in, then handed me two carrier bags of 'good stuff' which I was sure would bankrupt the NHS. I felt like a trussed leg of lamb... It was my first night's sleep in several weeks. I lay down and felt at peace, I was completely exhausted; there was no itching or burning,



Continued over...

just peaceful sleep through the night. I realize how important sleep is now and have come to regard it as a healing process for the body.

My Dermatologist told me 'Beware of Dr. Google', I was sure I would die and actually that seemed quite an attractive prospect, warnings on the internet about sepsis, prognosis not good – about a year with sufferers dying of sepsis, flares lasting between 2-5 years. I was resentful, then angry then defeated and sad. It felt like a modern day plague and I had been selected. A diagnosis of BP is considered a big stress factor in itself, but at first in my mind I refused to live with it; I decided I would take the easy way out.

Slowly but surely I have learned to manage my disease via steroids, Nicotinamide, Dermovate, Dermal and moisturiser (apparently Dermovate works better on moisturised skin). My skin looks like a battleground with all kinds of angry patches, mauve patches and half healed blisters and scabs and the track marks on my shins from the scratching worthy of any drug addict. It will take a long time to clear. Being alone it was difficult to reach my back 'Use the back of a spoon' said the Derm, obviously from the Bear Grylls School of Medicine! but it works.



It is a very time consuming illness and I am in full admiration of those who manage to work although I am retired now, but it still takes a lot of my time with daily management, pill taking, bathing, moisturising and hospital appointments etc. I would encourage everyone who is at a suitable place of their disease with perhaps some healing already taking place to bath rather than shower. I cover my body with Dermal first and then bath and usually I have a peaceful day after that. It is very soothing.



I have learned to manage my disease via steroids, Nicotinamide, Dermovate, Dermal and moisturizer. Meantime, I am winding down on my steroids and my Dermatologist tells me he will prescribe: Methotrexate, Azathioprine or Mycophenolate (watch this space!). It's been quite a year, it has been a true lesson in patience and acceptance – both of which I needed to learn but I didn't expect this means of learning. Now I can manage my condition with help from my Dermatologist, via PEM Friends and via medication and I am in a much better place than when this all started in March this year so I now have hope as well – that the reason, a cure, may be found. Somehow just sharing my experience and hearing from others who know better, helps enormously soin gratitude.

**Please let us know if you are
willing or able**



**to contribute a little
time to help PEM Friends.**

News in brief....



PV Numberplate

How do you like this then? Personalised numberplate of Ady, one of the members of PEM Friends. Many people use personalised number plates to display their name, initials, hobbies, interests, vehicle model or occupation. However,

Ady shares how he is raising awareness of Pemphigus Vulgaris.

Such a fantastic way to start a conversation and certainly adds a touch of individuality to his car! Well done Ady!

What is PV?

Pemphigus vulgaris affects males and females equally. It can start at any age but is most common in adults between 40-60 years old. It affects people of all races. The relative distribution of PV and PF varies in different countries.

Our immune system makes antibodies to fight infection. Normally these antibodies do not attack our own body. However, in an autoimmune disease, such as pemphigus vulgaris, the immune system makes antibodies

(autoantibodies) that work against tissues in the body.

The autoantibodies in pemphigus vulgaris attack proteins called desmogleins. These proteins are present on the cells in the outer layer of skin (the epidermis) and form the "glue" that holds the cells of the skin together.

When the autoantibodies formed in pemphigus vulgaris attack the desmoglein proteins, the cells in the skin and mucous membranes no longer hold together and

separate. This causes blisters and erosions that are typical of pemphigus vulgaris.

Although it is known that antibodies to desmoglein cause pemphigus vulgaris, it is still not clear why some people develop these autoantibodies.

Pemphigus vulgaris does not pass from generation to generation. However, genetic factors are thought to be important in combination with other effects.

"Our immune system makes antibodies to fight infection."

Did you know?

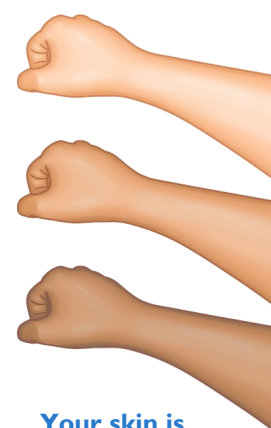
Your skin is your largest organ and plays a vital role in detecting hot and cold, regulating your body temperature and protecting your muscles, bones and internal organs from outside infection and disease. But that's just for starters. There is so

much more to your skin than you might think. Here are some of the most interesting facts about your skin:

- The average person's skin covers an area of 2 square meters.
- Skin accounts for

about 15% of your body weight.

- The average adult has approximately 21 square feet of skin, which weighs 9 lbs. and contains more than 11 miles of blood vessels.



Your skin is your largest organ

Pemphigus and Pemphigoid Patient Voice Publication

in collaboration with Trina Harris at PEM Friends



About Pemphigus and Pemphigoid

Pemphigus Vulgaris (PV) and Pemphigoid are rare autoimmune diseases, also called PEM diseases.¹ They are serious (potentially life-threatening) conditions that cause painful blisters to develop on the skin and/or mucous membranes.¹ There is currently no cure, but treatment can keep the symptoms under control.² The blisters are fragile and can easily burst and cause infections.²

In PV and Pemphigoid, the immune system mistakenly produces antibodies against specific proteins in the skin and mucous membranes, known as 'desmogleins'.² These are known as autoantibodies that attack the "glue" holding the cells together.²



"After taking Azathioprine for a while my liver enzymes became elevated and I was told to stop [the treatment]."

"The unpleasant appearance of blisters caused me embarrassment and discomfort, as well as the physical burden of care required to treat blisters. Since the diagnosis I have a poorer quality of life. I used to be a special needs teaching assistant, but I had to give that role up because of the impact of the disease on my life."

Burden of Therapy and Life Lost

Using corticosteroids for a long time, or in high doses, can cause serious side effects including diabetes, bone loss, thin skin, mood changes, high blood pressure, an increased risk of infection, stomach ulcers and a redistribution of body fat, leading to a round 'moon' face.³

Steroid-sparing immunosuppressant drugs such as azathioprine and mycophenolate help keep your immune system from attacking healthy tissue.^{2,4} They may have serious side effects, including increased risk of infection.

Many people get better with treatment, although it may take years. Others need to take a lower dose of medication indefinitely to stop their signs and symptoms returning.

Managing Symptoms

The aim of treatment includes stopping new blisters forming and healing broken areas of the skin.² Complete remission is possible with steroids and immunosuppressants.²

Steroids suppress the immune system and limits inflammation in the body.² Prednisolone is an oral corticosteroid that is used in combination with an immunosuppressant.² Topical steroids (like creams or ointments) are also used for the skin, and mouthwash is used for oral treatment.²

Immunosuppressants are also used to suppress the immune system.² Azathioprine is used initially alongside steroids to manage pemphigus; if azathioprine doesn't work then mycophenolate is used as a second-line treatment.² Rituximab is a B-cell antibody treatment that is currently used as a third-line treatment in the UK.²



Trial Participation

Many patients with PEM diseases eventually have to stop conventional immunosuppression and will move on to monoclonal antibody treatment (MAT). More research is needed into MATs which focus on B cells associated with PEM diseases. When developing potential new treatments, there is a need to focus on targeting autoantibodies.

Hopes and Aspirations

My life plan is to work with PEM Friends to support patients in the UK by:

- Supporting research studies and programmes relating to PEM diseases
- Promoting collaboration with other PEM groups outside of the UK to expand the information available to UK patients
- Advancing the awareness of PEM diseases to speed up diagnosis and improve treatment options



"I would like Pharma to research the efficacy of inducing remission using MAT drugs, such as rituximab, over immunosuppressant options."



"I have a number of hopes for future research activity, such as the relationship between the HPA (Hypothalamus, Pituitary, Adrenal) Axis and PEM diseases; food or environmental triggers; and the use of rituximab as both a replacement for long-term, high-dose steroids and as a prophylactic treatment during remission."



"There are gaps in treatment and inconsistency between treatment regimes due to lack of awareness of this disease by medical professionals. Pharma can address this by exploring with medical professionals the inconsistency and efficacy treatments across the UK."



Scan here to go to the PEM Friends website for more information



Scan here to find out more about Patient Voice Publications



1. Pemphigus Vulgaris. PEM Friends. Accessed at: <https://www.pemfriendsuk.co.uk/pemphigus-vulgaris>. Accessed 01 September 2021.
 2. Pemphigus Vulgaris. British Association of Dermatologists. Accessed at: <https://www.skinhealthinfo.org.uk/condition/pemphigus-vulgaris/>. Accessed 01 September 2021.
 3. Steroids. PEM Friends. Accessed at: <https://pemfriendsuk.co.uk/steroids/>. Accessed 01 September 2021.
 4. Diagnosis. PEM Friends. Accessed at: <https://pemfriendsuk.co.uk/medical-treatments/>. Accessed 01 September 2021.



What PEM Friends mean to me...?

"PEM Friends is the life line to everyone especially in the beginning of the journey with a PEM. It certainly helped me after finding it in 2017 when a nurse in the hospital told me there was a Facebook support group. Our weekly zoom meetings especially during Covid staying home trying to be safe. Chatting to everyone and hoping to support each other." **Maeve M**

"I found PEM Friends in May 2015 following my diagnosis of Pemphigus Vulgaris and then after further biopsy Pemphigus Foliaceous too. It was invaluable finding others who had been through what I was experiencing and who helped me through those early times of dealing with having a rare illness that few people understood. Having lovely caring people around to talk to was so comforting. Now 6 years on being part of this lovely group, having people to chat to, and welcoming new members too and hopefully giving back some support to others as I will never forget how terrified I was at the start of my journey and how helpful PEM Friends was to me. Loved the zoom calls in lockdown and being able to meet some lovely people 'face to face' for the first time." **Jacqueline S**

"It has been only 5 days since I was diagnosed with PV. I suppose I'm just about getting over the initial shock and thinking "why me?" Of course It is constantly on my mind, particularly what may develop over the next few months. At the moment I'm fine apart from a oral lesions (actually healing) and a sore throat. Thinking about it, I am very disappointed with the delay in arriving at a definitive diagnosis and the fact that the consultant who requested the biopsy did not bother to tell me the result until I saw him last Wednesday, despite knowing the results for over 6 weeks. I made several requests to his MS to get the results without success. In the end I contacted a dermatologist who I know quite well and he got me started on prednisolone. I've noticed several posts on the group about long delays in diagnosis and getting treatment, and whilst acknowledging the rarity of the disease and the pressure the NHS is in at the moment, feel that the Group may be able to focus and bring some pressure to bear on improving these delays. Enough of my ramble then. I am delighted to have the opportunity to talk to others who have this disease and hope to be able to contribute to the Group in some way over the next few months (as a new kid on the block as it were). All the best to you all." **Doug H**

"Quote: this experience is like riding a wave of pain. Yes we have autoimmune disease. No, we didn't do this to ourselves." **Jane S**

"I found PEM Friends in 1999 after initially being told about the P V Network by someone I met whilst at my dermatology appointment at Guy's. I used to go to their meetings in London. The group gave me a real sense of belonging and kindness. I have been in remission since 2004. Thank you PEM Friends." **Margaret Z**

"I found PEM Friends on Facebook two and a half years ago when I was desperately looking for some help, advice and support. This group is so special. People really care and give you practical advice and moral support. I think they're fantastic." **Nicola B**

"Well what can I say. 2015 was such a difficult year. I thought that was it, my time was up. Then eventually after months of being pushed from pillar to post I was given an appointment to see Professor Setterfield. After the scary words you have PV, I searched the internet for a support group and on the 23rd December 2015 I found PEM Friends. I received this message from lovely Isobel.. *Hi Trina. Welcome to the group. I'm sorry to hear about your Pemphigus diagnosis. It's a horrible disease and I'm sure you'll find that quite a few people in the group (or maybe all) have grappled with the prednisolone regime - you're still on a high dose, aren't you? But we can help with advice, support and a place you can vent. Such a welcome message! Such an amazing support group for the past 6 years. I've now become part of the 'fixtures and fittings'. I am so grateful for all your advice PEM Friends. Those early days were scary but knowing you were all there helped immensely. Thank you everyone!*" **Trina H**





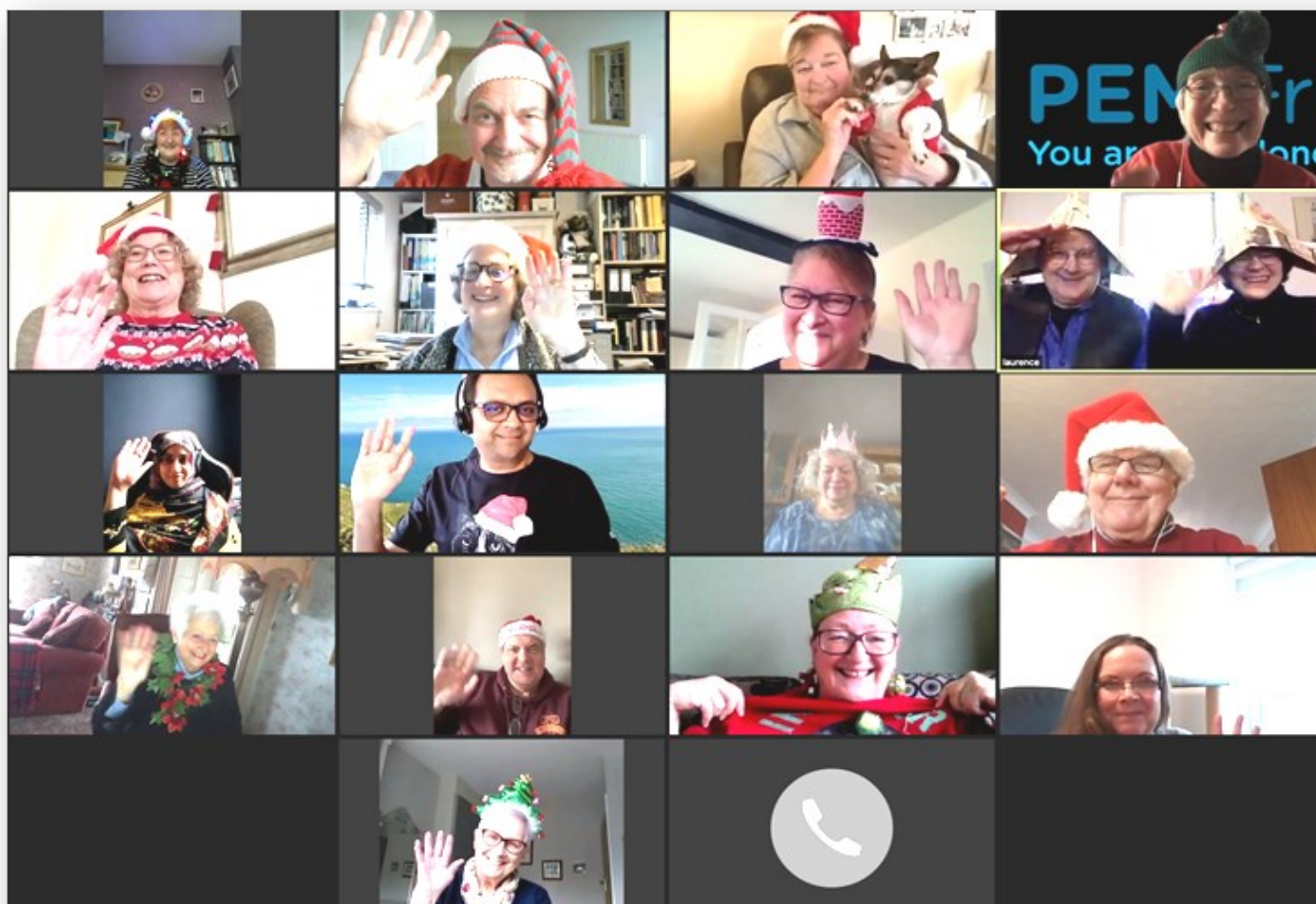
A Zoom is not just for Christmas!

By Andy Heath

Usually, our weekly gatherings on Zoom are a mixture of companionship, support, advice, practical help, and laughter. Our final Zoom of 2021 (shown in the picture) also included festive hats, a quiz, and a visit from the founder of PEM Friends. Carolyn reminded us why she started PEM Friends more than 20 years ago and said how our Zoom gatherings renew her original ambition for PEM sufferers to be able to get together and talk openly about anything and everything with others who really understand.

There is no agenda for these virtual get-togethers: sometimes we discuss coping strategies, sometimes we talk about medications, sometimes we chat about the issues of the day, and sometimes we just listen when someone needs to vent. At times people have a lot to say, other times they might say very little; some people come every week, others pop in when they can make it. Some people phone in, some don't have cameras and just have audio. We don't mind how people connect or what they wish to contribute. Zoom is pretty straightforward to use, and our combined experience is great at sorting out any technical hiccups should they arise. And new people are always made very welcome! I am very grateful for these regular Thursday virtual gatherings. I've learnt more about my PEM and my treatments by sharing stories with others. But I've also met a lot of new friends who have helped me to manage living with PEM. One of our Zoomers (is that a word?) posted a comment in the Facebook Group following the Christmas gathering which said, "Merry Christmas to all, I feel I have grown close to everyone because of the Zoom meetings".

If you are free at 12:30 on Thursdays for about an hour, then please do come and join us on Zoom. Just look for the invites that are emailed out on Wednesdays and posted on Facebook. Of course, I'm biased, but I think you'll be hard pressed to find a more welcoming bunch of people.



Silly hats and zoom quiz

Treatment of Pemphigus & Pemphigoid PSP

Treatment of Pemphigus and Pemphigoid Priority Setting partnership

Patient Setting Priority group

By Isobel Davies

In addition to our contribution in other areas, several of us patients have been involved with a Priority Setting Partnership.

This was set up in 2019 and some of you may recall being asked to tell us the top questions you would like answers to about pemphigus or pemphigoid. This was the start of the creation of a list of hundreds of questions that will be refined to create the top priorities for research or study into blistering diseases in future.

Questions also came from medics and professionals involved in treating our diseases.

It will ensure that clinical research is more focused on those areas that mean the most to those people affected by blistering disease.

The team is made up of several patients, including many from PEM Friends, as well as Dermatologists, Ophthalmologists, Nurses and Researchers. We have worked in close collaboration under the leadership of Karen Harman and the Centre of Evidence Based Dermatology and the James Lind Alliance who are experienced at managing these priority setting partnerships.

Details can be found at:- <https://www.nottingham.ac.uk/research/groups/cebd/projects/priority-setting-partnerships.aspx>

The work resumed in November following a long delay during the worst of the pandemic. By this time, the hundreds of questions had been categorised and a start had been made on grouping questions into "indicative questions". The work will continue into early in 2022 to confirm the shorter set of questions and then we will be looking for input to the final set of priorities.

Watch this space...

* What does the PSP involve?

The first stakeholder meeting was held in May 2019. A PSP goes through the following stages:

1. **Survey 1** will collect all uncertainties about treatments for bullous pemphigoid, pemphigus vulgaris and mucous membrane pemphigoid from patients and their carers, and healthcare professionals.
2. Submitted uncertainties collated and true uncertainties identified.
3. **Survey 2** will rank the most frequently raised questions about uncertainties in order of importance
4. Final **workshop** will involve patients with bullous pemphigoid, pemphigus vulgaris and mucous membrane pemphigoid, their carers/partners, and healthcare professionals to prioritise the submitted questions.

*source www.nottingham.ac.uk/research/groups/cebd/projects/5rareandother/pemphigus-and-pemphigoid-psp.aspx

PEM Financials

From Kalpesh Patel, our treasurer

Happy Holidays!!!, Wanted to start off by thanking everyone who has generously donated throughout the year, in particular families of Noel White and Roy Ives, who sadly died in 2021 and donated to us rather than send funeral flowers. Your continued support is very much appreciated,

In the year 2021, the net funds raised from Donations and Grants was £4776.71.00. A generous donation of £2500 from Richmond Pharmacology and a grant for £859.71 from IADPO! (otherwise known as GlobalSkin). £4205.75 was spent on Website improvement, Zoom Subscription, Google & Facebook advertising, Postage & Packaging, Leaflets/Posters and "Thank You" Vouchers and Gifts to our invited guests. The expenditure has surely raised our profile and awareness of our support group, which has reflected in the increased number of "friends" who have joined our group this year.

I would also like to pass on my sincere thanks to Isobel, Trina, Ingrid, Julie, Peta, Andy, Sharon and Umber for their hard work throughout the year to increase the awareness of PEM Friends, the PEM diseases and support services. Thank you again to those who keep us afloat. If you would like to make a one-off or regular donation, the details are:

Wishing you all Happy Holidays and Very Happy New Year !!!

Bank name:	HSBC
Account Number:	51504525
Sort Code:	40-08-33
Account name:	PEM FRIENDS



We are always very sad to hear about the deaths of people who are connected to our disease. None of the people we have heard about since our last magazine who have passed away died as an immediate result of their disease. But the consequences of PEM may result in related issues. And pemphigoid or pemphigus can add to the distress of someone already suffering from life threatening disease.

Our hearts go out to those who have lost loved ones for whatever reason.

ROY IVES

"Roy was taken too soon from his loving family on 27 October 2021 aged 78 after a second bout of Covid 19. He was one of the first Covid survivors back in March 2020. Covid had left him with serious health complications. He had also had BP for 2 years. Roy will be remembered by all who knew him for his great sense of humour, strong spirit & his kind & welcoming personality. He is greatly missed by his loving family."

Many thanks to Roy's family for asking that donations to PEM Friends replace flowers at his funeral.

PAUL BROOKS

Paul's story is told by his wife, Mila and son Rod. We are grateful to Rod for taking the trouble to tell us about the vascular implications of BP during what must be a very difficult time.

"Tragically, Paul died suddenly aged 67. We have just had the pathology report and he died of Pulmonary Embolism resulting from DVT. He had been suffering from breathlessness since release from hospital some weeks ago and had had high blood pressure and swollen feet and ankles intermittently for a long time but no investigation into DVT had been carried out. We feel the dermatologist and GPs missed a lot of warning signs. Recent research indicates that there is a much higher risk of DVT and Pulmonary Embolism in people suffering with BP. "

NOEL WHITE

We heard of Noel's passing when we received a cheque following his funeral. He wanted donations to go to a PV charity. No other details are available.

RAJU DHOOPER

We were also very sad to hear about the untimely death of our PEM Friend Suman's husband, Raju (Jag), who died at the far too early age of 52.

RAYMOND BLAIN

And, lastly, our condolences to Carolyn Blain who started this vital group, 22 years ago. Her husband, Raymond passed away a few weeks ago.

WE SEND CAROLYN AND THOSE FAMILIES WHO HAVE LOST LOVED ONES, OUR VERY DEEP CONDOLENCES.

Working with other groups

One of the marked changes in in the past 2 years has been the extent to which we have engaged with and been helped by other organisations.

Collaboration with other groups supporting people with PEM around the world has been facilitated by the [International Pemphigus and Pemphigoid Foundation](#) (IPPF) and in addition, our relationship with our French friends the [APPF](#) has grown from strength the strength. We are particularly grateful for their work representing our diseases on the [e-PAGS](#). These are the patient groups who work with the [European Reference Networks \(ERN's\)](#) – groups of medical specialists from across Europe whose aim is to pool knowledge to ensure better access for patients with rare diseases to highly specialised healthcare, improving diagnosis and care in medical domains where expertise is rare.



This type of collaboration can maximise the speed and scale of adoption and spread of innovations in medical science and health technologies. We are hopeful that, despite BREXIT, UK participation in these ERN's, particularly the one relevant to us on SKIN and Autoimmune Bullous Disease in particular continues.

The International Alliance of Dermatological Patient Organisations, better known as [GlobalSkin](#) (we are a member) has, with the IPPF, provided us with information, advice, links to other groups and access to potential new treatments and trials. Following the IPPF Conference, we have spoken to several Biopharmaceutical Companies who are doing great work on new treatments for Pemphigus and Pemphigoid.



[Findacure](#) in the UK has also helped a lot. They run training and information exchange sessions which keep us informed and building (some) expertise in important areas such as social media marketing – important if we are to grow awareness.



Several other organisations, including the [Coronavirus Community Support Fund \(CCSF\) Learning Hub](#) (a result of being provided with the National Lottery Community grant) have also provided us with much needed ideas, information, learning opportunities and an important boost to morale.



In partnership with
THE NATIONAL LOTTERY
COMMUNITY FUND

And last, but not least, we are very pleased to be allied with the wonderful Penny at the PV Network at last.

Pemphigus **V**ulgaris **N**etwork

Thank you to them all as well as to those not mentioned here!

www.pemfriendsuk.co.uk

PEM Friends
You are not alone

Support for patients with Pemphigus and Pemphigoid and their carers

About Us Types of PEM Managing PEM Resources

Resources Types of PEM

Categories of auto-immune bullous diseases

Texture of

The first international meeting of the world with specialist knowledge and outcome measures

Definitions and outcome measures

PDF version

Useful Links

Food and Nutrition

Azathioprine
Cyclosporin
Dapsone
Doxycycline
Mycophenolate
Methotrexate
Nicotinamide
Rituximab
Steroids

You will find a wealth of information as you look around the PEM Friends website.

Research

By Anne-Marie Leech

In our last newsletter, we discussed the importance of research into Bullous Pemphigoid and Pemphigus Vulgaris to better understand the disease and how many people are affected in the UK. Over the last three months, we have been working with Richmond Pharmacology, a clinical research organisation based in London, to raise awareness of clinical trials amongst our membership and invite you to participate.

General awareness of clinical trials has increased due to the COVID-19 vaccination programmes, but there are still misconceptions around safety and outcomes. Here, we explore clinical research and the potential benefits for rare diseases.



What are clinical trials?

Clinical trials are essential to aid the development of new treatments for medical conditions, including Pemphigus Vulgaris. Without clinical research, no progress can be made to improve treatments or find a cure.

To obtain a license for a new medicine, the pharmaceutical company must first conduct numerous stages of clinical trials. Every medicine must go through preclinical testing to assess the toxicity and safety before progressing to clinical trials in humans.

First-in-human trials (FIH) are typically conducted with healthy volunteers. However, with advances in therapies, such as gene editing, these FIH trials more commonly take place with patient populations where the effects of the treatment on the disease can be directly monitored earlier in the development journey.

The stages of early phase research

There are four phases of controlled testing in humans before it can be approved by regulators and prescribed by medical professionals.

Phase I

The earliest trials carried out on a small group of volunteers to establish the safe dose range of the medicine, the side effects, and see how the body absorbs and gets rid of the new treatment. Much of this information is known from the preclinical trials so the risks are controlled.

Phase II

Testing occurs with a larger number of patients who have the condition for which the new medicine is being developed. This phase tests for safety and efficacy and provides a sense of scale within the patient population.

Phase III

Tests involve a much larger group of patients to determine how safe and effective the treatment is. Phase III is the last stage of early phase research before the pharmaceutical company can apply for marketing authorisation from the regulator.

Phase IV

After the medicine is approved, phase IV reviews how effective the medicine has been in its wider use.

A progressive future

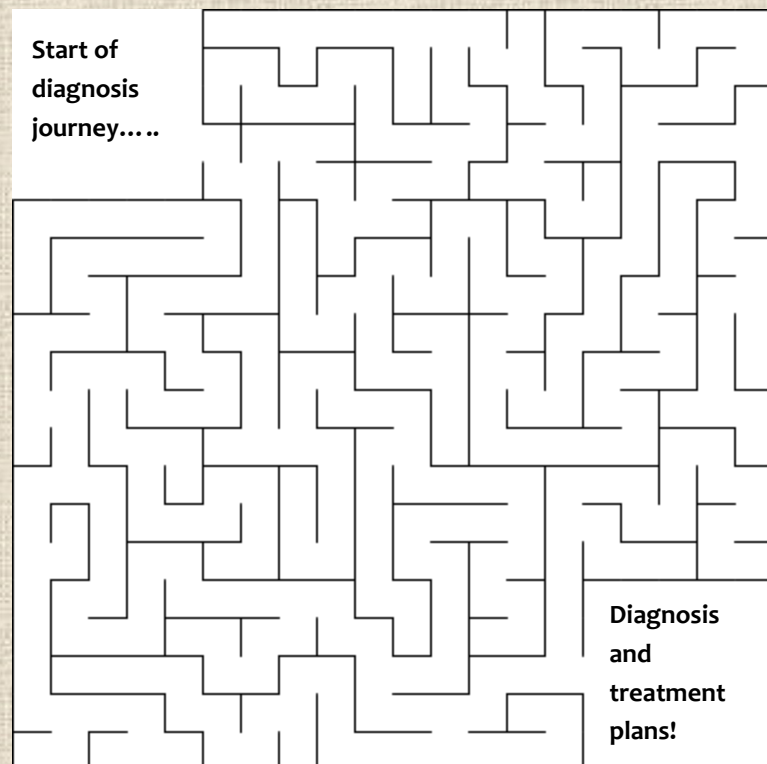
Thanks to these more advanced and sophisticated treatments, the future is bright for many patients, providing hope for patients with rare diseases.

The future of clinical research will reflect this with the development of treatments using novel therapies targeted at the specific genome. Richmond Pharmacology is working with a pharmaceutical company developing cutting-edge treatments for autoimmune diseases, including Pemphigus Vulgaris. These treatments target pathogenic immune reactions while preserving immunity so the body can defend itself against infection. The first-in-human trial is being conducted with patients. The study results may help improve the future treatment, symptoms, and quality of life of patients with Pemphigus Vulgaris.

If you would like to learn more about participating in clinical trials,
email pemvul@richmondpharmacology.com and ask for information about **C20028**.

The journey to diagnosis!

Can you make your way through this maze...



PUZZLE PAGE

A LITTLE
BIT OF FUN

Thought for 2022... this is
certainly true for all
PEM Friends

In true friendship,
one can express
anything and everything
without feeling ashamed
or afraid of being rejected.

Aparna Chatterjee

NEW YEAR'S WORDSEARCH, WHAT IS YOUR WISH FOR 2022?



**PROSPERITY - HAPPY - HEALTH
RESOLUTIONS - GOALS - JOY
TRAVEL PEACE - LOVE**



By Felicity

Links to other groups:

Rareconnect: <https://www.rareconnect.org/en/community/pemphigus-and-pemphigoid>

The International Pemphigus and Pemphigoid Foundation: <http://www.pemphigus.org>

NHS: <http://www.nhs.uk>

Findacure: <https://www.findacure.org.uk>

PV Network: <http://www.pemphigus.org.uk>

GlobalSkin: <https://www.globalskin.org>

CCSF Learning hub: <https://www.tavinstitute.org/projects/coronavirus-c>



January 2022

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