

The EHE Foundation (USA)
The EHE Rare Cancer Charity (UK)
The EHE Rare Cancer Foundation (Australia)



Quarterly Newsletter
for the EHE Group
July - September 2019

the pledge

Edition 18



Rowing the Atlantic for EHE

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Welcome

Welcome to “***The Pledge***”, our quarterly newsletter covering the activities of the EHE Group.

This is our 18th edition, for the third quarter of 2019. We hope that you enjoy reading it, and are inspired by the stories included within it.

As always we want to say a massive thank you to all our supporters for their contributions. Regardless of what type or magnitude of support and contribution you provide, they are all critical to our success and ability to manage and ultimately defeat EHE.



Highlights

Wonderful gifts from The Margie and Robert E. Petersen Foundation

The EHE Foundation received two amazing gifts from the Petersen Foundation comprising \$1 million dollars to support Dr Rubin's EHE research plus up to an additional \$1 million of matched-funding over the next three years to match funds raised by the EHE Group worldwide.

Private donation drives new EHE initiative at Sloan Kettering

A further \$1 million donated to the EHE Foundation by a private donor will be used to establish a new International Centre of EHE Excellence at Memorial Sloan Kettering in New York.

New EHE research underway in four countries

EHE-dedicated research, funded by our EHE community, is now underway in the USA, UK, Australia and Canada. This amazing expansion in EHE research has been made possible by the wonderful support and energy of the EHE community worldwide.

EHE bio-banking started in USA, UK and Australia

Bio-banking of EHE tissue and fluid samples to help promote and support EHE research has been initiated in the USA, UK and Australia, and we hope will be rolled out to include other countries in due course.

Membership of the EHE Group exceeds 1,700

When the EHE foundations were first established in 2015, the EHE Facebook membership numbered in the hundreds. Today we have over 1700 members worldwide, representing 68 countries. We believe that this is a unique rare cancer group providing support to patients across the globe.

Further details on these stories, and much more, can be found in this edition of *The Pledge*.



01 Patient Support and Advocacy

Supporting those diagnosed with EHE

The EHE Group strives to provide support to everybody diagnosed with EHE. At the same time we work hard to increase awareness and understanding of EHE amongst the general public and all other relevant groups. These two goals are at the core of the patient support and advocacy objectives that are common to all the EHE foundations. But we can only achieve them with the tireless energy and contribution of so many of our members, many of them being EHE patients themselves. So as always, we want to thank all our supporters who have contributed to this critical part of our activity, examples of which are provided in this section.

Mariana Coutinho participates in Youth Cancer Europe summit

Mariana Coutinho posted news about her participation at the Youth Cancer Europe summit that was held in Budapest in mid-July. *"I'm very proud to share with you all that I was invited to participate with 80 young cancer patients from all around Europe who gathered to discuss the issues that we face as cancer patients. This included cross-border healthcare which is one of the main topics in Youth Cancer Europe's white paper."*

Mariana continued, *"because of my story, they asked me to be one of the speakers and share my experience as a rare cancer patient living in Portugal and receiving treatment in London. I talked about the EHE Foundation, I talked about the importance of patient organisations, I talked about Nanoknife. And of course, about my rights as a rare cancer patient. Now they want me to go to the European Parliament with them to expose my story and bring awareness of the importance of cross-border healthcare for cancer patients, especially rare cancer patients and patients who need new and experimental treatments. It's an honor to do this, I feel like I can actually take*

something positive out of all the suffering I went through and am so happy that maybe sharing my story will actually change something."

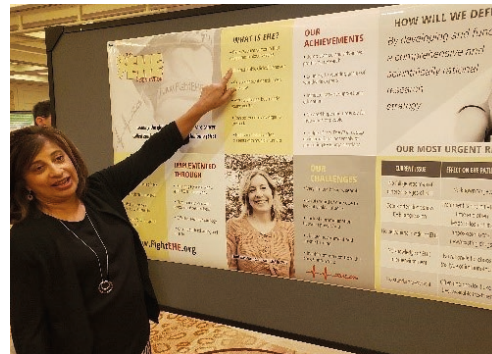
We want to congratulate Mariana for her determination and energy in describing the challenges that rare cancer patients face. It is so much more inspiring when patients who have faced and dealt with these challenges tell their stories. And of course we thank Mariana for spreading greater awareness of EHE as she has taken up this challenge.



The EHE Foundation represented at San Francisco Conference

Participating as a patient advocate in the prestigious AACR Scientist-Survivor Advocate Program, Medha Sutliff represented The EHE Foundation at the 12th AACR Conference on The Science of Cancer Health Disparities in Racial/Ethnic Minorities and the Medically Underserved in San Francisco, California. Over 800 researchers, clinicians and advocates gathered to understand, and ultimately help to eliminate, the disparities that represent a major public health problem in the US. Patients with rare cancers like EHE face significant disparities and challenges to medical care and treatment as well.

Speaking on advocacy at the plenary session and presenting a poster on EHE at the conference also helped Medha to stimulate awareness of EHE and foster networking among other patient advocacy organizations and researchers.



A cool story

Fiona Louise shared a lovely story about friendship and help. *"One of my side gigs (I am a high school teacher librarian) is volunteering to organize a literary festival in Brampton, Ontario. As a result I have actually become friends with several authors. An author reached out to me today and asked if she could base a character on me and my EHE journey to help raise awareness about the challenges of rare and orphaned cancers. I said yes, of course."*

It never ceases to amaze us how much people want to help when they hear about the challenges that our members face after receiving an EHE diagnosis. We love the idea that somebody wants to write a book about rare and orphaned diseases. And we cannot think of a better character model than our very own Fiona Louise.



01 Patient Support and Advocacy

True Cancer Bodies share Allana Parker's story

Allana Parker was proud to be able to let the EHE family know that her story had been used by **True Cancer Bodies** during their **Cancer Awareness Month**.

"I am honoured that the people at True Cancer Bodies shared my story and that of EHE during their rare cancer awareness month. This group are doing a fantastic job promoting the message that cancer isn't all pink and fluffy as shown in the media sometimes. Their birth followed a famous UK supermarket's advertising campaign that did not go down too well. They have had widespread newspaper and media coverage in the UK on the back of their standing against unreal images of cancer." Allana went on; "Please give them a 'follow' on Facebook and Instagram, I have a real cancer crush on these guys and their ethos."

Allana finished by noting *"Also an awesome bit of advertising for more EHE awareness"*. We completely agree Allana and congratulate you on telling your story so openly and honestly, and for raising awareness of EHE in the process.



 True Cancer Bodies
24 August · 🌐

Hi, I'm Allana (@allana_x_). I am 47, mum to 2 boys in their 20's.

In June 2018 my life blew up. After a year- long 'gym injury' I went to the doctors. On Friday I was sent for an MRI of my hip and on Monday my phone rang asking me to go to the hospital the following morning.

After a CT and biopsy I was diagnosed EHE Sarcoma. My first oncologist was not a specialist in EHE and initially I was given a prognosis of 1-2 years.

My brain was sludge and I spent hours on Facebook cancer groups asking for anyone else with EHE. I was found by a wonderful lady from the EHE foundation in America who gave me so much info and encouraged me to ask for a referral to Dr Jones at the Royal Marsden.

This man has become my life extender.

EHE is extremely rare, affecting less than one in a million but now with Dr Jones running the show my prognosis is 6 months - 40 years. Nothing can be specific with this cancer. There is no cure. It can sit dormant for many years with no symptoms, become aggressive at any time and also shrink itself.

I currently have tumours in my pelvis, liver, lung, skull - along with lesions in my hips, lower back, rib, quite a few vertebrae and bone around the side of my skull.

I've had 3 lots of radiotherapy, vertebroplasty to rebuild one of my vertebrae and now I am about to start chemo once a week for 6-12 months.

For me the hardest part of having an incurable rare cancer is there will never be the words 'in remission' or NED.

It has stripped me of so many things, especially my mobility. I went from being extremely active to a wheelchair within weeks.

Now a year on I am rebuilding a new life, with new goals and new EHE friends around the world. Physio has me walking with just a stick and I am aiming to be free of that soon.

I know I will live the rest of my life with cancer but I am going to make sure it is a good one.

#truecancerbodies #rarecancers #cancerawareness #cancerresearch #sarcomaawareness

Australia Launch new EHE-RCFA Facebook Page

The success we have in spreading awareness and engaging with the public, health services, government, and indeed all external groups, can be materially improved with high-quality materials, branding, and the messages they deliver. It was for these reasons that the EHE Rare Cancer Foundation decided to relaunch its Australia Facebook page, with new design and imagery developed together with Anna Wydro in the UK. This included a short video message designed to encourage everybody who came to the Facebook page to share it, to engage with the Foundation's objectives, and to create greater awareness of EHE.

We cannot do their video justice in a newsletter format. Here are just a few snapshots however taken from that video to give everybody an idea of the excellent graphics and simple messaging.

And if you want to see and share the actual video, it can be found at:

https://www.facebook.com/EHE-Rare-Cancer-Foundation-Australia-2051746421606077/?modal=admin_todo_tour



Support on the ice

Jenni Case Kovach posted a photograph of the top that her husband and a few of his closest friends were wearing when playing in the 'Hockey Beats Down Cancer' tournament in late August. *"They are proudly wearing EHE colors!"* noted Jenni. We think the shirts look amazing! We love the colors, and we love the strap line *"Beat Epithelioid Hemangioendothelioma (EHE)"*.



01 Patient Support and Advocacy

Drawing inspiration from a magical location

Australia is an awesome country with magnitude and extremes that at times are hard to believe. It is no surprise therefore that the belief systems of its indigenous peoples should have depth and texture that those of us outside it will never truly comprehend. This includes many sacred sites, and one of the greatest is Uluru, one of the world's largest monoliths, some 550 million years old, found in the Northern Territories.

It was from this sacred location that Jane Biddlecombe posted two brief video messages, one answering questions about the ISKS genetic research study (see our Research section) and the other to simply *"send healing energy and 65,000 thousand years of indigenous dreaming your way"*.

We love the messages Jane, and cannot think of a more inspiring location from which to share your EHE thoughts. We like even more the idea that in the not too distant future, EHE may become just a memory, perhaps to be likened to a vanquished evil spirit in our own story of *"the EHE dreaming"*! We can hear the didgeridoo playing as the sun falls in the east, and Uluru displays the ranges of colours that only it can produce at sunrise and sunset. ***Just Live.***



A mountain climbed!

We are always amazed at the ability of our EHE community to share their stories of challenges completed, despite living with EHE. These stories help inspire many as they deal with their own EHE diagnoses. Elise Halvorson-Maza, with over ten years

as a patient, did exactly that in a post to the support group which simply ended: *"On a side note - the other week I hiked Mt Humphreys which is the tallest point in AZ, it took me about 6hrs to go from 9000 feet to 12,100ft. **Just Live!**"* Well done Elise. ***Just Live!***

Family time shared

Carl Dickson was once again seeking to inspire and support fellow patients with more great photos. *"Another day out in nature to replenish the soul"* commented Carl *"with the best medicine known to man, and the least adverse side effects, your close family and sunshine."* Carl wanted to share the photos in the hopes that they would be uplifting for anybody who cannot get outside. We love the photos Carl, and the touching message that came with them.



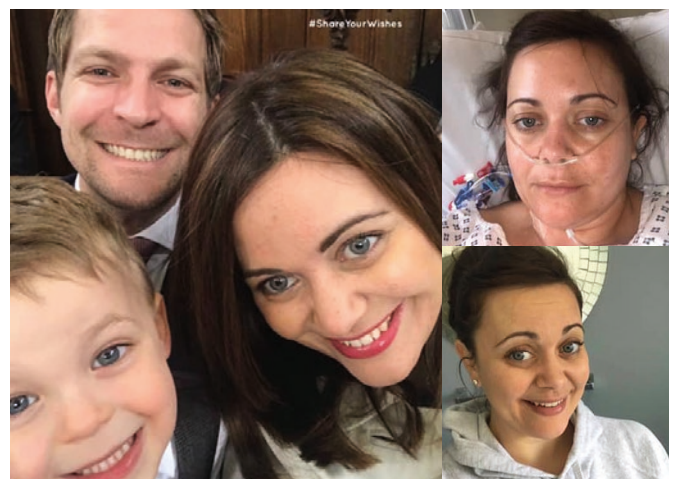
Nicola helps raise EHE awareness

In September, Nicola Henderson posted news that her story had been used to support organ donations.

"Hi guys, an organ donation charity asked for my story to help spread awareness of the importance of being a donor and sharing your wishes with loved ones. I also got to talk about EHE too so spreading some awareness there as well!"

The article started: *"Please meet 35 year old Nicola who earlier this year received the gift of life after being diagnosed some years before with a rare cancer. Nicola shares her story to encourage you to talk about your organ donation decisions."*

Both messages are so important to share, and we want to congratulate and thank Nicola and her family for sharing her story so openly to help such important causes.



01 Patient Support and Advocacy

Stunning art work

Whether our efforts to spread awareness are successful or not is inevitably influenced by several key factors. One of the most important is the visual impact of materials, as this is often the first 'hook' that will capture a person's attention. We are very fortunate to have several members who are extraordinarily talented when creating the artwork that accompanies our different strategies and campaigns.

Danielle Janisewski shared a number of her striking designs with the group and offered to assist anybody who needed help creating their materials. Here are just two of her pieces.

"Sharing these that I made if anyone would like to use" commented Danielle. *"Figured it'd be nice to help raise awareness."* We thank Danielle for her images, and encourage everybody who is interested to contact her for help in developing graphics to support their activities.



Why support is so important

Dealing with an ultra-rare cancer like EHE is extremely frightening, and very demanding as so little is known about the disease. Patient support is so important because it provides not only emotional care when needed, but also helps provide information that will allow every patient to make more informed choices. Our group is always touched when somebody shares a story that shows the value of such support.

Summer Ryan Keller shared a message in late July when her husband was undergoing IRE ablation. *"Thank you so much to you all"* posted Summer. *"My husband just went back for his IRE Nanoknife procedure with Dr Raj in Miami. Without this site we wouldn't be here today with all the knowledge that got us here. Thank you to those that have shared stories with me and personal messages. We are blessed to know those on this site."*

Thank you, Summer, for those kind words. We hope that the IRE ablation was successful and we all wish your husband a speedy recovery.



New Joint Sarcoma Research Centre holds first education day

We are always excited to see and hear about a new focus on sarcoma, such as the recent new Joint Royal Marsden-ICR (Institute of Cancer Research) Sarcoma Research Centre. Hugh Leonard, Trustee of the EHE Rare Cancer Charity UK was invited to attend the inaugural symposium of this new group, held at the Royal Marsden Hospital in September.

"It was wonderful to be able to attend the symposia" commented Hugh. "There is so much going on with sarcoma that people don't really see from outside these organisations. We are also delighted to be working with this new joint group in establishing a UK EHE biobank which started in July" said Hugh. "We hope that very soon EHE fluid and tissue sampling protocols will be

finalised and will start to be applied, initially for existing Royal Marsden patients and then other patients across the UK".

There is more news about the EHE biobanking project in the Research Section of this edition of **The Pledge**.



A Dad remembered

Each quarter we see photos and hear stories of people who decide to support our cause, and spread awareness, by getting a **"Just Live"** tattoo. This quarter Amber Guy Wilson did that in memory of a very special person. *"I got this last night in honor of my Dad who I lost on March 15 to EHE. His signature is on bottom!!! I miss him so much!!!"*. We think it is an admirable way to remember him, and we are sure that Amber's father would be proud to see her continuing the fight against EHE.



'Just Live' on a different arm

Many of our members have had the group's **Just Live** motif tattooed on their arms, both in support of loved ones, but also to spread awareness when people ask *"What does that tattoo represent?"*.

Shannon Estrem found a different arm to have the motif on when she was offered free engraving by the company providing her glasses. *"I didn't have to think too hard about what to put on them"* she posted. We love the idea that in the future people may ask her why she has **"Just Live"** on the arm of her glasses, and will then become educated on the subject or rare cancers!



02 EHE Research

Only through research can we answer questions and defeat EHE

We will ultimately defeat EHE because the questions that we do not have answers to today will be answered by the research we are funding, and the future research that we continue to promote and facilitate. This research will also lead to the generation of new drugs and treatments to manage and defeat this rare cancer. The following notes provide an update of some of the research we are involved with.

Petersen Foundation gifts will drive EHE research

Early in the quarter the EHE Foundation were delighted to report that two major gifts for EHE research had been received from the Margie and Robert E. Petersen Foundation. The gifts comprise an immediate \$1 million for Dr Brian Rubin's EHE research, and a further \$1m in matched funding over the next three years (see Fundraising section of this edition).

Dr Rubin's EHE research programme was started in 2010, and in 2011 his team discovered the WWTR1-CAMTA1 gene mutation that is believed to be disease defining as it is found in approximately 90% of EHE tumours. Today Dr Rubin's laboratory is working actively on drug and compound screening, the development of a GEM EHE mouse model, the generation of EHE cell lines, and the evaluation of secondary gene mutations that may be dictating whether EHE is indolent or progressive, and if progressive, then how aggressive the disease is.

The progress made by Dr Rubin has been partially due to prior donations made to his lab by the EHE Group. This includes funding in 2016 for a new post-doc researcher, Dr Che, for two years;

2017 biopsy funding relating to the clinical trial of Mekinist; and 2018 funding for proprietary equipment and compound library access for Dr Rubin's compound and drug screening assay.

We are delighted to see this extraordinarily generous funding being made available to Dr Rubin to place his laboratory on such a sound footing for the next three years. We are also excited that part of the funds can be used by Dr Rubin to support other EHE research, allowing him to coordinate and expand the overall EHE research programme in the USA.

International Center of Expertise in EHE established

A second major donation of funds for EHE research was received in September from a private donor (see Fundraising section). Jane Gutkovich has spearheaded this process and posted an update in September for the worldwide EHE community.

The extraordinary donation of \$1 million will go to the EHE program at the Memorial Sloan-Kettering Cancer Center in New York where a new program called *"Establishing an International Center of Expertise in EHE"* will be created, led by Dr William Tap,

Chief, Sarcoma Medical Oncology Service, and Dr Cristina Antonescu, Director, Bone and Soft Tissue Pathology at MSK.

Jane explained *“There are two tightly connected activity streams under development: clinical management of EHE patients and EHE research. The goal of the program is to use patients tissue, blood, and clinical observations to help us build a better understanding of EHE biology, develop useful models, screen for effective drugs and to then use this information to lead an individual-treatment approach for all EHE patients. Having these two streams under “one roof” will significantly speed up the process toward effective rational treatments.”*

So what does this mean in terms of the organisation and procedures that will be used within the new program? Jane confirmed that:

1. The whole team of EHE experts (oncologists, radiologists, pathologists, surgeons) will be teaming up together to discuss each EHE patient;
2. Every patient's tissue will undergo genetic analysis and a comprehensive mapping of the tumor microenvironment;

3. There will be ongoing correlation of a patient's data together with the patient's EHE behavior and response to different drugs;
4. The Sloan Kettering team will be collaborating with other institutions and researchers involved in EHE, allowing the best minds to combine and participate in EHE research, while also having access to state-of-the-art technology at Sloan Kettering.

We congratulate Jane and all those in The EHE Foundation who have worked so hard to secure this support, and look forward to sending out a lot of positive updates in future editions of **The Pledge**.

Canadian Research progressing

Fiona Louise was delighted to be able to report: *“CANADIANS: the research here is moving forward. Dr Razak is looking for tissue and blood samples. Please message me if you are ok with me giving him your name. I will need your name, current doctor/ oncologist and province”*. Fiona sent the message to all Canadians she knew and continued *“Please if I have missed you or don't know you yet, contact me.”*

The project is not free of course, and Fiona continues to raise funds while seeking support from other Canadian patients. *“Also, please contact me if you want to donate to this project or can help with fundraising”*.

Fiona was also able to share a description of the project provided by Dr Razak:

“Over the last half century, it has become well established that cancers can elicit a host immune response that can target them with high specificity. Only within the last decade, with the advances in high output gene sequencing and bioinformatics approaches, are we now on the forefront of harnessing the host's immune system to treat cancer. Immunotherapy is the platform being used to access this immune response.

Hypothesis: EHE carries a genomic complexity that will have immune signature and a neoepitope that predicts response to immunotherapy.

Objective(s): To characterize the immune signature and molecular profiling of EHEs.”

We congratulate Fiona on pushing this important and exciting project forward and hope that others may throw their weight behind tissue samples as well as fund raising efforts.



02 EHE Research

Understanding the impact of TAZ-CAMTA1 on Endothelial cells

In January 2019, Emily Neil started her PhD studies in the laboratory of Dr Valerie Kouskoff at the University of Manchester. Her project, funded by an EHE-RCC fellowship, aims to determine how the TAZ-CAMTA1 fusion protein, frequently found in EHE tumours, affects the biological characteristics of endothelium, the cell type affected in EHE. In this project, Emily intends to define how the expression of this aberrant fusion protein alters endothelial cell identity and behaviour such as proliferation, migration or interaction with other cells. It is hoped that addressing these questions will provide a better understanding of the cellular and molecular mechanisms underlying this rare vascular cancer.

To address this series of questions, Emily is taking advantage of a model system established in the Kouskoff laboratory. In this experimental system, embryonic stem cells are differentiated to form endothelial cells in which the expression of TAZ-CAMTA1 can be induced by the addition of doxycycline.

To date, a series of experiments have been performed to evaluate culture conditions for the generation of endothelial cells from differentiating embryonic stem cells.

Using selected optimal conditions, it has been shown that expression of the TAZ-CAMTA1 fusion protein at the early stage of endothelial formation had a strong negative impact on this cell type. Emily observed that upon TAZ-CAMTA1 induction, the expression of key endothelial proteins was significantly decreased.

Preliminary experiments using a TAZ-CAMTA1 mutant protein that cannot interact with critical molecular partners showed that the mutant fusion protein did not alter the expression of these key endothelial proteins.

Further work is currently ongoing to determine how TAZ-CAMTA1 expression produces its effect, whether it is through changes in proliferation, control of gene expression or induction of cellular death.

New EHE research makes exciting progress

In our previous edition of *The Pledge*, we were delighted to report that the EHE group had concluded arrangements to sponsor a grant application submitted by Dr John Lamar, based at the Albany Medical College. The research is looking to test the hypothesis that the TAZ-CAMTA1 fusion protein that is core to EHE is subject to regulation by Hippo-pathway independent

mechanisms in our cells, and which could be exploited to inhibit TAZ-CAMTA1 function and so treat EHE. Dr Lamar's first update after just three months provided some very encouraging results.

The project team have been able to establish and confirm TAZ-CAMTA1 expression in mouse cells. In these cells they can also demonstrate significantly higher transcription involving YAP/TAZ-TEAD when compared to control cells and have shown that this is due to the TAZ-CAMTA1 fusion protein.

The next step was to test a number of possible genes to see if they could repress the TAZ-CAMTA1 fusion protein. The team were delighted to find 6 genes that do appear to repress TAZ-CAMTA1 when those genes are overexpressed in cells. Ongoing experiments have also verified that the proteins encoded by these genes also repress the expression of genes that are regulated by TAZ-CAMTA1 (Ctgf, Anxa2, Igsf10, Mmp2, Plcd3). The focus now is on introducing these 6 genes into cells so that stable expression can be induced and so test if this will indeed repress TAZ-CAMTA1 functions using *in vivo* mouse models. These results suggest that existing processes in our cells can be used to repress the activity of TAZ-CAMTA1 which is essential for EHE progress. Dr Lamar's team will

then start looking at therapeutic compounds that will stimulate one or more of these 6 genes in the hope that this will lead to new treatment options for EHE.

In preparation for testing these compounds, Dr Lamar's team have been experimenting with the introduction of NIH3T3-TAZ-CAMTA1 modified cells in mice to see if they will form tumours. Their initial tests produced large tumours in mice, with ongoing refinement of this process now underway. They have proved therefore that they can create mice with tumours which can then be used to test the impact of the 6 'controlling' genes above, enabling the accurate measurement and monitoring of tumour formation and growth.



The Albany Medical College

47 New Scotland Avenue
Albany, New York 12208-3479

Finally, Dr Lamar's team are currently optimizing soft agar and proliferation assays which will be used to test the influence of their 6 candidate regulators on TAZ-CAMTA1 function. They are also working on experiments with endothelial cell lines and have begun experiments to stably modify these cells with the TAZ-CAMTA1 fusion protein.

Latest 'EHE Research Update Teleconference' approaches

The next EHE Foundation "Research Update Teleconference" is now being scheduled. Over the last 3 years the foundation has arranged annual teleconferences to provide research updates including a review of current projects, latest developments, new ideas and future plans. Jane Gutkovich who leads the call will also provide some basic information on EHE anatomy and biology for any new members who may attend. The call will be held on 2 November and we encourage all those interested in EHE research to attend. Watch the EHE Facebook page for further updates.

Brian Rubin makes exciting progress

Drug development - Dr Rubin's team have concluded one very large 50,000 plus compound screen, the purpose of which is to identify a compound that directly inhibits the actions of the TAZ-CAMTA1 fusion protein that is central to the inner workings of all cases of EHE. They identified a single compound that made its way through the complete screening pipeline, a series of tests designed to identify a compound with the potential to treat EHE. They are now

working with Dr. Shaun Stauffer, a medicinal chemist, to understand the mechanism by which our candidate compound works. This will allow them to modify their compound to make it more like a drug. Dr Rubin is also pursuing a larger format drug screen to identify other compounds that inhibit TAZ-CAMTA1.

Genetically engineered mouse model (GEMM) of EHE - Dr

Rubin's team have been successful in developing the first GEMM of EHE. After several years of planning, genetic engineering, mouse breeding and waiting, they recently saw the first EHE in their mouse model. These lesions appear to be very similar if not identical to human EHE in appearance and they are positive for vascular differentiation markers, confirming that they exhibit vascular differentiation, just like human EHE. Furthermore, these lesions arise in the same constellation of tissues that EHE occur in humans. The team will spend the next several months developing cell lines from these tumors as well as characterizing the tumors at the molecular (RNA and DNA) level. This is the first bona fide biological model of EHE so it will open up a lot of potential studies for Dr Rubin's team and their collaborators.



02 EHE Research

Australia driving ISKS



Jane Biddlecombe and Jonathan Granek, Directors of the EHE Rare Cancer Foundation in Australia, have both continued to work hard throughout the third quarter to promote the International Sarcoma Kindred Study. Jane posted "Calling all EHE patients. Help create a unique EHE genomic resource. The EHE Rare Cancer Foundation Australia is pleased to announce our new collaboration with the Garvan Institute under the guidance of Prof David Thomas, a great supporter of the EHE Foundation."

The International Sarcoma Kindred Study is a unique, free to participate, resource that enables rare sarcoma communities, like EHE, to study genomic risks relevant to their cancer type. But it can only deliver meaningful results if patients participate, leading to the creation of a one-of-a-kind resource that will be available to researchers across the globe.



And to help encourage everybody in the EHE family, Prof David Thomas provided us with an EHE-dedicated message for all EHE patients.

"My name is David Thomas, and I'm Director of the Kinghorn Cancer Centre and Head of the Cancer Theme of the Garvan Institute of Medical Research, and an advisor to the Australian Rare Cancer Foundation. I'm writing to make you aware of the International Sarcoma Kindred Study (ISKS), which aims to create a genetic database for all sarcomas, including EHE. All patients with sarcomas are welcome to participate in ISKS, and participation is free."



I encourage you to sign up to the ISKS, so we can create a unique EHE genetic database

for answering questions about why EHE happens to some people and not others, and to open up new ways to better manage EHE. Research of this kind is essential to improving the lives of those affected by EHE. Contributing to the study is easy. The EHE Foundations worldwide are committed to signing up as many EHE patients to the ISKS as possible, and your participation and support is key.

Please join the study if you can, and be part of the search for answers to EHE."

We want to endorse Prof Thomas's message. Just like the PROM QOL study we reported on earlier this year, EHE patients participating could be key to progressing important research, and again showing that the EHE community is worth engaging with. So please register your interest now!

Development of an EHE biobank in the UK

An important requirement for research into any cancer is the collation of viable tumour tissue and related fluid samples within a biobank, that can then be accessed by research teams who wish to undertake relevant research. Due to the extreme rarity of EHE, we do not believe that such biobanking of EHE tissue and fluids has been undertaken previously in any country in the world.

In July 2019, the EHE Rare Cancer Charity in the UK entered into a two-year funding arrangement with the Joint Royal Marsden – Institute of Cancer Research Sarcoma Centre. The funding provided is for the post of EHE Tissue Manager. We hope that this role will ultimately help deliver a number of key objectives:

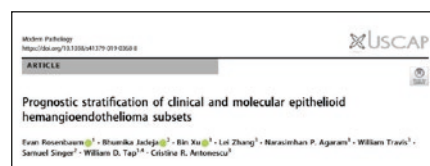
1. Collation and compiling of all historic tissue and clinical record sets from the past 20 years held at the Royal Marsden hospital (believed to be over 150 cases);
2. Development of ongoing tissue and fluid sampling protocols for all existing patients at the Royal Marsden (believed to be over 20 patients);
3. Set up and administration of the biobank facilities including all recording and storage in appropriate refrigerated conditions;
4. Expansion of the programme to include other key sarcoma centres in the UK;
5. Eventual inclusion of all UK oncology departments;
6. Reach out, engagement and enrolment of European centres;
7. Engagement and sampling access via the UK Posthumous Evaluation of Advanced Cancer Environment (PEACE) study allowing for detailed tumour sampling and analysis;

8. Engagement with research groups and the completion of tissue accessing procedures and protocols.

EHRC hopes that this programme will lead to a full and developing EHE biobank to support future research. We wish them luck with this important initiative and look forward to providing further updates in future editions of **The Pledge**.

Dr Antonescu publishes EHE Article

In late September Jane Gutkovich was thrilled to be able to bring to our group's attention the first EHE paper to come out from the team at Sloan Kettering, led by Dr Antonescu. "I am thrilled to provide a link to the EHE article that just came out this morning" commented Jane. "These are the findings of the first phase of the study sponsored by our EHE Foundation".



That is indeed exciting. We want to both thank and congratulate Dr Antonescu for her and her team's great work, and wish them ongoing success in their EHE research.

Making tissue samples available for research

Re-enforcing the message about tissue availability, Lisa De Young, Director of Patient Liaison at the EHE Foundation, contacted USA and Canadian members concerning access to EHE fluid and tissue samples. "First, I want to thank those of you who may have already willingly registered with pattern.org to provide fluid (ascites or pleural) OR fresh tissue from surgery (USA ONLY). Unfortunately, biopsy specimens are too small. It's a simple registration process and they coordinate via RCRF (Rare Cancer Research Foundation) with your doctor/surgeon to ship the specimen. Please allow at least 7-10 days PRIOR to your surgery for arrangements to be made."

We are also asking all our USA and Canadian patient members to consider making fluid and tissue samples available when they can. There is a video clip on the pattern.org website which will walk you through the registration process. And we also want to thank all those who have already transferred, or are in the process of transferring, their samples. It is these samples that will help in the ongoing EHE research we are promoting, and why every sample is individually so valuable.



03 EHE Fundraising

We are raising funds to support critical EHE research

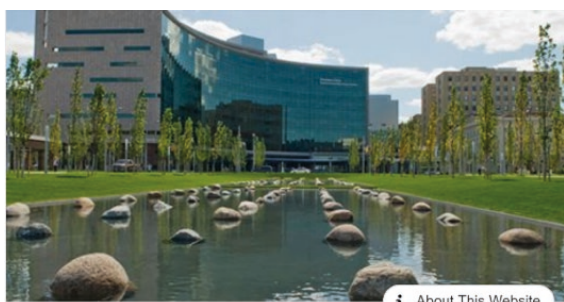
Today we are proud to have EHE-dedicated research being carried out in 4 different countries over three continents. The EHE Group remains totally focused on raising the critical funds needed to finance, drive and accelerate this existing EHE research programme, as well as the new and additional research that we hope will follow it. We thank all our members, their supporters and members of the public for their fundraising efforts, some of which are described below.

Two huge gifts from the Margie and Robert E. Petersen Foundation

In August, The EHE Foundation and the Cleveland Clinic announced breaking news that electrified the worldwide EHE community. The EHE Foundation and Cleveland Clinic had received a major gift from the Margie and Robert E. Petersen Foundation. One million dollars will be donated to the Cleveland Clinic to support EHE research conducted by Dr Brian Rubin, MD, PhD. Dr Rubin is a global leader in EHE research and this generous gift will support an ambitious research effort to develop effective therapies to treat the disease.

Medha Sutliff, Executive Director at the EHE Foundation stated *"After years of unwavering effort and commitment, The EHE Foundation Board of Directors are thrilled to announce a game-changing \$1 MILLION gift from The Petersen Foundation to the Cleveland Clinic!"* Medha continued *"I am incredibly proud of our Board and volunteers steadfast and laser-focused commitment to finding treatments and a cure for EHE!"*

However, this was only half the story, as The Petersen Foundation also agreed to match funds raised by the EHE group worldwide over the next three years, up to an additional \$1 million. *"This second gift, the matching facility, is so valuable to a cause like ours"* noted Julie Wahl, President of the EHE Foundation. *"It provides us with the ability to inform all our supporters that every dollar donated over the next three years will now be worth two dollars for EHE research"*. The matched funds will be added to the funding provided to Dr Rubin, who will continue to administer the Petersen gifts.



FIGHTEHE.ORG

The EHE Foundation and Cleveland Clinic Receive \$1M Research Gift from the Margie and Robert E. Petersen

"This new fundraising initiative is a win-win, as we can support two efforts simultaneously," said Dr. Rubin (pictured right), Chair of the Robert J. Tomsich Pathology & Laboratory Medicine Institute at Cleveland Clinic and a member of the Cancer Biology Program of Cleveland Clinic's Lerner Research Institute. "Support from people worldwide will go directly to our EHE research at Cleveland Clinic while also building the infrastructure of worldwide foundations that are raising awareness and additional funds for the research." Among those organizations are the EHE Foundation (U.S.), the EHE Rare Cancer Charity (U.K.) and the EHE Rare Cancer Foundation (Australia).

A heart-felt thanks to the Petersen Foundation for their transformational gifts. The EHE group will be working hard to ensure that the \$1 million sum available for matching is achieved, and are grateful for this additional gift to strengthen EHE fundraising, awareness and research across the globe.



EHE riders take on 100m ride in London

The 4th August saw 24 riders take on the Prudential-Ride-London-100 road event to raise funds for EHE research. Riders travelled from as far away as south Wales to take part. *"It's the second year we have had an EHE team riding"* commented Hugh Leonard.

"We could not be more grateful for their wonderful support, and raising over £10,000 was really amazing". Here are just a few photographs of the lovely EHE riders in their EHE riding jerseys!



03 EHE Fundraising

Cairns Clothes-Swap Party

On 31st August 2019, Angela Seirman from Cairns hosted her second annual EHE Clothes Swap Party. The day was a success with Angela raising \$1146 towards EHE Research.

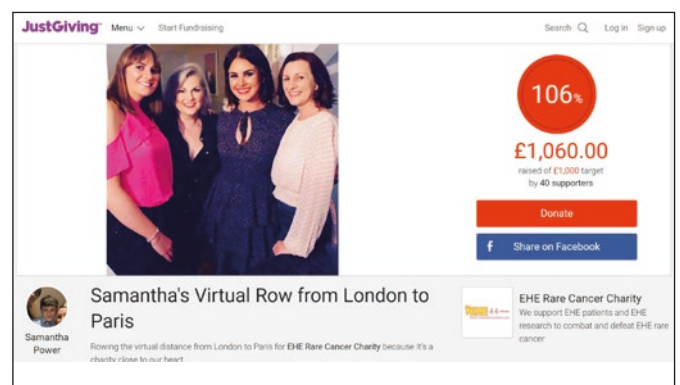
Thank you Angela for your continued support through this unique and fun event.

EHERelayRaisers still going strong

In our last two editions of *The Pledge* we reported the news of four great friends who are completing a 400-day Marathon in support of their friend, Allana Parker, while also raising critical funds for EHE research. Natalie, Samantha, Ellen, and Sophie are completing 100 days each. With legs 1 and 2 completed by Natalie and Ellen, raising over £2,400 between them, leg 3 is underway!

"My amazing wonderful Samantha has picked up the baton today and has started leg 3 of the relay raisers" posted Allana in July. "Not only is her life already full to the brim she's thrown herself in head first already today, rowing 6 miles of her 458 mile London-to-Paris-and-back virtual row! I'm so proud of them all and the work they are doing for this charity!"

We are also proud, very grateful, and continue to be amazed at the incredible effort and energy that the EHERelayRaisers have brought to their campaign to support Allana and EHE research. Well done ladies. We cannot wait to hear what Sophie's final leg will be. You are all EHE Warriors!!



Students' support for Fiona Louise

Fiona Louise was delighted when she was able to report more brilliant support from the students at her school. *"The students at my high school are actually starting an S.O.S. (Stomp Out Sarcoma) club in honour of me. We have already scheduled an event for next May and they want to spread it to other schools. Last year we raised about \$11,000."*

Congratulations to Fiona and her students for such an inspiring idea. It is because of groups like this that the EHE Group is able to make such good progress in the multiple research projects mentioned in the Research section of this newsletter.

Keeping up the shopping drive!

While we are rightfully celebrating the amazing gifts received this quarter from the Petersen Foundation and a private donor, we are also cognisant that we must maintain and encourage our grass roots fundraising which will continue to be the foundation of our fundraising efforts. One such method is the use of online shopping sites that offer donations to your charity of choice if you buy through them, with no extra cost to the buyer.

We encourage all our members and their supporters to use these shopping sites to help add 'free' funds towards EHE research.



amazonsmile
You shop. Amazon gives.

Same products, same prices, & same account (even with Prime!)

1. Go to www.smile.amazon.com
2. Sign in to Amazon Smile with your Amazon account
3. Choose **The EHE Foundation**, Hobart, WI as your charity
4. Shop on Amazon Smile and 0.5% of every eligible purchase will go to The EHE Foundation

Footy collection at MCG

Australia were delighted to be able to report news of the their 'tin-rattle' collection held over a two-hour period as the crowd arrived for an evening Aussie-rules football match at the Melbourne Cricket Ground stadium. Jonathan Granek wanted to congratulate and thank all those involved. *"Full credit must go to Jenny McDonald for championing the idea, organising the venue, date, volunteers, volunteer vests and tins. And huge thanks also to our volunteers: Ruby, Jim, Ros, Jackson, Reagan, Mikaela, Anny, Brooke, Shawana, Mia, Di and Don."*

The team raised \$538, which with matching from the Petersen Foundation will produce over one thousand dollars for EHE research! One of the two football clubs also kindly donated two team shirts autographed by the team. These will be auctioned by the EHE RCFA at a future event. We of course want to congratulate all involved for a great event.



The EHERCFA want to thank Odd Ball Pty Ltd who provided the tins and C&C Originals who provided the vests.



Odd Ball Pty Ltd
email: info@oddball.com.au
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www.oddball.com.au au-13 016 04 711

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03 EHE Fundraising

Private donation supports formation of new EHE team

With news of the Petersen Foundation gifts only a few weeks old, The EHE Foundation reported an astounding \$1million pledge from an anonymous donor to support additional EHE research. This incredible donation will be directed to Memorial Sloan Kettering Cancer Center in New York City to establish an International Center of Expertise in EHE Research. Further details of this can be found in the Research section of this newsletter.

Jane Gutkovich who has spearheaded this initiative noted *"As huge as \$1 million is, it will not cover the entire cost of the huge research plan that is still needed, but it will definitely lay the foundation for a very meaningful approach to patient care."* Jane continued *"And we have another million dollars arriving for EHE research at Cleveland Clinic, and another million that this community has already started raising to get a further million in matched funding to sponsor dozens of EHE projects. Who*

would have thought just a few years ago that we would be talking about millions of dollars going into EHE research?"

We join Jane in saying *"thank you to everybody for every big or small push in moving this huge mountain over the years! You made it happen!"*



Another great birthday fundraiser

The wonderfully generous trend of EHE family members using their birthdays to continue to raise funds for EHE research continued in the third quarter with Jane Biddlecombe launching her own version down-under.



Texel continue their great support

When we set up the EHE Rare Cancer Charity in 2015, our first corporate donation came from the Texel Foundation, which is the philanthropic arm of Texel Finance Limited, which specialises in credit and political risk insurance. Each quarter Texel organises a 5k or 10k fun run for the insurance community, with one run in London and the other in Singapore.

They use these events to raise funds for one of the charities they support, and in Q3 they raised funds for EHE research by supporting the EHE Rare Cancer Charity. We loved the thought that runners in London and Singapore will now be aware of EHE – that feels very good. And of course we send huge thanks to Texel and everybody who ran.

Emma's family fun-day a huge success

Emma Smith joined our EHE community in the middle of 2018. She quickly decided that she wanted to help in our battle against EHE, and together with her family and friends, threw herself into fundraising, starting with a **Justgiving** page. Emma then moved on to organise a family fun-day at her local pub, The Three Frogs in Wokingham near London. The event, held in July, was a huge success with great attendance from family and friends, and also from some of the pub regulars, and raised over £3,150, an amazing result. Emma's parents were there of course and told Hugh Leonard of the huge amount of work that Emma and friends had put into organising the event.

Emma's fun-day not only raised substantial funds for EHE research but also was reported in the local Wokingham Paper, providing more exposure for EHE amongst the public. Congratulations, Emma. You and the whole team rock!!



WOKINGHAM.PAPER.CO.UK

Wokingham pub's fun day helps raise funds for rare cancer charity - The Wokingham Paper



Motivation, focus and determination

Fundraising for EHE is a relentless process, but our group and all our wonderful supporters are equally relentless, driven by the understanding that EHE is a formidable adversary. We see and celebrate examples of this every quarter, such as Charleen Dardinski's post in September. Like many patients, Charleen is dealing with significant issues relating to her EHE, while exploring, understanding and managing her treatment options. Charleen shared about these issues, and simply added:

"The good news is that I was able to raise \$660 for The EHE Foundation so far :)."

It is another excellent example of how so many of our members, despite dealing with their health-related challenges, find the time, motivation and determination to still raise funds for EHE research. We thank and congratulate Charleen for her fundraising activities, and also all our members who continue to work so hard to raise the funds we need to ultimately defeat EHE. You are all awesome.



04 Upcoming events...

All Oar Nothing

In early August Sarah Medwin posted news of the latest fundraising day for the All Oar Nothing team, a four man-team that will be rowing the Atlantic Ocean this December to raise money for four charities including the EHE Rare Cancer Charity. Hugh Leonard noted *"what these guys are taking on is hard to get your head around. It is an unbelievable challenge. We cannot thank them enough for their wonderful support. The event is just around the corner, and we just want to wish all of them every success as they set out"*

And here's what the boat will look like with The EHE Rare Cancer Charity logo on the stern port side.

ALL OAR NOTHING
FUNDRAISING DAY @ TEIGNCREST GARDEN, SHALDON
SUNDAY 25 AUGUST 11:00 - 19:30
ALL OAR NOTHING ARE RAISING FUNDS FOR THEIR 3000 MILE UNSUPPORTED ROW ACROSS THE ATLANTIC OCEAN IN DECEMBER 2019 IN SUPPORT OF FOUR CHARITIES

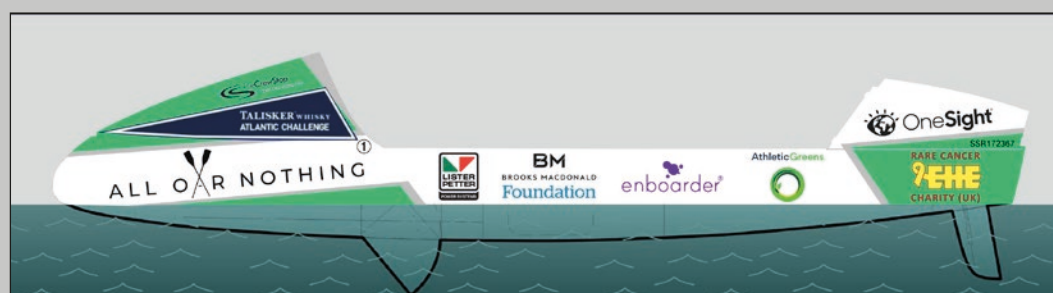
Come and see the boat, find out more about the row and enjoy games, cakes, raffle, prosecco and a rowing competition by **PB Strength & Conditioning**.

To enter the raffle, please see:
Sarah Mason
Katrina Drummond
Debbie Coyles
Amanda Redfern
£2 / raffle ticket

Raffle prizes include:
- A night in a Shaldon Beach Hut
- Cristal Champagne
- GHD Hair Straighteners from Andrew Hill
- £50 Clipper Voucher
- £25 ODE Voucher
Full list on the Facebook page.

IN SUPPORT OF:
OneSight Kidney Cancer UK EHE RIGHT TO PLAY

FOR MORE INFORMATION ABOUT THE ROW VISIT:
WWW.ALLOARNOTHING.CO.UK / @ALLOARNOTHING



EHE Masquerade Ball in February

Allana Parker posted news of the EHE Masquerade Ball that she and her friends are organising for next February. *"Posters have arrived for the ball so it's full steam ahead now!"* posted Allana. *"I can't wait to raise lots of wonderful money for EHE research."*

And as others have found in the past, the generosity of people when asked to help can be quite humbling.

Allana went on *"I received another fantastic auction prize from an old school friend for a week's holiday at her self-catering ski lodge in France! There really are some wonderful people in this world."*

We agree Allana, and cannot wait for this brilliant event.



05 And in other news...

A friend honoured

Dr Guy Weinberg is the founder of the Cravat Foundation, a regular contributor to the EHE support group, and also coordinates and promotes cancer research through the Telluride YAP, TAZ and TEAD Workshop. We are thrilled to share that Guy received the prestigious

Carl Koller Award by ESRA (the European Society of Regional Anaesthesia & Pain Therapy) for his outstanding lifetime contribution in the field of regional anaesthesia and pain medicine. Congratulations to Guy on this fantastic news.



TV Celebrity

Carl and Elizabeth Dickson shared a link to a TV advertizement, featuring their son, Broxdon, for the Denver Zoo. Great performance Broxdon, we think you are a star in the making! Broxdon is not the only TV star in the family, with his elder brother, Beckett, appearing in a commercial for a Denver law firm last year. Keep going boys, Hollywood is next!



A new arrival

At the end of August Bill Linda Cornell Eastridge posted news of a new arrival in their family. *"It's always nice to see good news. Our miracle EHE 12 year survivor meeting her brand new nephew. 2 well prayed for miracles. My heart is full".*

We also love these stories, Bill. Congratulations on the new arrival and thanks for the lovely photo of the new arrival with Aunty Brianna. We totally understand why this photo would fill your heart!



Getting together

We were delighted to see members of our group once again coming together to actually meet in person. Julie Wahl and Delaney both had the chance this quarter to meet JoAnna Jones and Cindi Titzer.



Julie Rivers Wahl posted *"I am so happy I was able to spend some time with this EHE warrior and her sweet mom! It is so wonderful to be able to sit and talk with someone who just gets it! So much love and safe travels to both of you!"*.

Delaney also spent time with JoAnna. Julie noted *"Don't let these cute faces fool you... these ladies are true EHE warriors!"*



Another 'new arrival'

The Pledge publishing team love to share happy stories of new arrivals when they are posted. Q3 is no different, although one new arrival this quarter is slightly different to previous posts. This is Hope who joined Adrianna Glennie in Scotland in August. We all agreed she is gorgeous and hope that she and Adrianna will share many brilliant times together.



05 And in other news...

Photo therapy

We know from many previous posts and past editions of ***The Pledge*** that photography can offer real peace and inspiration to the group. Recently we have seen many stunning photos posted by Anna Wydro and Artur Rozwalak. Here is just a small number of Artur's wonderful photos. Three of our favourites are of the deer in Richmond Park in SW London. Anna captured the mood perfectly with her accompanying posts *"Sharing calming deer encounter we had the pleasure to have yesterday at Richmond Park, especially thinking about those who are at home and need inspiration during treatment or recovery."*





Anna also posted some wonderful black and white images shot by Artur while they took the opportunity to travel around UK and discover some of its beauty spots. Anna also posed the questions as to whether others would like to join in and perhaps create an EHE calendar. We think that is a great idea, and with photos like this we know it will be a success.

Anna finished her post with a simply message to the worldwide EHE community. *"Wishing you a good day whatever time it is for you, and wherever you are!"*





The EHE Foundation (USA)

www.fightehe.org

The EHE Rare Cancer Charity (UK)

www.ehercc.org.uk

The EHE Rare Cancer Foundation (Australia)

www.ehefoundation.com.au