

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



Quarterly Newsletter
for the EHE Group
July - September 2018

the pledge

Edition 14



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Welcome

We warmly welcome all our readers to **“The Pledge”**, covering the third quarter of 2018.

As always we want to start by saying a massive thank you to all our supporters for their contributions. Whatever the form of support that you provide, and regardless of magnitude, they are all critical to our success and ability to ultimately defeat EHE.

Thank you to all of you, and we hope that you enjoy this newsletter.



Highlights

Valuable collaboration with National Cancer Institute progressing

The NCI is now actively engaged in EHE research and is collaborating with the EHE Foundation to explore additional ways to assist in understanding EHE.

EHE research in Australia progresses on several fronts

The EHE Rare Cancer Foundation Australia has seen significant progress with EHE research on a number of fronts.

Two EHE research projects contracted in the UK

The EHERCC has placed two research contracts this quarter. One is to develop an EHE Zebrafish model. The other will seek to explore how EHE affects endothelial cells.

EHE inclusion in Canadian research starts to firm up

Fiona Louise has engaged researchers in Canada who want to help identify appropriate immunotherapy treatments for EHE. Fundraising for this research is now a major focus in Canada.

PROM Quality-of-Life survey completed by over 100 members

The EHE Quality-of-Life PROM study being carried out in Holland has now secured 114 completed questionnaires, with more in the pipeline, and will close this phase of the project in early October.

Further details about all these developments can be found in the 'Research' section of this edition of The Pledge.



01 Patient Support and Advocacy

Supporting those diagnosed with EHE is one of our core objectives

One of our core objectives is ensuring that we provide support to all those diagnosed with EHE. We will do this, wherever a patient may live. At the same time we will strive to increase awareness of EHE amongst the general public, government and in some cases the medical and research community. We will also work hard to make clear what the impact that living with a rare and frightening cancer has on a person's life. We thank all our supporters who have contributed to this critical part of our activity, examples of which are provided in this section.

UK Cancer specialist engaged

In mid-July, Hugh Leonard met with Prof Peter Selby and Dr Tze Wah in Leeds. Prof Selby is a hugely respected member of the UK's oncology community. At the start of the meeting Prof Selby explained that he had asked the heads of each part of their MDT (multi-disciplinary team) structure for their experience, knowledge and thoughts about EHE. It was certainly a positive introduction.

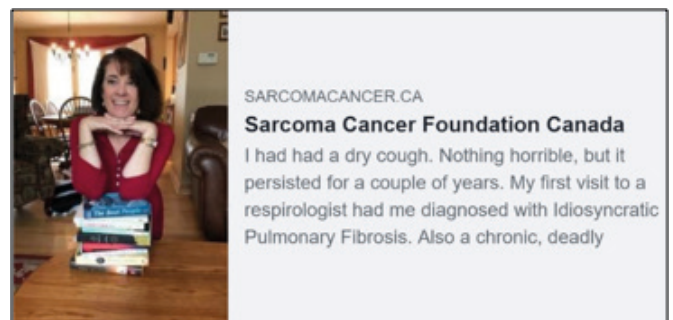
Prof Selby was very interested in all that we are doing, our worldwide coverage, and our ability to deliver a more meaningful data base of information from our patient group. He said at the end that he was impressed that we are "rattling cages" and getting people to talk about EHE because with ultra-rare cancers, unless somebody does this, then the reality is that not much happens.

Hugh provided an update on the ongoing research we are funding as well as our broader research ambitions. He also talked about the EHE PROM study being run out of Holland, our large retrospective clinical study being organised in the USA and the Treatments and Outcomes Case Series (TOCS) study being prepared in Australia. Hugh commented "It's a nice story as it really drives home our international collaboration and the strong patient involvement."

Overall it was a very constructive meeting, with positive endorsement from a recognised oncology 'heavy-weight'. Hugh noted *"I think the overall message for all our members is keep going; keep delivering our patient support and fundraising, advocacy and in particular any patient surveys when they come out. It's our global group effort that is so eye-catching to people, and will help us change how EHE is considered."*

Lifelong ambition achieved?

Fiona Louise, an English teacher, had always harboured a secret desire to be published. In August she finally achieved it with an article produced by the Sarcoma Cancer Foundation of Canada. "OK," as Fiona noted, *"not quite the way I want to be published, but here we are"*. We are sorry Fiona that it is not a No 1 Best Seller, but we love the fact that you are helping to get the message out there?



Just Live

A key goal of all three of the EHE foundations is to increase awareness of EHE. One way our members do this is by getting the EHE 'Just Live' heart-beat tattoo. Carla Renee joined that group in July and posted her own version with a simple message *"I woke up wanting a tattoo so why not! #JustLive"*. Holly Mcrae also posted a photo of her tattoo, celebrating 10 years of survival with EHE. Holly was about to complete her last treatment on the MOST trial. Holly accompanied her photo with a simple message: *"12 of the hardest months of my life. Stay strong my fellow warriors. Just live."* We love the fighting spirit, Holly, and look forward to the photo of the 20 year survival tattoo in 2028! We also love the emblem, and hope that where people have them, it may lead to questions, explanations and greater awareness of EHE in general..



Medical details beautifully presented

Adrianna Glennie posted a photo and message on the EHE Facebook page concerning a friend who owns a jewellery company. She wanted to provide Adrianna with the means to carry and present her medical condition and drug regimen in case it was needed if Adrianna was perhaps incapacitated. Mama Tiger Lily (her face book name) had made Adrianna a bracelet to wear and a card for her phone slot, providing some important medical information. The card has all Adrianna's drug details on the other side.

"I love it so much" said Adrianna. Others who might be interested can contact Mama Tiger Lilly through her facebook page at <https://www.facebook.com/mamatigerlily/>. Costs will depend on the amount of detail, but prices look very reasonable.



01 Patient Support and Advocacy

JCK rides Pelotonia

Jenni Kovach rode in Pelotonia, in Ohio, a bike ride where 8,000 riders gather and raise millions of dollars for their local cancer hospital and cancer research. Her 7th time riding the event, Jenni rode with her sister and her work friends *"It was more emotional this year now that I am a survivor. I thought of all of my fellow EHE warriors during the ride. Just Live!"*. We congratulate Jenni and love her EHE fighting spirit.



Canada brand is growing

Fiona Louise joined the EHE family in 2017 and has become an extraordinary campaigner for all things EHE. She has organised an EHE dedicated fund through the Sarcoma Cancer Foundation of Canada, as well as encouraging and securing the first EHE research in Canada. As part of this process our Canadian EHE group needed to establish their own branding. We were delighted therefore to see some simple but hugely effective Canadian logos produced by LeeAnn Deacon Conner. We love the impact of these logos and encourage everybody in Canada to throw themselves into the battle.



Laney goes off the rails... or more precisely off the road

Spreading the word about EHE sometimes just happens as a consequence of an event. Delaney Wahl's 'Make-a-Wish' experience at the 49th Annual Crandon World Championship off road races gave Delaney and her family a weekend to remember in Crandon, WI! The weekend included a chartered fishing trip, a parade with Delaney as the guest of honor, concerts, news interviews, extraordinary hospitality, and lots of off-road excitement as over 200 teams of trucks, open-wheel cars, UTVs, and karts battled for wins in their respective class. Delaney and her family gained many new friends, and as a bonus, lots of exposure for EHE. We know that Delaney had a great time, so we all want to thank the entire Clear by 1 Motor Sports team and their many sponsors for giving the Wahl family an experience of a lifetime and for spreading awareness for EHE.



Love, perseverance and a fighting spirit

The third quarter of 2018 was a tough period for our worldwide group, with several members of our beloved family losing their respective battles against EHE. Lisa De Young was just one of many who were deeply affected by these events, so when she saw the story of Justin and Morgan Singer she wanted to share it. *"I wanted to share their story which was just published by the Cleveland Clinic. It is about love, perseverance, and never giving up HOPE!"* The story can be found at: <https://newsroom.clevelandclinic.org/2018/09/04/husband-fighting-rare-cancer-after-wife-persisted-he-seek-second-opinion/>.



Jane wows the Rotary Club

On Thursday 30th August Jane Biddlecombe, a director of the EHE Rare Cancer Foundation Australia was invited to address the Darwin Rotary Club to talk about EHE, the Foundation, and the forthcoming 10,000 Steps Challenge. The Rotary Club is a worldwide organisation that brings together community minded business leaders whose aim is to promote goodwill and assist community groups to fundraise, and so they wanted to know how they could help? It was a great evening, with Jane once again wowing her audience. They closed with an impromptu whip-round and raised \$154 right there and then. We thank them for their help, and also have to congratulate them on great shirts!

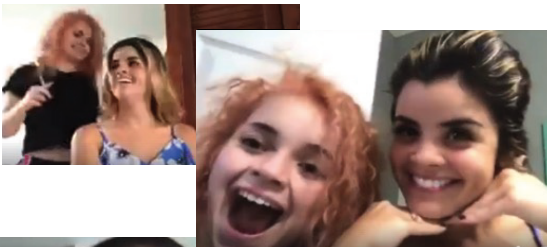


01 Patient Support and Advocacy

Still smiling!

In early September Noah Anthony posted a video of his cousin, Fransheska. *"Here's a quick little video of my sweet, little cousin, Fransheska Nazario, as she's getting ready to start treatments again. Great video. Cool music ♥♥"*.

We cannot replicate the video on this page, but we can try and capture Fransheska's great strength and determination, perhaps best characterized by her wonderful smile. Thank you for sharing such an inspirational video, and we of course want to send our very best wishes and love to Fransheska as she continues with her treatment plan.



Our own Caledon east woman in the news!

In early August the Orangeville.com website ran an article about Fiona Louise and her bid to raise the \$50,000 needed for EHE research. The article provided an excellent description of EHE, and the stress of living with an EHE diagnosis. It also described the need, in Fiona's eyes, for Canada to catch up other countries like the USA, UK and Australia in terms of EHE research taking place.

Fiona posted *"And here I am grasping onto my 15 minutes of fame. This is the "big city" of 20,000 people near me. I hope it brings in more donations or even a corporate sponsor of sorts."*

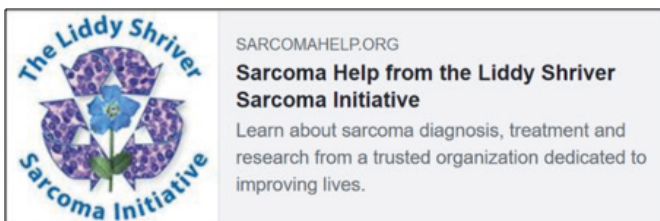
We congratulate Fiona Louise for her fantastic energy and determination, and we hope that the corporate sponsor turns up soon!!



EHE Outreach

Lisa De Young continues to represent the EHE Foundation in **SARC** (*Sarcoma Alliance for Research through Collaboration*) endeavours. For example, together with Jane Gutkovich, Lisa has been participating in teleconferences relating to the SPORE grant which Dr Brian Rubin and Dr Scott Schuetze are part of. Their project is one of three that make up the SPORE grant, which SARC has just resubmitted for grant funding approval. The SPORE (Specialized Programs of Research Excellence) program is a National Cancer Institute (NCI) grant program designed to promote collaborative, interdisciplinary translational cancer research. The Mekinist (trametinib) clinical trial is an example of this collaborative effort.

Lisa is also on a steering committee (through SARC) which is looking at the viability of taking over the Liddy Shriver Sarcoma Initiative website (www.sarcomahelp.org). Discussions are underway on how best to update this valuable resource and keep it well maintained.



A huge part of our advocacy objective is to increase awareness of, and engagement with, EHE. Lisa and others are playing a key role in this through their engagement with such important groups such as SARC. We see many expressions of deep and heartfelt gratitude for what Lisa does not only with SARC, but by supporting our EHE members wherever they are and whenever they need help, and we wholeheartedly endorse those sentiments.



02 EHE Research

Only through research can we answer questions and defeat EHE

The following notes provide a summary of key points relating to the EHE research that is already in progress, as well as projects that are in late-stage planning and those that are being discussed and considered.

Update from Dr Rubin

Drug screening

Dr Rubin's EHE team have now completed the first generation of their drug screen and they are working on their second generation drug screen. Once these screens have completed rigorous validation they will screen multiple drug libraries to identify molecules that could be pushed into drug development.

GEM model

Dr Rubin's genetically engineered mice (GEM) are being crossbred with mouse strains that should activate the EHE causing Wwtr1-Camta1 gene fusion. They have evaluated several early mice and have not seen tumors so far. However, their analysis indicates that they are able to activate the oncogene so they hope that they will begin to see EHE-bearing mice soon.

Others

Dr Rubin is very interested in determining what Camta1 interacts with in the Wwtr1-Camta1 protein complex. His team have started complex studies using mass spectrometry to identify proteins that bind to Camta1. Such proteins may represent additional drug targets for the treatment of EHE.



Dr Rubin

Great progress with the National Cancer Institute (NCI)

The third quarter has seen exciting progress with the EHE Foundation's engagement with the NCI. In August communications progressed culminating in a conference call with Jane and others from the EHE Foundation, Dr Rubin, and Dr Karlyn Reilly (a Director of the Rare Tumor Initiative) and Dr Brigette Weidenmann (A Director of the Childhood Cancers Program).

Dr Rubin updated all on the call about his research progress, and in particular on the challenges of working on: (i) targeted therapy for EHE; and (ii) the development of an EHE cell line or mouse model. Excitingly the focus of the meeting was on key issues such as how NCI experts can help with EHE; understanding why it behaves so differently in different organs; how we might attract more scientists to work with EHE; and finally, how to best engage with and involve our EHE community in the entire process.

Then in September Jane finally was able to meet face to face with the leaders of NCI Rare Tumor Initiative program, and was delighted to be able to report that EHE is now being studied by the NCI.



Jane will be providing a more detailed update on her next EHE Research Update videoconference on 13th October, but here are the headlines:

1. Research completed by the NCI on EHE tissue has identified a possible target for which there is an existing drug that is already in clinical trial.
2. The collaboration between NCI and the EHE Foundation will include engaging our patient community in the study "Natural History of EHE". The Foundation will be arranging for tissue and medical records to be sent to the NCI who will undertake detailed analysis, looking for patterns in both EHE behaviour and in EHE response to different treatments.
3. The preliminary analysis of EHE tissue suggests that some of the clinical trials currently being run by the NCI could be open to EHE patients.

These are just the immediate headlines, but Jane believes that other opportunities may present themselves as the collaboration grows. She has promised the NCI that we are very focused and able to assist with patient communications. Jane has also promised that she is available and can meet often if required.

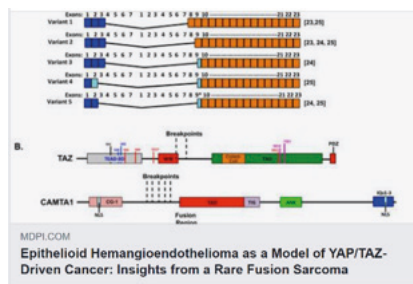
Throughout these discussions one key message came through loud and clear. Tissue is critical, and in particular, fresh or frozen tissue. Without it, so much research is blocked to EHE. This has to be a primary focus for all our foundations going forward, so please please let us know if you are facing surgery. We need as much notice as possible as hospitals have hugely bureaucratic procedures, so we need time to get ready.



02 EHE Research

Dr Guy Weinberg of Cravat Foundation publishes new EHE paper

The EHE community sent their congratulations to Dr Guy Weinberg of the Cravat Foundation (and member of our EHE community) on his excellent publication in July on EHE biology and the ways in which EHE research can lead to positive discoveries for other cancers. The possibility of positive impacts for other more common cancers is an important factor in promoting EHE research, and attracting funding. We are therefore extremely grateful to Guy for his time and dedication in producing this work which discusses and describes these benefits in a rigorous scientific paper.



PROM QOL Study update

On September 10th Marije Weidema, one of the researchers behind the PROM Quality of Life study being undertaken in Holland updated the group through the EHE Facebook page. The great news was that 114 patients have taken part in the study by completing the online questionnaire. This is quite a milestone for such a rare sarcoma, involving participants across Europe, Asia, Australasia and North and South America. The survey access is about to close, after which we will await the results. We do however want to register our thanks for all our members who took the time to complete the questionnaire.

Canadian EHE Research to progress

On 15th September Fiona Louise posted an exciting update from Canada. *“Looks like Canada is joining the EHE fray with real research. This is a write up from “our” lead doctor that is being brought forward to solicit more donations.”*

Epithelioid hemangioendothelioma (EHE) is a particularly difficult sarcoma to treat. In the past, this disease has been classified as one entity together with other soft tissue sarcomas. However, with the advances of science, we now know EHE is a distinct disease by itself and a ‘one size fits all’

therapy approach is ineffective. There is a huge need to find exact and effective treatment for this condition - ie: personalized medicine.

To overcome this challenge, Dr. Albiruni Abdul Razak, a Medical Oncologist at Princess Margaret Cancer Centre, Toronto, aims to profile EHE (once diagnosed) from an immune signature standpoint. He hopes to generate data so that exploration of immunotherapy in this disease could be carried out. Recently, Dr. Razak and his group were able to demonstrate why immunotherapy may be useful in patients with another type of rare sarcoma, called alveolar soft part sarcoma. In the past 5 years, his group has led and participated in several key studies involving targeted and immunotherapy in the treatment of sarcoma patients. Supported by collaboration with the pharmaceutical industry as well as money from philanthropy, Dr. Abdul Razak works closely with scientists and physicians at Mount Sinai, Princess Margaret Cancer Centre, and Sick Kids Hospital, as well as international collaborations. It's an example of how scientific collaboration traverses the boundaries of different institutions. *“Our work bridges Mount Sinai, Princess Margaret and beyond. We all rise above our institutions and work together in search of new treatments that improve patient care and benefit sarcoma sufferers.”*

In order to support Dr Razak's effort, Fiona Louise (Ross) is aiming to raise \$50,000 to start EHE research in Canada. Recently diagnosed with the disease, Fiona wants to get Canadian oncologists and research to a level where they have something to offer EHE research and treatment, using some of the best minds the world has to tackle this unique sarcoma. We want to wish Fiona and all our Canadian supporters success with their campaign to raise these funds.

As Canada strives to raise research funds, we also want to remind everybody that Canadians can now donate for EHE research in a tax efficient manner with tax receipts provided using the Sarcoma Cancer Foundation of Canada. Fiona explained *"write EHE in the online message box; or send it in via cheque with the donation sheet in old fashioned snail mail. Better yet make a monthly donation. Feel free to share with friends and family. Let's get Canadian research started and funded. #justliveeh"*.



SARCOMACANCER.CA

support

Every gift to SCFC, no matter the size, makes a positive impact on patients and families across the country, providing opportunities for research, awareness, and support. Whether you give once a

UK research update

1 EHE Zebrafish model

In the third quarter the EHE Rare Cancer Charity in the UK was delighted to sign a one-year research contract with the Bateson Centre at the University of Sheffield, the first dedicated EHE research project in the UK. Working in collaboration with Dr Rubin, the project will explore the feasibility of creating a valid EHE model using Zebra Fish, attempting to genetically modify the fish to express the TAZ-CAMTA1 protein in endothelial cells, and thereafter describe the phenotype associated with this model, focusing on the vascular morphology. TAZ-CAMTA1 is used as this fusion protein is believed to drive EHE.

September saw the start of the project and the first brief update from Dr Fredericus Van Eeden, one of the Principle Investigators, suggested a good start had been achieved!

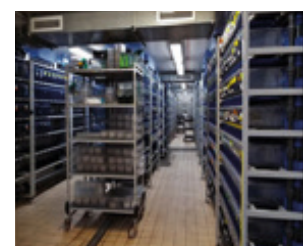
"I just wanted to give a quick update, as we have just started working on the EHE project. We received the EHE clones, and because it is a very essential "building block" on which most of our work will be based, we decided to recheck the sequence of the inserts and found them to be perfect; thanks Brian.

Eleanor has successfully cloned the gene into a plasmid that can be used to make mRNA for embryo injection. This will allow us to do global overexpression of the fusion gene in the embryo, and see its effect. This week we will hopefully do a first round of injections in a fish line where embryos have fluorescent blood vessels, so that we can see if there are major effects on their formation.

Also, a tricky PCR (polymerase chain reaction) that was needed to get it into the Gateway cloning system (that will be used to make transgenesis construct) has worked.

In short, we seem to have made a good start!"

We look forward to more good reports over the next 12 months!



02 EHE Research

2 Stem cell project

The second UK research project will be a PhD carried out at the University of Manchester, where Dr Valerie Kouskoff will be the PI. The EHERCC was delighted to sign the appropriate contracts with the University of Manchester at the very end of September. Dr Kouskoff hopes to start the PhD at the start of 2019 and is now advertising for a suitable post-graduate candidate.

This project will set out to address the following questions:

- i) How does TAZ-CAMTA1 expression affect the biological characteristics of endothelial cells?
- ii) How does TAZ-CAMTA1 expression modify the transcriptional landscape of endothelial cells?
- iii) Can TAZ-CAMTA1-expressing endothelial cells engraft in mice and provide an in vivo model of EHE?

Dr Kouskoff also hopes to create induced pluripotent stem cells using patient-donated tumour cells. These pluripotent stem cells will represent a unique and powerful approach to study the biology of EHE. The research will aim to understand better how the EHE cancer cells arise and proliferate. This research will also aim to generate and maintain enough EHE cancer cells in the laboratory to test drugs that could stop the proliferation or kill these cells.

Australian Advisory Board expands

EHE-RCFA's Director of Research and Clinical Development, Jonathan Granek, held meetings with Prof Kieran Harvey who recently joined the EHE-RCFA Advisory. Prof Harvey brings a wealth of expertise to the team, in particular through characterising the role of the Hippo pathway in human cancer using cell lines, patient samples and animal models.

Prof Harvey holds dual appointments. At Monash University he runs a laboratory in the Department of Anatomy and Developmental Biology and at the Peter MacCallum Cancer Centre he leads the Organogenesis and Cancer Program.

Prof Harvey has previously worked with sarcoma expert Prof David Thomas and more recently met with rare cancer expert A/Prof Clare Scott, Senior Research Fellow Dr Holly Barker and Jonathan to discuss the EHE Study efforts underway at Walter and Eliza Hall Institute of Medical Research (WEHI-EHE; see more below) and additional research opportunities.

Gaining a Better Understanding of EHE

In a collaborative effort, Prof Kieran Harvey's lab together with research labs at Canada's Mt Sinai Hospital and USA's University of Iowa have been studying EHE's TAZ-CAMTA1 and YAP-TFE3 fusion proteins and how they lead to and influence cancer progression in EHE.

Using transduced cell lines and xenografts (mouse models), the data from the study indicates that the cancer causing properties of these fusion proteins can be explained by their nuclear enrichment, dysregulation of the upstream Hippo pathway, and interaction with key transcriptional regulatory proteins.

The team is confident that future studies will help identify additional ways in which these fusion proteins transform cells and initiate and promote the development of EHE, potentially resulting in new therapeutic targets.

WEHI Stafford Fox Rare Cancer Research Program's EHE Study (WEHI-EHE)

EHE research under the leadership of rare cancer expert A/Prof Clare Scott and Senior Research Fellow Dr Holly Barker is starting to take shape. The study is open to EHE patients throughout Australia and involves:

- Collection of patient data;
- Biobanking (archival blocks, fresh tissue, blood, etc);
- Development of pre-clinical research models (cell lines, organoids, patient-derived xenografts); and
- Identification of novel EHE treatment options.

During the last quarter, WEHI-EHE saw the first of its EHE patient enrolments, data collection and specimen collection from a patient's surgical procedure.

Please contact the EHE-RCFA (info@ehfoundation.com.au) if you have any queries regarding this study, wish to participate, or to notify us of a recent/upcoming surgical procedure. For upcoming surgical procedures, please provide as much notice as possible (minimum 24 hours). Over the coming months, the EHE-RCFA will be updating its website to help streamline patient notification of upcoming surgical procedures and enrolment in the study.

Australia's Innovative Molecular Screening and Therapeutics (MoST) Program

Sarcoma expert Prof David Thomas and his team have developed MoST, an innovative clinical trials program, to evaluate the effectiveness of new treatments and help patients with high-mortality cancers like EHE.

MoST builds on partnerships across multiple academic research organisations and cancer centres, the public health sector, and the pharmaceutical industry, including the NHMRC Clinical Trials Centre (University of Sydney), the Peter MacCallum Cancer Centre, the Kinghorn Cancer Centre, Chris O'Brien Lifehouse, AstraZeneca and Pfizer.

In the MoST program, tumour samples from patients undergo genomic/molecular screening. Patients are then assigned to clinical substudies of targeted therapies based on their specific genetic variations. The MoST program looks to understand how targeted therapies work and find new biomarkers that can predict which patients will benefit from these treatments.

The MoST program is also conducting clinical studies to test novel immunotherapy drugs in patients with high-mortality cancers like EHE. The first MoST

immunotherapy substudy looks at so-called 'immune checkpoint inhibitor' drugs that take the brakes off the anti-tumour immune response so immune cells can attack cancer cells. This three-year clinical trial is open to patients who cannot be matched with a targeted treatment. A second immunotherapy study will combine a targeted treatment with a checkpoint inhibitor and will recruit patients with specific genetic abnormalities in their tumour.

Although immunotherapies are proving to be effective in many cancer types, they do not work in all patients. Prof Thomas and his team are looking to find biomarkers that can predict which patients will benefit from specific immunotherapy treatments to better understand how they work to fight cancer for a more personalised approach for patients.

Several biomarkers have been suggested to predict response to treatment with immunotherapies, including the presence and change of certain immune cells in the tumour and blood, patterns of immune-related gene expression, proteins released by immune cells, inhibitory proteins displayed on tumour cells, genetic changes in tumour cells and inherited genetic changes in immune genes.

The EHE-RCFA continues to maintain contact with the EHE patients enrolled in MoST and wishes them well with their treatment.



02 EHE Research

Treatments and Outcomes Case Series (TOCS) Questionnaire

The Treatments and Outcomes Cases Series (TOCS) Questionnaire is a EHE-RCFA joint-venture study under the leadership of sarcoma expert Prof David Thomas and Oncology Fellow Dr Rajiv Shinde. It involves a 15-minute online questionnaire for patients from across the globe to complete. The study aims to:

- Provide insights into patient demographics, disease presentation, progression, treatments and outcomes;
- Identify hospitals / institutions / professionals with expertise in various aspects of care;
- Contribute to a growing international body of knowledge to support patients, clinicians and researchers now and into the future; and
- Identify priority areas for new research.

EHE-RCFA directors Jane Biddlecombe and Jonathan Granek met with Prof Thomas and Dr Shinde on 30th August 2018 to finalise the questions for the upcoming TOCS study, including input from UK and USA-based foundations. The proposal is currently undergoing Human Ethics Approval and we hope to receive approval very soon. The intention is for the study to go live in Q4 of 2018 and present the findings in time for ASCO 2019.

Empowering the EHE Community to Enable Research through the Centre for Analysis of Rare Tumours (CART-Wheel)

Over 10 years ago, Cynthia Pollak, the parent of an EHE patient in Melbourne, Australia, began her own registry of patients with similar cancers (the H.E.A.R.D. Support Group Registry) in response to the lack of information and treatments available. Cynthia was able to gather data from over 200 voluntary international participants over the duration of her work. Clinicians and researchers saw the value of such an approach but without the involvement of the research community, it lacked the necessary Human Ethics Approval required for the data to be collected, stored, and used for research.

Under the leadership of rare cancer expert A/Prof Clare Scott and with Cynthia's assistance CART-Wheel was developed in Australia to help overcome these challenges and facilitate research into rare cancers such as EHE. Through CART-Wheel, EHE consumers (patients, guardians of patients, and next-of-kin) can now be actively involved in the research process, making it possible for researchers from all over the world to readily identify and access quality patient data and cases for their studies in accordance with Human Ethics requirements. Recently EHE-RCFA

director, Jonathan Granek, has been working with A/Prof Scott and the rest of the CART-Wheel team on further developments including the recent addition of an electronic/online participant consent feature.

With the support of the EHE-RCFA, the CART-Wheel team invites EHE consumers from all over the world to participate by registering and filling out the privacy-protected, internet-based questionnaire. Participants have control over their personal data by providing consent to different uses of their data including permission from participants to:

- Allow their treating clinician to enter, view or edit their data;
- Contact their doctors to obtain histologic report(s) and medical details to confirm their tumour type; and
- To be contacted regarding the possibility of participating in other ethically-approved research projects studying EHE. Other such projects may, for example, seek to collect more detailed information from you about your case; may inform you about a clinical trial that may be relevant for you; may request a blood sample from you if that is convenient; or may request access to the use of biospecimens previously collected and stored in a biobank.

CART-Wheel is an initiative endorsed by the Clinical Oncology Society of Australia, Victorian Cancer Agency, Melbourne Health, Walter and Eliza Hall Institute of Medical Research, BioGrid Australia, Rare Cancer Australia, Cancer Voices Australia, Picchi Brothers Foundation Inc., and EHE Rare Cancer Foundation Australia.

The EHE-RCFA encourages EHE patients from across the globe to support current and future research initiatives by visiting the CART-Wheel website and registering (<https://www.cart-wheel.org>). Please contact the EHE-RCFA (info@ehfoundation.com.au) if you have any queries on this study, or are a researcher with an ethically approved research project looking to access the data.



03 EHE Fundraising

We are raising funds to support critical EHE research

We will ultimately manage and eventually defeat EHE through a dedicated and focused research effort that will deliver answers to the many EHE questions that cannot be answered today about this rare cancer. The EHE Group is completely focused on raising the critical funds needed to finance, drive and accelerate our expanding EHE research programme. We thank all our members, their supporters and members of the public for their fundraising efforts, some of which are described below.

Aiming high to beat EHE!

In early July Melissa Clark McPherson posted on Facebook news of her 5K hike as part of the US Virtual 5K campaign. *"Take that EHE- 5 mile hike to the summit of Mt. Battie, Camden Hill State Park, ME. It took me two hours to get to the top but I did it!! Just live!♥"*. Congratulations Melissa on a stupendous effort..



Cold, wet and tired, but still smiling!

Last year, Claire Coddington became the first of our EHE group to ride the Prudential RideLondon100 cycling event to raise funds for EHE. This is a 100 mile ride from central London, out into the Surrey countryside, and back to central London. This year we had a four-man team enrolled and so on 29th July, Scott Milligan, Dominic Steinitz, Nic Brown and Mark Schaeffer rode the race in their EHE cycling jerseys to raise awareness of EHE, and at the same time raised about £3,500 for the EHE Rare Cancer Charity. Here are some pictures taken of the race, from start to finish.

We want to thank Dom, Scott, Nic and Mark, and Steven Barr who helped organise them, for supporting us on a day where the 100m ride was through heavy rain and strong winds. It was an awesome performance!!



Mission possible!

Fiona Louise is a woman on a mission, and that mission is to beat EHE. Fiona has already set up a collaboration with the Sarcoma Cancer Foundation of Canada, creating a dedicated EHE fund that will allow Canadians to donate to EHE research within a tax-efficient not-for-profit structure. She has also found a Canadian researcher who wants to work with EHE.

Fiona has also been on the Fundraising trail and in Q2 reported raising over \$10,000 for EHE research. She has not stopped there however. In July Fiona updated our worldwide EHE family on two new initiatives; her bottle drive and PaintNite, both in Caledon in Canada.

Her bottle drive had people donating empty bottles so that Fiona could recover the deposits for EHE. She estimated that she could raise \$300, but soon surpassed this total. On the day Fiona posted *"We're at \$532 and bottles are still being dropped off!! We have another \$415 in donations and a local restaurant has offered to donate all their empties for the next year"*.



Her Paint Nite was also great fun. *"Great paint night fundraiser with friends, colleagues and neighbours. Thank you so much. Another \$300 for EHE research."*

Overall Fiona had raised well over \$1,000 from these two simple ideas alone. We again congratulate Fiona for her fantastic efforts and we send a huge thank you to everybody who took part donating bottles or painting some great pictures



03 EHE Fundraising

Mabel's Labels

Fundraising comes in many different forms, and it is not just the 'very big' that matter. Lots of smaller sums all add up. Fiona Louise was signed up by a friend to the Mabel's Labels Fundraising site. You just need to organise a sales event and the cause of your choice will get 20% of your sales. You can also buy directly and nominate a cause. Fiona Louise called on all Canadians with children to use Mabel's Labels and donate to the EHE section of Sarcoma Cancer Foundation of Canada (EHE). We hope that many will answer her call and also spread the word.



John Lamb's call for assistance

John Lamb wanted to help Fiona Louise with her fundraising for her Canadian research project. He raided his piggy bank, treasure chest and Mason jar and had \$37.63 to send. He also called on his fellow Americans to do the same to *"help our friends up North with their research for EHE"*. We love the attitude and we also love the idea that all our members might be collecting loose change in jars, bottles or piggy banks to help EHE research!!



Washing cars to raise funds

In early August, Fiona Louise's daughter, together with her theatre troop friends, threw themselves into the EHE fundraising effort, running a charity car wash. It was a warm and sunny day and the event got great support with lots of people bringing their cars to be cleaned. They raised nearly \$700 for EHE research which was a fantastic effort. We thank each of them for joining our cause and helping to raise EHE research funds that are critical to understanding and defeating this disease.



Italy calls for help

Alberto Battaglieri, one of our EHE family based in Italy, contacted the group in August asking for help on how to carry out fundraising events. We love Alberto's ambition and were delighted to feedback ideas. We hope to report on Italian fundraising success in future editions of The Pledge.



Jenni's Sunday Funday

Jenni Kovach and her supporters, based in Ohio, held their Sunday Funday in late September, in aid of the EHE Foundation. The event, which included great music, drink specials, and great company, was supported by over 250 people. With door donations, a live auction and a silent auction, this fantastic event raised over \$20,000 for EHE research.

"I never thought it would turn out so well but I realized that telling my story, educating people on EHE and asking for help really goes a long way. I know that we will find treatments that work through additional research. We just need the funds to make this happen!" said Jenni.

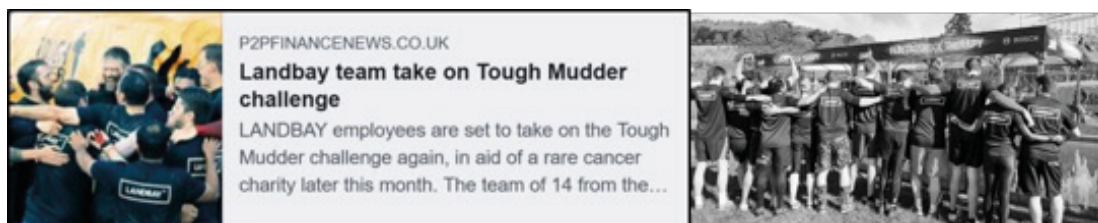
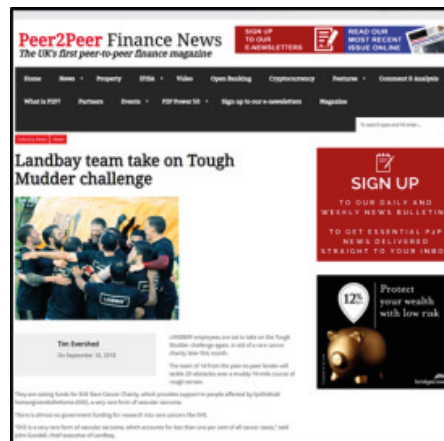
Equally exciting for Jenni was to meet a fellow EHE patient, Cedric Gegel, who turned up to support the event with his parents. We love the support, we love the sentiment, we love the fight, and we love the funds raised. We send our thanks to Jenni not only for hosting a fabulous event, but for recently joining the U.S. EHE Foundation's Board of Directors as the Director of Development.



03 EHE Fundraising

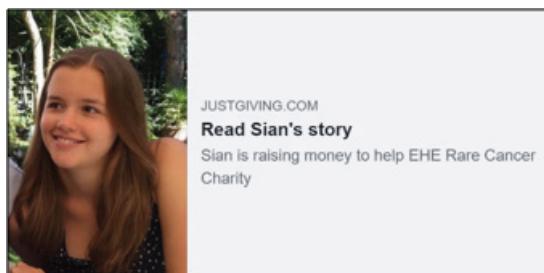
Landbay get muddy for EHE

At the end of September, a team from Landbay, a specialist financing group based in London, took on the Tough Mudder assault course to raise money for the EHE Rare Cancer Charity, and in support of their colleague Artur Rozwalak, an EHE patient. The group completed the SE London course and raised an amazing £1,735 with further donations to come. Landbay had also publicised their fundraising in industry communications, giving EHE even more exposure. We thank them for their fantastic support, and hope that they got really muddy in the process!!



Running for Isabelle

In September Claire-Anne Escoffey shared Sian Smith's fundraising page with our EHE members and asked them to share this as much as possible. Claire-Anne's daughter, Isabelle, lost her battle with EHE in February, and Sian, a close friend, wanted to help fund research by running the Ealing half marathon at the end of September. We cannot thank Sian enough for what she has done, raising over £1,000 for EHE research. We think Isabelle would have been very proud and touched by Sian's fantastic support.



Shave, dye, tattoo

Gareth Miller, living in the UK, wanted to help raise money for EHE research. So what do you do when you have a head full of fantastic, thick brown hair? Some would say 'brave the shave', but Gareth wanted more. So he set his support network a challenge - £500 to see the head shaved, and another £500 to see his eyebrows dyed red and an EHE tattoo on the back of his head! Well Gareth raised over £1,800 pounds and so, on September 21st, in his local pub, the deed was done, as these photos prove. Gareth, we cannot thank you enough for this wonderful event, and more than that, for your on-going love and support.



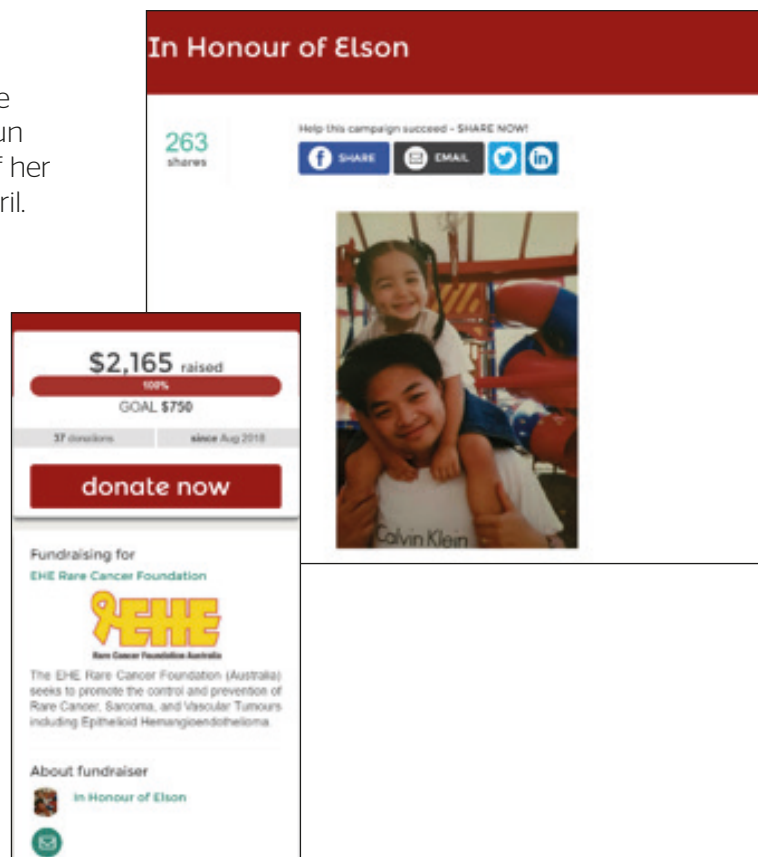
Tea and clothes

Angela Siermans, from Cairns in Australia, wanted to help in the battle against EHE, hosting not one but two events in July. Her first was a workplace morning tea; while the second was a clothes swap party. Angela raised a whopping \$1,000. Her wonderful workplace also agreed to match this, donating another \$1000 to the EHE-RCFA! We thank them all for their fantastic support.



Eden runs in memory of her father

Eden Woolard posted news in August that she would be running the 2018 City to Bay Fun Run on 16th September in honour and memory of her father Elson who lost his battle to EHE last April. Eden wanted to do this to raise awareness of EHE, raise funds for the EHE-RCFA, but most of all for her Dad. Eden also hoped that others in the Adelaide area might run with her. Her campaign was hugely successful, raising over \$2,160 for EHE research. Eden was amazed at the support she received. *"Thank you guys so much for the ongoing support, words can't even describe how lucky we've been, Dad would be so proud!"* We are sure that it was an emotional day for you, Eden. We congratulate and thank you for doing this, and we too think your Dad would have been very proud of what you achieved.



03 EHE Fundraising

Art for fundraising (and therapy?)

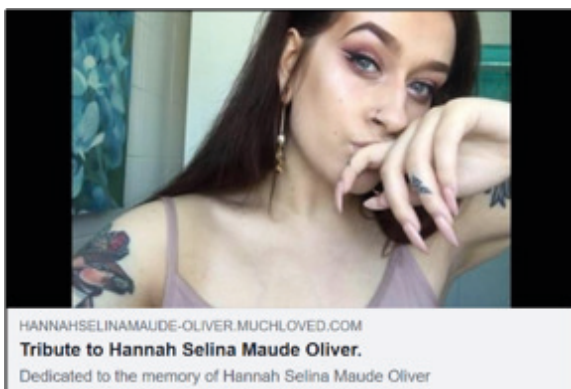
Fundraising does not need to involve huge events and days of organising. Fiona Louise decided to organise her second art evening with her friends. It was a fun night out with lots of laughter and some great paintings at the end. She thought it took about 2 hours to organise, and raised \$400 for EHE research in the process. Just think if all 1500 of our supporters did this once a year, that would raise \$600,000 per year for EHE research. That sounds good!



Tributes to a special young lady

It is always with sadness that we report events and support for our EHE cause that involve the loss of one of our members, and that is true for the inspiring reaction to the tragic passing of Hannah Oliver in August. Hannah was a young lady with great spirit, a 'never say no' attitude to life in general, and a 'never-give-up' attitude to her condition. And she was not alone in this fighting spirit which has shone so brightly in her family and friends since Hannah passed away. Fundraising to fight EHE started at Hannah's Celebration of Life Service which was a terribly sad but also inspiring event.

Hannah was building a growing reputation internationally for her work as a tattoo artist. Shortly after her Celebration of Life, Hannah's colleagues at Infinite Ink in Coventry, UK organised a 'flash day' where all proceeds would be donated to the EHERCC. This amazing event alone raised over £2,000 for EHE research.



Jess Oliver, Hannah's cousin, was also determined to join the battle and become a fearsome EHE warrior. She decided to Brave the Shave as her first event. *"This cancer has torn a hole in my family for where Hannah should be thriving she is gone! We hope to raise a significant amount of money for EHE rare cancer charity, who without vital funding could not do the research so desperately required! I will be shaving my noggin box style full skin head on 23rd September to help raise some of those vital funds! Where Hannah may not be here in person her fiery spirit is within us all that she left, and we will help in any way we can to make a difference!"* We love that fight and

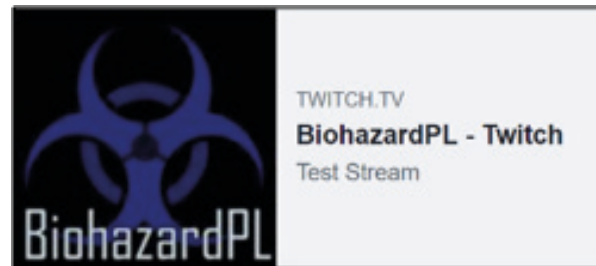
determination and we thought Jess braving the shave was an awesome trail-blazing event.

Hannah's Mum, Dee Evans, posted her thanks with words that we think leave nothing else to be said. *"We are continuing to raise funds to go towards research against this disease. At this heart-breaking time for all of Hannah's family and friends, we give our heartfelt thanks to our EHE family for all the love and support you have freely given to us. Keep fighting, keep winning and although 'just live' is the EHE moto, I would add to that 'Make every day count' 'love Dee'".*



Gaming for EHE

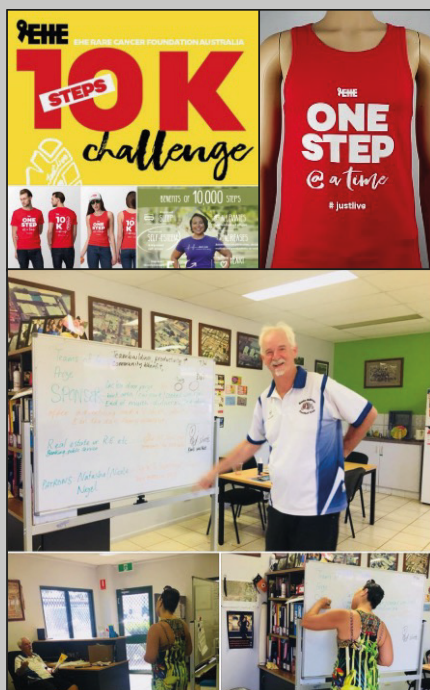
We are always delighted to see new ideas for raising funds and awareness of EHE. On September 22, 2018 Jan Bartkowiak of Wisconsin hosted a one-of-a-kind 24 hour gaming marathon to raise funds for EHE. Jan is a Team Laney supporter and in her own words, had the *"help of some awesome people"*. The event involved streaming live games on twitch.tv/BiohazardPL. We love the live interaction that this provides, and thank Jan and all her fellow gamers for their support, which raised an amazing \$540 for EHE research.



04 Upcoming events...

The '10K Steps Challenge' is back!

Last year one of the big EHE fundraisers for EHE research was the EHE Rare Cancer Foundation's **10,000 Steps Challenge** in Australia. Well its happening again! Starting on October 1st, EHE supporters across Australia will be setting out to do 10,000 steps every day for a month, armed with their free pedometer and campaign tee shirt. Of course these sorts of events don't happen on their own, so Jane Biddlecombe was thrilled to introduce Simon Day to the EHE-RCFA team. Simon has taken on responsibility for organising this year's event, and as Jane said, using Aussie Rules terminology *"he has been kicking goals!!"*.



Jane Biddlecombe, EHE-RCFA Director, was also delighted to be able to introduce and thank Natasha Fyles MLA, Attorney General, Minister For Health and Leader of Government Business for becoming the Patron/ Ambassador for the 10k Steps Challenge. Natasha has been a firm supporter of the EHE-RCFA since it was founded in 2015; through volunteering her time to collect money tins on a Sunday; including articles about EHE-RCFA events in the electorate newsletter; attending EHE events, and assisting with information when required in regards to NFP grants.



We love the idea of combining EHE fundraising with an event that promotes a healthy life style. There will be lots more about this event in the next edition of The Pledge, but here is just a taster from the first organised 10k walk held on 30th September. We think it is another great effort from our Australian members.

We leave them with the words of the famous soccer anthem *"Walk on, walk on, with hope in your hearts, and you'll never walk alone!"*

Just live



London half marathon runners doubled

After the success of the 2018 London Landmarks Half Marathon, the UK were quick to apply for additional places in 2019, over and above the 21 they had this year. This was driven by the wonderful support for the first event and several runners asking for more places next year. So Hugh Leonard was delighted to be able to confirm that 50 places have been secured, with a further 4 obtained in the public ballot. *"So the EHE21 of 2018 will be passing the baton to the EHE54 of 2019"* said Hugh. *"We hope that this will be even bigger and better for us and raise some serious funds for EHE research. We'll be keeping you all updated as things progress."*

Charity Masquerade Ball being planned

Nicola Henderson was delighted to be able to post news of a major charity ball being organised by her and her supporters for next February. *"I am so excited to share the date of this event with you guys! Tickets will be on sale this Saturday. It's going to be amazing!!!"* she said. Nicola is also hoping that other members living with EHE will attend the ball. *"It would be great to get some fellow EHEers at the ball – it's going to be such a great event!!!"* We wish Nicola and her friends every success with the organisation, and are already looking forward to a great evening.



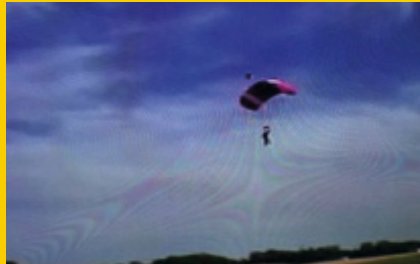
05 And in other news...

Something different

Sometimes it's good to just forget about EHE and do something fun! Fiona Louise took her youngest to the water park! *"Went down two water slides (first time in years)! #justlive"*



Carl Dickson meanwhile took to the air! *"In order to get back into the right mind set in order to heal my soul and my physical self I needed some clean air and a great view. Remember it's what we make of our time, sometimes ignoring the pain, that will bring us the inner strength to live. Just Live!"*



"There are many ups and down in our fight with EHE but every once in a while we must push past what we can to bring back what truly is a part of us."

Great photos

We love the photos people post to provide some inspiration. Here are just a couple from this quarter.



Robinson Ortiz Junior: Never forget how far you've come. Everything you have gotten through. All the times you have pushed on even



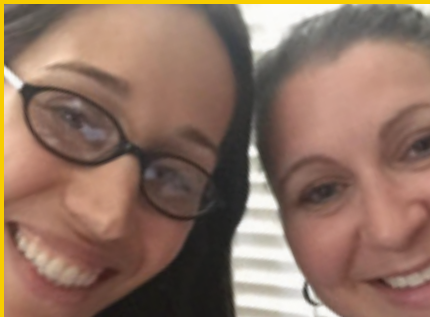
when you felt you couldn't. All the mornings you got out of bed no matter how hard it was. All the times you wanted to give up but you got through another day. Never forget how you kept moving forward one courageous step at a time.



Carl Dickson: *"Now, night is approaching, I sit and watch as I feel fall is certainly in the air and the night brings a calmness to my soul. I hope you all find something that brings this calmness to your soul as you live with EHE."*

Getting together

We also love posts and stories about people within our diverse EHE family getting together to work, to play, and sometimes just to support each other and feel connected.



Julie Rivers Wahl getting EHE work done with Sarah Bright Yaneza.



Jenni Kovach and Jane Gutkovich doing EHE Foundation work.



Fiona Louise and Karen Starr-Lang meet in Canada #justlive #EHEwarriorsEh

A new addition

Amanda Holland delighted us all with some lovely news. *"I forgot to announce to my EHE family! In April, Madilyn will be a big sister! Now just prayers that the pregnancy doesn't cause my EHE to come back...."*

We send Amanda and her family our congratulations and wish her well indeed through the pregnancy





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