

The EHE Foundation (USA)
The EHE Rare Cancer Charity (UK)
The EHE Rare Cancer Foundation (Australia)
EHE Italia - Non solo Laura
EHE Canada



Quarterly Newsletter for the EHE Group
April - June 2023

the pledge

Edition 33



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Front cover:

Jennifer Ness's team supporting the EHE Foundation 2023 Fun Run and Walk.

Welcome

Amazingly **this is the 33rd edition of The Pledge**, the quarterly newsletter of the EHE Group, and covers the second quarter of 2023. **The Pledge** includes stories relating to the key activities of the global EHE community.

As always we want to say a huge thank you to all EHE supporters for their amazing energy, drive, fundraising, time given, and the many other forms of assistance that they contribute. They are all critical to our success and ability to manage and ultimately defeat EHE. We hope that you enjoy this edition of the Newsletter, and that you are inspired by all that is happening. ***“Just Live”***.



Highlights

Tammy Silverthorne joins The EHE Foundation

The Pledge is thrilled to welcome Tammy Silverthorne who joined The EHE Foundation as Executive Director in June, bringing valuable experience to The EHE Group and its activities.

EHE 360 Global Patient Conference a huge success

The EHE Foundation hosted yet another hugely successful patient-focused conference in April with over 200 participants registered from 21 different countries. Guest speakers and panel participants included some of the world's leading EHE research and clinical experts.

Jonathan Granek joins prestigious Committee

The excellent contributions and depth of knowledge of Jonathan Granek, Founding Director and Chair of the EHE Rare Cancer Foundation Australia, is recognised, leading to his invitation to sit on the Peter Mac Community Advisory Committee.

Two major EHE databases are launched within weeks of each other

There was huge excitement in Q2 as both the European prospective observational study of EHE patients coordinated by INT in Milan, and the EHE Global Patient Registry coordinated by The EHE Foundation in the US, were launched.

The 2023 ASCO Annual Meeting includes EHE presentations

Denise Robinson who attended the 2023 ASCO Conference was delighted to see EHE data presented and to meet key research and clinical specialists.

Major fundraising events in the UK, USA and Canada

Q2 included the London Landmarks Half Marathon and Ride London 100 events in the UK; the USA's annual Fun Run and Walk; and the launch of the Annual ***Just Live*** Fun Run in Canada.

Further details on these stories, and much more, can be found in this edition

the
pledge Edition 33



01 Patient Support and Advocacy

Patient Support and Advocacy represent two critical objectives of the EHE Group and the worldwide EHE community. Indeed, the foundation of all that the Group has achieved is the original support and advocacy that a handful of people started when they found each other on the internet in 2013.

Patient support within our global community largely comes in the form of advice, support and encouragement provided by EHE patients to each other. Advocacy is about promoting greater awareness of EHE, and striving to bring about better care and better outcomes for EHE patients everywhere.

We hope that the following stories will give you some idea of the activities that are taking place, thanks to the tireless energy, support and contribution provided by so many of our global EHE community.



Tammy Silverthorne joins The EHE Foundation

In June, Tammy Silverthorne joined The EHE Foundation as Executive Director. Jenni Kovach, The EHE Foundation President, said:

“We are very excited to have Tammy. Her strong leadership abilities, passion, and experience in the rare cancer community are a perfect match in helping us work toward our vision: to live in a world where Epithelioid Hemangioendothelioma (EHE) is easily diagnosed and treatable. We are confident that under her leadership the Foundation’s success and impact in achieving its mission will continue to flourish!”

Tammy has very significant experience of the challenges faced by the EHE patient community, having dedicated many years to the growth and development of the Chordoma Foundation, Chordoma also being a rare sarcoma. With over twenty-five years of experience in the leadership and growth of small organizations, Tammy is

distinctively qualified to guide The EHE Foundation as it continues to build on a strong track record of successes. Her extensive experience, particularly in the context of growing an organization from a small, volunteer-led operation to a well-regarded leader in its field, provides the skills and insight the Foundation needs as it looks to build long-term sustainability. Her experience will help build the growing collaboration between EHE patients, clinicians, and researchers, as well as maintaining and expanding The Foundation’s relationships globally with all other members of the EHE Group.

Tammy is hugely excited as she takes on her new role. Tammy commented:

“The EHE Foundation has shown impressive growth in a short period of time, and I am thrilled to have the opportunity to work with such a dedicated team. I am excited to combine my passions for growing small organizations and improving healthcare options for those with rare diagnoses with the momentum in the tight-knit EHE community to advance progress in treatment options and support for those touched by EHE!”

Tammy is a native of North Carolina and a graduate of Duke University. She enjoys traveling, spending time outdoors, attending sporting events and concerts with friends and family, reading, and one day hopes to train a therapy dog, a fact that will resonate with many of our patient community who regularly post about their own dogs and the comfort and therapy they provide.

Everybody at **The Pledge** also wants to send Tammy a huge welcome on joining **The EHE Foundation team** as well as our **EHE global community.** ■



01 Patient Support and Advocacy

Jonathan Granek

"It's very rewarding being part of a diverse yet collaborative committee that's truly focused on identifying and addressing needs, continuously improving experiences and outcomes, and striving for world class comprehensive cancer care as the standard."



Recognition of Jonathan Granek's Contribution

June saw congratulations being sent from many to Jonathan Granek, Founding Director and Chair of the EHE Rare Cancer Foundation Australia, for his invitation to sit on the Peter Mac Community Advisory Committee.

Jonathan provides expertise across a range of areas due to his own experience as a rare cancer survivor of 12+ years, Director of the patient-led EHE Rare Cancer Foundation Australia for Epithelioid Hemangioendothelioma (EHE), and since 2015, as a consumer representative collaborating with researchers at multiple institutions in Australia including WEHI, Peter Mac, Garvan, and elsewhere.

Jonathan has experience working across common and rare cancer research programs and projects, and has been invited to speak on the importance of cancer, rare cancer, and sarcoma research in various forums. Jonathan is currently also a member of the VCCC Precision Oncology Consumer Reference Group and a mentor for other consumer representatives.

Jonathan's journey has instilled in him a passion to help develop improvements in cancer control and support for patients and carers, especially for those with rare and less common forms of cancer, and from under-represented communities more broadly.

Jonathan's key interests include health equity, genomic medicine, precision oncology, therapeutics development, quality of life outcomes, health economics and big data.

Hugh Leonard, Chair of Trustees of The EHE Rare Cancer Charity (UK), was just one of those who wanted to congratulate Jonathan on his new role, and also recognise Jonathan's passion and depth of experience. Hugh commented:

" I think I will speak for everybody in our community who knows and has worked with Jonathan when I say congratulations Jono for your new role with Peter Mac. We are so lucky to have this brilliant man on our side. Jonathan has great knowledge and understanding of the patient representative role, and a depth of understanding of the hugely complex issues around oncology, sarcoma, EHE, the delivery of improved care for EHE patients everywhere, and the extraordinarily complex integration of EHE research with other key areas and studies. I always love spending time talking with Jono and usually leave feeling a little in awe of him. It is simply great to have him 'in our corner', fighting for our community. This appointment is such a deserved recognition of all those skills and his huge contribution. Congratulations Jono, so deserved! "

The Pledge wants to join Hugh and many others in extending our congratulations to Jonathan for his new role, and to say a huge thank you for all that he does. ■

Getting Together Always Helps

We know our EHE community always reacts positively to news of patient members meeting up. This is not that common when you think that this is a 'one-in-a-million' club. These meetings don't only help those involved, but helps patients feel less isolated wherever they live. So we love reporting the stories when posted by the community.



Michelle Lynn was one patient who posted news of such an occasion when she met Malda Sherikheh in Canada. Michelle was thrilled:

“Can you believe it? I met another one of us!! So grateful for the powers of social media. Today I had the pleasure of meeting someone just like me. A ‘zebra’ as they say in the rare cancer world. A fellow EHE cancer patient! I received an email from Malda, explaining that she was on my little island for two days, and I knew I had to find a way to ensure we met. I shook my head after I read it. What are the chances... literally less than 1 in a million!”

EHE treatment options can be limited, with procedures that are being utilized in other parts of the world not necessarily being readily available in Canada. So Michelle was excited and super impressed with Malda's drive and determination.

“I was in awe meeting her today and hearing her story. She wasn't taking no for an answer! She's finding her own way, but unfortunately that comes at a cost; a financial one. But when it comes to your life, your survival, is there really a dollar amount? I'm so proud of her and look forward to seeing her results on this new, minimally invasive technique that's proven to kill liver tumours.”

It was also clear that the respect and heartfelt gratitude was reciprocated, as Michelle also explained:

“We had such a powerful conversation, both of us wanting to move research forward not only for us, but for those future Mommas. She's looking to pave the way, advance cancer treatment in Canada, be our Guinea pig, per se. But she also thanked me for being the voice; raising awareness, and I thanked her too. We all play a part in this. We have to. It's just us... maybe only 20 of us in Canada, So we stick together and move forward, hand in hand, and with hope in our hearts and a mindset like no other.”



Denmark were also posting on the theme of coming together. Tina Biehl Nielander posted this wonderful photo in June with another simple but great message:

“The Danish dynamite! Here are Rikke - the 'lion mum' to Rosa, Lilli and Tina. We are living! This is the EHE group meeting today in Roskilde, Denmark. All because of our EHE Facebook group. Thank you”

We cannot be anything but inspired by these messages, and the amazing strength, support and determination that so many of our patient community demonstrate. They truly embody the spirit of the EHE community motto **“Just Live!”** ■



01 Patient Support and Advocacy

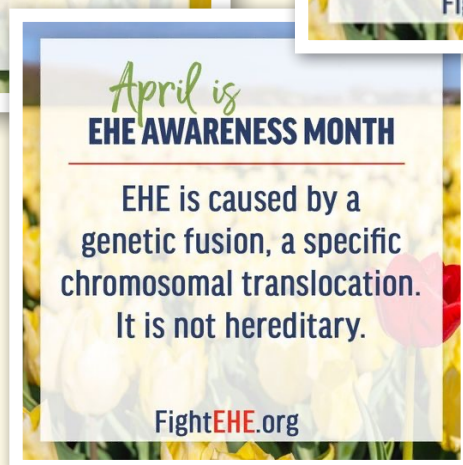
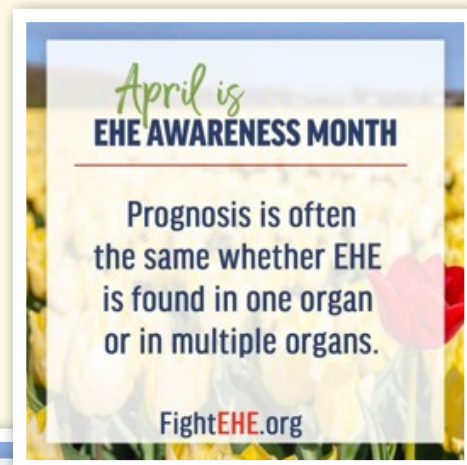
EHE Awareness Month

The EHE Group established April as EHE Awareness Month shortly after the inception of the first three EHE foundations in 2015. The EHE Foundation (US) celebrated EHE Awareness Month by hosting the annual EHE 360 Global Patient Conference and by opening the registration for the largest EHE Fun Run event in history. Articles on both these events can be found in this edition of **The Pledge**.

Each April the foundations also use social media to promote awareness of the disease, and coordinate this with different events. 2023 was no different with both The EHE Foundation in the USA and The EHE Rare Cancer Foundation Australia launching vibrant social media campaigns.

Both Foundations used the month to provide supporters with facts about EHE, including what sort of cancer it is; that the disease can be asymptomatic at diagnosis; and prognosis is often similar whether EHE is found in a single organ or multiple organs. This is contrary to the prognosis of most other cancers and why staging is often disregarded with the rare cancer, EHE.

Another key area addressed by both Foundations was that of the genetic fusions or chromosomal translocations that are now known to be the cause of EHE and are considered to be disease defining. This occurs due to simple chromosomal rearrangements. The TAZ-CAMTA1 fusion affects about 90% of all EHE patients, while the YAP1-TFE3 fusion affects approximately 10% of patients.



The requirement to find new drugs or cures, the exceptional research growth we have seen in the last ten years, and the impact of that research were also addressed. Sadly, because EHE is so rare, research relies on patients diagnosed with EHE participating, if they possibly can. The EHE Group want to thank EHE researchers, clinicians, and patients for advancing EHE science.


Examples of the excellent graphics used by the two foundations are shown opposite and below. You can also check out the awareness posts at: <https://www.facebook.com/Ehefoundation> for the US and: <https://www.facebook.com/EHERareCancerFoundation/> for Australia. ■

EHE RARE CANCER FOUNDATION AUSTRALIA
#Be Aware of Rare

WHAT IS EHE?

Epithelioid Hemangioendothelioma (EHE) is a rare and often deadly vascular sarcoma that affects men, women and children of all ages.

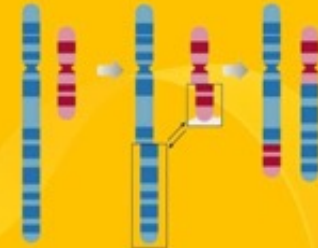
There is **NO CURE** for EHE, which can turn from indolent to aggressive with little indication and fatal results.

WWW.EHEFOUNDATION.COM.AU 

EHE RARE CANCER FOUNDATION AUSTRALIA
#Be Aware of Rare

WHAT IS A TRANSLOCATION?

A translocation is a swapping of genetic material between two genes.



WWW.EHEFOUNDATION.COM.AU

EHE RARE CANCER FOUNDATION AUSTRALIA
#Be Aware of Rare

WHAT CAUSES EHE?

EHE is caused by a genetic malfunction, a specific chromosomal rearrangement known as a translocation (swapping of genetic material) called the 'WC Fusion.'


This genetic alteration results in the production of a fusion gene, and a fusion protein. It is suspected that this translocation, which causes the fusion protein causes the cells to grow without normal regulation.

It is this abnormal system that could theoretically be targeted for therapy.

WWW.EHEFOUNDATION.COM.AU 

EHE RARE CANCER FOUNDATION AUSTRALIA
#Be Aware of Rare

THE IMPACT OF RESERCH



RISK FACTORS & PREVENTION
UNDERSTAND RISKS & WAYS TO POTENTIALLY AVOID OR REDUCE RISKS

TREATMENTS
DEVELOP MORE EFFECTIVE TREATMENT OPTIONS & WITH LESS SIDE EFFECTS SUITED TO THE INDIVIDUAL

IMPROVED DETECTION, DIAGNOSTICS & PROGNOSTICS
IDENTIFY EHE AS EARLY AS POSSIBLE, UNDERSTAND ITS CHARACTERISTICS & BEHAVIOUR TO GUIDE IT'S MANAGEMENT

SURVIVAL
IMPROVE LONGEVITY & QUALITY OF LIFE

BE PART OF THE CHANGE - DONATE TODAY

WWW.EHEFOUNDATION.COM.AU



01 Patient Support and Advocacy

Australia Holds Meet and Greet

The EHE Rare Cancer Foundation Australia was delighted to hold an online Meet and Greet session in early April for Australian and New Zealand group members, as part of EHE Awareness Month.

“With so many new Australian and New Zealand faces joining our global EHE community in 2022 - 23, we want to hear your story, and what you (patients or carers) need better support with and what information has been useful and what information is lacking.”



The EHE Rare Cancer Foundation Australia not only wants to help patients introduce themselves but also hopes that their Meet and Greet event will lead to face-to-face meetings of patients on a state-by-state basis across Australia in the future, re-enforcing the EHE patient mantra, **“ALONE WE ARE RARE - TOGETHER WE ARE STRONG”**. ■

2023 EHE 360 Global Patient Conference: Empowering Patients



This year's annual EHE 360 Global Patient Conference was held on Saturday, April 15 featuring an international group of clinicians and scientists presenting the latest clinical and research developments in EHE. The theme of the conference was *Empowerment*, providing an opportunity for everyone to learn and grow in their knowledge of EHE.

The conference was organized by The EHE Foundation with the generous support of volunteers from the EHE community - including patients, advocates, clinicians, and researchers who gave their time to make this event a success. A highlight of the day was the Ask the Expert panel, which remains a popular session among patients and clinicians. They love having this opportunity to answer patients' questions!

Over 200 people representing 21 countries registered to attend the live event. An amazing audience! All sessions are recorded and available to view at <https://fightthe.org/2023-ehe360-global-patient-conference/>.

Prior years' presentations are also available on The EHE Foundation's [YouTube channel](#).

The EHE Group want to thank all the speakers, shown on the page opposite, who gave their time to the EHE360 event and who were instrumental in making it so successful. ■

Featured speakers



Denise Robinson
THE EHE FOUNDATION

Patient-Led Research: Power in Numbers



Brian Rubin, MD, PhD
CLEVELAND CLINIC

EHE 101: What You Need to Know



Breelyn Wilky, MD
UNIVERSITY OF COLORADO CANCER CENTER

Systemic Treatment Options and Triaging EHE



Ajaybabu Pobbati, PhD
CLEVELAND CLINIC

Why TEAD is a Convincing Target for EHE Treatment



Denise Adams, MD
CHILDREN'S HOSPITAL OF PHILADELPHIA

Sirolimus as a Treatment Option for EHE



Hugh Leonard
CHAIR OF TRUSTEES | EHE RARE CANCER CHARITY UK

Patients' Perspectives on Sirolimus



Michael J. Wagner, MD
FRED HUTCH CANCER CENTER

Indolent or Aggressive? EHE Outcomes and Monitoring Strategies



Vinod Ravi, MD
MD ANDERSON CANCER CENTER

Ask the Expert Panel



Abha Gupta, MD
PRINCESS MARGARET CANCER CENTER

Ask the Expert Panel



Tamara Vesel, MD
TUFTS UNIVERSITY SCHOOL OF MEDICINE

Ask the Expert Panel



William Tap, MD
MEMORIAL SLOAN KETTERING CANCER CENTER

Ask the Expert Panel



01 Patient Support and Advocacy

Canadian *Just Live* Clothing Campaign Launched

The second quarter saw the culmination of Canada's newest foray into raising awareness about EHE while also fundraising. Michelle explained:

“Our *Just Live* Clothing Campaign is a new Canadian fundraising campaign that is near and dear to my heart, as well as being an opportunity to spread the *Just Live* love. Why am I doing this? Because what I didn't know when I was diagnosed with Stage 4 Incurable Cancer was that it would be up to us, the patient community, to raise the research dollars. That's because ultra-rare cancers like EHE don't get the vast funding that more common cancers attract!”

Michelle pours her heart and soul into raising funds, but not necessarily for herself. For Michelle, it is also about helping future EHE patients as they face their own EHE diagnoses. Michelle is particularly desperate to help all the other 'Mommas' who will follow her, for the Momma 10-20 years in the future.

“If I can play a part, even the smallest part, to help them feel less alone, and if we're lucky, somehow stop the cycle of young Moms hearing the words “incurable cancer”, then that would be truly amazing.”

So that was why Michelle was super excited to post news that her *Just Live* Clothing Campaign had gone live. The initial sale was just for Canadians and would be open for 14 days, with The Sarcoma Cancer Foundation of Canada matching ALL PROCEEDS raised. Michelle is also looking into being able to open access for the USA, and eventually international locations also.

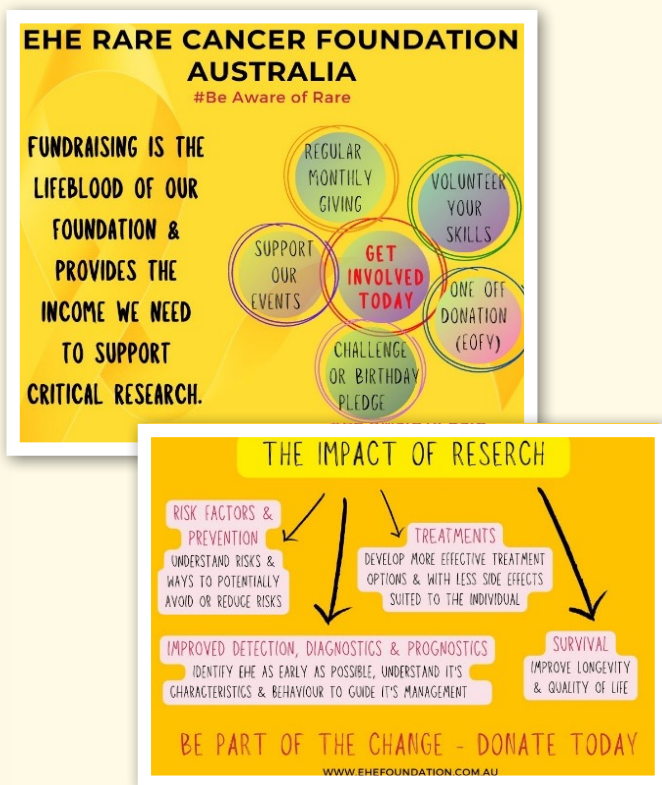
She was even more excited when news of the total sale proceeds came in, with an amazing \$5,000 raised. With matching funding, this meant a huge total of \$10,000 of new funding would be available for EHE research.



Above are just some of the garments that were available during the sale. ■

Explaining the Fundraising-Research Imperative

The EHE community sees many messages posted by members who are fundraising to allow the EHE Group to fund critical research that will lead one day to better treatments and hopefully even a cure for EHE. The EHE Rare Cancer Foundation Australia explained why the fundraising is so important, different ways that people can contribute, and the impact of the research and what it can deliver. This was achieved using simple but powerful graphics that were posted on their Facebook page, website, and also shared with the global community.



Huge congratulations to the EHERCFA for this brilliant and informative campaign which we are sure had the desired effect and helped many understand why it is imperative to raise the funds and drive the research. ■

The pursuit of a new treatments!

We want to thank to David Casimir, PhD, JD for his informative patient-level presentation in June that addressed the questions: What is the Hippo pathway and TEAD? How do they relate to EHE? How are they being used to develop EHE treatments? How do these treatments move from the laboratory, through clinical trials, to patient care?



The EHE Group strives to empower patients to advocate for their care through various types of continuing education and support. This includes offering programs to help patients understand complex medical topics.

The Hippo pathway is a critical transcriptional signaling pathway that regulates cell growth, proliferation, and organ development. Combined with Transcriptional enhanced associate domain (TEAD), this can easily become a topic that is difficult for many patients to fully understand.

David was able to simplify these topics using an analogy of something most people are familiar with: the postal system. Hugh Leonard said:

“ Well done, David! I will for evermore look at postmen and postal trucks and think of the Hippo Pathway! ”

You can view the video of David’s presentation and other Community Connections sessions at: <https://fightehe.org/community-connections/> ■



01 Patient Support and Advocacy

Canada Drives EHE Awareness

The second quarter saw Michelle Lynn, with help from Fiona Ross, driving a new EHE awareness campaign involving an updated running shirt, a 10km run and a Subaru film crew. Michelle explained:

“Hi everybody! Kimberly Young gathered a list of names of those with EHE (living or deceased) to put on the back of her Race Shirt for her marathon back in 2021. I feel it's time to add more members to this shirt, so I'm designing a new running shirt for a big race I have coming up, and I'd love to add to the names I have. I'd like to represent everyone; take you all along with me on this journey.”



The new design of shirt looked fantastic in a vibrant pink, with all the names provide on the back. Michelle ran her race with so many names of our EHE community on her back, as Michelle explained:

“I carried every single one of you over the last two days during an incredible opportunity to bring more awareness to our beast of a disease. Subaru Canada also sponsored a video featuring my journey with running as a Momma with incurable cancer. We have a voice. Use it loud, and PROUD. I sure do. **Just Live**”

Congratulations Michelle on another great event. ■

Get Help with Out-of-Pocket Expenses for Travel for Clinical Trials

Many patients don't have the resources to travel for participation in clinical trials.

Jordan's Dream Fund was established in 2017 in memory of Jordan Andrew Singer, who was 28 when he passed from cancer. Through Jordan's Dream Fund, Sarcoma Foundation of America grants up to \$5,000 four times each year to cover out-of-pocket expenses for sarcoma patients participating in clinical trials. Learn more at:

Sarcoma Foundation of America grants up to \$5,000

4x per year to cover out-of-pocket expenses for clinical trials

THE EHE FOUNDATION

<https://www.curesarcoma.org/sarcoma-resources-patient-resources-jordans-dream-fund>

If you want to learn more about clinical trials for EHE patients then go to The EHE Foundation website at: <https://fightehe.org/ehe-clinical-trials/>

If you are looking for more resources to help with finances then go to: <https://fightehe.org/additional-resources/> ■

Our Global EHE Community Continues to Grow

One of the most common accolades the EHE Group receives from clinicians, researchers, regulators, donors and other stakeholders, concerns the EHE patient community that has been established globally, and who contribute so much to patient support, to research and to fundraising. Much of this comes from the community's EHE Support Group page on Facebook, set up by Dawn Scott in 2013.



The growth has been extraordinary, and in June this year passed the 2,600 member mark, representing patients from nearly 80 countries. While the Facebook page could and does help hugely with patient support and guidance, those same veterans recognised that a Facebook page alone could not drive the key issues of research and finding new treatments which ultimately needed funding. This led in 2015 to the EHE community setting up their own EHE foundations in the USA, Australia and the UK to act as the focal points for fundraising and EHE-specific research. Canada and Italy have followed more recently, while other countries have established EHE patient networks.

Now as we look back, we can see that over the last 10 years, the EHE community, initially established through that first Facebook page, has helped in advancing awareness and knowledge about EHE; driven an active and very successful fundraising programme; and jump started and advanced EHE research which has expanded from Brian Rubin's small team in 2013 to multiple research projects with major international collaborations today.

So as the EHE Support Group page reaches its 10th birthday, we want to thank Dawn Scott for setting up that initial page, and all those who have contributed so much to create the growth in our global EHE activities to where they are today. There are too many to list individually, and lists always run the risk of somebody being missed, but we think you all know who you are.

It has indeed been an amazing journey, with phenomenal progress for such a rare cancer. All of that has only been possible thanks to the unbelievable support of the EHE global community, and so most of all we want to say a huge thank you to every individual member of our EHE community who have provided unconditional support to hundreds of EHE patients and caregivers, and contributed to the growth in our research programmes.

At this point we also want to remember all those members of our patient community who contributed so much but who sadly left us far too early. They are a major part of the reason why we all spend so much time and energy working to find new treatments and a cure for EHE. We owe that to all those who are no longer with us. ■



02 EHE Research

EHE research is absolutely critical to our ability to find new ways to treat and manage EHE. We will ultimately achieve these goals because of the ongoing research we are funding today, and the new research that we will promote, encourage and fund in the future.

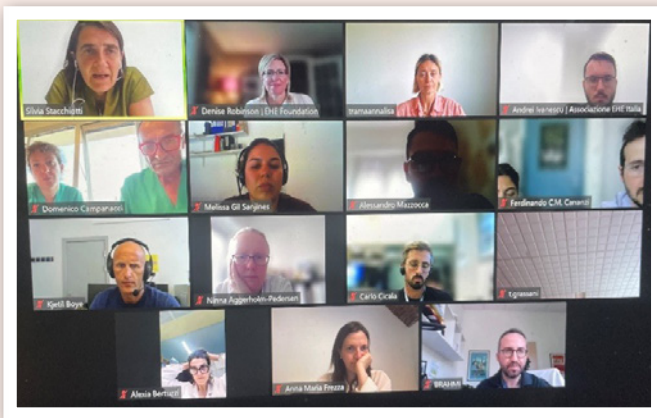
It is this research that will provide the answers and understanding we need; that will lead to new treatments; and that will allow doctors to one day stabilise and ultimately cure the disease.

This section of the newsletter provides an update on the EHE research that the EHE Group is involved with, as well as newly published papers and information.

European Prospective Study is Launched

We have reported on several occasions news on the progress being made to establish a prospective observational study of EHE patients in Europe and the UK, based within the EU's major STARTER project. Further information on the project and the people behind it can be seen in the following pages of this research section where we are focused on the collaborative research being funded by the EHE group and involving by Silvia Stacchiotti and the teams at INT in Milan.

This quarter also saw the first briefing session of European hospitals that may be interested in joining the study. A virtual zoom meeting was held during which Dr Stacchiotti, Dr AnnaMaria Frezza and Dr Annalisa Trauma lead a presentation of the study and invited expressions of interest.



The Project is being partially funded by The EHE Rare Cancer Charity UK, and Hugh Leonard participated in the call, representing the EHE patient community. Hugh commented:

“It is really inspiring to see experienced and senior clinicians across Europe participating in such a positive discussion about this study. People are keen to be involved which is great to see. There will be a face-to-face ‘kick-off’ meeting in Milan in September where I hope to meet many of these clinicians and have the chance to talk to them about our EHE Group and the global EHE programme we are driving. Ultimately we hope that this registry will lead to a better understanding of the disease and also help identify and drive research in existing and new areas.”

Hugh was not the only EHE Group member to take part. Andrei Ivanescu, President of Associazione EHE Italia, also participated, representing both the Italian patient community as well as all European patients. In addition, with the EHE Foundation having just launched the EHE Global Patient Registry, and with a clear desire for these different databases to be harmonised so they can be more easily used within research going forward, Denise Robinson was also invited to participate. Denise felt that taking part had been very useful indeed:

“I was so grateful to be included in the meeting about the EHE prospective registry. Having heard about this project over these past couple of years, this meeting helped to give me a greater understanding of the project. I want to congratulate all those working to bring this registry forward! The registry team’s offer to share their CRF (the actual data questionnaire) with us is also fantastic as it will help us maximise the chances of being able to harmonize the European data with our own Global Patient Registry in the future.”

We are all looking forward to seeing the results and **The Pledge** wants to echo Hugh’s and Denise’s congratulations to all those involved with setting up the registry. ■



02 EHE Research

The EHE Foundation Launches the EHE Global Patient Registry

A Natural History Study of Epithelioid Hemangioendothelioma (EHE)

The EHE Foundation and the National Organization for Rare Disorders (NORD®) have launched the EHE Global Patient Registry, a natural history study that consists of electronic surveys to collect information from people diagnosed with EHE about their experiences and EHE progression. Patients, or their caregivers or guardians, can enter information from anywhere in the world. The data is confidential and stored securely in the IAMRARE® online portal.



**EHE Global
Patient Registry**
EHEregistry.iamrare.org

JOIN TODAY

This study creates a platform for patients around the world to share information about EHE. Its purpose is to build an international resource to be used by scientists in future research. The EHE Foundation may share data with individuals or institutions conducting research or clinical trials, as approved by the study's governing board that includes scientists, doctors and patient advocates. Of course, no data will be shared until it has been completely anonymized.

Denise Robinson, Director of Research and Principal Investigator of the Registry said:

“the EHE Patient Registry opened in May 2023 and by the end of June over 130 people from 14 countries had joined the registry platform! From published data, it is estimated that <1/1,000,000 people are living with EHE which leads us to believe that there are several hundreds of people around the world living with the disease. Just think of all the things we can learn if EHE patients all over the world come together to share their EHE story. Every person with EHE can make a difference for the future.”

Jenni Kovach, President of The EHE Foundation added:

“Today, there are no large studies or datasets describing patients' experiences with EHE. Patients, doctors, and researchers struggle to understand the various presentations of EHE and courses of disease progression. Because EHE is so rare, the road to find treatments and a cure relies on patients joining together and this registry truly empowers patients to improve our understanding of this ultra-rare disease. I am living with EHE, and I want my voice heard in the research community. By raising our collective voices, we can envision a future where EHE is more easily diagnosed and treatable.”

The EHE Foundation has launched the study in collaboration with NORD, an independent nonprofit that built its natural history study platform as part of its mission to help identify and treat all 7,000 rare diseases. The EHE Foundation is a member of NORD, and the two organizations work together to eliminate the challenges that rare disease patients face.

For more information about the EHE Global Patient Registry, visit fightehe.org/registry or contact Denise anytime for questions at registry@fightehe.org ■

Highlights from ASCO 2023

The EHE Director of Research, Denise Robinson, attended the American Society of Clinical Oncology (ASCO) 2023 Annual Meeting and the Sarcoma Alliance for Research through Collaboration (SARC) Semi-annual Meeting in Chicago, IL, in June. Over 40,000 people joined together in person to conquer cancer!

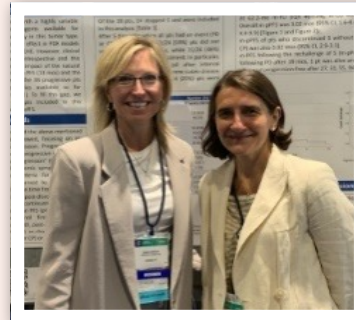


ASCO is one of the world's leading organizations for oncology, bringing together clinicians, patient advocates, and professionals to learn and discuss research and programs that aim to change the lives of people with cancer. SARC supports research to prevent, treat and cure sarcomas - of which EHE is an ultra-rare subtype.



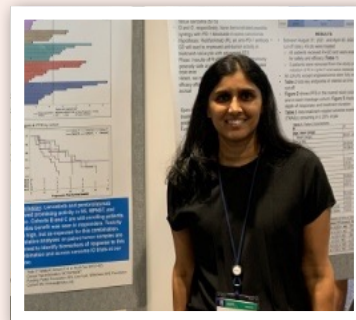
Dr. Bill Tap, MSKCC, gave an empowering talk at the SARC meeting challenging all stakeholders to expand our approach and thinking on rare cancer research. His talk, **Sarcoma - the**

evolution of care in rare cancers, highlighted the importance of real-world data (RWD) in rare cancer research - specifically calling out the patient-led research initiatives in the EHE community. The take-aways for doctors and advocates - there are lots of data out there that can help us better understand and study rare cancers. Data from medical records, registries and clinical trial results can be synthesized to improve our understanding of rare diseases like EHE. We thank Dr. Tap for championing EHE and rare cancer research!



Dr. Silvia Stacchiotti (right) and colleagues from the IRCCS National Cancer Institute Foundation presented a poster entitled **Outcome of patients with advanced epithelioid**

hemangioendothelioma (EHE) after failure on sirolimus. This data reviewed the outcome of patients with advanced EHE from a prior retrospective study who discontinued sirolimus for any reason. The conclusions highlighted that those patients who discontinued sirolimus for reasons other than disease progression benefitted from sirolimus re-challenge (restarting treatment), which suggests that in advanced EHE there is a subset of long-responders to sirolimus. Dr. Stacchiotti is a force of nature in EHE research. Stay tuned, she and her colleagues will continue to bring forward new data!



Dr. Sujana Movva, MSKCC, presented data from **A pilot study of lenvatinib plus pembrolizumab in patients with advanced sarcoma**.

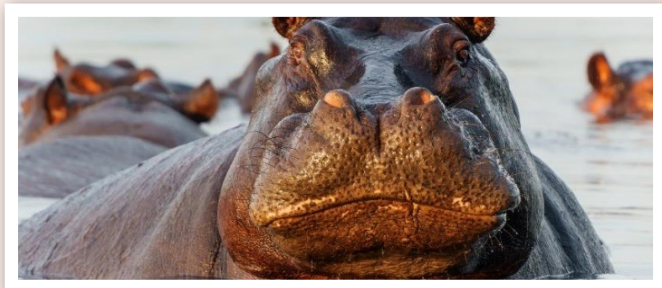
This study is ongoing and currently enrolling at MSK, including EHE patients. Many thanks to patients who have participated in this study, and to generous donors in the EHE community who have helped make this study possible. Finally, a great many thanks to Dr. Movva and her MSK colleagues who continue to provide expert care and consultation to EHE patients worldwide!

<https://fightshe.org/highlights-from-asco-2023/> ■



02 EHE Research

Exciting Data @AACR 2023 - Hippo (pathway) Makes a Splash



The annual meeting of the American Association of Cancer Research was held in April, and the Hippo pathway made a big news splash. Exciting data was shared from multiple drug developers targeting the Hippo/Yap/Tead pathway. Two investigative drugs in clinical trials including EHE patients are featured in the article - Ikena Oncology and Novartis are sponsors of those trials. A list of other companies and compounds in development also reported data - all hopeful news, and excellent progress for people living with EHE. Read the full article online at: <https://www.evaluate.com/vantage/articles/events/conferences-trial-results/aacr-2023-hippo-makes-splash> ■

2023 EHE Research Grants Cycle Update Funding Impactful Projects That Support Our Mission

The EHE Group seeks to support and fund the most promising basic, translational, and clinical research that will expand our current understanding of EHE and accelerate the development of treatments, and a cure for EHE.

Each year, The EHE Foundation initiates a new Research Grants Cycle, inviting researchers and clinician scientists from around the world to submit proposals that support our mission. Research proposals are reviewed and scored by expert reviewers including the Foundation's Advisory Board and complemented by experts from the global research community.

Last year The EHE Foundation funded three research projects totalling \$541,000. You can read more about those research grants [here](#). This year The Foundation are grateful to have received five letters of intent and four full proposals for review. Pending final scoring and approval, the recipient award announcements will be made in September. Stay tuned for updates!

The EHE Foundation are grateful for your financial support which allows the EHE Group to fund impactful research proposals. Every donation, large and small, contributes to the growing body of knowledge about this disease, and enables the Foundation to purposefully advance their and the EHE Group's mission. Your support makes a difference!

To learn more about projects previously funded, go to <https://fightehe.org/ehe-research-grants/> ■

IRE Ablation Available for EHE in the UK

Tyler Davis posted positive news in May concerning the possibility to have IRE ablation for

EHE in London. This is very encouraging as previously IRE ablation had not been available for EHE in the UK as the procedure is not formally approved by NICE (National Institute for Clinical Excellence) for EHE treatment. Tyler explained, his comments particularly directed to fellow patients being treated under the London and Southeast Sarcoma Network (University College Hospital, The Royal Marsden Hospital, and The Royal National Orthopaedic Hospital, Stanmore):



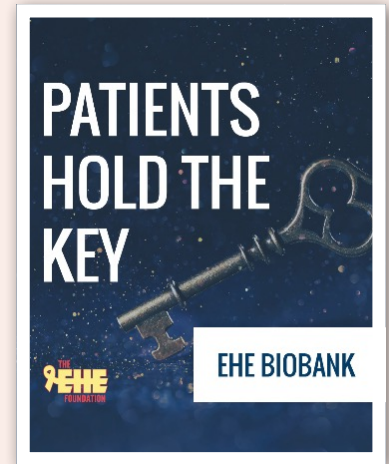
“ I had a long conversation last week with the Head of Interventional Radiology at UCLH, regarding the pros and cons of IRE vs Microwave for liver mets. He confirmed IRE CAN be offered as an alternative to Microwave ablation on the NHS at the insistence/ request of the patient, and is a procedure which has been performed for the last 10 years at UCLH. However, it is only typically used if the tumour is close to another organ, such as the gallbladder or an artery within the liver. UCLH's default position is that Microwave ablation is more effective / successful as a treatment to destroy liver tumours than IRE ablation, which he stated is supported through years of research. However if a patient with EHE insists that they want IRE, as the alternative, UCLH will not say “no”! ”

Tyler wanted to post the news in the event that it helps someone, as the prior understanding is that Nanoknife is not available in the UK on the NHS for EHE treatment.

Well done Tyler for obtaining this clarification from UCLH. This may open the door for other patients to ask for IRE ablation in the future. ■

You Can Donate Tumor Tissue to Help Find a Cure for EHE

The EHE community has now established EHE biobanks in the USA, UK, and Australia, and hopes that other countries will soon follow. These biobanks empower people living with EHE to advance research by donating their tumor tissue or body fluid. How can you help?



- If you are planning a surgery, a transplant or need to have fluid drained from your lungs or abdomen – please get in touch with your nearest EHE Group foundation, even if you do not have a scheduled procedure.
- If you have had tumors removed in the past, we may be able to use tissue stored at your hospital.
- Leave a legacy by planning a posthumous tissue donation.

Advance planning is helpful for the biobank teams to coordinate all donations. Please get in touch to learn more. Use the following links to learn more about how you can donate tissue:

In the US email biobank@fightehe.org or visit <https://fightehe.org/ehe-biobank/>.

In the UK email: rnh-tr.ehebiobank@nhs.net or visit: <https://www.ehercc.org.uk/national-ehe-biobank-uk>

In Australia email: info@ehfoundation.com.au ■



02 EHE Research

Visit Your EHE Library

Whether you are newly diagnosed or want to stay updated on EHE publications, continue to visit the **EHE Library**, located in the Research section of fightehe.org. There are now over 150 publications listed including case reports, retrospective studies, and some highly scientific research articles that illustrate the research ongoing around the world to fight EHE. Note, not all publications are freely accessible. If there is something of interest to you that you cannot access, please reach out to research@fightehe.org for assistance.

Here are several new recent additions to the library that you might want to read:

- [Current Model Systems for Investigating Epithelioid Haemangioendothelioma](#)
- [Outcome of patients \(PTS\) with advanced epithelioid hemangioendothelioma \(EHE\) after failure on sirolimus \(S\)](#).
- [New Molecular Insights, and the Role of Systemic Therapies and Collaboration for Treatment of Epithelioid Hemangioendothelioma \(EHE\)](#)
- [Prognostic Factors in Epithelioid Hemangioendothelioma: Analysis of a Nationwide Molecularly/Immunohistochemically Confirmed Cohort of 57 Cases](#)
- [Loss of CDKN2A Cooperates with WWTR1\(TAZ\)-CAMTA1 Gene Fusion to Promote Tumor Progression in Epithelioid Hemangioendothelioma](#)

In addition to the excellent EHE Library described above, every quarter a number of new papers are high-lighted on the EHE Patient Support Facebook page, often with a simple insight as to what the paper is addressing for members of our community who may have limited scientific experience. These papers often provide interesting and positive updates in terms of clinical and treatment performance, as well as EHE research. We have included some of those posts in this section for those who may have missed them on the Facebook page. ■

Successful immunotherapy for advanced EHE:



At the current time we don't have any predictive biomarkers that can tell us who is likely to respond positively to immunotherapy. That does not mean however that immunotherapy can't work. This paper, published in the National Library of Medicine in the USA, concerned a patient diagnosed with EHE in multiple organs and aggressive pathology, but who responded well to immunotherapy. It is excellent to see this positive report and to know that immunotherapy can be part of the 'clinical arsenal' for treating EHE.

The paper can be found using this link:
<https://www.ncbi.nlm.nih.gov/pmc/articles/PMC10063782/> ■

Update on systemic therapies for EHE:

New Molecular Insights, and the Role of Systemic Therapies and Collaboration for Treatment of Epithelioid Hemangioendothelioma (EHE)

Silvia Stacchiotti ¹, William Tap ², Hugh Leonard ³, Nadia Zaffaroni ⁴, Giacomo G Baldi ⁵

A new paper providing an updated summary of systemic therapies for EHE was also published in the second quarter. This was a collaborative paper involving European and US doctors, but which also included our own EHE community presented by Hugh Leonard, Chair of Trustees of The EHE Rare Cancer Charity UK. The collation and presentation of this data was largely possible due to the ongoing focus driven by our EHE community and the research and awareness created by the community's ongoing activities. Jane Gutkovich commented:

“ It is a first step. Of course we need more research, more data, we need better drugs, but we know now how to drive the process, and we'll do it! ”

This paper can be found in the EHE Library. ■

Excellent response of bone EHE to radiation



In May, a new article was published reporting on two cases where an excellent response of bone EHE to radiation was achieved. Jane Gutkovich noted:

“ Our EHE community sees some similar responses reported but unfortunately (and authors of this paper admit it) there are no published studies on the role of radiation in EHE treatment, just rare case reports. I speak with many EHE patients, and it is so frustrating to see that even doctors experienced with EHE, including radiation oncologists, are often hesitant to offer radiation as a treatment. This is largely because there are no published studies, and sadly nobody is looking at single cases because doctors need to see studies. I really hope that we will be able to quickly collect good data from our global patient registry and be able to publish it! ”

The paper can be found using this link:
<https://www.ncbi.nlm.nih.gov/pmc/articles/PMC10134116/> ■



02 EHE Research

EHE models described in new paper



We were also delighted to receive news in April of a new paper published by Emily Neil and Dr Valerie Kouskoff entitled “Current Model Systems for Investigating Epithelioid Haemangioendothelioma”. This paper incorporated information from Emily’s four-year PhD that was funded by The EHE Rare Cancer Charity and a follow-up piece of research funded by The EHE Foundation, and provided a comparison of several different EHE models that are now available for research.

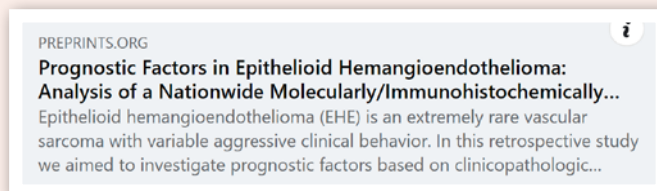
Jane Gutkovich was also excited by the progress in models that this paper high-lighted. Jane commented:

“ This paper provides real proof of how our EHE community moves EHE research forward. Although it is very technical, a paper like this can create a big leap in the development of effective treatments for EHE. How does this happen? This paper summarises and most importantly highlights the existence of models for studying EHE and for testing drugs. This in turn will be seen by many small and big pharmaceutical companies, as well as cancer research labs, as they search for cancers they can target with their products, and so will include EHE in their thinking! The important thing to note here is that EVERY model described in this paper was developed thanks to funding and/or tumor tissue provided by the EHE community. That support includes the PhD and research undertaken by Emily and Dr Kouskoff, the authors of this paper. So there are hundreds and hundreds of contributors to all this model research from our community, but whose names are not there. But that is fine, we are just glad that we have been able to have such a huge impact on EHE research up to now, and will continue to do so in the future. ”

This paper can be found in the EHE Library. ■

A new study of 57 EHE patients from the Netherlands

Researchers (including a good friend of our community, Dr Marije Weidema, who had previously published a study on the Quality of Life of EHE Patients involving data provided by 114 of our patient community) tried to confirm previously proposed prognostic factors for EHE using data from 57 EHE patients. While some prognostic factors were found for single EHE tumours, little was found for EHE patients with multiple tumors in either a single or in multiple organs, except for pleural EHE which is recognised to have a poor prognosis.



Jane Gutkovich noted:

“ The authors emphasized that since EHE is an extremely heterogeneous disease, more definitive answers could only be obtained by analysing a much large group of EHE patients. That is why we have launched our global EHE registry. If we can collect information from 400-500 patients around the world then we will have created a hugely powerful stream of information. We will also be able to keep updating the registry, adding new treatments, following long term outcomes of different procedures, learning how EHE is treated in different parts of the world. The first entry of data does not take long, and thereafter it’s an easy 10 to 15 minutes process twice a year to update your data. The data base the registry will create will be such a powerful tool for understanding EHE, so if you haven’t done it yet, please register through The EHE Foundation website. ”

This paper can be found in the EHE Library. ■

Regional Updates:

European Collaboration

Each of the editions of **The Pledge** this year will focus on the key research taking place in different regions of the world. Our Q1 edition was focused on the UK. This edition is focused on our European Collaboration. Q3 will focus on the US and Q4 on Australia.

The European collaboration including Istituto Nazionale dei Tumori (INT) Milan, Italy; the Institute of Cancer Research (UK); and the Royal Marsden Hospital, London

The overall research program entitled “Evaluation of Cytokines and Hormones as Biomarkers for EHE” is a multi-center collaborative project that comprises a number of key objectives which at a high level include the identification of novel prognostic and predictive markers; to better describe the natural history of the disease and its radiologic features; to collect prospective data on quality of life and the activity of systemic agents administered in EHE; as well as to comparatively assess the activity of drugs in newly generated preclinical models of EHE directly obtained from patients’ tumor specimens.



FONDAZIONE IRCCS
ISTITUTO NAZIONALE
DEI TUMORI

ICR The Institute of
Cancer Research

The ROYAL MARSDEN
NHS Foundation Trust

The different objectives or projects, can be summarised as follows:

- i) the identification and validation of novel circulating and tissue biomarkers to inform patient management (prognosticators and predictors of response to medical agents) as well as potential therapeutic targets **(Project 1)**;
- ii) the development of patient-derived xenograft (PDX) models **(Project 2)**;
- iii) the assessment of the activity of drugs relevant for the disease **(Project 3)**;
- iv) the generation of PDX-derived cell lines to investigate the cellular and molecular determinants of drug activity and to be submitted to a CRISPR whole genome screen to identify new therapeutic targets **(Project 4)**;
- v) the identification and evaluation of miRNAs **(Project 5)**; and
- vi) the description of the radiologic characteristics of EHE and their correlation with the clinical outcome **(Project 6)**.

Updates of these different projects are provided overleaf.



02 EHE Research

Project 1:

The assessment of circulating cytokines, hormones (and miRNAs), and ER α , Er β and GPER expression etc and the identification of a novel biomarker for EHE;

Using a protein array able to simultaneously detect the expression of a hundred different cytokines in plasma samples of EHE patients (n=15) and healthy individuals (n=6), a small panel of inflammatory cytokines was found to be differentially expressed. Among them we focused on Growth and Differentiation Factor-15 (GDF-15), a member of the TGF- β super-family, which plays multiple roles in a wide variety of cellular processes. Using a specific ELISA assay, we looked at the concentration of circulating GDF-15 in a retrospective series of 23 EHE patients and observed a statistically significant association of GDF-15 levels with EHE aggressiveness. This result was confirmed in a second cohort of 21 EHE patients prospectively collected within the currently ongoing observational study. The assessment of circulating hormones and tissue expression of ER α , Er β and GPER is ongoing.

The assessment of miRNAs is undertaken in Project 5.

Project 2:

Development of additional PDX models;

Thus far we have only one fully established EHE PDX model, whose ability to reproduce the characteristics of the originating clinical tumor has been confirmed in terms of histo-morphology, presence of the WWTR(TAZ)::CAMTA1, overall transcriptomic profile (as detected by RNA-seq) and presence of CDKN2A homozygous deletion. A cell line was established following disaggregation of the EHE PDX.

Project 3:

Assessment of activity of drugs;

The PDX model was used to comparatively assess the activity of doxorubicin and sirolimus (at different doses). Doxorubicin showed almost negligible activity while sirolimus caused a dose-dependent tumor volume inhibition in treated mice and induced the down-regulation of mTOR signaling. The PDX is currently being used to assess the activity of inhibitors of the Hippo pathway, such as the TEAD family of transcription factors.

This PDX and the corresponding cell line were exploited to provide further evidence supporting the value of GDF-15 in EHE. GDF-15 was detected in the medium of the EHE cell line as well as in the blood of EHE PDX but not in healthy mice or in mice carrying another sarcoma type, confirming that the cytokine was released by the EHE. Interestingly, we found that sirolimus decreased the abundance of GDF-15 in our in vitro and in vivo EHE models.

Project 4:

Use of CRISPR in cell lines to help identify genes that confer drug resistance or sensitivity;

The EHE cell lines will soon be provided to Dr. Paul Huang (ICR, London) to start the CRISPR-based experiments.

Project 5:

Identification and evaluation of miRNAs

The expression profiling of plasmatic miRNAs has been initially carried out using the OpenArray Technology (which evaluate the expression of 754 different miRNAs) in the retrospective series of 23 EHE patients. Six miRNAs were found differentially expressed between patients with indolent and aggressive EHE. In order to evaluate the overall miRNome (global profile of expressed miRNAs), we repeated the analysis on both retrospective and prospective series of patients by miRNA-seq. The results are currently being analyzed by a dedicated bioinformatician.

Project 6: Establishment and assessment of a radiological data base.

The clinically-annotated radiologic scan collection on the study platform has been started. Response assessment is also collected by REDcap CRFs. The data analysis is planned after the end of the study.

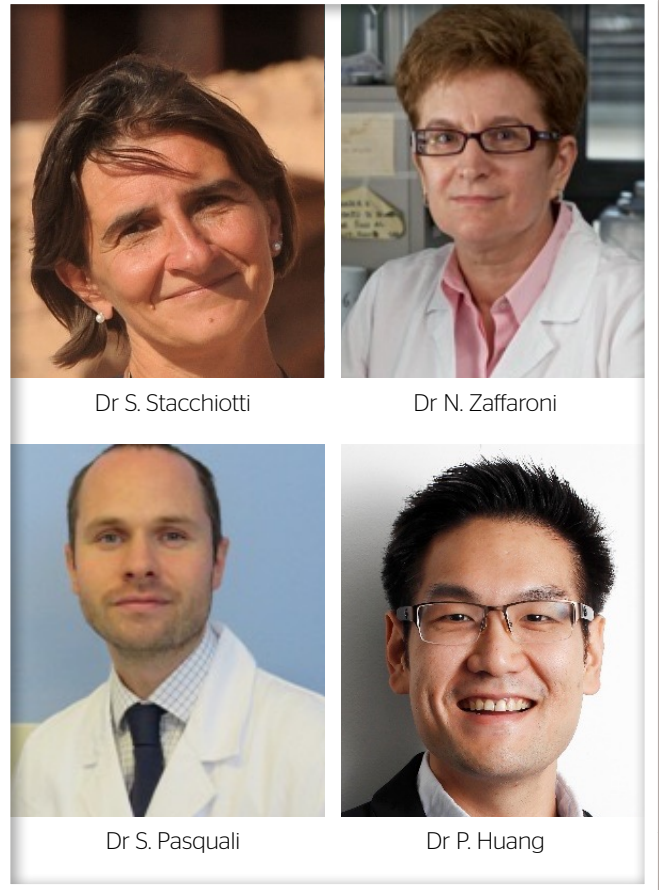
In summary

Overall the project is progressing well.

- The study is currently open to enrolment in Milan and, as from the fall 2022, in London. It has so far included 31 EHE patients (29 in Milan; 2 in London), from which clinical and radiologic data, blood samples and tumor specimens have all been collected.
- We already identified a circulating cytokine significantly associated to disease aggressiveness and validated the results in an independent patients' cohort.
- Using the newly generated preclinical models of EHE we have demonstrated the superior antitumor activity of sirolimus compared to doxorubicin.
- The search for additional prognostic/predictive biomarkers is ongoing on tissue and blood samples.

This is exciting as within a few years, we believe that data collected within this study will provide us with:

- 1) a better picture of the disease to inform the decision on how to treat patients with EHE and to define the prognosis based on disease presentation; and
- 2) preclinical and clinical data to identify new treatment options and design better prospective clinical studies.



Dr S. Stacchiotti

Dr N. Zaffaroni

Dr S. Pasquali

Dr P. Huang

Dr Silvia Stacchiotti and Dr Nadia Zaffaroni are joint PI's of the research taking place at INT, while Dr Paul Huang is PI for the ICR, London. Dr Sandro Pasquali is also a key member of the INT research team. ■



02 EHE Research

The EHE pan-European prospective observational registry within STARTER

The EHE Rare Cancer Charity (UK) is funding the setting up of the pan-European EHE prospective observational registry that will be managed and led by Dr Annalisa Trama and the team at INT in Milan, Italy. The registry will be maintained within the European STARTER project and patients will be enrolled and data contributed by clinical providers at the participating hospitals/centres. EHE is the first cancer within this important new data gathering initiative.

What is a cancer registry?

Generally speaking a cancer registry is an information system designed for the collection, storage, and management of data on persons with cancer.

There are two major types of cancer registries: population-based registries and hospital-based registries. Cancer registrars are the people who collect and report cancer data.

Population-based registries record all cases in a defined population (most frequently a geographical area such as a country (e.g. The Netherlands, Italy etc), a region (e.g. Tuscany) or a metropolitan area (e.g. Milan). Population-based registries are designed to provide answers to questions such as:

- How many cases of a specific cancer (e.g. lung cancers, sarcomas etc.) are newly diagnosed in a specific country, or in other countries?
- Is the number of newly diagnosed cancers increasing or decreasing?
- What is the prognosis? Is it ameliorating?

Hospital-based registries maintain data on all patients diagnosed with cancer at a particular healthcare facility. The focus of the hospital-based cancer registry is on improving patient care. These registries often provide informational opportunities for those who want to learn more about specific cancer types and support for those who may suffer from it. In other words, these registries are more clinically oriented and developed to support research and improve care for specific cancers.

Why a registry for the European Reference Network in rare adult solid cancers (EURACAN)?

The registry was developed considering that gathering data on rare cancers will support research to increase their understanding. The new evidence will help to develop and/or ameliorate clinical practice guidelines, to support multidisciplinary discussion and consultations for patients with rare cancers and ultimately to improve quality of care across Europe.

What did the STARTER (Starting an Adult Rare Tumour Registry) project achieve?

In the end, the project:

- defined the registry model and set up the appropriate IT infrastructure
- defined and implemented the legal and ethical framework
- developed the registry governance
- initiated 3 registries, the first one on EHE

Current status

We have finalised the tools for the data collection which will be tested by participating centres in the next few months.

What is exciting you about this work and the potential results?

We believe that this EHE registry will improve our information on the natural history of EHE, on the prognosis of patients with EHE based on disease presentation, on the activity (effectiveness) of systemic agents currently available for the disease, and provide information (real world data) which may be used in the future as comparison external data for non-randomised clinical trials. Moreover, we are working so that this data collection can in the future "talk" with other EHE data bases from extra-EU countries (e.g. US, Canada, Australia etc). Finally, we think that the registry will also help in harmonizing the treatment of the disease across European countries.

The EHE Registry project is being led by Dr Annalisa Trama and Dr Silvia Stacchiotti. Dr Anna Frezza is also a key member of the research team. ■



Dr A. Trama



Dr S. Stacchiotti



Dr A. Frezza



03 EHE Fundraising

The EHE Group remains focused on raising the funds needed to finance, drive and expand the existing EHE research programme that is so critical to finding new ways to treat the disease.

Highlights of those fundraising activities can be found in this section of our newsletter. We thank all our members, their supporters and members of the public for their fundraising efforts, some of which are described below.

We could not have achieved the progress we have without your wonderful contributions.

100 Miles Cycled for EHE Research

Each year the EHE Rare Cancer Charity UK secures a small number of places in the Ride London 100 sportive where participants cycle 100 miles on closed and empty roads. The event starts in central London and goes north east out into Essex before returning to London to finish on the iconic Tower Bridge.

Why do people ride 100 miles? Simply to raise funds for one of a huge number of fantastic causes, all of whom are desperate for funding. This year EHERCC had 20 places in the event and had a great turn out. Hugh Leonard, Chair of Trustees of the EHERCC, took part again together with his son Sam, all in support of Hugh’s wife Sally Baker, an EHE patient and Trustee of the Charity.

Sam Leonard said:

“Wow, that was fantastic but very very tiring! The last 10 miles coming back into London were very hard indeed with a series of flyovers to get over. But it was definitely worth it as we have been able to raise a lot of funding for EHE research.”

Paul Dean was again the star for the EHE team, as he was in the London Landmarks Half Marathon. On his bike, Paul completed the course in 4 hrs 48 mins, averaging more than 20 miles an hour over the whole ride. Unbelievable.

We want to congratulate all the riders for their brilliant performance and for helping to raise such badly needed funding for EHE research. ■



03 EHE Fundraising

2023 Fun Run Event in US Breaks Record

Thank you to every sponsor, participant, and donor, for helping make The EHE Foundation's 2023 EHE Fun Run and Walk the LARGEST in their history with 1,156 registrations and more than 60 teams! Registration for the event went through the month of April and supporters participated in the virtual event across the country mostly in May and June. Some teams were strictly virtual, while others gathered many people to complete the run/walk together. Other teams are planning to gather later this year

A variety of all ages and abilities participated in this family fun event. Together, they raised over \$60,000 to support EHE research. Those who registered received a "Shine a Light on Rare Cancer" t-shirt, bracelet, and "Just Live" temporary tattoo. Many people also supported the event by simply donating to a team of their choice. Some of the group events were huge, with one such group appearing on the front cover of this edition of **The Pledge**.

It is proven that rare cancers create awareness and attract science to their cause with grassroots fundraising and events. A special thanks to the 2023 EHE Fun Run and Walk Sponsors:



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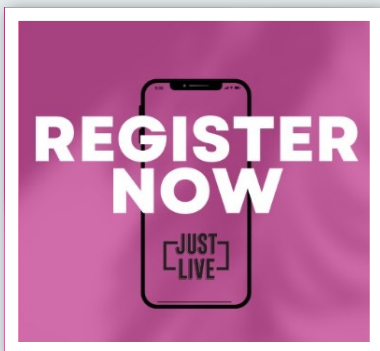
Check out more pictures and top teams at:
<https://fightehe.org/2023-fun-run/>



03 EHE Fundraising

Canada Launches its own EHE Fun Run!

We were delighted in June when Canada posted news that registration was open for the inaugural **Annual Just Live Fun Run**. There was an incentive to be



quick as the first 200 registrants for the 10/5 km run and the first 100 registrants for the 1 km run would receive a fantastic race kit and a **Just Live** race shirt! There was immediate interest with the first day taking \$8,000 in registrations. That equates to an amazing \$16,000 for EHE research as all proceeds will be matched by the Sarcoma Cancer Foundation of Canada. The organisers would really love to see as many people as possible take part if you'd like to make the trip to PEI, Canada!!

This is the latest fundraising event set up by Michelle Lynn under her 'My Journey to **Just Live**' brand. It will take place on 20th August which happens to be the 2 year Cancer-versary date of Michelle's diagnosis with EHE.

Michelle started running in January 2022, 5 months postpartum, and 4 months post her diagnosis with EHE; she never had run before. Her goal was to set herself up for success, and was hopeful for the best possible outcome to tackle EHE. Michelle is now often seen running, pulling and/or pushing all 3 of her children; Juliet (7), Adeline (4) and Hatton (1), just for fun. This event will have 'family' and 'running' at the heart, and with hope to raise funds along the way.

This event will be an Atlantic Chip Timing Event for the 5km and 10km Walk/Run; sponsored by Arsenault Bros. Construction and SFX Transport. It will take place at the Town of Cornwall with the route taking you along a picturesque waterview, and throughout

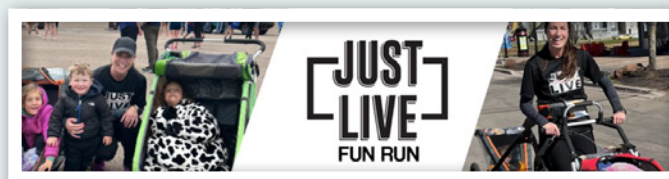
the trails within the woods. This is a looped event; with a clear, well-marked, breakout section for the 5/10km route change. There will also be a 1km accessibility Walk/Run for all abilities and ages. This route will include Mascots, Disney Princesses, and Super Heroes to add that extra fun for the kids!

Each runner/walker will receive a finish "medal," a token of accomplishment, "**I DID**" to add to the back of their race shirts that read, "**I CAN, I WILL.**"

Michelle was super excited by the funding arrangements which ultimately mean more funds for EHE research. Michelle said:

" This event is shaping up to be a FULLY SPONSORED event making every dollar count, thanks to our generous sponsors; so let's come together and support a great cause! "

We want to congratulate Michelle and the whole team behind My Journey to **Just Live**, and we wish you guys every success on 20th August. And registration is still open, so if you want to join in, you can register at:



<https://raceroster.com/events/2023/76100/just-live-fun-run> ■

A Small Group Making a Big Difference

In early June, Jennifer Mulligan shared news about a generous donation for The EHE Foundation. Jennifer commented:

“ Today our family accepted a donation from Briarcliff High School Pediatric Cancer Foundation Club. Joe accepted the check and my older son, who is an officer of the club, gave a beautiful speech! Julie and Laney Wahl were on video and said some beautiful words to the kids! This is the second annual donation! They hope to donate every year, too. ”

The donation, from fundraising carried out through the year by the Club, was made in support of Joe Mulligan (Jennifer’s son), who was diagnosed with EHE and has become a champion of our cause.



We want to add our thanks also to the Briarcliff High School Pediatric Cancer Club for their amazing support for Joe and The EHE Foundation. ■

Throwing Punches at EHE!

Many of our EHE global community use a boxing glove or clenched fist emoji when they post about their fight with EHE. Personalising the disease gives it a targetable characteristic. So we all smiled when Kelly Denton posted news of her latest fundraising idea:

“ Hey Guys, believe it or not I have a signed Tyson Fury boxing glove which I am going to raffle for EHE, so please buy a ticket if you can. ”

There was certainly a lot of interest because there will always be excitement when such an iconic item becomes available for a current reigning world heavy weight boxing champion. Kelly was certainly not disappointed with the final result:

“ Amazingly Tyson’s glove helped me raise £500 for EHE research! Let’s hope that money helps towards hitting EHE harder than a Tyson Fury punch! ”



We totally agree. Well done indeed for this amazing fundraising idea Kelly. And we also want to thank Tyson Fury for donating the glove. ■

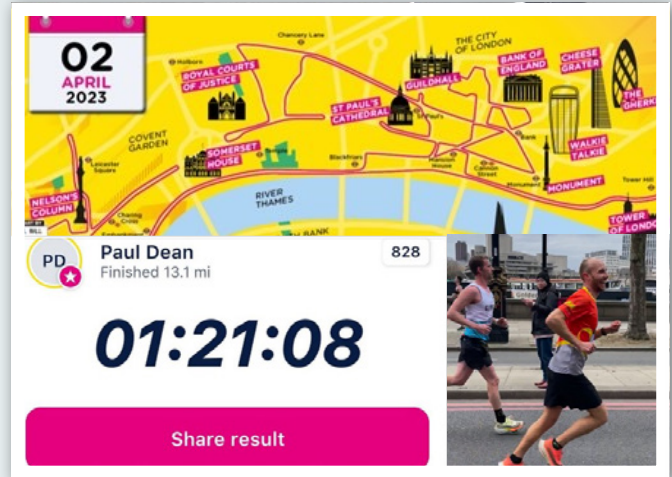


03 EHE Fundraising

2023 London Landmarks Half Marathon

The 2023 London Landmarks Half Marathon took place on the 2nd April, and the EHERCC had a team of 50 people running for the Charity, both to spread awareness of EHE and to raise critically needed research funding. Sally Baker, an EHE patient and Trustee of the Charity commented:

“ We had two teams of 19 within our 50 runners. Paul Dean, a fellow EHE patient, had a team of 19 runners, while Gingers Fitness, an outdoor fitness group from south London run by Paul Preston also had 19 runners, supporting Kelly and her daughter Neve, also an EHE patient. The other 12 who made up our 50 were all running in support of UK patients. ”



It was a fantastic day, not too hot and the rain stayed away. Paul Dean was the star of the show running the course in an extraordinary 1 hour and 21 minutes. Amazingly Paul was disappointed as he had set his heart on being sub 1.20.



Each year the Charity has to set up a cheer station on the course and this becomes a focal point for people who want to come and watch. Hugh Leonard loves the spirit of the event:

“ I find it inspiring to see so many people running for so many fantastic causes. And having 50 of those runners representing our EHE global community is really special. We cannot thank them enough. ”

As we go to press, the EHERCC is excited to already have 50 runners for the 2024 event. Bring it on! ■



03 EHE Fundraising

Mia is Running 100kms for 'her Zaza'



It is always heart-breaking to lose a dear friend to cancer, but that is particularly true when the friend is just 18. But that was what Mia Sallet faced when her dear friend Isabelle died in February 2018 from EHE.

Isabelle's family and friends were utterly devastated, but over time Mia realised that she did not want to forget Isabelle. Far from it, Mia remembers her every day.

Not only does Mia remember Isabelle, she wanted to do something to honour her friend and decided that a great way to do that would be to raise funds for EHE research through the EHE Rare Cancer Charity. Mia explained on her Facebook page:

“ Hello everyone, and thank you for visiting my page. I will be running 100km in a month in memory of my best friend Isabelle, gone too soon. My Zaza, my Isabelle, I still think of you every day, more than once a day, I miss you just as much as the day you left us, and will always have a hole in my heart, a deep sadness that will stay with me forever. I am doing this for you, for all the wonderful years we spent together, and all the memories I hold on to so so tight, it was too short.

I hope you'll be proud, because quite frankly, running is not my favourite sport in the world, except maybe in 6ème with the cross country ;)! I have been wanting to do something in your memory, and to celebrate you for a long time. Thank you to Andy, for asking me to run the Asics 10k with him, which will be part of my 100km challenge within a month. You've been gone 5 years, and it feels like yesterday. I am doing this for you, for your family, for all those who love and miss you and for EHE.

Miss you forever mon Isabelle, this is for you. Mia ”

Mia started her challenge in early June and plans to therefore finish her 100kms of running in one month in early July. We want to thank Mia for her wonderful support, and wish her every success over the last days of her challenge. We also look forward to reporting about the end of Mia's challenge in our next edition of **The Pledge**. ■

Encouraging Discussion about EHE and Sarcoma

As we know, EHE is a very rare form of cancer. It is one of many different rare cancers that are collectively referred to as sarcomas, and are cancers of connecting tissue in our bodies. While our EHE Group is of course focused on EHE specifically, and funding EHE-specific research, we also want to support sarcoma awareness.

Jane Biddlecombe, a Direct of The EHE Rare Cancer Foundation Australia wanted to highlight sarcoma as we approached July, which is designated as Sarcoma Awareness Month. Jane decided to do this using lovely bracelets, as she explained:

“ I am selling my handmade with love Pearl and gold bracelets to remind us to spread awareness of EHE and sarcoma throughout July. I only have 100, so for those of us that want to support research and spread awareness this is a fashionable way to do it! I always get compliments when I wear the bracelet, hence it gave me the idea, why not use it as a conversation starter to talk about Sarcoma and EHE. Plus the funds raised will be allocated to research currently taking place in the USA. ”



The Australian EHE Rare Cancer Foundation Australia is also launching a series of posts through July, which they hope will provide the tools and the language to allow people to talk confidently about sarcoma and EHE. Jane noted:

“ I know for me, being stable, sometimes its hard to talk about EHE when I look well having fully recovered from Chemo and radiation therapy. So talking more broadly about sarcoma is a great way to segway into talking about EHE. ”

Jane also explained how the bracelet represents EHE:

“ The pearl represents the rarity of EHE and the unique shape of each pearl represents the individuality of each patient - were no two cases are the same. Sarcoma is symbolised by a yellow ribbon, so the gold beads represent sarcoma and EHE. ”

Jane has been delighted with the interest in the bracelets and was delighted to post a photograph of bracelets packed and ready for shipping to Melbourne.

If you want to purchase a bracelet then just follow the link below. There you can make a donation of A\$50 to purchase the bracelet. For those living overseas the process is slightly more complex so please contact Jane Biddlecombe to check on details for delivery.

Follow the link and donate \$AU50 (tax deductible) <https://s4g.co/b/8ATUgB>

We want to congratulate Jane on another imaginative strategy to help spread awareness of EHE and sarcoma in general. ■



03 EHE Fundraising

Stretch for Sarcoma

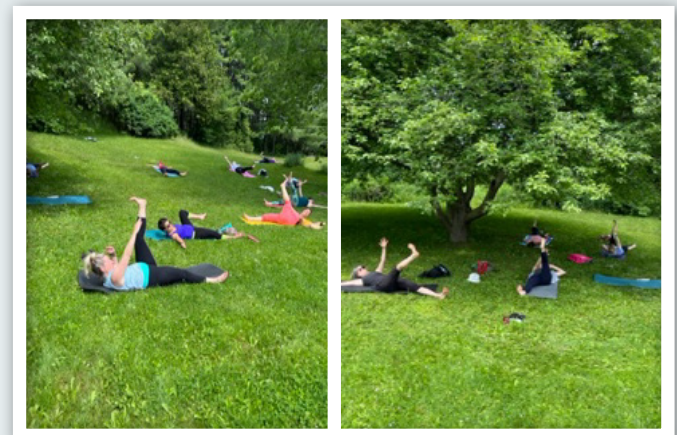
Fiona Louise was excited in May to announce a new opportunity to participate in exercise sessions while raising funds for EHE rare cancer research. Fiona explained:

“ A former colleague and science teacher, who also teaches yoga, approached me about the idea of using yoga as a fundraiser. She created a poster and a google doc (teachers know how to organize) and away we went. We hosted our first event on June 25th and have a second planned for July 16th. ”

The events will be held outside, weather permitting, which was lovely for the participants at the end of June. In addition to the yoga, a few local people donated some 'door prizes': make up, bracelets, beauty items and an incense burner as incentives for people to sign up. The team charged \$25 for a one hour session with \$5 going to the instructor and \$20 (with tax receipt) going to the Sarcoma Cancer Foundation of Canada (Pro Care EHE). After one event the team were well on their way to their target of raising \$500 for EHE research.

We love the idea of our EHE supporters using relaxation as a way to raise funds for research while at the same time doing something positive for their general health and wellbeing.

Well done guys. Keep it going! ■



The OMA Project is launched

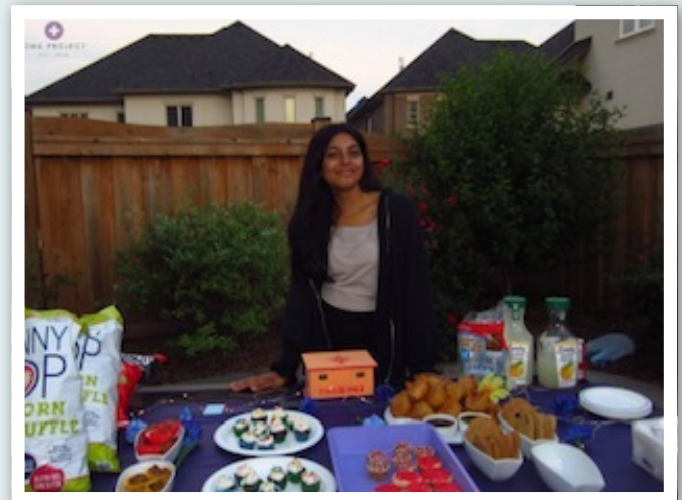
When Fiona Ross spoke to her former high school about EHE, she was approached by a small group of three young women after her talk. They asked if they could help spread awareness and fundraise for EHE? Fiona explained:

“ These students are in the International Baccalaureate (IB) programme. This programme includes the “Community Action Service” (CAS) component under which the students are required to commit 120 hours of voluntary time to a suitable cause, and they have selected EHE!

The group have called themselves the OMA Project. According to the medical dictionary the suffix “oma” means a swelling or tumor. Many words in medicine end in ‘-oma’. Some examples include adenoma, atheroma, carcinoma, fibroma, and of course hemangioendothelioma. ”

On July 14 they organised a backyard movie viewing party. They spoke about EHE and rare cancer research and provided healthy snacks for purchase. They also had a lot of fun, and raised an amazing \$373.55. This wonderful trio are planning other fundraising and awareness events already.

We want to join Fiona and the Canadian EHE community in saying a huge thank you to the OMA Project team. ■



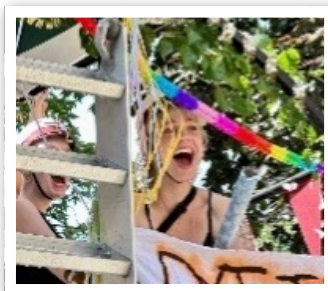
04 And in other news...

Every quarter we include messages that have been posted by our EHE community that are not necessarily related to EHE, but reflect the *Just Live* spirit of this wonderful group! Here are this quarter's contributions

Celebrating Graduations

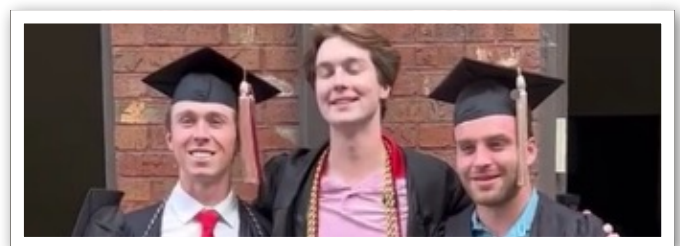
This part of the year is a tough time for many students as they face key exams at school, high school or university. So, we love it when people share news of their kids graduating.

Rikke Pedersen posted news of the graduation of her daughter Rosa at Malthe Bruuns Vej, Frederiksberg, Denmark. Rosa graduated 18 months after her diagnosis and attended school every day even while doing 8 rounds of doxorubicin. Rikke commented:



“ My daughter Rosa’s Graduation Day, 14 years of school done, so proud of her. ”

Lael Bellamy also posted news of her son, Foster, graduating. Lael provide a lovely video of the day and said:



“ Foster (in the middle) was diagnosed with EHE second semester of his freshman year in college. He was coughing up blood. Long story short, I found this group. He started Sirolimus. Covid happened. He road-tripped across the country and surfed in Malibu. He seemed stablish so got his wisdom teeth out, which nearly killed him because he had to go off Sirolimus. Bounced back later that summer and learned to dive in Key West.

Had a completely normal college career. Graduated yesterday summa cum laude from the honors program at the University of Georgia. He’s spending the summer as a camp counsel in Hawaii—surfing and diving. Might take a gap year in Spain. Might go to law school. *Just live.* ”

We love these stories and want to congratulate every graduate who is part of our EHE community. ■

Mothers' and Fathers' Day Celebrated

When a life is affected by a rare cancer like EHE, the questions and uncertainties about the future may feel overwhelming. On Mother's Day, the EHE community celebrated every mother who has had to worry about these uncertainties for themselves or for their children.

“ Finding a new normal after a rare cancer diagnosis is a challenge, but I try to find beauty in every single day especially on special days like Mother's Day, ”

said Stephanie Kennedy, who is a mother of two and grandmother of six (with one baby on the way).



Happy Mother's Day to those who give so much to their families day after day. ■

Jane Gutkovich shared her annual Mother's Day message:

“ I post this picture every year on Mother's Day. To all moms in this wonderful family. WE GOT IT! HAPPY MOTHER'S DAY! ”

We think this picture also symbolizes the strength of our EHE community. As individuals, people may feel small and relatively weak, but inside our community, we each have real strength behind us, supporting us, and looking out for us.

Powerful picture... Fear nothing in front of you because of who is behind you !!!!



But it's not just about mothers. Jane also posted her own message for all the fathers in our community,

“ Happy Father's Day to our brave, gentle, dedicated -to-family men. Hope you all are having a wonderful celebration! ”

Join us in celebrating all the mothers and fathers in our community. You are all truly awesome! ■





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

EHE Italia - Non solo Laura
website not yet available

EHE Canada
website not yet available