Patient Stories • Conferences • Achievements

PEM Lives

Issue No: 6
December 2022

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

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PEM Friends
You are not alone

INSIDE THIS ISSUE:

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 the festive edition of the PEM Lives magazine.

NOTES ON WHAT SEEMS LIKE A VERY SHORT YEAR

How the time has flown this year. It's nearly Christmas! It does seem as if we should be battening down the hatches, doesn't it? The news seems filled with pessimistic stories about climate change, wars, pestilence, economic crises, etc. Many of us have the additional trauma of dealing with chronic diseases. I am concerned to see the increasing number of youngsters filling our PEM Friends ranks.

However, this edition of our PEM Lives magazine is jam packed with a lot of good news. It's been a phenomenally busy year (as you will see) and we have accomplished a huge amount. We can't measure the levels of awareness we have managed to raise, but with nearly 600 PEM Friends in the UK, and an increasing number of medical contacts involving us in major research projects, we think we have had a far bigger impact than our small organisation could hope for. We are also increasingly approached by pharmaceutical companies wanting input from patient groups.

The list of activities we have been able to do through funding from the British Association of Dermatology and GlobalSkin has been a great boost. It enabled us to attend the European Association of Dermatology and Venereology Congress, and we have kicked off a huge amount of work that has thoroughly drained the energy banks of our volunteers, but we think the Photo Library will be a major contribution to faster diagnosis and, with the journal, improve each owners ability to manage their disease.

Attendance at the British Association of Dermatology Annual Meeting and our presence in their Patient Support Group has helped us a lot in getting the PEM message across.

In fact, work on both the Photo Library and the Survey are going to be hugely beneficial to the work of PEM Friends in raising awareness of pemphigus and pemphigoid but these have stretched our meagre resources (sorry to call you meagre, Andy!) to the full. We should also acknowledge Ingrid's vital work on the website and mention that we hope to add a section early in the new year that will be specifically for Clinicians and other medical experts.

Many of us have been excited to have participated in research projects. As well as the work going on in Birmingham into Ocular Pemphigus and Pemphigoid and our very small involvement in research into new treatments for PV and BP, we have been a big part of the Priority Setting Partnership for Autoimmune Blistering Diseases. This will inform the future of research into pemphigus and pemphigoid and PEM Friends has played a major part in creating and prioritising the areas for investigation down to the top 10. As we go to press, we are awaiting the publication of the results.

We also continue to work with the Centre of Evidence Based Dermatology in their ongoing research into Bullous Pemphigoid.

We have been fortunate during the year to have received donations from the wonderful Ives family as well as sponsored runs from Mark, Simon and David. I hope you will sponsor Mark in his valiant efforts on our behalf in the next London Marathon.

There is more about our financial resources also on the following pages. It doesn't take an enormous amount of money for us to have created all of this and so much for our PEM Friends. With a little more man/womanpower and enough cash to exploit the work we do, we could move mountains.

On a more practical note, we are given a reminder by Gail about the importance of keeping our own data available.

Our own little princess, Felicity has given us her Christmas wishes and, in this edition, she is joined by Lily, a fellow young sufferer. We are seeing a rather frightening growth in smaller heroes with various types on pemphigus or pemphigoid.

I hope you enjoy reading this edition of PEM Lives. We hope to show the important journeys that some of our PEM Friends have been on as well as all the developments in the sphere of Autoimmune blistering disease. Even the excitement of Trina's trip to A&E doesn't escape notice.

I hope that 2022 concludes a year of ongoing recovery and wish you the very best season's greetings from me and the PEM Council, Andy, Ingrid, Julie, Kal, Mark, Peta, Trina and Umber.

PEM Council Annual Meeting by Trina Harris

From left to right: Ingrid, Carolyn, Isobel, Trina & Andy

"This was the first time some of us had ever met face to face!"

On the 25th June 2022, the PEM Council were invited to hold a PEM Friends annual meeting at The Stables, Aston Under Lyne, in Manchester. We decided to make it a weekend visit, because it was such a long way for some of us to travel. 'The Stables' belongs to Carolyn Blain, the founder of PEM Friends who generously allowed us to stay in the beautiful cottage free of charge. Isobel, Trina, Ingrid, Andy (and guests, Simon, James and Ashley), arrived Friday evening and left on Sunday after lunch. Sadly, due to either ill health or last minute plans Umber, Kal, Julie and Peta couldn't join us and we did miss their input.

This was the first time some of us had ever met face to face! It was so lovely to meet each other and share with one another.

Saturday morning we woke bright an early and shared breakfast together before getting on with 'business'. We started the meeting with a fun getting to know you exercise. Together, we discussed many things such as, finances and who would represent PEM Friends at the various conferences for the year. We also debated the PEM Friends survey, social media responsibilities and website administration. There was much enthusiasm amongst us when talking about the new Photo Library project and the help and support of a medical photography student. We discussed the constitution of PEM Friends and finished the day at a lovely Italian restaurant, and Carolyn was able to join us. After the long day of discussing 'business' it was great to chat and share ideas about the direction and future of PEM Friends.

We scheduled our next regular meeting for July to discuss progress of the Journal, Photo Library and Survey projects. We agreed that the PEM Council would have a planning session in November. We also agreed that we would like to hold a virtual AGM for all members of PEM Friends in February 2023 via Zoom. We confirmed our aim to complete a stakeholder map detailing the organisations we wished to closely work with.

Overall, we had a very productive weekend and many exciting plans and future ideas were shared amongst the team.

If you would like to see a copy of the full minutes please email Isobel and she will send them to you.

EADV Congress Milan 2022

By Isobel & Trina

The MiCo Conference Centre.

PEM Friends has attended the European Dermatology and Venereology (EADV) Annual Congress 3 times now. Twice in person and once virtually. We were able to attend this conference to further our aims of building awareness of our diseases, learning more about the development of treatments for autoimmune blistering diseases and collaborating with other patient organisations thanks to the Global Impact Fund granted by GlobalSkin (the International Alliance of Dermatology Patient Organisations) of which we are a member.

Trina and Isobel attended this congress in Milan from the 7th to 10th September, along with their long suffering husbands, Phil and Simon. Isobel was there for the first half of the conference and Trina for the second half because Trina was at her daughter - Jess's graduation on the Wednesday morning and Isobel was at a wedding on the Saturday. Since we were only allowed 2 free tickets, Phil was Trina for 2.5 days and Simon then became Isobel. Confusing or what?

Chatting to a delegate

Scheduling talks and booth time was a bit of a nightmare as was the planning overall - we had to master the technology and the conference programme was huge – so many sessions - although not quite so many that were relevant to our own blistering diseases.

Cheap flights were late at night and meant we had to arrive one day earlier to allow us to get to the Conference early for our booth on Wednesday morning. It also made arrival times very, very late. Trina got to bed at 3am on Thursday morning.

We had quite a nice Italian AirBnB, although we had an exciting late evening on Thursday trying to gain entry through the various security obstacles, only to discover we were trying to get entry to number 3 – we were staying at number 5! We can't imagine what would have happened if someone had opened their door to us trying to get into their apartment!

It was very interesting to travel to and from the conference centre on the Lilac line on the Metro. The station was a 15 minute walk through the streets of Milan and past the huge crematorium.

The conference centre was absolutely HUGE. It looked like an armadillo draped across a boulder from the outside and was vast inside. Our booth was on the lower ground floor and we were a little disappointed that we were out of the way of the large number of Dermatologists travelling to the talks and the exhibition area.

PEM Friends were given booth time for 9 hours over the 4 days, but ended up helping man the GlobalSkin and IPPF sessions too.

We attended a GlobalSkin reception on Wednesday preceded by a series of presentations by other patient groups about some of their projects. We learnt a lot and came away with some great ideas. The buffet and prosecco was pretty good too. Thank you GlobalSkin!

Isobel and Trina manning the Booth

The Congress

Our friend Marc Yale from the IPPF, enabled us to access a talk by Janssen about a new treatment "Exploring the world of bullous pemphigoid: unmet needs and new therapies". This talk, which included DeeDee Murell, Joost Meije and Prof Joly (all very eminent internationally in the field of bullous diseases) was extremely interesting and the new treatment being developed by Janssen sounds very well worth further investigation. I also spoke to the Janssen lady who had produced an excellent animated video. She also told me that they hoped to produce a slightly less technical version for patients.

Isobel and Marc Yale (IPPF) in discussion with a Dermatologist from Ukraine.

There were a disappointing number of visitors but those who did stop and chat were very interesting. The people involved in the various journals were particularly good to talk to and we are hoping we might be able to help with some articles from a PEM patient perspective.

Thursday night was spent in conversation with the IPPF and we managed to find a restaurant on our way home and had a very enjoyable and very productive dinner. Thank you for supper, IPPF. A new experience too – queuing for an ice-cream at 11pm. It was worth it though.

Towards the end of the congress, there were many more talks that were relevant and it was hard to choose the most interesting ones.

What does shared decision making look like for patients?

While one of us manned the PEM Friends booth (from 8.30am – 10.30am), the other attended a session by Marc Yale, **"What does shared decision making look like for patients?".** This was an informative session but it was disappointing to see how few Dermatologists were present, despite many Patient Support Group Leaders represented there. Marc explained how shared decision-making with patients leads to a shared responsibility as well as the patient being involved in medical management decisions. This allows for collaboration between the patient and patient's family as well as the doctor and the care-giving team. He explained that when there is engagement between the patient and doctor a better relationship is developed, and this cultivates trust. This eventually leads to a better treatment plan, based on the values of the patient. In addition, when patients are actively involved in decision-making, they may be more motivated and compliant with the treatment plan.

Patient Association Working Group Roundtable

The final session of the conference was the **"Patient Association Working Group Roundtable".** This interactive roundtable was developed by the EADV Patient Association Working Group (PWG) together with the International Alliance of Dermatology Patient Organisations (IADPO-GlobalSkin). This was a workshop style patient centred session.

The EADV and IADPO-GlobalSkin are addressing these areas and will be in touch in the future with a report and will identify the most popular areas that will be addressed.

Trina and Isobel were exhausted by the end of the week and had probably walked many, many kilometres.

The Patient Association Working Group Roundtable

My blistering disease

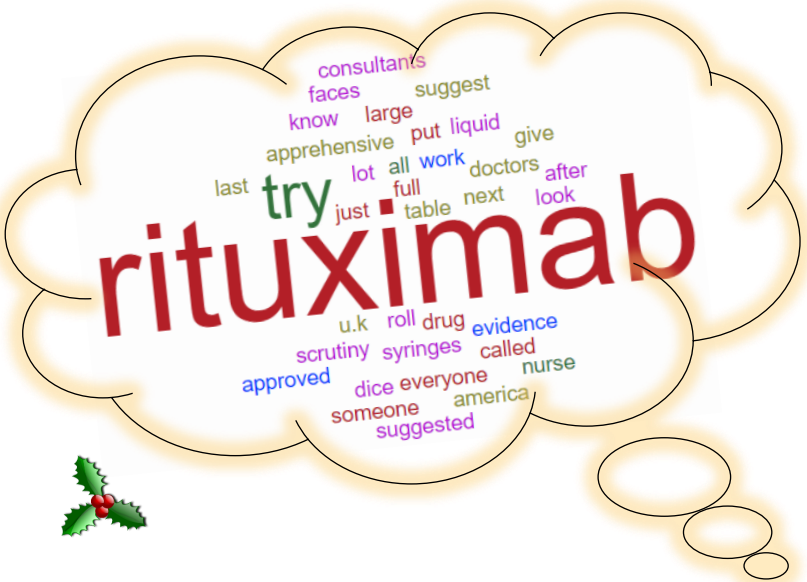
By Mark Noble

Mark was around 36 years old when his eyes suddenly became very sore, irritated, and inflamed. He put it down to a bout of conjunctivitis. Here he shares his 'journey'.

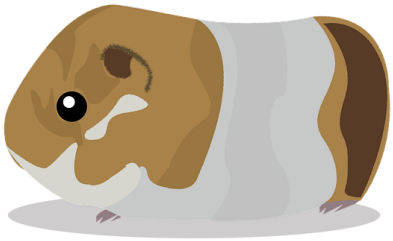
It's autumn 2006 and the Bristol rheumatology department is open especially today with only one patient and it's me!

Only five months ago I woke up with what seemed like conjunctivitis and a few blisters in my mouth. Now, after spending almost the whole summer in hospital, I'm here, blind in one eye, my body covered head to toe in blisters and on a cocktail of medication that could knock out a horse. The clever ophthalmologists and dermatologist confirmed I've got 'Severe Mucus Membrane Pemphigoid' and it's running a mock. Steroids, antibiotics and chemotherapy aren't getting it under control.

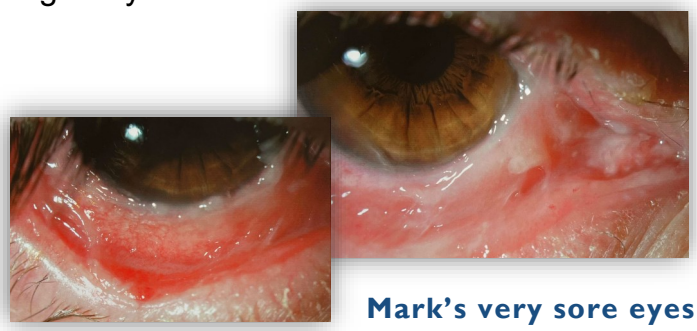
After a lot of scrutiny I've been approved to try a drug called Rituximab. All I know is someone in America has suggested to the U.K. consultants to "give it a try". There's no evidence it will work, but it's the last roll of the dice and by the look on the doctors faces and the large syringes full of liquid the nurse just put on the table next to me suggest everyone is apprehensive. All I can think about it the scene in Pulp Fiction where John Travolta rams the adrenaline shot it into Uma Thermal's chest to restart her heart.



Rituximab is a treatment that, in my IT role for a bank, I'd describe as forced reboot of my body's immune system. It targets and kills off b -cells, hoping they behave better on reforming some 6 months later. It's been used for non-Hodgkins lymphoma and nowadays is a standard treatment for rheumatoid Arthritis, but for MMP I was about to be the guinea pig, the first British one anyway.



By this point my eyes felt like they had shards of broken glass in them every time I blinked, caused by the cornea scarring and conjunctiva shrinking. I was desperate, but with implicit trust in the NHS specialists this was the right thing to try.



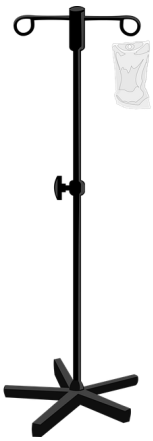
Mark's very sore eyes

The infusion itself was very straightforward. It was administered very slowly over a period of about eight hours. Copious amounts of coffee, a very kind nurse for company and even some Marks & Spencer shortbread even if that lovely buttery sugary taste was tarnished by the copper tinge from the blood weeping from my gums.

I was asked to stay in the hospital for three days, under constant observation , but despite all of the concern I didn't have as much as a headache and even had a very exciting ride home in an ambulance. The second dose, the drug is administered as two doses a fortnight apart, was slightly quicker and thankfully just one night in the hospital.

You'll often hear medics tell you that you know your body better than anyone and although the expectation is several months to know if it has

worked I could tell within a few weeks that the miracle had happened. By Easter I had no blisters anywhere and was in remission. It didn't spell the end of my medical journey but I'm absolutely convinced that without the Rituximab I would be totally blind and possibly not here at all.



Fast forward 13 years and I had another pair of infusions in late 2019 on signs remission was waning and the only thing on my checklist was food and entertainment for the two days hooked up to the drip. This time I was sat next to a gentleman having Rituximab too, for his Rheumatoid Arthritis. A far more relaxed experience as the medical world has realised side effects are very rare and can be largely mitigated by simple checks before administering. If you're reading this because you're about to get an infusion I'd suggest you relax, take a deep breath, pack sandwiches and snacks and download a box-set or film. The expectation is much more worrying that the reality.

Autoimmune diseases?

By Trina Harris

If you get diagnosed with a rare autoimmune disease you probably have never heard of it before, let alone met another person who has it. Which is why PEM Friends is here! We aim to support one another in our Pemphigus and Pemphigoid journeys, with this rare disease! It is hard for us as patients to explain it to others in the general population, let alone our own families.

How many autoimmune diseases are there?

Currently there are more than 100 autoimmune diseases that have been identified and this list is sadly growing.

What is an autoimmunity illness?

The immune system in the human body is comprised of an intricate network of cells, tissues and organs. These all function in an organised fashion in order to protect the body from detrimental organisms. So what is an 'autoimmunity' illness? A healthy immune system defends the body against disease and infection. But if the immune system malfunctions, it mistakenly attacks healthy cells, tissues, and organs and causes confusion in the body.

So what causes an autoimmune disease?

Many researchers are still unsure what the causes are of many of the different autoimmune diseases, but there is evidence to suggest that past infections, genetic factors and different environmental factors of could be responsible.

***Although rare diseases may be individually rare, they are collectively common, with 1 in 17 people being affected by a rare disease at some point in their lives.** *(*source https://www.gov.uk/government/publications/uk-rare-diseases-framework/the-uk-rare-diseases-framework)*

www.freepik.com

Update '10k for PEM Friends'

By Trina Harris

On an overcast bank holiday Monday (2nd May 2022), **MARK RANSON-THOMPSON, DAVID BARRETT** and **SIMON HARRIS** took to the streets of London and ran the 2022 Vitality London 10k, to raise much needed funds for PEM Friends.

Trina attempted to get a photo of Mo Farah but he was just too fast!

The London Vitality 10,000 had returned after a two-year absence due to the covid pandemic. Thousands of runners spent their May Day bank holiday running through the streets of London, battling their potential against top athletes. This race, which took place around many of the capital city's landmarks, featured top athletes including Olympic champion, Mo Farah, English 10k champion, Nick Goolab, and Eilish McColgan!

The assembly area was in Green Park, and participants were given various colours which reflected their different start times. David, Simon & Mark set off at a few minutes apart, with a different colour starting every eight minutes. The last colour set off roughly an hour and 15 minutes after the first colour started. The men's finishing times were incredible: Mark 01:09:44, David 01:18:47, Simon 01:23:15!

CONGRATULATIONS GENTLEMEN - YOU DID IT! The photo below says it all! PEM Friends are so proud of you! We truly appreciate every single donation no matter how small. On behalf of everyone who supported you in this huge achievement, we want to say THANK YOU! Direct donations were £70.00 and via JustGiving it was £2,158.44 from **105 supporters**. **The total amount raised was an amazing £2,228.44!**

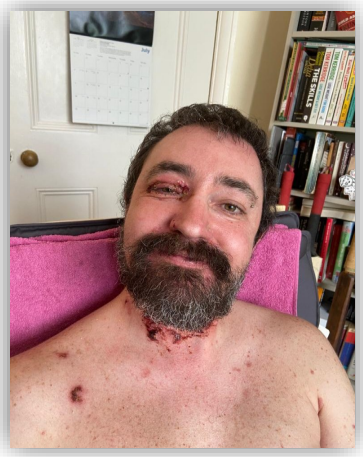
Look at those proud faces!

Erythema Multiforme or Pemphigus Vulgaris? - Mark's story

By Mark Ranson-Thompson

Hi PEM Friends,

My name is Mark. I am 42 and in January 2021 I started experiencing the horrific symptoms of Pemphigus Vulgaris. Little did I know that these small, random blisters appearing on my shins would worsen, take over my whole body, and set me on the journey I am now on – a diagnosis of Pemphigus Vulgaris.



Mark unwell July 2021

I believe there was some soft palate involvement, which I wrote off as a cold, but the first time I knew something really wasn't right was when a few red dots appeared on my belly, swiftly followed by blisters on my shins. Being January 2021, I was well into lockdown Christmas (and also birthday mode, being a January 2nd baby!)

I didn't want to bother the NHS but made good use of calling 111. Fortunately, I spoke to a doctor who knew something wasn't quite right.

My GP – the Jenner practice – also were quick to realise this was no ordinary skin condition, and I was fast tracked to dermatology at King's in South London.

I was originally diagnosed with Erythema Multiforme, but only for a short while. My 2nd biopsy soon revealed my true diagnosis – Pemphigus Vulgaris. I was mis-medicated for a while, which worsened my condition. I was soon using a walking stick to get about due to severe blisters on the soles of my feet – being barely able to walk lasted for the best part of 10 months – a hindrance when the pubs reopened in April 2021!

My wife Chrissie found PEM Friends on Facebook. Unusually, for such an extremely rare condition, she saw her colleague was in the group - who provided priceless support and information, along with everyone else who was part of PEM Friends, of course.

Thanks to the support of PEM Friends, and especially my wife's colleague, I was able to make first contact with the dermatology team at Guy's Hospital in London Bridge, where I am now seen by Dr Groves and his team.

Dr Groves has prescribed both ample medication and a course of Rituximab, which I feel has massively improved my condition. I fortunately responded very well to Rituximab and have been told that I will be having this again within the next 8 months.

In December 2021 I was truly feeling better again when disaster stuck 2 days before Christmas – my wife noticed I had an unusual cough and I also had a headache, and she sent off for a PCR test. I had covid for Christmas and was therefore housebound yet again! Fortunately, I was eligible for the new antiviral drugs which helped me shake covid unscathed.

Mark and his mum with their walking sticks

I was always keen on sports at school but was more of a sprinter rather than a distance

Continued over...

runner (undefeated in the 200m sprint by the way!) After a brief-ish hiatus, possibly distracted by me discovering the joys of a good pub, I returned to running thanks to Parkrun - after going to watch the London Marathon in 2019 I was inspired to go along to my first local Parkrun in Catford.

After the ups and downs of covid and life with Pemphigus Vulgaris I gradually returned to running, and previous readers may remember I ran the Vitality London 10k in May 2022, alongside Simon Harris and David Barrett, all raising money for PEM Friends. I was back enjoying my long-distance running. My official time was 1hr 9, 44s. We raised a grand total of £2,228.44, especially thanks to the fantastic publicity of Trina Harris!

Mark after his 10k, hoping to meet HRH!

For perhaps the 15th time of applying since my early 20s for the London Marathon I have finally secured a ballot place and will be taking part in the 26.2 miles course on 23rd April 2023. I am delighted to announce that I will be raising money for PEM Friends again, and training has already started!

Please donate if you can but please don't worry if not. I appreciate times are very difficult at the moment. I will just be glad to be raising much needed funds for our support group, the wonderful PEM Friends.

Mark

JustGiving™

You can donate to support Mark and ultimately, PEM Friends who will be running The London Marathon on 23rd April 2023, by going to the JustGiving page. (*Just type the following into your browser:)*

https://www.justgiving.com/crowdfunding/mark-rt-marathon?utm_term=rz7vw3p55

Please share with your friends and family too!

Covid 19 Guidance from www.gov.uk

Last updated 7 October 2022

Immunosuppression means you have a weakened immune system due to a particular health condition or because you are on medication or treatment that is suppressing your immune system. People who are immunosuppressed, which could include most PEM sufferers, or other specific medical conditions, may have a reduced ability to fight infections and other diseases, including COVID-19.

For the latest updated guidance for COVID-19 (This guidance only applies to people living in England) **www.gov.ukgovernment/publications/ovid-19-guidance-for-people-whose-immune-system-means-they-are-at-higher-risk/covid-19-guidance-for-people-whose-immune-system-means-they-are-at-higher-risk**

The Things We Do for Love

(made possible with money!)

This is a summary of phone, WhatsApp, email and face-to-face conversations between Isobel Davies and Andy Heath about what PEM Friends has achieved in 2022 and how it was funded.

Isobel
Hi Andy, are we going to talk about what PEM Friends has achieved in 2022?

Andy
Hi Isobel, yes, I've been looking at the numbers as well, so we can cost our activities.

Isobel
PEM Friends is a pocket rocket! We do a huge amount, despite our small size and the fact that the core management group (the PEM Council) is entirely voluntary. All the council members grapple with their own disease so every achievement is far greater than you would expect.

Andy
I've estimated the total time the PEM Council has volunteered over 2022: it's roughly about 1,750 hours. If we paid for that time at the minimum wage that's over £16,000!

Isobel
Blummin' 'eck! But don't forget the support of our members, they go above and beyond to help each other, on Facebook, helping others 1 to 1 and elsewhere. We don't know how much time they put in. One of the principles that we hold close to our hearts is that we do not charge people to be a PEM Friend.

Andy
I imagine that puts the total value of volunteering over £20,000. Wow! It's a shame I can't include that sort of "in-kind" number into our budget/forecast.

Isobel
Why do you call it a budget/forecast?

Andy
It's just a fancy spreadsheet really but I've included lots of calculations so that it can predict our cash flow. It just means we can forecast our finances in advance when we do our planning. Oh, and the charts are useful for the PEM Council to quickly seeing how we are doing.

Isobel
And the budget is kept updated with the bank statements I send you every month?

Andy
That's right. Of course, that means I don't know all the details about what's happened this year that didn't involve income or expenditure.

Isobel
There's a lot! Running the Facebook private group and the Facebook page, organising expert talks, managing the various email groups, participation in PSP and Bullous Pemphigoid surveys and research, BAD PSG and DCSE membership, and attending meetings of the European Dermatology Group and inputs into the BAD and Nursing initiatives. We also wrote an article for the Primary Care Dermatology Society Magazine. And we're involved in planning the FDA/IPPF January 2023 meeting. We also manage a Twitter account. And don't forget the survey project!

Andy

I certainly remember the survey! That took a lot of time. And there's more work to do with the survey results.
But I didn't realise quite so much had happened. Obviously, I know about the website maintenance, the email hosting, the Zoom meetings, the Photo Library project, the Journal project, the magazine, the leaflets, posters, tee shirts, badges, hats and pens, and attendance at conferences because these all need money and are in the 2022 budget/ forecast.

Isobel

We've been extremely busy this year but by being so involved we can grow awareness of our diseases to help those who experience them directly and to educate people based on what we can learn. And the more our medical professionals know about us and the diseases we suffer, and the impact of those diseases and the treatment options, the better the outcomes will be.

Andy

Because you've just sent me the latest bank statement, and we're near the end of the year, we can forecast the remaining income and outgoings pretty accurately. In 2022, the income to PEM Friends will be approx. £8,080 and the expenditure approx. £8,470.

Isobel

Even though we have spent more than has come in are we OK?

Andy

Yes, we are OK right now. We benefited from the very sad and untimely death of Roy Ives in 2021. We've been able to do so much due the enormous generosity of the Ives family.

Isobel

Their donations were huge. Margie Ives is still raising money for us through her activities like car boot sales. We'd not be in the position we are in without these donations.

Andy

We also received a substantial donation from Richmond Pharmacology in November 2021 after we helped them find patients to participate in their trials for a new treatment for PV. All those donations meant we started 2022 with a decent bank balance.

Isobel

This year Simon, David and Mark did the Vitality event in London and ran miles for us. They raised over £2,200 in sponsorship. It made a big difference to our ability to deliver our awareness campaigns.

Andy

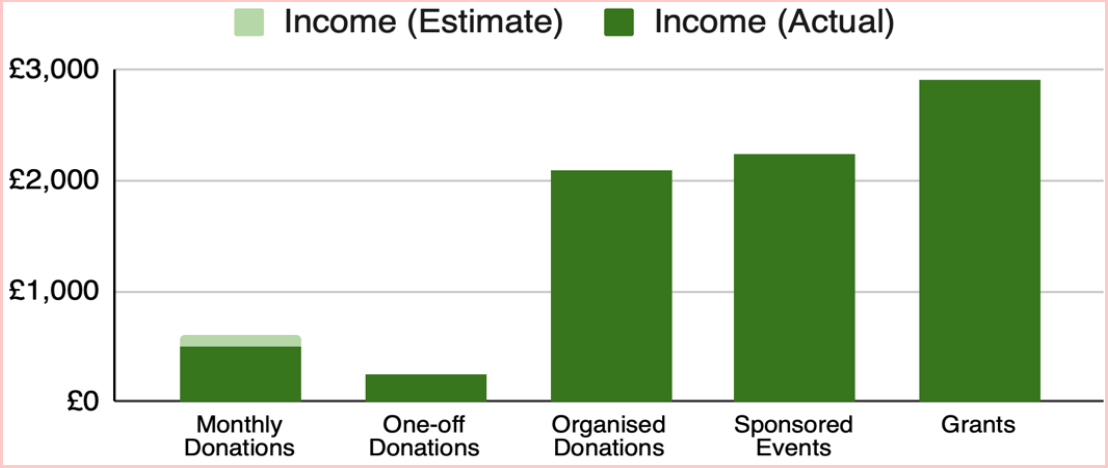
I was so impressed with that. I couldn't run around the garden. The people who sponsored them were very generous. I think Mark might be in the London Marathon next year. What could I do? Do you think people would sponsor someone creating spreadsheets?

Isobel

😂

Andy

Our income this year has come from three main sources: donations, the sponsored run and grants. Here's our income chart from the November budget/forecast:



Isobel

The grants came from the British Association of Dermatology for the Photo Library and the Journal projects. And a grant from the GlobalSkin Impact Fund to help us attend the EADV Congress in Milan.

Andy

The grant for the Photo Library was £900 and then £720 for the Journal Project. The GlobalSkin grant was $2,000 Canadian Dollars, so about £1,290 after it was exchanged into sterling.

Isobel

Without that we couldn't have attended the European Dermatology and Venereology Congress. It was a really important event because it allowed us to understand more about developments in the treatment of our diseases, make new contacts and talk to Dermatologists about our work. The grant didn't cover all costs though, the total was over £1,700. PEM Friends also contributed.

Andy

You and Trina paid some of the expense out of your own pockets! The British Association of Dermatologists conference in Glasgow cost £930: that feels core to the work of PEM Friends.

Isobel

It is. It was a great opportunity to speak to UK Dermatologists about our diseases and our patient support activities. Having access to so many professionals and making them aware of pemphigus and pemphigoid is paramount to reducing diagnosis times. Also, both the conferences give us a chance to mingle with other support organisations. We learn from them and sometimes we can offer advice. We make connections and improve our support network.

Andy

I used to attend a lot of conferences when I was working. They are hard work, especially if you are lugging materials around, manning a stand and travelling a lot.

Isobel

It's very useful though.

Andy

Did our customised pens go down well at the conferences? They cost about £60 for 50 pens.

Isobel

The pens were incredibly popular with doctors. They will remember PEM Friends when writing a note or signing something.

Andy

Another big expenditure is the magazine. It averages over £600 per issue, and we print 3 issues annually. That figure mostly includes the P&P but it's a hefty sum, and the costs have increased recently. Mostly I'm a digital person but I do like the paper magazine.

Isobel

I think that the magazine on paper enables us to tell our story to so many people. Sending a hard copy does seem to ensure a more guaranteed readership but the absence of addresses for many of our targeted doctors means we must send them the magazine as a PDF.

Andy

Another substantial cost is for the internet services we use. Like the email and website hosting, and the Zoom license which enables us to hold the Thursday meetings.

Isobel

How do our finances look for 2023?

Andy

Well, all the planning isn't in there yet of course but we can forecast that our basic running costs will be a little over £2,200. That will cover 3 issues of the magazine and the internet services we need.

Isobel

What money do we have coming in?

Andy

Our funding bedrock is the generosity of our PEM Friends. We receive monthly donations via direct debit from PEM Friends members which totalled £600 this year. We also get one-off donations. But we don't know what other income might, or might not, come in.

Isobel

We are so thankful for these donations. Without them we wouldn't be able to run the website, or the Thursday Zoom calls. And the magazine would have to be a digital only, and there would be no leaflets for clinics.

Andy

We wouldn't be able to do lots of the other activities though.

Isobel

Absolutely! And we wouldn't be able attendance conferences which enable us to learn more about pemphigus and pemphigoid and contribute to the growing awareness of these horrible diseases.

Andy

We've achieved a huge amount this year partly because we have been fortunate with income and all the time given by volunteers. And because of your efforts pulling it all together.

Isobel

As PEM Friends has grown, we have more responsibility, and we have got busier and busier. We frequently get requests for help from patients and organisations. I'd welcome more volunteers and more donations; one-off or, even better, regular donations make a huge difference.

Andy

Well, PEM Friends certainly isn't showing any signs of slowing down!

We would welcome any contributions *to PEM Friends no matter how small. Regular payments are even more appreciated.*

Donations can be made to:

HSBC Bank

Account Name: PEM FRIENDS

Account Number: 51504525

Sort Code: 40-08-33

Andy and Isobel deep in discussion...

PEM Friends needs YOU!

Run on the work of a small group of volunteers (The PEM Council).

We need a few more willing and able volunteers to:

- **join the PEM Council**
- **do various activities on ad hoc projects.**
- **"Befriend" someone who can't access Facebook or needs some additional help.**

IF YOU ARE INTERESTED AND WILLING TO GET INVOLVED IN ANY OF THESE 3 AREAS, THEN E-MAIL US, WITH A SHORT SUMMARY OF THE SKILL AREAS YOU CAN OFFER AND THINGS YOU WOULD BE INTERESTED IN, AT mail@pemfriends.org.uk

Did you watch 24hours in A&E? *(Series 28 Episode 2)*

24 Hours in A&E shown on Channel 4, is a British documentary programme, set in a teaching hospital in inner London. Filmed inside St George's Hospital in Tooting. Cameras film round the clock for 28 days, 24 hours a day in A&E (Accident and Emergency). This offers unprecedented access to one of Britain's busiest A&E departments.

Trina was diagnosed with Pemphigus Vulgaris in January 2015. In October 2020 she was taken by emergency ambulance to St George's Hospital following a huge flare of Pemphigus. This was her 3rd flare like this and each episode caused admission into hospital. This was the worst flare she had ever experienced. She was finding it difficult to breathe after an eruption of blisters in her airway. She spent a further 4 days in hospital receiving IV steroids and antibiotics to treat the blisters.

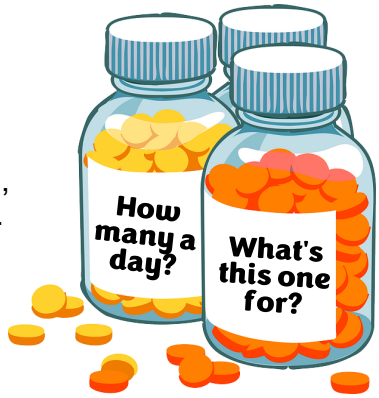
The episode featuring Trina was aired on Monday 12th September 2022. You can still watch it here: https://www.channel4.com/programmes/24-hours-in-ae/on-demand/70071-022

ICE (IN CASE OF EMERGENCY) PICKS

By Gail Windows

We wanted to raise the importance of carrying your medical information with you. For those of us on steroids, it is particularly important to let a medic know if you have a medical crisis.

In our update about the journal project, we make the case for keeping an ongoing record of your symptoms and medication. This is a chronic disease, after all, and can last for many years. However, if you are involved in any medical incident that threatens your wellbeing you will need to convey information about your medical circumstances.



Gail's story:

Are you 100% sure what your meds are, what dosage they are and why you take them? I suspect many of us including myself know most of what we take, why we take it and how much, especially regular and longer-term medication. I have taken some for many years but I don't know the dosages of them all! I'm only now beginning to realise, since being diagnosed with Pemphigus and being autoimmune, the importance, highlighted by an event several years ago, which I'd like to share.

Since being diagnosed with Pemphigus and learning about autoimmunity I'm only now beginning to realise the importance of knowing about your medication which was highlighted to me at an event several years ago. I'd like to share this with you.

I have suffered severe depression and following a family bereavement, my dad's dementia and possible skin cancer, I struggled and the GP prescribed anti-depressants. Within a few days I started to get shakes/dizziness building to what felt like heart palpitations. One day, out shopping, my heart felt like it was going to burst out my chest. In a panic, I phoned my husband at work. Realising I was near my GP's surgery, he said GO THERE NOW! I did, only to be told I was just having a panic attack, "Keep on with the tablets and it will 'settle down'". However, it didn't improve so I eased off them and soon the 'attacks' stopped, but I kept the partly used bottle.
Subsequently, we moved to a different area. My dad was diagnosed with Dementia and I became carer for both parents. There were also other ongoing 'family issues' and my depression started again. I knew I needed help. I couldn't just carry on 'alone'.

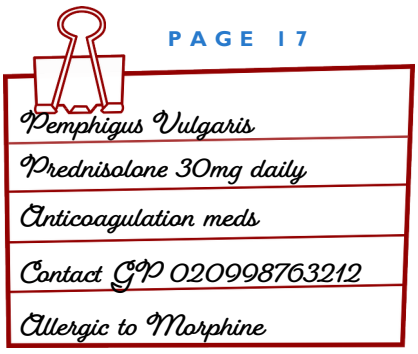
I went to see a new GP, a trainee doctor. He was great! On reviewing my historical medication he asked about amitriptyline and I advised I took it for migraines. He explained a medication he wanted me to take and, after checking the bottle at home, I confirmed it was the same previous medication I'd had issues with. A senior GP joined the conversation and I explained my experience of what felt like heart palpitations, but that they had been dismissed as a panic attack. I was shocked when they said that Amitriptyline and this medication conflict and caused heart palpitations! I had been very lucky to stop when I did. I could have had future serious heart problems. Shocked is an understatement.

This new GP had done something EVERY GP should do and check for drug interactions. My own stubbornness of 'you can conquer this Gail' had, without me realising it, saved possible future heart problems.

Over time, I became blasé about my meds and nothing really changed. My life became a constant rush between caring for my parents and my job, which I eventually had to give up . Then I started getting a skin problem on my scalp. There followed more GP and hospital visits until a dermatologist referral. After two years of misdiagnosis, and many different treatments, including 3 lots of cryotherapies, I was diagnosed with acantholysis. It was like BOOM! I was referred to a specialist.

My GP was not aware of Pemphigus (he is now!) There is a lack of awareness of our condition. I researched acantholysis and during that, the word Pemphigus came up, leading to me finding PEM Friends and my specialist Karen Harman, realising my life was going to change for me and my family.

I now appreciate the importance of knowing what medication we take. In an emergency, however unlikely, carrying an alert card stating your specific condition, medication name and dosage, is important and will assist paramedics and medical staff and keep you safe.
It's kist a small card but it could just possibly save your life one day! Whether it's a card you make yourself (you can use the template I have provided), get from GP, hospital or pharmacist, please carry a card.



Comments from PEM Friends:

If you would like to check the interactions between your drugs, PEM Friends recommends Drug.com, https://www.drug.com which enables you to list all your medications and see what the possible interactions might be.

There are many different ways to personally carry ICE information:

- If you would like to get a copy of the card that Gail has designed for the purpose, then please get in touch.
- Alternatively, you should ensure that you hold emergency data on your mobile phone. Mark advises:- "Increasingly many of us carry a personal mobile device with us everywhere we go and it's simple to store ICE details electronically in your phone."

  Emergency services will be aware with a click and a swipe on your screen they have access to your emergency contact and medical details such as your blood group, medical conditions and current medication.

  There are many helpful web pages and YouTube videos if you search "how to add ICE information to a mobile phone" Click this link or type into your browser for an example of one of these videos: https://medium.com/@n.craig_62483/how-to-set-up-ice-on-your-iphone-cbf6ca250ff0
- Or, finally, you can subscribe to MedicAlert. Their products (mostly bracelets and necklaces which are at pulse points) hold a minimal amount of data and a phone number. The medical team can call the number listed 24/7 and obtain detailed information about your condition and drugs. There was an article on them in the July edition (number 3) of our magazine.https://www.pemfriendsuk.co.uk/_files/ugd/ab0c71_0bf041f0e60c4fe69e3b3c7db3afae83.pdf

PEM Financials From Kalpesh Patel, our treasurer

What a year it's been!! We've managed to raise funds through various methods such as regular donations, one-off donations, a 10K run, car boot sales, funeral donations, and grants. Thank you to each and everyone who has donated and help raise funds throughout the year, your continued support is very much appreciated and vital to our work. Without you, we would be unable to continue in the way we have.

If you can spare even a few pounds every so often, it would enable us to offer our support to more people.

Funds Raised

Donations and grants - £7,980.50 (£5,069.28 raised/donated by supporters of PEM Friends and £2,911.22 in grants (BAD - £1,620.00 and GlobalSkin - £1,291.22).

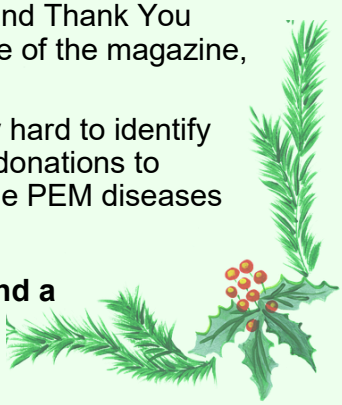
Expenditure

So far this year £5,619.27 has been spent on two issues of the magazine, conferences, projects, and internet services, with more modest amounts spent on marketing materials, admin, and Thank You vouchers. We have further commitments still to come this year, not least on this issue of the magazine, and anticipate ending the year spending slightly more than we have raised.

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

PEM Council members are all working very hard to identify and implement effective ways of spending donations to increase the awareness of PEM Friends, the PEM diseases and support services.

Wishing you a joyous Holiday Season and a most prosperous and healthy New Year.



ERN & ePAG's.

IMPORTANT ACRONYMS FOR US ALL TO KNOW

Laurence Gallu (APPF)

November 7 2022

European Patient Advocacy Groups (ePAGs) were created in 2017 in parallel with the European Reference Networks (ERNs). ERNs are health care networks dedicated to rare diseases. There are 24 ERNs and ours is the ERN SKIN - https://ern-skin.eu/ Each one is divided in sub-groups also called thematic groups; ours is the AIBD – the Auto Immune Bullous Disease group and it is chaired by Prof Pascal Joly (Rouen, France) and Branka Marinovic (Zagreb, Croatia). There are 19 participating Reference centers in our ERN SKIN, including Guy's and St Thomas' NHS Foundation Trust – Dept of Dermatology, with Jane Setterfield as its official representative.

ERNs are networks of Centres of Expertise that connect experts and researchers, who share the same interests in a specific rare disease or highly specialised treatment, across the European Union and the countries of the European Economic Area region.

ERNs are first and foremost healthcare networks that aim to improve access to diagnosis, care and treatment by sharing their expertise, knowledge and resources, guiding care and treatment. They do this:

- by connecting experts from different countries to form a critical mass of expertise, knowledge and data sharing
- by transferring expertise and knowledge through clinical case discussions and peer learning

Expertise is therefore made to "travel", not the patient, through virtual clinical information dissemination tools. It is therefore of utmost importance that ERNs remain centered on patients' needs. ERNs give clinicians and patient organisations from different countries the opportunity to co-design better healthcare services for people living with rare diseases.

Each of the 24 ERNs focuses on a particular disease area and has its own ePAG. Right now, I am the e-PAG SKIN representative for our AIBD thematic group. I think Luigi (from the Italian Pem patient organization) is involved as well but in the background.

e-PAGS are voluntary and have to be endorsed by a patient organisation based in a European country.

Our e-PAG SKIN meets about 6 times a year. Our **role is:**

- **to work** with our national and European networks in order to facilitate a 2 way communication between the ERNs and the patient community
- to work in partnership with the clinicians to support the development of clinical practice guidelines and other clinical decision support tools and contribute to research, education, information and awareness activities.

Other than translation work, the e-PAG SKIN AIBD group has prepared and collaborated on the PV **Patient journey** with PEM Friends, with the German and Italian patient organisations as well as with our team of doctors who checked and endorsed the "treatment" section. This "journey" goes through a timeline from diagnostic delay to diagnosis, to treatments and relapses and concludes on the burden of our diseases. At each step, we listed the difficulties we experienced & what we would have wished as help – what did we get vs what we would have wished to get as help, what were our needs & necessities and were these met. Doctors use it as a frame of reference. We need to continue working on the BP & MMP ones now – we have started but will need more input/ feedback. These journeys are on-going since (and as long as) research is on-going.

As an e-PAG SKIN, I've worked also on the evaluation questionnaire given to patients after a "visit" to an expertise ERN center and on the patient journey on common needs, shared by the 9 ERN SKIN thematic groups.
All in all, working as an e-PAG is a great experience because you feel less isolated in your disease!

This article was written by our good friend, Laurence Gallu, who works for Association Pemphigus Pemphigoïdes France (APPF). We are grateful for her contribution to PEM Friends, including keeping us updated on the great work being done by the APPF, as well as representing Pemphigus and Pemphigoid patients on the e-PAG for our diseases. The Pemphigus Patient Journey is available on request.

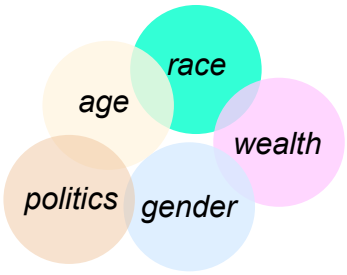
THE STUDENT VOICE
COLLABORATING WITH FUTURE MEDICS

In the last edition of PEM Lives, we published an article by one of the students who we sponsored to compete in an essay competition about rare diseases. There were 2 young students, Devni and Safa who took an interest in Pemphigoid/Pemphigus. It was Devni's turn in the last magazine and now we are delighted to publish Safa's essay. The essay competition is sponsored jointly, by Medics4Raredisease and Beacon (formerly known as Findacure).

The Student Voice Prize Patient Group Pairing Scheme gives medical students, nurses and biological sciences undergraduate and masters students the chance to be paired with a rare disease patient group to learn first-hand about their condition and patient experience. It benefits students because it introduces them to rare disease early on in their career and helps them to understand the patient experience.

The scheme also benefits patient groups because it allows groups to share their story and experiences with a doctor, nurse or scientist of tomorrow who may never have considered working with rare disease before.

Rare Diseases and Intersectionality:



By Safa Iqbal

A wide range of factors, such as race, wealth, and gender, can impact an individual's experience of healthcare and society in general. This is known as intersectionality. Use a case study to demonstrate the specific challenges faced by patients with rare diseases on a daily basis. Discuss how intersectionality can mean that these people experience greater disadvantages.

The humans behind the statistics.

This essay describes my reflections when speaking to four patients from the PEM friends support group, in the age range of fifty years to seventy-five years old. Female W has mucous membrane pemphigoid (MMP), female X and Y have bullous pemphigoid (BP) and patient Z, a fifty two year old male, has ocular mucous membrane pemphigoid (OMMP), respectively. Female preponderance was analysed in studies associated with this disease, the female-to-male ratio ranges between 1.04 and 5.1 but several studies also show the rate of the disease seemed to be higher in women until the age of seventy-five, but thereafter the rate of this disease increases in men.

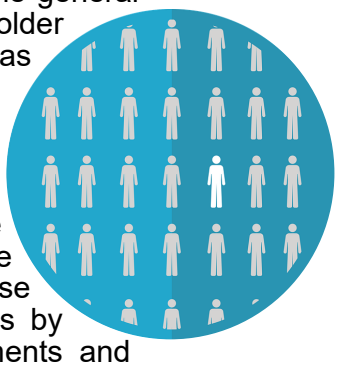
I will discuss how intersectionality affects their life, the challenges patients face in their everyday life and obstacles they have had to overcome when embarking on their patient journey in the healthcare system. Intersectionality can be defined as an approach that explores the interactions of different factors, such as: race, ethnicity, gender, wealth and geographical locations, which can disadvantage people. In the context of rare diseases, intersectionality can have significant effects on patients with rare diseases as hindrance can occur when trying to achieve a diagnosis- thus prolonging the discovery of crucial treatment plans. In this essay, I will attempt to recognise factors that contribute to intersectionality and how lack of recognition increases the challenges and obstacles faced by patients, with rare diseases in particular, on a daily basis.

..."I will attempt to recognise factors that contribute to intersectionality"

Although having similar effects on declining patient well-being, this group of autoimmune diseases has varying symptoms that are displayed- depending on factors such as age, gender, and severity of the disease. Autoimmune diseases arise when the immune system attacks the body's own healthy cells, causing damage whilst simultaneously making the body susceptible to opportunistic infections- about 25 percent of patients with autoimmune diseases have a tendency to develop additional autoimmune diseases. The rare bullous autoimmune disorder usually affects the skin and is characterised by the development of red rashes and clusters of fluid filled blisters found across the body. Patients with MMP commonly have blisters residing in the mucous membranes. MMP often starts in the mouth upon developing painful ulcers and can progress to infect the nose, throat, genital areas and anus. Blisters leave scars which cause long term damage, infecting the skin barrier. Severe untreated MMP can develop further into OMMP, experienced by patient Z, affecting the eyes that can result in partial or full blindness. Patient W experienced great levels of discomfort due to her nose constantly leaking and undergoing three surgeries to correct her inverted eyelids caused as a result of MMP. Patients X and Y had red, itchy rashes,

Continued over...

commonly confused with eczema or hives. The itchiness was described as unbearable by both patients and can remain constant in that form. However, BP is more likely to evolve into large painful blisters filled with transparent fluid or blood that can spread across the arms, legs, abdomen, mouth and eyes. The annual incidence of BP is estimated to be between 2.4 and 23 cases per million of the general population but it is rising exponentially to 190–312 cases per million in individuals older than 80 years. A higher annual incidence of 42.8 cases per million population was reported in the United Kingdom.



On a global scale, around 1 in 17 people will have a rare disease at some point in their lives. Studies show that rare disease diagnosis can take 5 to 30 years, depending on the disease and healthcare setting. In the UK, it can take an average of 5.6 years, eight clinicians (including four specialists) and three misdiagnoses before the correct rare disease is identified. The only way to find the best treatment for rare diseases is to increase the amount of research and funding that goes into these diseases. All four patients had been dismissed and misdiagnosed for 6-12 months by general practitioners, despite booking multiple appointments. Many creams, ointments and steroids were used but zero improvement was seen as a range of different medications can have adverse effects that worsen a patient's condition. Furthermore, every patient is different- perhaps in ethnicity or colour causing skin conditions to show differently on people of colour versus white skin- further increasing the difficulty when clinicians attempt to diagnose. Having experienced the shortcomings in the healthcare system, Patient Z and Y decided to undergo private treatment, in which a diagnosis was quickly achieved. Intersectionality is evident in this scenario as these patients were fortunate enough to afford private healthcare thus raising questions about the prolonged suffering for those who cannot. Moreover, patient Z was told he had conjunctivitis, perhaps due lack of awareness that this disease can infect younger age groups, causing the disease to spread over time resulting in severe eye problems therefore contributing to intersectionality and further disadvantage faced by patients. The lack of awareness of the rare diseases caused limited treatment options and patients were given many steroids that were seen as quick relief but caused greater disadvantage as the long-term severity and discomfort of the disease was not solved. Patients not receiving the right treatment at the right time, lacking reassurance from clinicians causing mistrust in the healthcare system further implies that intersectionality causes disadvantages until a proper diagnosis is achieved.

All four patients went through specific challenges on their patient journey and even after a diagnosis was achieved- directly impacting the individual's experience in the healthcare system and society as a whole. All patients had to change their way of living to accommodate their disease. For example, patients had to sleep in individual beds and experienced lack of sleep and also change their bedding constantly due to ruptures of the blisters and the material of certain bedding not suiting their skin. Patients had to accept change when elements of their previously normal daily life were no longer suitable for them. They had to use different clothing materials, changing their diet and different shampoos in order to try and combat the spread of the disease. The patients also faced mental and physical challenges that affected their personal and social lives. Patients described their feelings being hurt when constantly turned away and facing inequalities from the healthcare system- primarily designed to help everyone equally. For example, patients W, X and Y are still shielding as a result of the global pandemic Covid-19. When checking for antibodies, these patients did not have any despite taking the Covid vaccine which meant they had to continue to shield due to their weaker immune systems. The concept of intersectionality does not address the deteriorating mental health of rare disease patients who have had to restrict social gatherings and not see their loved ones. The lack of understanding exacerbates poor mental health. Patient Z in particular describes himself as becoming more introverted. Patients' families were also impacted as their partners had to take on more responsibilities such as doing the grocery shop by themselves- decreasing the independence patients had. Furthermore, Patient Y had to give up a much-loved hobby of group photography and now has four glasses, one of which are used to edit photos due to BP affecting her eyesight. All patients experienced extreme fatigue and caused patient Y to take many breaks when going on a short walk from her house to her garage. As a result, the patients had to overcome challenges caused by intersectionality which put them at a greater disadvantage when being challenged both mentally and physically.

Intersectionality affects rare disease patients as they are forced to live with debilitating systems whilst constantly feeling misunderstood. Statistics and intersectionality highlight the shortcomings in the healthcare system and hinder the ability for clinical staff to understand the impact of this rare disease on people's everyday lives. Campaign group Rare Disease UK found health professionals' attitudes played a major role with 88% of patients and carers saying poor

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awareness of rare conditions by health professionals had a negative impact on their mental health and 80% were negatively affected by health professionals not believing them.

The misunderstandings that are associated with more common diseases, such as dermatological skin issues, also contribute to the challenges patients face. To conclude, studying the concept of factors that contribute to intersectionality highlights that more awareness needs to be achieved to prevent putting people at a greater disadvantage which decreases the quality of their life.

References:

https://resyst.lshtm.ac.uk/sites/resyst/files/content/attachments/2018-08-21/Resilience%20and%20intersectionality%20brief.pdf https://www.thelancet.com/journals/lancet/article/PIIS0140-6736(20)325137/fulltext#:~:text=Medical%20institutions%20need%20to%20turn,7

https://www.gosh.nhs.uk/conditions-and-treatments/conditions-we-treat/chronic-bullousdisease-childhood/ https://www.ncbi.nlm.nih.gov/pmc/articles/PMC6109638/ https://www.ncbi.nlm.nih.gov/pmc/articles/PMC3150011/ https://www.msdmanuals.com/en-gb/home/skin-disorders/blistering-diseases/bulloustypes/pemphigoid https://www.pemfriendsuk.co.uk/bullous-pemphigoid https://imperialcollegehealthpartners.com/new-report-reveals-undiagnosed-rare-diseasepatients-cost-nhs-excess-3-4-billion/ https://rarediseases.org/for-patients-and-families/information-resources/patient-andcaregiver-resource-center/undiagnosed-rare-disease-patients/ https://www.nottingham.ac.uk/research/groups/cebd/projects/blistering-diseases/informingtrial-development-in-blistering-diseases.aspx https://www.nottingham.ac.uk/research/groups/cebd/projects/5rareandother/pemphigus-andpemphigoid-psp.aspx

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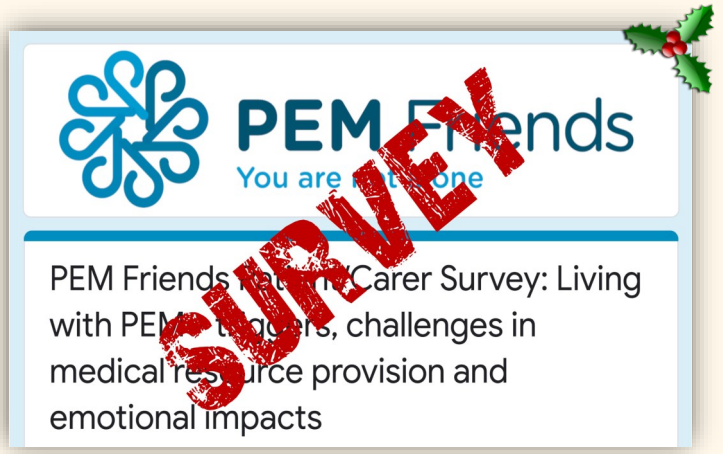
PEM Friends Patient/ Carer Survey - Update

By Andy Heath Ph.D.



This is a quick update about the survey we conducted in the spring. The response was excellent: 101 people participated. The survey was anonymous, so although I don't know who you are, I want to thank you again. There were 35 questions, some in several parts. The outcome was an impressive spreadsheet containing over 4,000 pieces of information. I analysed the data to create a (mainly) graphical report, uploaded it to our website, and sent out a link. I hope you've been able to have a look at the report.

The motivation for conducting the survey was to gain a deeper understanding of our PEM Friends members. The PEM Council has considered the survey report in depth, including a long discussion during our June meeting in Ashton-under-Lyne. We concluded that we had learnt a huge amount but there is more work to do with the existing data and more focussed information to gather. Don't panic! We won't be asking you to complete another huge survey, but we would like to dive deeper into some issues by asking members to complete a few "mini-surveys" at some point. Each of these would be short, probably including only two or three multiple choice questions, and, of course, would be completely anonymous.

I could fill pages about the results, but space is limited so I've highlighted a few points:

Half of respondents reported they received their PEM diagnosis from a dermatologist, but substantial numbers were diagnosed by maxillofacial professionals or dentists. Two thirds said their PEM was initially misdiagnosed and named 35 different initial diagnoses. Only 3% had their PEM diagnosed by a GP. We initiated our Photo Library project partly in response to this very low percentage (developing the Photo Library has taken up a lot of my time which is partly why the survey project is yet to move any further forward than it has).

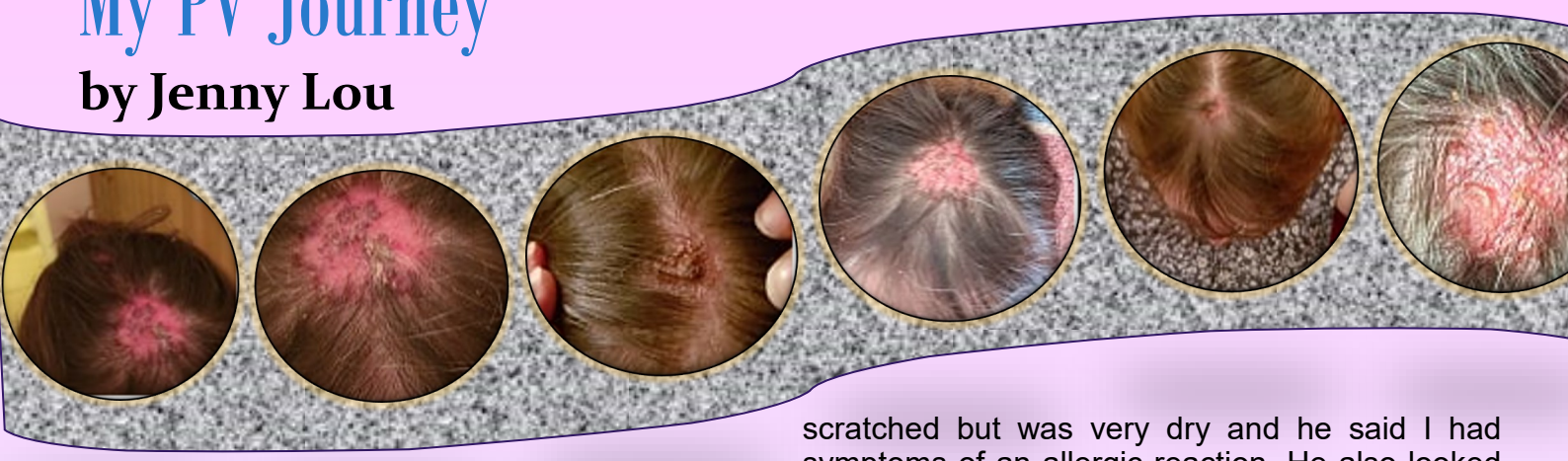
I plotted the approximate locations of respondents on a map. Unsurprisingly there was clustering around some of our bigger cities, the largest cluster being around Greater London. The greatest surprise was that 92% of respondents lived in England. PEM Friends members who responded appear to be very rare in Northern Ireland, Wales, and Scotland.

Most respondents (91%) said their PEM care was overseen by a hospital. Nearly 50 hospitals around the country were mentioned but by far the most popular (23%) was Guys and St Thomas' in London. Other popular hospitals used were Leicester Royal Infirmary, RVI Newcastle, Birmingham Dental Hospital and Oxford Churchill Hospital. Perhaps 'popular' is not the right word to use!

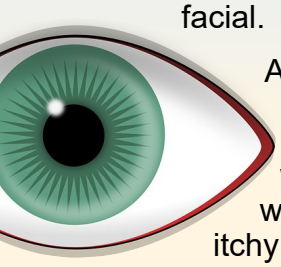
The survey results are proving to be a great resource that allow PEM Friends to better understand how our members live with their illnesses, and tailor what we do to address the challenges. The results also strengthen our voice within the organisations we are associated with. Thank you again to everyone who has participated so far. And I promise any future surveys will be "mini"!

My PV Journey

by Jenny Lou



I wanted to share my journey as it is a positive one and I want to give hope to others. I am not entirely sure when my immune system started to 'play up' – but my body did start to misbehave when I had my third child: I was covered in red itchy spots which perplexed the doctors – was it hormones, stress, bed bugs, herpes, gluten? Blood tests came back clear so they settled on scabies. I couldn't understand this as I shared a bed with my husband, had two children and no one else in the house had any symptoms of scabies. As I was pregnant, they couldn't treat me, so I was left covered in spots and sores and full of anxiety: I didn't want to give birth in a hospital as I didn't want to infect the nurses and I didn't want to hold or breast feed my new-born, as I didn't want to infect her either. Eventually, a lovely midwife made me realise that I didn't have scabies and immediately I felt less stressed and anxious and started to enjoy motherhood. The sores started to heal, the spots disappeared, and I laid the whole experience to rest. Then in 2018, I experienced a bereavement and the itchiness started again: I found myself clawing at my shins and arms and this time, along with the spots, I had mouth sores and lesions on the inside of my cheeks, mouth ulcers that would not heal, sores on my lips, red wounds on back of throat, sore throat (pain when swallowing) and a hoarse voice. The GP suggested it was stress related or hormonal and I was signed off work for two weeks. This made no difference. I decided to try the dentist for advice – and she agreed that it could be 'hormonal' or old age (I was 40) and to rule our mouth cancer referred me to maxillo-facial.



A month later, I went to the optician as my eyes felt dry and it felt like I had something in them. There were blood shot patches on the whites of the eye, and they were itchy. I had been to a pharmacist and tried conjunctivitis medication but had no relief. The optician said the eye was clear and was not scratched but was very dry and he said I had symptoms of an allergic reaction. He also looked at my mouth and said that these were symptoms of a cat allergy and prescribed eye drops to lubricate the eye and antihistamine. I immediately rehomed our cat as coincidentally we had bought a kitten in September this was when all the other symptoms had started. The kids were so upset at losing the family pet – but I was just so desperate to feel better. I still attended the maxillo-facial appointment when it came through and explained that I thought that the symptoms were because of our cat which I had now given away. The consultant asked if I could get the cat back as he thought it was viral and prescribed steroids: 30mg tapering dose over 6 weeks. The spots cleared, my mouth cleared, and we managed to get our cat back just in time for Christmas!

In January 2019, once I stopped taking the steroids, the mouth symptoms returned and this time I developed a small lesion on the top of my head which started to grow!

I again went to the GP who prescribed antifungal treatments in case of ringworm. I then tried a steroid cream, an antibacterial cream and even anti-biotics – nothing worked, and it kept getting bigger and more painful. By April, everything had worsened: receding and red gumline, lesions front of mouth, sore throat, red sores on roof of mouth and back of throat. My Achilles tendons were inflamed, and I had open sores on my breast and arms and lesions on my vulva and inside the vagina. I was always tired and lost my voice. With desperation, I went to see a private rheumatologist who thought it might be sarcoidosis. He prescribed steroids but a chest x-ray was clear, ruling out this diagnosis and a test for Lupus was also negative. I went to my GP again and she said she would refer me to a dermatologist so that they could look at my scalp and take a biopsy. Finally, in July 2019, I was diagnosed with Pemphigus Vulgarus. Steriods were prescribed (60mg), along with calcuim and vitamin d (evacal) omeprazole, alendronic tablets and doxyciline.

In September 2019, the hair loss had worsened and I started on Azathioprine, but ended up in hospital with severe diarrhea and liver problems and so was taken off this medication but had started to suffer with the effects of prednisone: moon face, joint pain, insomina, mania, hyperactivity, weight gain and facial hair and repeated water infections. In November, I was started on mycophenolate and was able to get the steriods from 60mg to 30mg. It was an improving picture until March 2020 when I experienced my worse flare to date.

As a result, the mycophenolate was increased and this caused a resting heart rate of 148 beats per minute and I was hopsitalised again. I was also experiencing diarrhea (6 to 8 times daily), leg cramps, pins and needles and tremors. So this medication was reduced and the steriods increased again however this caused fluid in the lower legs, weak muscles, decreased kidney function, back pain and joint pain which was so bad that I couldn't even do the school run with my children. I just wanted to be a mum to my girls and felt completely broken. Through PEM friends I had heard about a consultant in Leicester and so I asked my dermatologist to refer me.

This appointment changed by life. I was offered Rituximab and three months following this treatment I was blister free and finally able to get off the steriods with out blistering. Fast forward to today and I am only on a minimal dose of mycophenolate and my most recent blood test has shown that my PV antibodies are now very low and so I hope to be tapered off this medication also. My moonface has gone, the joint pain has gone, my hair has grown back and I am completely blister-free!

So that is my journey: it bought me so much pain and unhappiness but has taught me so much more. I am certainly less vain and a more grateful person. I look after myself more and I appreciate those people who showed me love and loyalty through this difficult time. I am now able to put all of my energies into my children, my marriage, my career, my home, my art and everything that I hold dear. It does get easier and there is an end!

YOUR PEM JOURNAL, YOUR FRIEND

by Julie Berger Martin

When I was diagnosed with Pemphigus vulgaris on my birthday in 2011 I was already logging the painful 'mouth ulcers' that just wouldn't heal months before. The diagnosis was bittersweet, but at least I had the answer, and as I started my pem journey my journal was my constant companion where I noted my progress, my test results, my flares, my emotional wellbeing, and of course my meds which was a constantly changing cocktail of tapering and increasing steroids and immunosuppressants.

Jump forward eleven years and I am in remission...drug free since my birthday in 2020! My journal still plays a big part of my journey, so much so that I wrote an article about keeping a pem journal on our PEM Friends website. I urge everyone to keep a journal. Please have a read of the article as it gives handy hints on what to include in yours.

Don't have a journal? PEM Friends has been awarded a grant by the British Association of Dermatologists to create our own dedicated journals for our members. We're working on the design now and plan to offer them to our PEM Friends in the new year. If you are interested in having a PEM Friends journal please let us know as we'd appreciate a show of interest in this project.

Improving Diagnosis One Photo at a Time

by Andy Heath

The PEM Friends Photo Library is a resource that has the potential to help medical professionals diagnose pemphigus and pemphigoid (PEM) diseases. The idea for the library originated in a PEM Council meeting while we were planning this year's activities. We were debating if there were any ways in which PEM Friends might be able to ease diagnosis of our diseases. We realised we might be able to leverage a very useful resource – a potential gold mine of high-quality images from our members showing pemphigus and pemphigoid symptoms at varying stages of disease progression on different parts of the body.

We decided our aim should be to create a searchable library of anonymous PEM images on the web. For instance, a GP presented with a patient displaying blistering on an arm should quickly be able to find all our images of arms. We needed to collect photos, and information about each photo, and devise a way to use the information to search for specific images. My science background was full of software projects, and I love designing this kind of thing.

But creating the library required a lot more than software design. We formed a small project group comprising Isobel Davies, Ingrid Thompson, and myself. None of us knew what a doctor expected from an image in terms of technical quality, or exactly how medical privacy should be respected on the internet. We had ideas about what image information to collect but were not sure how to prioritise it. Subsequently our first decisions were that we needed expert input and resources.

In April we wrote a grant application to the British Association of Dermatologists (BAD) for funding and Isobel approached the Institute of Medical Illustrators (IMI) to enquire if they could suggest someone to help us. We received good news on both fronts: the BAD funded our application and the IMI provided information which led to us to (virtually) meet Sophie Kibbler, a medical photographer based in Cardiff. After a few minutes in to our first Zoom meeting, we realised Sophie was experienced, talented and a perfect fit to work with us. We were very fortunate to find Sophie and she joined our photo project team.

We were still debating what information we required to accompany each image and decided it was best to seek the advice of dermatologists. Isobel's attendance at the BAD conference in Glasgow provided an opportunity, and we received suggestions such as body site, diagnosis, age range, etc from delegates who kindly completed our short questionnaire.

We wanted to be sure we were remaining true to the objective of the Photo Library, which is to aid diagnosis. We contacted Dr Tim Cunliffe, a former GP and consultant dermatologist at Tees Valley Hospital and Dr Sonia Gran of the Centre of Evidence Based Dermatology at the University of Nottingham. PEM Friends has worked with Dr Gran on other projects and Dr Cunliffe is the primary author of a website developed to aid GPs with dermatology, including a general dermatology diagnostic tool. We were very grateful for their feedback: they suggested some subtle modifications but confirmed our ideas were sound.

Our photo project team had many Zoom meetings. Sophie developed a protocol document

that guided patients in how to provide the best photos and gained appropriate consent. She explained how she would ensure privacy by cropping out identifying features from photos if necessary. To ensure the photos would be sent in anonymously, I created a page on the PEM Friends website where people could enter information and upload images. I also did some research into various websites that could manage, filter, and display the selected images and information. It turned out that our own website provider offered a good solution and we upgraded to use some new features.

With everything now in place, we advertised for volunteers to donate photographs in the PEM Friends private Facebook group and via our mailing list. There was a good response and around 50 people replied to Isobel who emailed them Sophie's document and the address of the upload page on our website. The photographs started to come in and after a week the first batch of images and related information was passed to Sophie. Sophie made the selections, edited photos where necessary, and returned the images for me to populate our new secure customised database on the website. After a little more feedback from our experts, we decided how to display the images and which search filters to apply, and the initial version of the Photo Library was published.

We have repeated this cycle several times and at the time of writing the library contains 110 images. The next batch of photographs is currently being processed. Sophie applies the same selection rules to our photos as she does to her professional work. To date, less than 45% of the photos have been chosen for inclusion in the library but it is vital that the images meet standards such as size, focus and quality required by doctors to aid diagnosis.

The Photo Library continues to grow. In addition to the kind efforts of PEM Friends members, we have also received contributions from both Pemphigus Africa and the Association Pemphigus Pemphigoide France (APPF), and we hope that more sources of photographs will become available. We are proactively seeking to extend the already diverse range of images to strengthen any under-represented categories. We are also focused on the task of publicising the library to doctors and will seek advice from our experts and the BAD on how best to do this.

We really appreciate the enthusiasm of the participants who have engaged with this project, and the generosity of the experts. Thank you everyone.

The library can be visited at **http://www.pemfriendsuk.co.uk/photo-library**

FUNDED BY
BAD
BRITISH ASSOCIATION OF DERMATOLOGISTS
EST. 1920 - HEALTHY SKIN FOR ALL

Talking to the regulators about PEM and what it means

By Marc Yale

Advocacy & Research Coordinator at International Pemphigus Pemphigoid Foundation and President of the Board of Directors of GlobalSkin.

We are excited to be holding an Externally Led Patient Focussed Drug Development Meeting. https://www.fda.gov/industry/prescription-drug-user-fee-amendments/externally-led-patient-focused-drug-development-meetings

These meetings were initiated by the Federal Drugs Agency in the USA to help them, and other drugs regulators across the world, to understand the challenges of having a rare disease and what the drug options and difficulties are in treating them. The intent, of course, is to speed up the process of approval for new ways of treating pemphigus and pemphigoid.

We are currently putting together a packed agenda for the workshop on 25th January 2023 so we can best communicate all the challenges of our diseases and are pleased that our friends in PEM Friends are fully involved in creating what will be a very full and interesting day.

Registration will start on 1st December so do attend if you can.

(NB Editors comment) Our own PEM Friends will be speaking too. Don't miss it.

SETTING PRIORITIES FOR RESEARCH INTO PEMPHIGUS AND PEMPHIGOID

By Isobel Davies

It's nearly a year since we updated you about the Blistering Diseases Priority Setting Partnership.

It's been a long journey, largely due to interruptions due to covid-19, but the result of the works will hopefully be published soon. The PSP has been led by Karen Harman, Consultant Dermatologist at the Centre of Evidence Based Dermatology, University of Nottingham, and with the support of the James Lind Alliance,

As we go to press, we are awaiting confirmation that we can pass on these results to you.

The paper is the culmination of a group of medical professionals from different disciplines, and a bunch of patients to agree the top priorities for blistering autoimmune conditions.

US!!

Six of us were a part of the steering committee and many of you have been involved in the input to the list of questions and filtering these down, alongside inputs from clinicians, nurses, GPs, etc. Other PEM Friends joined us in September for a workshop at which we further ranked a set of 17 questions to produce a Top 10. These will be the basis of future priorities for research and will influence future grant applications.

University of Nottingham
UK | CHINA | MALAYSIA

James Lind Alliance
Priority Setting Partnerships

I think those who attended the workshop would concur with me about the level of collaboration, detailed discussion and quality conversations that characterised the workshop. We were told that the level of agreement about the top priorities was very impressive.

You can obtain further information about the Pemphigoid and Pemphigus PSP here.

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

Very soon, though, we will be issuing the letter after it is published.

OUR YOUNG MEMBERSHIP IS SADLY GROWING.

Another lovely little PEM princess joins our Felicity (age 7). Lily is 10 and both pass on their Christmas messages to everyone.

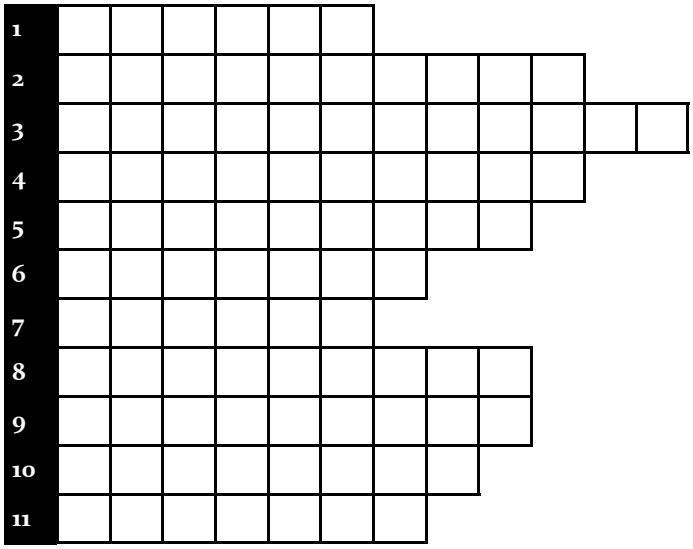
A Christmas wish...

Hi, my name is Lily, I am 10 years old and I have Pemphigus Foliaceus. I would like to wish that everyone has a happy and itch free Christmas!

My wishes for Christmas are that I can reduce my steroids down to none by Christmas, I also wish that everyone with pemphigus gets better or feels better at Christmas. I hope you all are, or start to feel better at Christmas!

MERRY CHRISTMAS! ***From Lily***

Achievement



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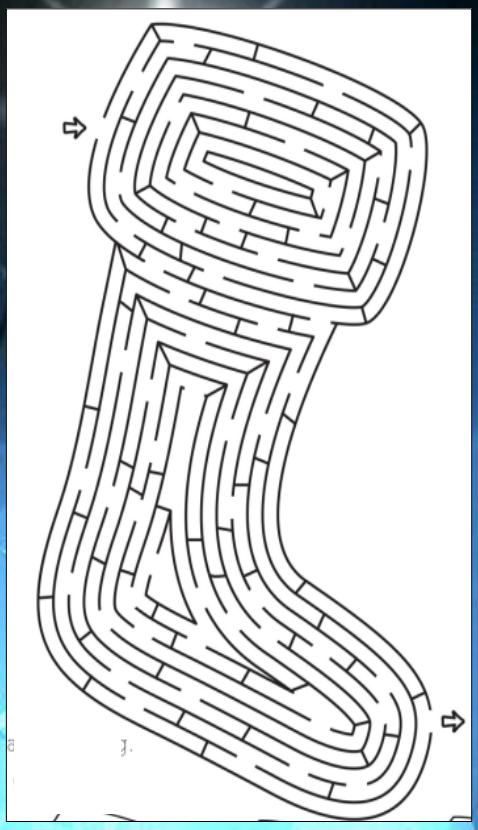
8) Cenelhagl
9) osMlnitee
10) oPrsgesr

Can you unscramble the words?



Can you spot the difference in these two fun Christmas images?

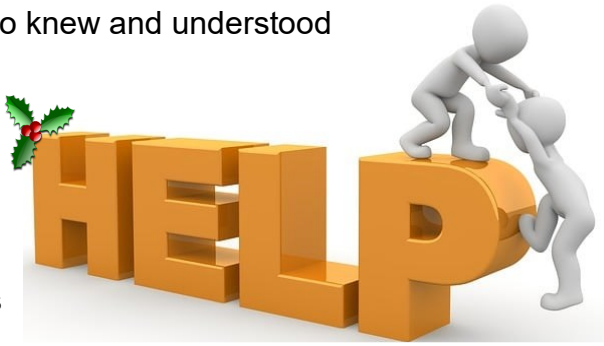
Christmas Quiz

1. Which country started the tradition of putting up a Christmas tree?
2. How many ghosts show up in *A Christmas Carol*?
3. In *Home Alone*, where are the McCallisters going on holiday when they leave Kevin behind?
4. Which country did eggnog originate?
5. How many gifts were given in total in 'The Twelve Days of Christmas' song?
6. What year did Mariah Carey's 'All I Want For Christmas Is You' come out?
7. In which Christmas movie does Tom Hanks play 6 of the characters?
8. In which country is it tradition to eat KFC for Christmas dinner?
9. In the movie *Love Actually*, which character sang 'Christmas Is All Around'?
10. How many of Rudolph's fellow reindeers' names start with 'D'?

QUIZ Answers
1. Germany. 2. Four: former business partner Jacob Marley, and the spirits of Christmas Past, Present and Future. 3. Paris. 4. Britain. 5. 364. 6. 1994. 7. *The Polar Express*. He plays Hero Boy, Father, Conductor, Hobo, Scrooge and Santa. Claus. 8. Japan. 9. Billy Mack, played by Bill

Being a friend to another PEM Friend

By Helen Robinson



I joined PEM Friends to find a "buddy" for myself. Someone who knew and understood how I felt having pemphigus. I was introduced to John who has PV and we have spent the last few years e-mailing each other each week.

It felt like I was thrown a lifeline, someone who understood my fears and my condition.

I then joined PEM Friends Facebook group to find other people with PEM. I realised I knew and understood how new members felt and I could help them, in turn.

Helping others with their questions helps me feel better and useful myself. You don't have to know everything to be a "buddy", but just be able to provide help and guidance. Being there just to listen goes a long way.

Could you be a "buddy" to someone too?

NEWS IN BRIEF...

DERMATOLOGY REFERRALS

We heard a very encouraging presentation at the Dermatology Council for England by Dr Julia Schofield. The process for referrals for Dermatology cases is being completely re-vamped from start to finish. We are hopeful that diagnosis and treatment for our conditions will be considerably speeded up.

More details to follow.

NEW TREATMENTS

We are very encouraged by the discussions we are having with pharmaceutical companies developing new treatments specifically developed for pemphigus and pemphigoid. These are at different stages – some are almost ready to recruit patients for their trials and others are just starting and keen to talk to patients. We will be assisting with all of this, as having drugs and biologics which can resolve our symptoms is absolutely vital.

Noel Mudibo

THANK YOU AND GOOD LUCK

We are grateful to Noel Mudibo in Kenya, who is valiantly trying to set up a patient support group there. Noel has been very supportive in trying to get photos for our photo library of people with darker skin.

An update on research in the area of bullous pemphigoid

Dr Sonia Gran

A team at the University of Nottingham led by Dr Sonia Gran are currently looking at the association between medications/ vaccinations commonly prescribed in older people and bullous pemphigoid. The results are interesting and will be published in 2023. It is hoped that the results will raise awareness among healthcare professionals of the risk of bullous pemphigoid following the administration of certain medications and help patients make informed decisions.

The team involved in this research is multi-disciplinary and includes pharmacists, dermatologists, healthcare of the elderly physician, GPs, methodologists as well as patient partners: Mikolaj Swiderski, Yana Vinogradova, Roger Knaggs, Karen Harman, Rowan Harwood, Vibhore Prasad, Carron Layfield, Grazziela Figuredo, Sue Norris, Ingrid Thompson and Isobel Davies.

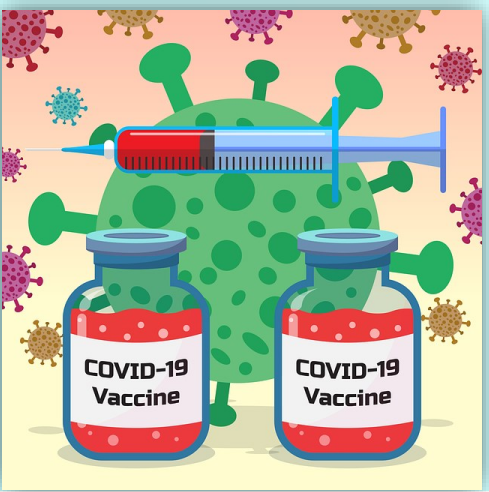
Please watch this space for some interesting results!

Future research

Dr Gran is awaiting the outcome of funding for a project to explore the reasons for delayed, and barriers to, diagnosis in bullous pemphigoid from the perspective of GPs as well as what tools could help GPs recognise bullous pemphigoid earlier.

We will interview 15 GPs, on a one-to-one basis for one hour online, from different parts of England, to explore their views on reasons and barriers associated with diagnostic delay and how to help them identify bullous pemphigoid earlier. We will make sure the GPs come from different backgrounds, types of practices and have different levels of experience. Patient co-applicants will help with writing the interview questions.

From this study, we hope there will be greater awareness of diagnostic delay for bullous pemphigoid amongst GPs, patients and dermatologists. The findings from this study will also contribute to a larger piece of research, involving patients and dermatologists, to help identify bullous pemphigoid earlier. Earlier diagnosis is important as it may mean disease control is possible with less aggressive treatment and improved wellbeing, for example by reducing discomfort from blisters.



Dr Gran is also keen to do some work on the association between the Covid-19 vaccine and bullous pemphigoid. She has applied for some funding and we are awaiting the outcome. An update will be provided in the next news letter!

She would like to thank all the patient partners who work with her and inspire her to do important research that matters to patients. If you are interested in becoming a patient partner and working with Dr Gran and her team please feel free to contact her: sonia.gran@nottingham.ac.uk

Working with other groups

One of the marked changes in in the past few years has been the extent to which we have engaged with and been helped by other organisations. Our relationships with (and gratitude to) many of these friends continues to develop.

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. This is explained further in the article in this edition of the magazine, by Laurence Gallu of the APPF.

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. GlobalSkin also has a European network to which we are active contributors.

Beacon (formerly known as 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.

PEM Friends are also a part of the BAD PSG (British Association of Dermatology Patient Support Groups) and Dermatology Council of England groups, which enable us to keep up to date on changes in Dermatology practices in the UK and also give us access to their grant funding, another benefit for which we are very grateful.

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

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

December 2022

Editors: Trina Harris & Isobel Davies
Design and layout: Trina Harris
Proof reading: Peta Howell

Website: www.pemfriends.org.uk
Facebook: https://www.facebook.com/groups/

Contact Information for next issue:
Isobel Davies: isobel@pemfriends.org.uk
Trina Harris: trina@pemfriends.org.uk

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Wishing all a Happy Christmas & a healthy Happy New Year