

PATIENT STORIES - CONFERENCES - GRIDD SURVEY

PEM Lives

ISSUE No: 7
SUMMER 2023

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

Inside
Finances
Research
Patient Stories
Kids with PEM
Advice from the experts
Priorities for research
London Marathon 2023
And so much more...



PEMPHIGUS AND PEMPFIGOID

PEM Friends

You are not alone

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PEMPHIGUS AND PEMPHIGOID
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 summer edition of the PEM Lives magazine.

WELCOME

...to our Summer edition of PEM Lives.

Many thanks to Trina for her perseverance and talent in putting together another riveting read, as well as our gratitude to all the contributors who, I hope, will make you smile, laugh, cry and want more.

In this edition, you'll find updates from some of the many things PEM Friends have been up to so far in 2023.

We can all take heart from some of the stories relayed by people like Roshni, Karen and Ana-Paola, as well as articles about the role that families and friends play in supporting us through our diagnosis and treatment. As both Gail and Helen say, it's often our long suffering partners who help us find the path forward. We should celebrate their support in particular. One such partner is Chrissie who has been there, cheering on the heroic Mark in his Marathon challenge.

I only have to read the letters from little Felicity and Lily to see how positive and courageous these little stars are (not to mention their Mums).

We are increasingly helped by other groups and individuals too, such as the work done by the Centre of Evidence Based Dermatology at Nottingham and some very dedicated and knowledgeable experts across the UK and beyond. All we need now is a few more of them!

Last but not least, I would like to celebrate the contribution you all make. I am so proud of what we have achieved together when I look at the Facebook posts or attend a weekly Zoom. Your compassion and care for each other is exemplary.

So I hope you'll all try to make it to our inaugural Annual Meeting on 9th September, whether by Zoom or Face to Face. I can't wait to meet more of you then.

By the way, if you were wondering, I'm a doggy person

Happy reading.

PEM Council Report

By Isobel Davies

I hope you enjoy our most recent PEM Lives magazine. As ever, it is packed with all sorts of information and chat. Don't forget you can get a hard copy version if you let us know your address via subscription – details of how to do this are below.

2023 has been as busy as ever, despite saying at the start of the year that we would minimise our

activity due to the challenge of resources. We always pack a punch, despite having only a small band of volunteers who are struggling with their own diseases and little incoming funds.



On the last note, we are in the debt of several people who continue to donate to our work. An extra special thanks goes to Mark, who ran the London Marathon and collected over £2200 for us. We are also very grateful to the lovely Marjie who has done car boot sales and donated from those and from her own reserves. Sue and Derek also participate in a European patient group who are working with Janssen on their drug development programme on Bullous Pemphigoid and have generously donated their fees for this work to PEM Friends. We owe a debt of gratitude too, to the mourners attending the funeral of Kath Lonergan who donated to PEM Friends thanks to Kath's daughter, Sarah.

We also appreciate those PEM Friends who donate on a regular or ad hoc basis. All this helps us to produce the magazine, attend conferences, run Zoom meetings, keep the website going and do the work to make these awful PEM diseases more easily diagnosed and treated with greater care and understanding.

£ Andy has helped us get a clear view of our incomings and outgoings and we have a good idea about our financial capacities. However, most of what we do is done through the hard work of members of the PEM Council and others. We have a packed website, thanks to Ingrid's and Andy's efforts, a very effective Facebook group which is carefully managed by Julie, and Kal has been trying to get our new bank arrangements set up. We are moving from HSBC to NatWest, who offer a much lower cost service.

We are getting ourselves into a position that we can operate as effectively as any charity but

without the bureaucracy, costs and other overheads to slow us down. As part of this, we spent many hours at the start of this year, ensuring we are compliant on General Data Protection Regulations. You may have seen us ask for those of you for whom we hold addresses on a secure Excel spreadsheet to add their details onto their subscription so we can delete all records not held on that master list. We will still hold a small number of contact details for PEM Friends with no access to a computer.

We are also grateful to Mark for his review of our Facebook Group membership. Auditing our Facebook membership from time to time ensures we can safeguard our member's privacy and security, although we are very careful about admitting new PEM Friends into the group.

Another accomplishment this year is the fulfilment of an ambition I have held for many years. We now have a PEM Advisory Group.

Following the advice of the wonderful Professor John Dart, we approached a number of medical and other specialists with trepidation, inviting them to provide us with advice and guidance.

They were all thrilled to be asked. We had an introductory meeting last month and the next session is planned for August, just before our annual get together of the PEM Council, followed by a UK wide PEM Friends bash. The latter is yet to be designed but will be the first time we have been able to invite every single PEM Friend to come together to discuss topics relevant to us all. This meeting will be both via Zoom and face-to-face for anyone wanting to join us in Croydon.

We are hoping to win a National Lottery Community Grant to fund this meeting which will be held on 9th September.

On the subject of grants, Mark was instrumental in helping Professor Rauz at Birmingham Eye Hospital when he provided Sight Research UK with a detailed article about his Ocular MMP.

On the conference front, Trina is attending the GlobalSkin

Annual meeting in Brussels, which gives us a chance to learn what other organisations are doing and share



Continued over...

LIVERPOOL



British Association of Dermatologists
103rd Annual Meeting
27th-29th June 2023

ideas and connect. At the end of June, several PEM Friends - Martin, Amber, Alison,

Roshni and Pauline have volunteered to 'man' the stand at the Annual British Association of Dermatologists (BAD) meeting in Liverpool. This is a great opportunity to build awareness amongst the many UK Dermatologists who will be attending.

One of the highlights of this year was the production of the Photo Library. This project was funded by the BAD and we had the great pleasure and luck of working with a young Medical Photographer, Sophie, who used the opportunity to further her own qualifications. We were also helped by the support of Dr Tim Cunliffe, who chairs the Primary Care Dermatology Society (PCDS) and who also wrote the diagnostic support element and connected us to his own organisation's diagnostic tool. Sonia Gran, a good friend of PEM Friends from the Centre of Evidence Based Dermatology, also provided very welcome support, as always.

Andy has worked tirelessly on the Photo Library and many of you have input your images. There are over 160 rather scary images on the pages but we would still welcome more, especially from MMP patients and people of colour.

Dentists, names and addresses provided by you. Please let us have more contacts who should know about the library. These link to the website and will, we hope, help in reducing diagnostic times by showing medical experts what PEM looks like. Andy was invited to write an article about the Photo Library for the next PCDS Bulletin which is due to published soon. The bulletin has an audience of around 6,000 medical professionals.

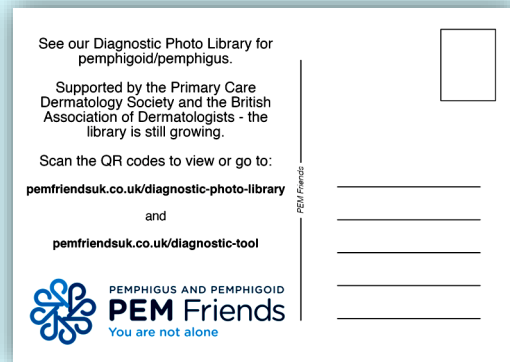
Julie is working on a PEM Friends journal, also funded by the BAD, that we will provide to new members and to anyone who wants to ensure they keep a record of the progress of their disease. Despite receiving BAD funding, we are challenged by the cost of producing something that is of the quality we aspire to.

Our regular Zoom calls on a Thursday continue to offer friendship and advice to new and 'old' callers, and we hope to see more new PEM Friends join to discuss matters that help deal with the challenges our diseases present.

Possibly the most important contribution to all we do though, is the effort that goes into producing these PEM Lives magazines. It is no mean task to collect articles, collate and design them, get them published and then distribute these documents. We do know, though, that it is worth the effort. So, a HUGE thank you to Trina who manages to get these done despite her own challenges. Well done, Trina.

To subscribe (or edit your subscription details), go to <http://eepurl.com/hoye09>.

If you already subscribe, it should recognise you and tell you how to edit.



We sent around 150 postcards out to Doctors and



Please let us know if you are willing or able to contribute a little time



to help PEM Friends.

Summer of 2020 - 'all change'

by Roshni Maher

It was summer of 2020, the height of Covid and lockdown. I was sat in my gynaecologists office in West Middlesex Hospital, 7 months pregnant, covered in painful blisters and crying.

"Why isn't anyone helping me?" I said. I was already going through a difficult pregnancy, with fibroids causing me to bleed every day.

My symptoms had started a good 10 months prior to this, but because of Covid there were no face to face dermatology appointments and my GP just kept prescribing different creams, not knowing what the problem was.

Luckily that particular gynaecologist got on the phone to dermatology straight away and demanded a face to face appointment and biopsy for me. Once this was done I was finally diagnosed with Pemphigus and little did I know it was the start of a very complicated and painful couple of years. They started me on a very high dose of prednisolone straight away and gave me Dermovate cream to help with the itching. I was lucky it was lockdown and I was working from home as it was so painful for me to wear any kind of clothing that aggravated the blisters. Showering was also painful, my skin felt like it was on fire every time the water hit it. The effects of the prednisolone were challenging, they caused me to develop steroid induced gestational diabetes and I then had to go on a strict diet and was prescribed metformin to reduce the blood sugar levels. We were also in the middle of renovating and moving house during my pregnancy and lockdown, the combination of everything really did test my stress and patience!

My daughter Anushka was born healthy and happy on 18th August 2020 and was showing no signs of Pemphigus. We were in the new born bubble and my blisters had started to ease. However every time I tapered down my prednisolone, I would get a new flare. Six months after giving birth I had an operation to remove my fibroids which were huge and still causing me to continuously bleed. They measured 11cm and 9cm when removed and the effects of the operation caused another flare. Later in the year my dermatologist put me on

immunosuppressant medication. This caused me to feel constantly unwell with coughs and colds as it had further weakened my already delicate immune system. Fast forward a few months into 2022 and I had developed what I thought was a really sore throat. I looked into my mouth and my skin was red raw, it was painful to eat or drink anything except for water. My GP thought it was an infection and put me on antibiotics but this didn't help. The soft tissue inside my mouth was falling off every time I brushed my teeth or tried to eat and the skin in my genital area had also started falling off, making it very painful to go to the toilet or even walk or sit for long periods. It was at that moment I realised the Pemphigus had spread to my mucus membranes. Due to not being able to eat properly, I lost 7kg and was very weak. I made an emergency appointment with my dermatologist who ramped up my prednisolone again and discussed the option of Rituximab treatment with me. She also recommended that I don't go to work and luckily I had very understanding management who reduced my hours and allowed me to work from home.

I had done a lot of research on Rituximab and read about positive experiences from PEM Friends and decided to go ahead. I had my first infusion in April 2022 and my second infusion two weeks later. I knew it would take a while for the medication to start working. However in late May I was feeling very unwell, my temperature was hitting over 40 and one day I just couldn't get out of bed. A call with 111 instructed me to go to A&E for a check up...I came out of hospital 6 days later! I had developed neutropenic sepsis, possibly because of the Rituximab or possibly just a coincidence. I remember the doctors looking at me asking how I was still walking and functioning, my neutrophils were at 0. This was when I realised how dangerous and life threatening sepsis really is. They had done every single test but couldn't find the cause of infection. I was being given antibiotics every 4 hours via a drip and my blood

"because of Covid there were no face to face dermatology appointments"



Continued over...

tested several times a day to check whether the neutrophils had gone back up. I really did feel depressed in hospital, the rituximab was supposed to help me, not make me more unwell. Five months later in October lightning struck twice, I was in hospital again with the same neutropenic sepsis and this time it caused me to develop health anxiety. I was constantly scared the same thing would happen again and if I felt slightly unwell I would check my temperature and heartrate every hour. I feared I wouldn't be able to look after my daughter properly or live a normal life. I got sign-posted to IAPT for some CBT for anxiety and this really helped to calm me down and see things from a different perspective.

Despite the sepsis, Rituximab is the treatment that really did put my blisters and pain at bay and I really do hope it stays this way. I currently have no blisters or skin involvement and I am not on any medication. I am also 15 weeks

pregnant with my second child which really is a blessing and miracle considering we had agreed to have no more children when I was so unwell and in pain. PEM Friends has been an absolute lifesaver for me. The knowledge, advice and friendly ears have been second to none and it really helped me knowing that I wasn't suffering alone. For anyone who is starting their journey with Pemphigus please remember that you aren't alone, we are all here to support you and never think you are asking a silly question, we have all been there!



**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

Helping family understand

By Gail Windows



I was finally diagnosed with Pemphigus Foliaceus just under a year ago, after being misdiagnosed for 5 years. After many months, on 26th January 2022, the health company I had been seeing, told me I had Acantholysis. Whilst researching Acantholysis I found Pemphigus and read that it can be fatal. You can imagine my husband's worry seeing that! Mine also! The same day my husband found PEM Friends, who I can honestly say have been amazing for advice and support. They directed me to the specialist I now see. We are both so glad that we found PEM Friends!

I didn't speak to my family about my diagnosis until I saw the specialist and had it confirmed. My father had skin cancer many years ago, and everyone presumed (as I did) that it was the same for me. Because the treatment causes immune suppression, I told my sons. I really didn't want to do this because I did not want to worry them. However, I knew that if they were to pick up an infection then that could have serious implications for me. I have 6 grandchildren, I didn't want to worry them especially because we sadly had 4 family losses in last few years. Because of Covid, my 9 year old granddaughter was worrying about illnesses and whether we could die, so we decided not to tell her. Instead we told our grandchildren that I had a 'nasty rash' on my head and this is why I wore hats all the time.



It's not easy telling family or friends. My Mum still does not really understand it. She is 88 & I used to be her carer. Due to her disabilities and constant infections I decided to get paid carers. This was very difficult for her to understand at first and it's taken some time for her to accept it. She often asks when will I get better and when will it go away? My Dad did everything for my Mum when he was alive, but when he passed away that baton was passed to me. There were many upsets and arguments between us as she would get angry at me for not doing the things I used to, this would cause me to stay away altogether for several days because the stress of it all caused flare ups. I told her I couldn't help her like before because I'm constantly exhausted. She still struggles to understand, but has accepted it. Other close members of the family did not believe it, to the point they checked it out! To this day they have not asked how I am or if they can help with Mum in any way. However, my Aunt & Uncle are very supportive and always check up on us both.

My son's? Well, they know I have this disease but don't talk to me about it. I think it's been hard for them. We lost my father-in-law, then my niece & her baby, and then my dad all within 3 years. Suddenly their mum is diagnosed with this. My son, who lives locally, does more with my Mum. He visits often, takes her shopping and spends time with her, which gives us a break. My other son, who lives in Devon, brings my granddaughter up more to see us, knowing how tiring it is for us to keep travelling there. Unfortunately, when we were staying with them just after Christmas I had bad side effects from the first treatment I had. This worried them and made me feel guilty that it upset all our Christmas plans. Now I tend to put on a 'happy face' so they don't worry.

The main person affected is my husband. I see the concern in his face. On bad days I can't do much. I've always been an active person and I felt young. I decorated, gardened, played with the kids, in fact many didn't believe I'm 68! This illness has made me feel older. My husband sees the changes in me more than the family do. He worries about me and in turn I worry about him worrying about me! If I have a bad day I get tearful and escape to my computer room to do some work. I will have a little cry. He knows I do this but it's something we don't talk about. I hate being the cause of his worry. He tries to do too much for me, in a way. I understand why he does - he sees how tired and breathless I've become - but I want my own independence too. If I can do it I will! We have learnt to alter our ways.

On a positive note it's made us relax more! We sit in our garden more & enjoy the birds that nest and enjoy our new lodger Harry the Hedgehog! My husband still shakes his head over the newly installed hedgehog house & hedgehog highway I got him to make! But these things have become our new way of life, a shared time together that we can enjoy & not be thinking of the blood tests I have to have & the visits to the hospital that on other weeks seem to dominate our lives. He recently has had to have a few hospital appts of his own & today said that he now understands why I get so fed up with it!



Continued over...

People react in different ways when it comes to their loved ones having illnesses, some can deal with it, some can't but over time we all learn our own ways to deal with it. My sons may not talk about it, but they help without stating it, quietly doing things that they know help my husband and me, without making it obvious. Being an independent woman, I appreciate that!

There is a saying 'not all disabilities are visible'. It's the same with Pemphigus in a way, unless the blisters are in visible areas, then you look 'fine', nothing wrong with you! Not everyone will understand your condition, but as long as your family & true friends do, that's all that matters!

My holiday plans this year

By Alison Hand

Hi, I'm Alison and I have MMP - luckily for me it's always been described as 'mild' (although it didn't feel that way at the start when I was unable to eat due to the pain, but apparently it was!) It's currently well under control without any meds other than the odd dose of steroid mouthwash now and then. Thankfully this also means I don't have many restrictions on my life, which is a good thing, as I'm a single mum working full time in the NHS with a pre-teen child, a dog and 3 guinea pigs to look after! It's tiring work, so I very much look forward to my summer holidays each year, especially our annual camping trip.

I'm part of a single parent travel club, and every year in August approximately 100 of us (single parents and children of all ages) head down to a farm site near the sea in Dorset, fully loaded up with pets and camping gear for a week of community living. The kids are always desperate to see their friends from previous years and disappear off into the haystacks to play as soon as we arrive, while the grown ups help each other put up tents and provide much needed wine and adult company.

In the daytime there are trips to the nearby beaches, visits to local tourist attractions like National Trust properties or cider tasting tours, water park or activity centre sessions and even long distance hikes to join for the people feeling energetic (those ones aren't for me!). The campsite itself has activities such as circus skills for the kids, yoga and massages for the adults, a sauna and Turkish bath, a large shop to buy groceries and camping supplies, and a food court with plenty of locally sourced food options if you don't feel like cooking.



In the evenings, groups get together to eat tea (although you'll often find your kid has already managed to get themselves fed at someone else's tent), and as it goes dark the campfires start to spring up - there are always many to choose from, with marshmallows to toast, gin to drink, films shown on projectors, shooting star-spotting, and friendly faces all round keeping an eye on the kids so that everyone can just relax and have a good time.

It can be hard going on holiday as a single parent, especially with illnesses thrown into the mix, but when surrounded by people who know how that feels and are happy to offer a helping hand, there is nothing better than a stress-free week in the seaside air to keep the PEM at bay!



Saturday 9th September 2023



Our first PEM Friends get together.

We will be sending a formal invitation nearer the time, but please put the date in your diaries.

You can attend in person or via Zoom.

The venue will be in Croydon.

We hope to have plenty of discussion about our plans for the future as well as a chance to talk about topics that are very relevant to many of us. It will also be an opportunity to see friends from our group in person (or as near as zoom can offer!)

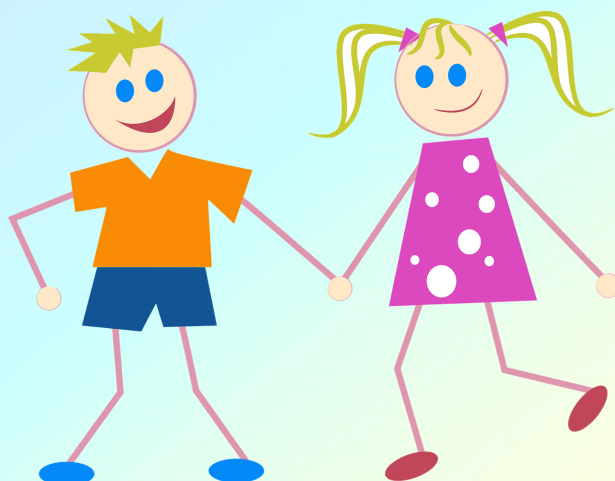
If you would like to be involved in designing the event or simply have a suggestion about what you would like to see on the agenda, then please get in touch. It's early days yet, so plenty of time to input your thoughts.

Children with Pemphigus or Pemphigoid

Pemphigus and Pemphigoid are such awful diseases, but it seems even worse when they are encountered by children. We seem to be seeing more of these cases just recently.

Since these are rare diseases and even rarer in the young, the guidelines for their treatment are unavailable and Dermatologists are left to deliver treatments that, even in adult bodies, can incur trauma. We recently heard about one young adolescent who has been diagnosed with steroid related osteoporosis.

We are trying to find a Dermatologist who has some interest in this area but, in the meantime, to help the parents of these youngsters share information and support, we have set up a Facebook group called "Parents of Children with Pemphigus or Pemphigoid". It is only small and has members from several countries including 3 of our own UK PEM Friends. We hope it will provide some help.



Letters from our Kids with PEM...

Felicity explains in her own words the difference between dogs and cats.

"I love cats and dogs they are both my favourite because I really love animals. I love my dog Eddie. He wags his tail and tries to talk to me! He likes to go to the park and eats his dinner really fast!"

Love Felicity x

Hello my Pem friends, it's me Lily. I made this super last minute because I've not been feeling myself lately. Is it just me or do any of you get blisters in your inner thighs from where your legs rub together?

I'm getting worried about my skin and blistering in the really hot weather because one of my key triggers is the sun because that is what started it all for me last summer.

My doctors have really looked after me, I got my PF diagnosis really quickly, Saleem (my dermatologist) when he met me did a biopsy straight away and sent it for special testing for pemphigus. Mum and I think he is amazing and a superhero. My skin is now clear, except for a few outbreaks and my hair still has 'crusties' the skin on my scalp is a bit scaly and yellow.

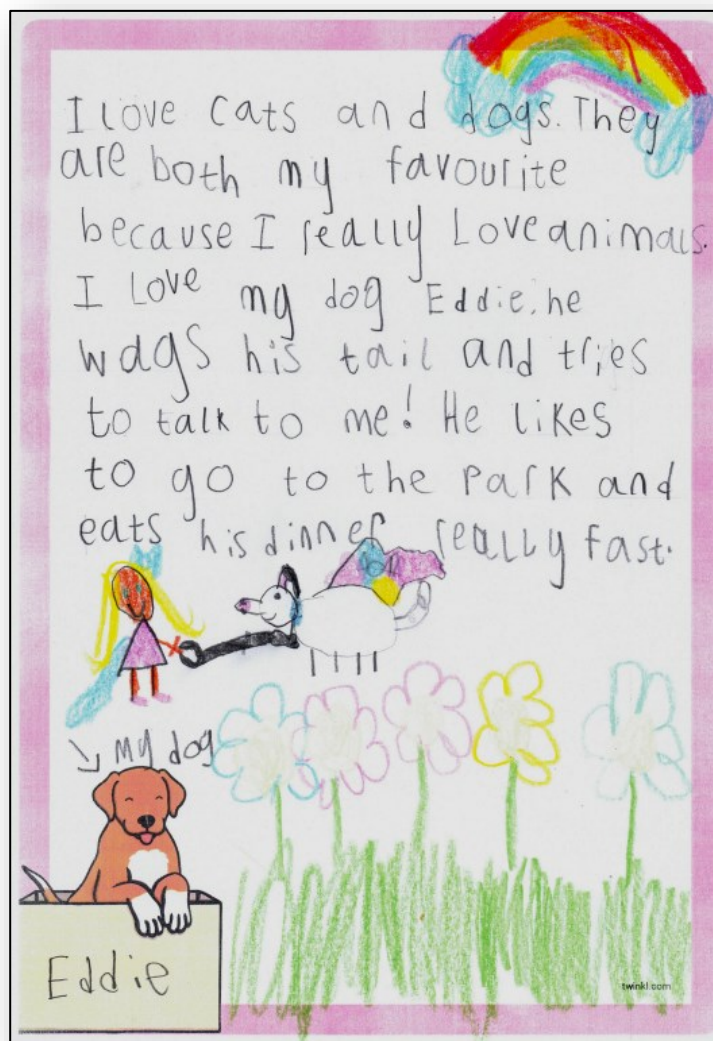
Saleem started me on lots of prednisolone (40mg) and Methotrexate (10mg) weekly. I'm now down to 1mg prednisolone every other day and MTX weekly. The problem now is it is making everything hurt, especially walking. I'm having an MRI on Thursday to see if they can see what the problem is.

I'm still going to school every day that I can, and am very happy that it's half term holiday. In school they let me wear my own special uniform that is comfy - leggings and a soft t-shirt so that it doesn't rub. Normally we have to wear shirt and tie or a summer dress now.

On World Book Day we had to go as a scientific word, I chose 'Pemphigus Foliaceus' and dressed as a Doctor/Dermatologist, what I want to be when I grow up. I like raising awareness and making people know about us. Oh, I also got my official diagnosis for autism in March, what a year!

What are your wishes for this summer? I know mine definitely is to be blister free, to go on my holiday to Disney that was cancelled last year when I was diagnosed and for all of us to not get anymore blisters.

Lots of love from Lily x



PEM Friends meets the US Food & Drugs Administrators

By Isobel Davies

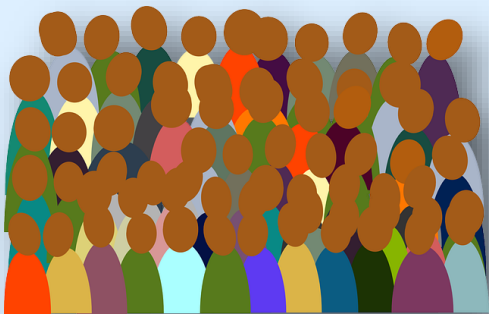


In 2022, we were invited to join the IPPF in setting up a conversation with the US Food and Drugs Administration, the regulator and approver for new drugs in the USA.. This was an event going by the natty name of an Externally Led Patient Focussed Drug Development meeting, an EL-PFDD. The intent of these sessions was to be able to explain to as many members of the FDA as possible about our diseases, how they affect us and the implications of current treatments on these. We also talked about our hopes for future medications. The outcome is that any new treatments will be proposed to a regulatory audience who understand the context.

We leapt at the opportunity to participate, as the US FDA is recognised as a world respected regulator of new drugs and our views that this matters in the UK were confirmed in the recent budget, when Jeremy Hunt said that the rapid approval of medicines will be introduced, confirming that the MHRA (Medicines and Healthcare products Regulatory Agency) is *"exploring partnerships with trusted international agencies, such as in the US, Europe and Japan to provide simple, rapid approvals for medicines and technologies that have received their approval from 2024"*. The chancellor stated that the MHRA would move to a new model to allow *"often near automatic sign-off for medicines and technologies"* that have already been approved by regulators in jurisdictions such as the US, Europe and Japan.

Preparing for the EL-PFDD which occurred on 25th January, 2023 took a lot of time with the organisers, the IPPF, supported by a representative from the FDA and input from the APPF (our French friends) the Japanese PEM Patient organisation and ourselves. There was also a survey to which many PEM Friends contributed.

The meeting had inputs from many patients about their disease and the treatments they received for them. Our own Andy, Mark and I did short videos and Trina and Ingrid presented "live".



In the audience were patients, FDA representatives, drugs companies, doctors and anyone with an active interest in pemphigus, pemphigoid and treatments now and in the future.

What a day it was! It was uplifting, distressing, sad, joyful and a very long but worthwhile day. We left knowing that the FDA had heard everything we can say about the traumas of Pem. It wasn't easy to know how many PEM Friends watched the sessions or the recordings which followed, but I hope there were many among the hundreds in the audience.

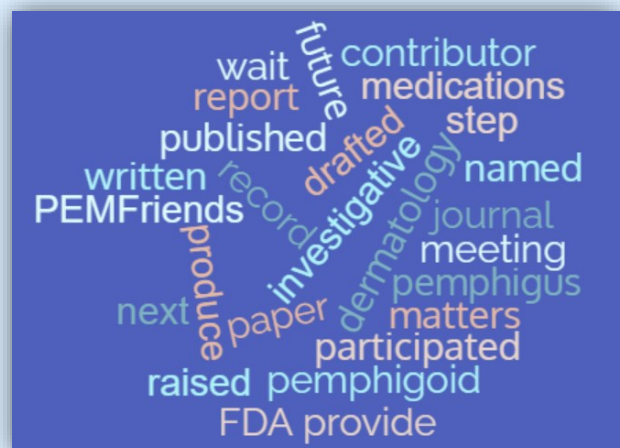
Additionally, there were presentations by 4 eminent experts in pemphigus and pemphigoid from around the world.

Aimee Payne MD PhD and Victoria Werth MS MD are from the USA, Professor Pascal Joly is from France and Professor Dedee Murell from Australia all spoke about the diseases and the present and future medications for Pemphigus and Pemphigoid.

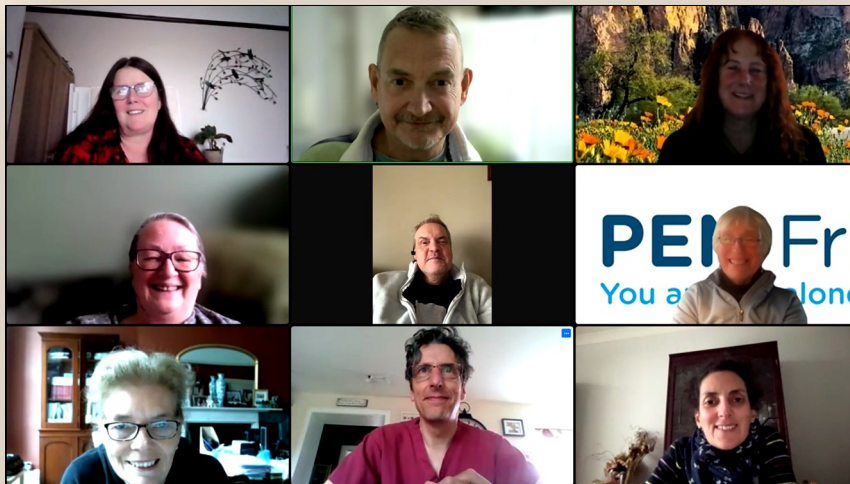
Following the meeting, these 4 specialists have participated in a paper, drafted by Aimee Payne, which is to be published in the Journal of Investigative Dermatology. PEM Friends has been named as a contributor and we wait with bated breath for it appearing.

The next step is to produce a report, currently being written, which will provide the FDA with a record of all the matters raised in the meeting.

You can watch all or some of the EL-PFDD here.
<https://www.pemphigus.org/el-pfdd/>



By Isobel Davies



From left to right: Trina, Andy, Julie, Ingrid, Mark, Isobel, Professor Jane Setterfield, Dr Tim Cunliffe, Dr Sonia Gran

As mentioned in the PEM Council review, we now have a group of experts who can provide advice and guidance.

There are occasions when we haven't known exactly what to suggest for a specific troublesome case or when we have seen a pattern emerging that would benefit from knowledgeable input. Likewise, we develop our plans on the basis of our best understanding of our PEM Friends' needs. We hope that the overview of people with a medical or research perspective will add a much better context to our objective setting.

Members of the PEM Advisory Group are:

- Professor Jane Setterfield
- Professor John Dart
- Dr Tim Cunliffe
- Dr Sonia Gran
- Dr Roddy McMillan
- Mel Westmoreland
- Mr Guri Sandhu

There will be more detail in our next issue of PEM Lives magazine with a short profile of the PEM Advisory Group members.

Supporting One Another

by Helen Robinson

Having an auto immune condition that no one has heard of, for me Pemphigus Foliaceous (PF), isolates you. You go through strange and painful happenings to your body, thin skinned blisters all over your body that constantly burst, soaking your clothes and having a very red and crusty face for everyone to see. All the time being itchy and in constant pain, feeling like a burning sensation. I was on high steroids for 5 to 6 months, my symptoms calmed down and they took me off my steroids really quickly and my body suffered the effects. Constant pain both mentally and physically and by the end of my first year of having PF all of me couldn't cope and I had to leave my preschool which I loved, and at the age of 55 that was the end of my working life.

After 3 months off steroids my PF came back with a vengeance and it all started again.

My husband found PEM Friends and they very kindly found another member with PV for me to email. Without him I honestly don't know how I would have survived. We constantly messaged each other and found out about each others symptoms and treatments, how they effected us and how we were coping with life, doctors and hospital visits. We met up in a cafe on our journey from north to south to visit our family for new year. It felt so good to meet in person and not feel totally alone with our condition and we talked and talked.

That was 5 and a half years ago and we still message each other every week.

A while after meeting him, Mark helped me join the Facebook page and I met a few other people with PF like me and lots of other people with PEM, it was like having a big family to share everything with and is still wonderful. I then joined the zoom meetings which has really helped too.

Over the years I have been able to help other people with PF by sharing my experiences and knowledge of how this works and have learnt from them. It's wonderful to be part of this PEM family.

Research in autoimmune blistering diseases starting this summer!



By Dr Sonia Gran

Dr Gran and team at the University of Nottingham have recently received funding from the National Institute for Health Research's School of Primary Care Research for a project to explore the reasons for delayed, and barriers to, diagnosis in

Autoimmune Blistering Diseases (AIBDs) from the perspective of GPs as well as what tools could help GPs recognise AIBDs earlier.

We will interview 15 GPs, on a one-to-one basis for one hour online, from different parts of the UK, 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 AIBDs. 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.

The team includes Dr Laura Howells, Dr Vibhore Prasad, Dr Karen Harman, Mrs Ingrid Thompson and other members of PEM Friends.

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 team please feel free to contact her:



University of Nottingham
UK | CHINA | MALAYSIA

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My most positive hospital experience

by Helen Robinson from Alston, Cumbria

Six years ago I started with Pemphigus Foliaceous, PF. At first a small patch on my shoulder and on my scalp. My local doctor was really helpful and tried lots of things which didn't work. So he sent me to Carlisle hospital, I had a biopsy and saw 3 dermatologists over the next few years, who all left. I was put on steroids and Azathioprine and I was told I would be seen at a nurse led clinic, which didn't ever happen. I was in a bad way, physically, mentally and emotionally and had to leave work. My husband Mark found PEM Friends for me and to begin with I emailed with another person with PV. After a while Mark persuaded me to join the Facebook group which led to me joining the zoom meetings. On my first zoom meeting I met Caroline and she said to go to the RVI in Newcastle and my doctor referred me straight away.

My most positive hospital experience happened on my first appointment with Dr Phil Hampton at the RVI. After sharing my experiences of the last 4 years and showing him my photos. He told me I was going to have Rituximab infusions, after having my Covid vaccinations and asked me to stay. I was photographed, had 12 vials of blood taken, a skin biopsy taken from my back and a chest x-ray. 2 hours later, exhausted and my head spinning, I was back in the car with Mark not quite believing what had just happened to me. The tests were fine, my local doctors gave my vaccinations and I had my Rituximab infusions in Newcastle. **That was 2 years ago now and I am blister free and so happy to have met Caroline and the wonderful Dr Phil Hampton at the RVI Newcastle.**

T	N	S	A	N	D	S	I
S	G	F	I	S	H	T	J
H	U	H	W	A	V	E	S
E	D	O	L	P	H	I	N
L	Z	F	E	T	D	P	L
L	S	E	M	S	U	N	N
S	Z	B	W	H	A	L	E
J	H	G	B	E	A	C	H

A day at the beach

Can you find these words in the wordsearch?

BEACH	SAND
DOLPHIN	SHELLS
FISH	SUN
WAVES	WHALE

“A smile is like a sun ray. As the sun ray gives life to a seedling, so does your smile gives energy and good feelings to the people you meet.”

Summer Quiz

1. Which British seaside attraction is 158 metres tall?
2. At 1.34 miles, where is the longest pleasure pier in the UK?
3. Which group had a Top 10 hit with Summer Sunshine in 2004?
4. What was the name of the ITV holiday show fronted by Judith Chalmers which ran from 1994 to 2003?
5. Thunderstorms are most likely to hit the South East of England. True Or False?
6. When does summer official start?
7. The month of July is named after which famous historical person?
8. What British show starts every year in August and ends just before Christmas?
9. Which month in Summer are most babies born in Britain?
10. 'The First Day of Summer' is an annual public holiday celebrated in which European country in April?

Sudoku

The goal of Sudoku is to fill a 9x9 grid with numbers so that each row, column and 3x3 section contain all of the digits between 1 and 9.

As a logic puzzle, Sudoku is also an excellent brain game.

If you play Sudoku daily, you will soon start to see improvements in your concentration and overall brain power.

		3	5	7		2		
2	9		1		3			6
					4	1	9	
	8	2		3			1	7
			7		8			
9	1			4		3	6	
	2	1	4					
7			8		1		2	5
		5		9	2	6		

QUIZ Answers

- 1.Blackpool tower 2.Southeast 3.The Corrs 4.Wish you were here 5.True 6.21st June 7.Julius Ceasar 8.XFactor 9.September 10.Iceland

Managing my Pemphigus Foliaceous

By Ana-Paola Luna

After losing my Dad in 2019, the world faced a pandemic, and I thought 2020 would be one of the toughest years in my life. Little did I know that 2022 would be literally be like 2020 too, but more of a life changer for me.

In January of 2022 an irritable rash started to appear in my scalp. After several visits to my GP, calling the 111 medical service, and visiting the Hospital A&E department, I was finally given a diagnosis of shingles. I was told there was not much I could do but wait until it cleared up. It didn't!

By May, the 'rash' had extended all over my body. When I say all my body it was exactly that - especially my chest, back and face. With growing pain, I sought a private dermatologist appointment which was difficult due to lack of availability close to where I live. When I finally saw one, the dermatologist said my condition was very serious and referred me as an emergency to his team at Guys Hospital in London the very next day. Following many tests and biopsies, a swift diagnosis of Pemphigus Foliaceous was given - an autoimmune condition

I had never heard of before (and which I happily could have lived my life without knowing what it was!).

I was prescribed medication straight away: A lovely cocktail of Prednisolone, Mycophenolate and other drugs which for sure have helped, but of course have brought other side effects and have changed my lifestyle

and quality of living. Yet here I remain on my first anniversary with Pemphigus, a person battling a rare autoimmune condition, writing a few things about what has helped me to manage thus far.

My first advice is to listen to yourself and your body. It is okay to be 'pushy' in order to get seen by doctors and taken seriously. It is okay to say you need time for yourself and it is okay to ask for help. Keeping a positive attitude is very hard when you look in the mirror and you don't recognise yourself anymore, and even worse when you are seeing that through constant pain.

Due to the breakout of lesions on my body, my first priority was to get rid of the sensation that insects were crawling in my body and eating me alive. My skin was in such a state of deterioration that even applying water onto it was painful. I found the best approach was to just lay

in bed covered from head to toe with Hydromol ointment. Not easy to relax right?

My mind was thinking in overdrive. Why me? How did I get Pemphigus? What I am going to do? How long will I suffer? None of the usual distractions like TV, music or podcasts seemed to help because intrusive thoughts brought me back to the same questions, and more. A friend then recommended I listen to a YouTube video called 'sacred numbers', which is basically a repetition of the numbers in a sequence a certain amount of times. Following the repetition was strangely calming and I listened to them for several hours during the day.

Being a Mum of two is not easy and even less when you have Pemphigus and have no extended family around to help. My husband and myself were very honest and open with the situation with our children. We explained to them about the illness and the side effects of the medication, such as mood swings. One of the more bizarre results was that, because of the flammable ingredients of the skin creams, it wasn't ideal for me to be cooking next to the hob. As you can imagine, I cooked very simple meals for some weeks and embraced the convenience of preparing frozen meals.

Having young kids with a husband working full-time also means that I need to do school drop-off and pick-ups. With Pemphigus my clothing was not suitable as modern clothing is made from more synthetic fabrics, which hurt and stuck to my blistered body. In response I bought lots of cotton basic tops, baggy lounge trousers, sarongs and tunics, and no underwear for a few weeks as the discomfort was simply unbearable.

At one point my blisters were so numerous they appeared in my eyelids and because of this, and one of the side-effects of medication, reading became very difficult. Even now sometimes I feel like the letters on the page jump around and make me feel dizzy. Luckily I found several audio book podcasts on Spotify, the best of which made me laugh. Laughter plays a big role in feeling better, when it is otherwise hard to find reason to laugh.

Internal swelling in the throat caused difficulty swallowing and restricted air flow. At night the



**“Laughter
plays a big
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better..”**

feeling was so bad I felt that I couldn't breath at all. Those nights I sleep almost in a seated position and sprayed the room with a lavender fragrance to relax .

My scalp remained bad as well. I lost lots of my hair and the remaining hair became dry and frizzy. I went for a big chop and

most of the time now I held it in a bun. To reduce the impact I bought lots of cotton bandanas and even a cheap wig and a fake fringe from Amazon for special occasions.

The first shock, when recovery begins and you start to see beyond your pain, is the image of yourself.

I still don't recognise myself in the mirror. When I started with cortisone, I read so much about the weight gain that it was one of my first questions to the doctor. I asked what to eat or not to eat to reduce this. His answer: “you are having such a bad time that just eat what makes you feel happy. At the end we are all different and it is impossible to predict which side-effects you will get or not”. It was a response that you may think is not very helpful. However, it has largely worked for me because although I have put some weight, I am still eating whatever I want and I don't feel anxious about food. I do try to eat as healthy as possible and make adjustments. I eat more protein to avoid feel hungry and over-snacking and I have also prepared many smoothies from frozen fruits to keep myself hydrated with flavour. The so called ‘moon face’ effect of facial bloating has been bad for me, so for the worst days I do use an ice pack for a few minutes to try and relieve the swelling.

Many people recommended exercising, but for me this felt impossible when in so much pain and I also have been diagnosed with medicine-induced osteoporosis, so I don't want to risk further damage. I didn't even attempt it. They say a dog is man's best friend and, with Pemphigus, I would very much agree. Taking her for walks has helped me greatly, not only to have some mild exercise, but also to be outside and unwind my mind. She has been a fantastic companion during my darkest moments.

Recently, once on the road to recovery, I have been following ‘wall yoga’ videos. They are very mild, short and easy to follow.

Support. Support. Support. I learnt years ago how critical a support network is when I went through post-natal depression. As much as you may want to believe you are unique and special,

that's not true, and for sure someone in the world has felt - or is feeling - the same. I am Mexican and in my culture we openly talk about everything to everyone – it's normal and expected.

Such communication helps a lot, not only because you are expressing yourself, but also because of contacts. Maybe someone knows someone who knows a specialist doctor, or about different medicines, clinical trials, or new advice that can help you? The culture in the UK is more closed, so working hard to find a support group is worth it and plays a big role when you are in a low point in life. Luckily you can now find support groups for nearly everything online, so go on social media and you will find amazing groups like PEM friends, where you can find information, empathy, care and lovely people.

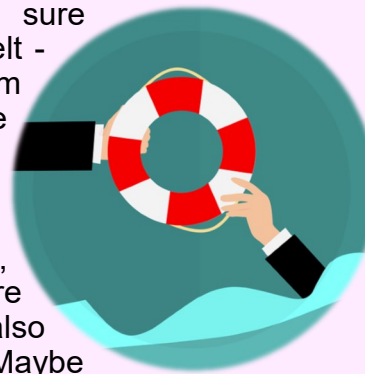
At the end recovery is all about love. It's a cliché but probably I wouldn't be able to manage this without the love and support of my husband, my kids and my dog. It wasn't always the kind of support you see in the movies - like my husband telling me how much he loves me, or holding my hand, or my kids trying to be independent and not nagging. They have been here for me and do amazing things, just small things, every single day that make me want to be here and fight for a happier life despite any illness or condition.

To accept this illness it has been very important to receive love and for those I love to accept ‘the new me’. My body, my face, my hair, and my mind have all changed. Some changes I would say worse, some better.

I do feel now more empathetic with others. I care more about quality and not quantity from simple things such as clothes. I have learnt to pay attention to my body. When there are signals of fatigue I won't work. We will try to eat out because I don't feel like cooking and cleaning.

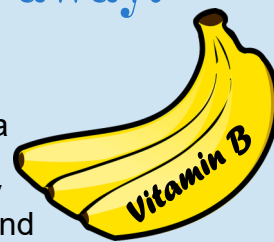
Living with pemphigus is not easy but is not impossible. There are terrible days, but also days where you even forget you have a horrible condition.

So I guess we just have to live life as best as we can, taking it one day at the time.



My Pathway to Diagnosis: Just an ambulance ride away!

By: Karen O'Hara (Kilkenny, Ireland)



February - early March 2022. I was developing a mouth ulcer or two and also a couple of small scalp lesions. The mouth ulcers appeared to go away and I just assumed that the mouth ulcers may have been caused by a Vitamin B deficiency from the weight loss diet that I was on. I wasn't that concerned about my scalp and didn't want to risk picking up Covid from my GP's tiny and usually brimming-with-patients waiting room; but, my daughter recommended that I should get the scalp lesions checked out so I took her advice.

My GP seemed rather interested in my scalp more than I thought she would! She prescribed a cacao ointment to put on the scalp for an hour and then a medicated shampoo to wash it off with, to be applied for 3 consecutive days for a couple of weeks. The treatment didn't work! By this point, I had now developed a loud, tickly, suffocatingly type cough that eventually made me lose my voice. I thought I had picked up a bug from the waiting room. After several days of laryngitis, I rang my GP. It took her a while to understand what I was saying but eventually she prescribed antibiotics. Several days later, I still had a cough and no voice so I returned again. She switched antibiotics and decided to try steroids to see if it would bring my voice back. A few days later, success! However, as the cough was becoming more productive and I seemed to be healing, I noticed that the mucous was tinged with blood. This had never happened to me before.

I was delighted to have my voice back; but my mouth was starting to get really sore when eating. Back to the GP, she nodded her head and said the antibiotic destroyed all the good bacteria leaving the thrush/fungus to prosper in my mouth. *"I will prescribe Mycostatin and that will definitely clear it up! You also should start taking probiotics as well."* A week later, my mouth wasn't feeling that much better. Although Mycostatin didn't do much for this "fungal" infection, I finally realised I was leaving a portion of the dose in the dropper, so I wasn't actually taking the prescribed amount. I wondered if the GP had under-prescribed the dose. So, back to the GP and she prescribed a higher dose.

I also had a little white growth inside one of my cheeks, the doctor suggested an Oral Surgeon should examine this. The bottom of my tongue also looked much bigger on the left compared to the right and very white as well. Was it a tongue tumour? The GP advised me to continue the regime of ointment and shampoo but to give it a break for a few weeks then try again.

The results from the now ramped up dosage of Mycostatin did feel about 70% more effective when I ate, but eating was difficult because of the tongue sores, my mouth felt like it was on fire.

I visited my GP several times after this and she prescribed different antibiotics regardless of whether the possible fungal infection in my mouth and scalp would be made worse and which more than likely wasn't going to do much. She even prescribed me a one-time-only anti-fungal pill called Fluconazole that was supposed to be super-effective! But, unfortunately, Fluconazole did absolutely nothing! The GP advised she would try to get me an appointment with a Dermatologist to have a look at my now increasingly growing scaly scalp, but not in my County, because they didn't have any Dermatologists but maybe in Dublin or Waterford. Even though my scalp was becoming more unsightly it wasn't painful or itchy. Unlike, the fireball of pain that was going on in my mouth and no longer just when I ate.

Around that time, I had noticed that the insides of my nose were constantly filled with dried, bloody mucous "crusts," and I couldn't blow my nose. The GP prescribed me a soothing antibiotic cream called Naseptin which provided a very quick fix of relief. It was probably one of the most effective drugs she ever prescribed for me. Interestingly, at this time, my husband and daughter (Frank and Dalilah) and I had contracted Covid for the first time. We all had mild enough symptoms but I actually had no symptoms! Maybe it was because of all the antibiotics and antifungals I was taking? Maybe I just had great antibodies because of the Covid boosters?

June 2022. I started getting sores on the skin of my nose, forehead, upper back and around the neck and shoulders. I was into the fourth month of visits to the GP, and I don't think she was even connecting the dots and thinking that all of my ailments may be related or under the umbrella of a particular fungal infection. I took a little break from my GP and visited my dentist instead to get more clarity on my situation. He didn't see anything too sinister going on in my mouth, but he thought an oral surgeon perhaps should look at the white growth-like structure on the bottom of my mouth. He suggested it could still be Candida and suggested that I rinse my mouth with salt water as much as I can.

Continued over...

July 2022, I was living on a limited variety of food: soup, fruit juice, soft boiled eggs, cupcakes with a bit of peanut butter and mashed broccoli. I also was still experiencing an occasional tickling cough which would still produce a bloody sputum. 11th of July, the day we were scheduled to fly out to visit friends in St Briac, France. I knew eating would be very difficult and the mouth soreness was progressing much worse with a now constant hot mustard sensation in my mouth all the time, but I would be stimulated in other ways by our friends, scenery, sites, etc. Despite my mouth, scalp and skin situation, I still had plenty of energy for getting around that traveling requires. We would still go!



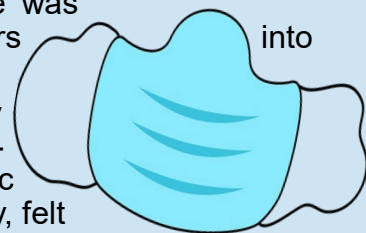
It was Day Four of our trip to France and Frank and I noticed that my lips were very sore encrusted with darkened blood, this was painful and a new type of pain to deal with. I could barely smile let alone put my lips on the rim of a glass. What were our polite friends thinking of my situation? Each day, the food looked so good with plenty of effort put into it by our friends, but I could barely touch it! The wasted dinners, the coughing up of bloody sputum in their car and the fact that my face was starting to look like I was hit by a bus. All of these symptoms from a fungal infection? Did they think we were we better off staying home?

It was a beautiful sunny day when we finally arrived home. My two daughters were shocked by my very sore lips. We went out for dinner for my daughters birthday that evening. I contemplated not going but went anyway. I couldn't even swallow the pureed vegetable soup or drink the wine. A few days later, the skin on my nose developed a large dark square, it looked like little mosaic tiles to match my blood encrusted lips!

My GP checked me for diabetes and took blood tests, the results were normal. Meanwhile, I had a scheduled dental appointment which I decided to keep. The dentist had seen me just two days before I went to France and had thought my possible Candida was okay enough for me to still go. I took my mask off, the dentist was horrified! While I was sitting in the dental chair, he mumbled behind me, "You should be in a hospital!" and noted that my scalp "looked awful!" He then called his more experienced partner into the office and they were both thinking, "It has to be systemic!" He took photos of my face, mouth and scalp for a referral to the Oral Medical Specialist based in County Cork.

My dentist wasn't the only one who thought I should go to the hospital. Dalilah's boyfriend, Sean, thought I should go to the local A&E in Kilkenny. I promised my family that I would go to A&E the next day. I was dropped off by around 7:30pm on Friday and walked into A&E. I was seen by a mature, senior male nurse that was doing the triage and who was not interested in seeing what my face looked like behind my mask. It wasn't till the wee hours of the morning on Saturday that I was finally seen by a young, male doctor. When I took my mask off he said that he was surprised that I was triaged so low, which explained why I was waiting for hours into the morning.

This doctor was very kind and empathetic to my condition. He took very detailed information. He had a hunch that I had something called Stevens-Johnson Syndrome indicated by my encrusted lips, which is a severe allergic reaction to antibiotics. According to another Dr, who had this syndrome recently, felt it couldn't possibly be that! Yet another doctor also wanted to examine me and take detailed information. I was a medical mystery to her and she took photos of me for my file so that other doctors and interns could learn from my case.. The doctor told me I should be admitted to hospital to have the best chance of meeting with a Dermatologist and for sustenance. He also said it would be quicker for me to see a Dermatologist if I were an inpatient.



All that this hospital could really do for me, without the guidance of a Dermatologist was to make me more comfortable and help me anaesthetise my mouth with unlimited amounts of mouthwashes, Mycostatin, Bonjela, protein shakes for energy and sustenance while monitoring me and giving me daily blood tests, a chest x-ray, a CT-Scan, etc. The CT-Scan turned out normal and the daily blood tests showed that I didn't have any auto-immune conditions. It was helpful to have the protein shakes because I couldn't eat much. However, the morning porridge was the best porridge that I ever had in terms of taste and texture! Better than restaurant standard. I was treated very well in this hospital and the staff were lovely, encouraging me to use the abundant supplies of Mycostatin, Bonjela and Difflam.

A few days into my stay, I received a phone call from my Dentist's receptionist asking me how I was doing and pleased to hear I was now an inpatient and getting some treatment. Meanwhile, the junior doctors in the hospital worked incessantly trying to connect me up with a Dermatologist.

On my sixth day in this Kilkenny hospital, one of the junior doctors came over to me to inform me that

Continued over...

I would have an appointment the next day with a Dermatologist in Dublin. This meant being taken by ambulance and unsure whether I would be admitted there or brought back to Kilkenny? This was the news I was yearning for!

12th of August 2022, I was so excited! I was dressed, packed and had had my porridge for breakfast super early and then was greeted at my bedside by the driver and paramedic from the ambulance crew. The Dermatologist and Nurse met me and I was escorted into his office and noticed that he had had a medical file on me. He just simply said, "You had mouth sores and scalp lesions since March.... Do you have Ashkenazi Jewish Ancestry?" Flabbergasted, I said to him, "How do you know that?" He then replied, "You have an auto-immune skin disease genetically linked to your Jewish Ancestry called Pemphigus. Did you ever hear of Pemphigus?"

When he said I had an auto-immune disease, I was actually elated as I now had an explanation for my suffering and learned that all of my symptoms were coming from the same source and not as individual ailments, and also learned how Pemphigus is treated. My frustration was now lifted! I was the first person in Ireland that the doctor had seen with Pemphigus. He then performed several biopsies from my back, scalp and took blood for Rituximab screening, in case I needed to have it but explained that it took several days to get the results to confirm the Pemphigus diagnosis. He handed me some leaflets which explained Pemphigus and Rituximab.

I was admitted to that hospital by the Director of Rheumatology who explained that I would be given steroids as a quick fix and explained I needed Rituximab. Immediately, I was started on the IV steroids. I read the Pemphigus Vulgaris and Rituximab leaflet and learned that I actually had the same exact symptoms in the same exact time frame as what was described in the Pemphigus leaflet. I was *classic* Pemphigus Vulgaris!

After two days I was able to eat some real food again without my mouth feeling that it was being cut up, Minced meat and mashed bacon and cabbage, and apple crumble for dessert! Oh, my! I wasn't able to eat like this for months! A few days later my biopsy came back positive for Pemphigus Vulgaris and we would now be able to proceed with the Rituximab infusions. In addition, the lab was able to grow some bacteria from the swab taken off of my scalp, which meant I was on antibiotics for a week, and required the hospital to move me to a private room with my own shower.

During my eight days spent in this hospital in Dublin, their heroic and dedicated Dermatologist checked in on me daily to see how I was doing. I also received frequent visits from his team. His wonderful nurse came to see me several times to help structure my routine using the various topicals and helped me even to wash my hair. I received Rituximab on day eight and had a 2nd infusion treatment two weeks later in a cozy, hospice-style infusion centre in Dublin.

A couple of weeks after I was home, I decided to ring my Dentist's office to inform him of my diagnosis since he so kindly rang me up to find out how I was doing while I was in hospital. His receptionist said that Bernard would call me back because he had interest in diseases. And, he did. When I informed him that I had an auto-immune skin disease called Pemphigus Vulgaris, it turned out that he was quite knowledgeable about Pemphigus and Pemphigoid and what part of the dermis they affected. Hopefully, this is a signal that more medical and dental personnel will become better acquainted with our various PEM and Mucous Membrane skin diseases in the near-future.

The nurses in the infusion centre where I have my periodic Rituximab treatments never heard of Pemphigus and just assumed I was there to treat my Rheumatoid Arthritis. Through me, they are now being introduced to our type of auto-immune disease and how we differ from other auto-immune diseases.

HOW
ARE YOU
DOING?



..they are now
being introduced to
our type of auto-
immune disease..

In Memoriam

We received this sad note from Kath's daughter, Sarah, following Kath's untimely death in February this year. Kath Lonergan was an amazing women of strength and love.

She was a mother to 3 and nana to 4. She loved us all dearly and taught us to love others and never judge anyone. She taught us that we could reach for the universe

Unfortunately she started with Pemphigus Vulgaris last year and got sepsis from her sores. She managed to beat that but again sepsis caught back up with her and she lost her fight on the 1st February.



* HOT OFF THE PRESS *

Setting priorities for research into Pemphigus and Pemphigoid

By Isobel Davies

I was about to tell you that the much anticipated paper on the results of our work to prioritise research work in the UK on Blistering diseases is still awaited. Today I received the announcement that it is published.

Many of you have inputted to the prioritisation process and we will, we hope, benefit from the focus it offers to anyone seeking funding for research into PEM. It's been a long journey, largely due to interruptions due to covid-19, and it is very good to see the outcomes made public at last. 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. Karen has ensured that the PSP article has been published and we are grateful to her and the rest of the team for their patience and perseverance.

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.

As I mentioned in our last PEM Lives magazine, six PEM Friends were on 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.

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 see the article published by the British Journal of Dermatology if you are a member, but we hope to replicate it in our next magazine.

Drum roll.....

The top 10 priorities voted for by medical specialists and patients are:

1. How effective, safe and cost-efficient is Rituximab (or similar biologics) in BP/PV/MMP compared to standard steroid/immunosuppressant use, when should it be started and should it be a 1st line treatment?
2. Are outcomes for patients with BP/MMP/PV better if treatment is started earlier and with 'stronger' treatments, such as an immunosuppressant or biologic, rather than escalating from 'milder' treatments if they do not work?
3. How should persistent mouth lesions be best treated in Pemphigus and Pemphigoid?
4. What is the best treatment for preventing and repairing scarring in MMP (medical and surgical)?
5. Is it possible to identify drugs that block the specific immune pathways for BP/MMP/PV rather than treat them with broad immunosuppressive drugs?
6. What are the risks and benefits of the different tablet and injection treatments used to treat BP/MMP/PV? (such as Azathioprine, Mycophenolate mofetil, Methotrexate, Cyclophosphamide, Chlorambucil, Nicotinamide, Dapsone, Intravenous Immunoglobulin, Plasmapheresis)
7. What factors predict relapses in BP/MMP/PV, how can the risk of relapse be reduced and how are relapses best treated?
8. What is the best/most effective dose to prescribe for steroid tablets in BP/MMP/PV including the starting dose, when and how quickly to reduce the dose, and when to stop?
9. Can we predict the response to treatment in BP/MMP/PV and what factors affect this?
10. What is the best way to treat skin wounds in BP/MMP/PV including how should blisters/ erosions be best washed and managed and does treatment vary according to body site?



Marks Marathon

By: Mark Ranson Thompson

My name is Mark. I am 43 and in January 2021 I started experiencing the horrific symptoms of Pemphigus-Vulgaris. You may recall my previous article about running the London Vitality 10k from issue no. 6 of Pem Lives from December 2022. I have a little update on my running journey...

I wrote in detail about my early Pemphigus symptoms and diagnosis in my previous article but I now want to tell you all about my journey in training for the 2023 London Marathon!

In 2019 I went along to watch the London Marathon as a spectator and was so inspired by the runners that I went along to my local Parkrun (Catford Parkrun). I was a regular there until the Covid 19 Pandemic, and then later my running was scuppered when my Pemphigus diagnosis struck.

After a successful response to Rituximab I gradually returned to running, starting in January 2022 and ran the Vitality London 10k in May of the same year. This was the longest distance I had ever run.

Training for the marathon started in January 2023, following a bout of Covid the week before Christmas, immediately followed by a lengthy, even nastier cold. I was to run three times a week, increasing in distance.

Early on in training, I found my ribs were hurting occasionally during runs. Not ideal, but I was able to run through the pain. Aside from the odd ache and pain, including some slight twinges in my right knee, training went smoothly and regularly. At times, it was very difficult to motivate myself to go out in the cold and dark following a day's work, even for a short run, but it was worth pushing myself through those dark evenings. Other occasions led me to last minute rescheduling due to complete fatigue – I blame the prednisolone!

My last big run before the marathon was just over 18 miles - from where I live in Lewisham to Greenwich with some laps of the local parks en route. My first big worry appeared during that run: after approximately 10 kilometres I felt incredibly sick. Fortunately, I learned my lesson in fuelling my body properly during a run, and not just to eat only beforehand!

The final hurdle in my training – attending a wedding as a guest 1 week before marathon day! I still managed to have a great time celebrating and even got in a short run during this tapering off period around the beautiful town of Great Malvern. There was also a small hike most of the way up the North Hill despite my fear of heights!

Finally, the day of the Marathon came – I set off alone and nervous in the drizzle towards Blackheath. Fortunately, I am local and knew my journey on the trains to the start line - My family and friends had gone on ahead to secure good spectator spots along the course.

I had my runner number pinned with a big red cross (denoting that I have a medical condition) and all my gear – a banana to eat before the start of the race, a protein bar, energy gels, electrolyte water, a knee support on my right knee, orange running shoes that could be seen from space, and of course – my PEM Friends running top! After a bit of time hanging around in the heavier than anticipated rain, it was my turn to set off at 11.15.

I enjoyed a fantastic start from Blackheath down to Greenwich - I was pacing well and was ahead of my anticipated time without having pushed too hard, when disaster struck! At mile 7, coming around the bend by The Cutty Sark, I suddenly felt a sharp pain in my left knee (of all the knees!). At the time I wasn't overly concerned and hoped to 'run it off'. However,



Pre Marathon
Mark & Chrissie



Big Ben Official
©London Marathon
Photo



Buckingham Palace ©Official London
Marathon Photo

after a few more miles, I had a horrible feeling that the pain wasn't going to stop and this was perhaps a more serious injury than I first thought.

It's amazing what the cheer of the crowds can do for the runners. I kept going! There was so much noise, music, and pop-up parties that it felt like a constant party for the whole race.

I can't remember exactly where, but I had to scramble to get the knee support off whilst wearing running tights to change it over to the now agonising left knee. I also had to stop to ask St John's Ambulance for some paracetamol (so many forms to fill out!!)

Not long after Mile 17 is where I saw my first familiar faces in the crowd: My wife and 2 of my good friends had come to cheer and give jelly babies out to the runners. This is where it was first pointed out to me that I had some quite substantial nipple chafing! I was completely oblivious due to the knee pain!

Sadly, the pain from my knee was so bad that the last 10 miles went by in a bit of blur. I had more friends and family waiting for me at mile 25 but I couldn't stay for too long otherwise I wouldn't have carried on. I was in so much pain that it was very tempting to stop and stay when I saw their friendly faces, but I battled on after they shooed me away.



Finish Line ©Official London Marathon Photo

Towards the finish line, I checked my watch, knowing I was getting close to 6 hours. I put in the best sprint I could manage for the last 50 metres, and came in just under, at 5 hours, 59 minutes, and 57 seconds. The 'sprint' was worth it!

After collecting my medal and goody bag, it was very difficult trying to find my supporters at the end with scrambled phone signals and physical barriers up around The Mall, but finally I found them and was rewarded with a beer and some jelly babies. The joy was short lived, my dream of having a few pints in my favourite West End pubs was dashed by my knee pain and sheer

exhaustion. I just wanted to get home – fortunately not a long train ride for me from Charing Cross to Lewisham. A few pints over my local instead (The Blythe Hill Tavern) and 'cheering' a couple of fellow marathon runners who were also there, and I was ready for bed.

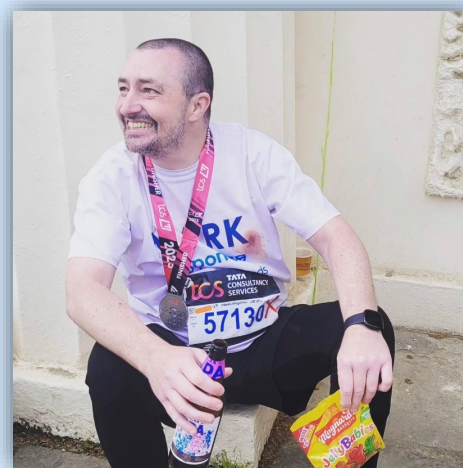
'Lucky' for me I had a folding walking stick in the drawer from where I suffered horrible blisters on my feet from my worse Pemphigus-Vulgaris days. Unluckily, however, my knee is still causing me problems, and I am still using the walking stick over a month later, whilst waiting for a physio assessment. This is still true at time of writing at the end of May!



The gin, a thank you from PEM Friends

As horrible as running a marathon sounds from this personal account I've given you, I absolutely loved it and am incredibly proud of myself. I have been applying for a ballot place for the London Marathon for over 20 years and can finally tick it off my list. More importantly, I raised £2236.50 for PEM Friends (after fundraising page fees) and want to thank anyone and everyone who sponsored me. A special thank you to PEM Friends for the wonderful bottle of gin they sent me. What a lovely surprise!

Would I run another marathon? Maybe...



©Sarah Maxwell

By Andy Heath (on behalf of Kalpesh Patel, our treasurer)

Thank you so much, you are amazing!

Period Beginning of January 2023 – End of May 2023

People are so generous. Sue and Derek have participated in a drug development programme with Janssen and donated their fees to PEM Friends, we received some very handsome one-off donations at the beginning of the year, and Mark raised over £2,200 in sponsorship for running the London Marathon. Some new people joined our 'bedrock' group of donors who give to PEM friends every month by standing order. In total, you have contributed over £3,300 so far this year. And thanks to her generous daughter, we also received a donation from the mourners at Kath Lonergan's funeral.

Thank you all so much, you are amazing.

So far this year we have spent just over £950 on internet services, thank you vouchers, office costs, marketing and publicising the Photo Library. Although they are only a small token of our appreciation, we like to say 'thank you' to the kind people who help PEM Friends by giving their skills and time. We keep the office costs to a bare minimum, with most of this expense so far this year going on buying stamps and public liability insurance, which is a requirement for us to attend the British Association of Dermatologists (BAD) conference. We have focussed our marketing to date this year onto one thing: branded pens. The pens are very popular at conferences, especially with doctors, and will remind their new owner of PEM Friends and our conditions long after the conference. We printed and posted out around 150 postcards to publicise our Photo Library to medical professionals using contact details provided by you.



Attending conferences is important to PEM Friends. It gives us exposure, provides opportunities to network and make new connections, and we learn a lot from medical professionals and from other patient support groups. Anticipated expenditure in the near future will include the travel, accommodation and subsistence costs for our delegates attending the GlobalSkin (in Brussels) and BAD (in Liverpool) conferences. Some of these expenses will be helped by a bursary for travel and accommodation (GlobalSkin) or reduced through subsidised accommodation in Liverpool (BAD).

One of our main expenses is the magazine. We love sending hard copies to you if you have given us your address, and to your medical experts. All this printing and postage does cost money though, so we are doubly grateful when we have help to pay for some of it.

Finally, news hot off the press: we have been awarded a grant of £4,000 by the National Lottery Community Fund to allow us to hold a hybrid (face-to-face and virtual) meeting in September. This is an ambitious (and expensive!) event that will provide PEM Friends with an opportunity to get together like never before. Many thanks to the Lottery but also a huge thank you to Isobel for having the foresight to apply and the eloquence to write such a clear and persuasive proposal.

Next time, we will tell you our new bank details. We are switching from HSBC, who charge us for their services, to NatWest who won't. Our HSBC details still work, and all new and existing standing orders will automatically transfer to NatWest when the switch is complete.



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



Meet the PEM Council - Mark.

What do you do as part of the PEM Council?

I'm very much the newbie on the council. My career was 30+ years in financial IT and as a trustee of a charity for 9 years I've got what I hope is a good broad skill base and ideas to bring to the party.

Briefly describe your PEM journey.

I've had Ocular MMP since 2006.

What's your funniest PEM related moment?

I thought morphine was addictive so declined it after a big operation, instead going for a very nice sounding pill called Tramadol. After 8 of those I apparently had a lengthy conversation with my hospital bedside lamp, believing it was a nurse.

And the strangest thing a medic has ever said to you?

Oh, I have a lot of those! Two that spring to mind are:-

On the morning of a complicated operation the NHS surgeon (who I trust with my life) asked how I was feeling. I confided in him that I was absolutely terrified and he replied, whispering, that he was also terrified of what he was about to try!

I also remember vividly an ENT professor explaining to me that the probe covered in anaesthetic about to go far down my airway would make me convinced I had stopped breathing and to "try not to panic" before asking the nurse to get an adrenaline shot ready in case I did.

Dinner party with 3 guests, one living, one passed, one fictional?

Kimi Raikkonen, Freddie Mercury and Homer Simpson.

You're on a desert island with everything you need to stay alive. What two luxuries do you choose?

An iPad with the whole box set of Breaking Bad downloaded and a jar of Branson Pickle.

If you had a 10 square metre area on earth you could never leave where would you choose to be?

The Sky Bar, the roof terrace of the Grand Central Hotel in Barcelona.

If you have, or could have a tattoo what would it be?

I'd have a QR code tattoo'd on my right buttock that directed people to my wife's Instagram page, just in case I was found lost without my phone.

Last question. Cats or Dogs?

Ouch, one to divide the room. I'd say cats, because dogs are hard work. My wife and I had brother and sister kittens when we bought our house. We named them Mulder and Scully, she made it to an amazing 19 years old.

Interestingly my daughter, when I first had eye problems and then 3 years old, was really excited I might go blind from the MMP, because she really wanted a golden Labrador guide dog as a pet. Kids eh?

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!



www.pemfriends.org.uk



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

European Reference Network (ERN) update...

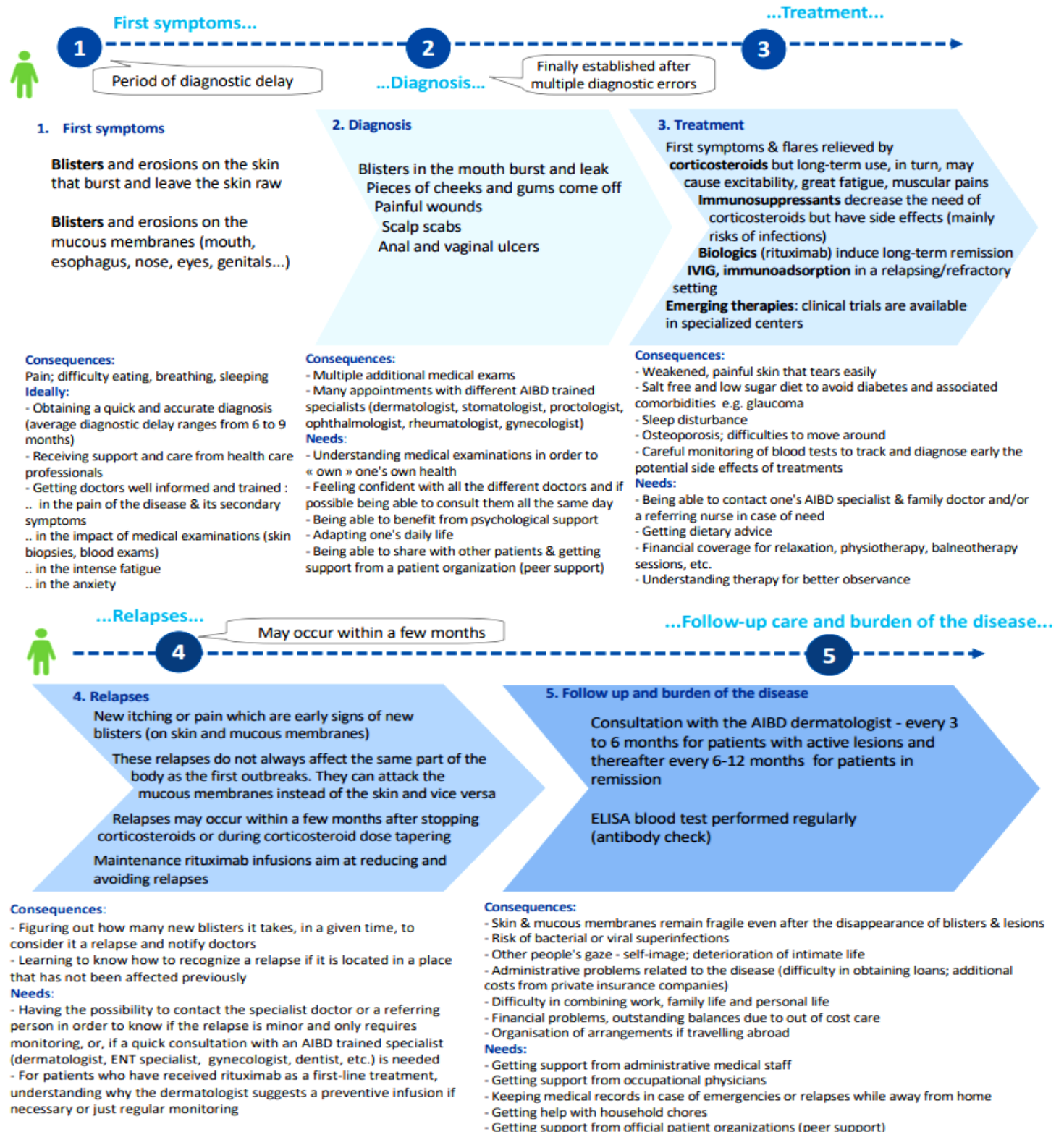
In the last issue, you read about the background of what the European Reference Network Skin (ERN Skin) does, its structure and functions. Our European Patient Advocacy Groups (EPAG) work has been concentrating on the development of patient journeys for each of our diseases. We work on these collaboratively, involving as many patients as possible through our patient organizations. This collaboration includes our Auto Immune Blistering Disease (AIBD) team of Health Care Professionals.

Here is the Pemphigus patient journey – the Bullous Pemphigoid one is almost complete and we need to start working on the MMP one. Anyone interested in participating? If so please contact us!

Pemphigus Vulgaris (auto-immune blistering disease - AIBD)

PATIENT JOURNEY


A rare skin and or mucous membrane autoimmune disease that affects people of all ages but primarily those over 50



GlobalSkin - *HOT OFF THE PRESS*


The Global Research on the Impact of Dermatological Diseases (GRIDD) project

DERMATOLOGY PATIENTS NEEDED FOR GROUND-BREAKING GLOBAL GRIDD STUDY

WHY?	WHO?	START NOW!
<p>You are the expert & your opinion matters.</p> <p>By participating you will be helping to show clinicians, researchers, and policymakers the true impact of living with your dermatological condition.</p>	<ul style="list-style-type: none"> ✓ You have a dermatological condition ✓ You are aged 18 or over ✓ You can understand & read one of these 17 languages <p>English, Arabic, Bengali, Simplified Chinese, Danish, Dutch, French, German, Hindi, Italian, Japanese, Portuguese, Russian, Serbian, Spanish, Swahili, or Vietnamese.</p>	<p>To Participate in the GRIDD Study, just scan the QR CODE to start! The online survey will take 10-20 minutes to complete.</p>  <p>ONLY UNTIL September 28, 2023</p>

LEARN MORE AT [GLOBSKIN.ORG/RESEARCH](https://globalskin.org/research)

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LEARN MORE AT [GLOBSKIN.ORG/RESEARCH](https://globalskin.org/research)

As a PEM Friends patient, you are invited to participate in ground-breaking research that is being conducted by the International Alliance for Dermatology Patient Organizations (also known as GlobalSkin) in collaboration with researchers at Cardiff University (UK) and University Medical Centre Hamburg-Eppendorf (Germany).

As a Member of GlobalSkin, PEM Friends UK is supportive of this important project. You are the expert and your opinion matters! By participating, you will be helping to show the true impact of living with your dermatological condition which will help to inform areas of need for improved care, better treatment options, and more affordable medicine for dermatology patients globally.

GlobalSkin is looking for 10,000 people like you to take part in the study by completing a 10–20-minute online survey, and possibly one six weeks later. To participate in this important research please follow this link: <https://globalskin.org/GRIDDStudy>

"The GRIDD Study takes my voice into account as the expert on my dermatology disease."




DERMATOLOGY PATIENT

Take the Survey Today!
Learn more at: globalskin.org/research

The online survey is open from June 5th to 28th September 2023

"Our voices matter because we are the experts on living with our skin conditions."

- DERMATOLOGY PATIENTS



Take the Survey Today!
Learn more at: globalskin.org/research



Summer 2023

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