

ISSUE TWELVE
MARCH 2016



Living for today....
Planning for tomorrow.

Newsletter

HAE UK is an Association of HAE Patients, working together to improve the situation for all HAE Patients in the UK

Welcome to our Spring 2016 newsletter

Welcome one and all to the first of our newsletters for 2016. Winter at last seems to be loosening its soggy grip and the days are lengthening which is always heartening. Sadly, some of you have had several attacks over the Winter, with most often viral infections being the trigger, but hopefully the fine weather will help us all to put a 'Spring' in our step!

Since Christmas Rachel and I have had a busy time keeping up with all the good news that has been coming in! So this issue of the newsletter is largely about our various fundraisers: Dana Shapiro climbed Mount Kilimanjaro, following in the footsteps of Ed Price who did it a few years ago. Rick Talbot did his Boxing Day Swim, both raised a staggering amount of money for HAE UK. And clever Izzy Sealey and her friends raised money by taking part in a project at their local school.

See all of these stories later in the newsletter.

THE QUARTERLY QUESTION:

Where does my C1 come from?

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By the time you receive this newsletter, Samantha Jones will be just one day away from running the Cardiff Half Marathon and Furkhanda Haxton will be nearing the end of her 'Healthy March' refraining from all her favourite treats, including chocolate!

On 14th and 15th May Danny Owen will be taking part in the London Revolution, a 185 mile cycling sportive around London. Both these two will be wearing our new HAE UK T-shirts (see later!)

We cannot tell you how grateful we are to all these people and the many of you who support HAE UK. Finances are always a difficult area for small charities and we rely heavily on individuals. As a small and niche charity, it is also difficult to raise awareness with the wider medical community let alone the general public. With HAE Day coming up (16th May) we are going to send all of you two posters and would very much appreciate it if you could take one to your local A&E and one to your GP practice. Ask if they will put them up in waiting rooms or common rooms so that we can get our message out there!



One of my dearest aims is to establish a youth wing to HAE UK, but my initial idea of a specific meeting has rather fallen through because of the difficulty of getting everyone together. So the Trustees have agreed that we will go at the project a slightly different way:

We would like to sponsor a person under the age of 18 (and a parent or guardian) to go to the HAEi Global Conference in Madrid this May. The meeting has a specific 'Youth Track' and we would like our delegate to go and meet other young people with HAE and join in all the lectures and discussions

so that they can come back to the UK and be our official 'Youth Ambassador', working with us to create and manage our HAE UK Youth wing. This will not just be an honorary position, we will expect you to work! See more details later in the newsletter.

Finally, I make no apologies for this newsletter being mainly about celebrating the wonderful fundraisers. But all of you can help – we are very happy to receive donations through Payroll Giving or why not set up a Standing Order through your bank account for a small amount each month? Please 'HELP US TO HELP YOU!'

HAE UK Patient Days 2016

Bristol:

We are pleased to announce that the 2016 HAE UK Patient Day 2016 will be held on Saturday 19th November at the Mercure Bristol Holland House Hotel and Spa.

This year the meeting will be led by Dr Mark Gompels and his colleagues from Bristol. We also hope to be joined by HAE International's Henrik Boysen and Tony Castaldo.

Perth:

We are also pleased to announce that we are holding a patient meeting in Scotland. Due to everyone's diary commitments we have plumped for Saturday 1st October, in Perth.

The meeting will be led by Dr Richard Herriot.

More details about both of these patient days will follow in the next newsletter.

hae day :-) - 16th May 2016

For hae day :-) we will be sending out posters to all of our members: please take one to your local A&E department and one to your GP. Particularly with the A&E depts. ask if they can be placed somewhere staff can see them, because we hear too many tales of people going to A&E and, even if they have letters from their consultant, not being believed about their condition. If they want more information, pass their details on to laura.szutowicz@haeuk.org or rachel.annals@haeuk.org and we will be happy to send them more details.

If you have changed address and don't think you have updated your details with HAE UK, please e-mail: rachel.annals@haeuk.org

T-Shirts

Rachel has designed HAE UK branded T-shirts for our fundraisers and also for anyone who would like to support us. We are planning to wear them at the Global Conference in Madrid so we can be spotted easily by HAE UK delegates. If you would like a T-shirt yourself, please contact us.



Izzy Sealey and her friends Catherine and Lily



Izzy, Catherine and Lily took part in a Maths Business project held at lunchtime in their school. They were tasked with making items and selling them to make a profit.



In total the girls raised £200.46. Izzy was only diagnosed with HAE in March last year, so her friends agreed their profits should be donated to HAE UK. *Shown left to right: Catherine, Izzy and Lily.*

Dana Shapiro

The first time I saw Kilimanjaro was in November, 2013, and I knew immediately that I wanted to attempt the climb. I had been trained to administer my own C1 during autumn of 2013. During that trip to Africa, I self-administered my second IV successfully in the Serengeti, despite my intense fear of needles. During 2014, I had 44 HAE attacks, most of which were treated with self-administered C1 or Icatibant.

I first discussed the idea of climbing Kilimanjaro with my consultant in January, 2015. I was not sure if I could make it to the top – not everyone who attempts actually makes it – but I wanted to try. To my surprise, my consultant was supportive, and suggested a few preventative measures. She made me aware of the risks of the climb, most notably that altitude causes



increased fluid leakage through the walls of the blood vessels which means that HAE attacks are more likely at higher altitude.

As part of my preparation for the trip, I began taking the steroid Stanazolol daily as an additional precaution with the hopes that I would not have any HAE attacks while

climbing. I had terrible side-effects from the pills for the first 2 weeks, but I persevered until my body could tolerate them. In the months leading up to the climb, I trained my body for the exertion by walking and eventually running to work. My final endurance test was to run the 6k to work consecutively for 14 days straight. I had my doubts that running the flat lands of London might not be the best preparation for trekking a mountain for 8 days, but was confident that my improved cardiovascular

health and endurance would still help.

The morning I began the climb, I administered 2,000IU of C1 in my hotel room in Moshi, Tanzania. The first few days of the climb felt easy to me. I was even able to completely enjoy the gorgeous and ever-changing scenery. The air was still thick with oxygen. I managed to sleep well. I was on altitude medication that kept draining my body of fluids, so I drank litres upon litres of water every day. I felt strong, capable and excited as the temperatures steadily decreased. There were 10 other trekkers climbing with me, and I was one of the lucky few who got very limited altitude sickness symptoms. But my fears centered around the 6th day of my trek, when I would need to administer another C1 IV at 4,600 meters above sea level. This would be at near freezing temperatures with no running water, after being unable to bathe for 6 days.



I had been sleeping with my C1 and Icatibant in my sleeping bag at night to ensure it did not freeze as the temperatures dipped below freezing during our ascent. I did not sleep on the 5th night of the trek, and it is hard to know if it was from the altitude or because I was so petrified of giving myself an IV on the mountain the following day. In the afternoon of day 6, I sat with one of my friends in the dining tent and managed the IV with more ease than I could have imagined. My friend, who happens to be a doctor, was encouraging and supportive the entire time. Having her there made me feel less nervous, and it turned out I was able to give myself the IV without her physically helping.

We went to bed early that night, but sleep was hard to come by. For me, the toughest part of

the climb had happened in the tent earlier in the afternoon, but I eagerly anticipated the part of the journey that I had not thought would really be possible.

At 11:30pm, we awoke to begin our ascent to the summit. While I was still in my sleeping bag, I gave myself a subcutaneous Icatibant injection as another preventative measure and carried an additional Icatibant jab and C1 dose just in case I needed it at the summit. We started trekking up the last 1,000+ meters to the Kilimanjaro summit at midnight, under the cover of the most incredible stars I had ever seen.

“Poli poli,” slowly slowly, our guides repeated through the night, and encouraged us to keep taking our tiny steps towards the top. I was lucky: I didn’t have any HAE attacks on the trip, and I made it to Uhuru Peak at 5,895 meters (19,341 ft) above sea level with 8 other members of my group.

The lack of air pressure at the top of the mountain makes you feel like every breath you take is a shallow, quarter-gulp of air. But I felt giddy with accomplishment after making the heavy footsteps to the top. As

I looked out over the vast planes of Tanzania, I reflected on all the times when people had told me that I couldn’t do something because it might make me sick, or that something was not a good idea because I had HAE, or the times when I had to sit on the sidelines because I was too sick to be in the game. Determination, will power and the right type of support made my crazy goals a reality.

As the fatigue of the day intensified, and I trudged back down past the receding glaciers at the summit, I was overwhelmed by the enormity of what I could accomplish when I believed it was possible and was surrounded by people who said, “Yes, go do it!”

Dana raised an incredible £2,482.46 for HAE UK.

Rick Talbot

Rick Talbot, a fellow sufferer of Hereditary Angioedema (formally known as Angio Neurotic Oedema) successfully took part in the Annual Boxing Day Sea Dip organised by Llandudno Lions Club in the Irish Sea on Boxing Day 2015.

Rick, along with in excess of 100 hardy souls, the majority of which were in fancy dress and supporting various charities of their choice, joined the annual dash into the sea, swam around for a couple of minutes then waded back in to the shore. In the process Rick raised a sum in excess of £1,800 for HAE UK.

After the swim Rick said that it was in fact not too



cold, but this may have been because the rain was pelting down almost horizontally, which made the sea feel quite warm (it is far colder in April after the winter months). Also because of the atmosphere amongst the contestants, everyone was buoyed along, in fact the comradie amongst everyone including the 100's of spectator's and members of his family, made it all worthwhile.

Rick also said a big thank you to all those who supported him both on the day, and also financially, in order to make a healthy sum for HAE UK.

Supporting HAE UK - Help us to help you

Thank you

Payroll Giving



Payroll Giving is a flexible scheme which allows anyone who pays UK income tax to give regularly and on a tax free basis straight from your wages or pension. This happens before tax is deducted from your income.

To set up a regular donation to HAE UK please complete the Payroll Giving form: www.givingonline.org.uk/HAE-UK_2407/index.html

Setting up a monthly standing order

It is easy to set up a standing order direct from your bank account to the HAE UK account - we are happy to provide you with these details upon request.

Shopping online or booking your summer holiday online?

....why not register with **easyfundraising** and nominate HAE UK as your chosen charity? By doing this, each time you make a purchase online with one of the 2,700+ participating retailers, a small donation will be made to HAE UK, with no cost to you!

Retailers include Booking.com, Hotels.com, First Choice, easyjet, Amazon, John Lewis, Argos, eBay, Next and many many more. To register see: www.easyfundraising.org.uk

HAE Global Conference in Madrid, 19th-22nd May 2016

'Creative Advocacy for Expanding Access to Therapy'

This exciting conference, which takes place every two years, offers the opportunity to hear from some of the world's top HAE specialists about developments in HAE management.

The conference is open to patients and their families, clinicians, nurses and research scientists – and this year there will be a separate track for young patients.

The organisers expect to have 600 attendees taking part from all over the world.

All available travel grants have been allocated for the conference now, but places are still available if people would like to attend and the registration rates are very reasonable. We have over 20 people attending the conference from the UK.

We look forward to seeing you there!

In the week before the conference, and as part of hae day :-) HAEi have also organised the HAE Camino Walk, walking part of the legendary pilgrimage route El Camino do Santiago. This event takes place from the 15th to 17th May and there are 80 people registered to take part so far. If you are interested in this fantastic opportunity please see the HAEi website.

HAE UK Young Ambassador

We are inviting applications from a diagnosed HAE patient under the age of 18 who would like to be our HAE UK Young Ambassador and attend the Global Conference. The conference registration, accommodation and flights will be paid by HAE UK for the Young Ambassador and parent/guardian.

Young Ambassador applicants are requested to submit a short essay (up to 500 words) titled 'What I will do as HAE Young Ambassador' to outline how they will use the experience at the Conference to go forward and help the HAE UK Team to establish and manage a useful support group for younger HAE patients. The Trustees will then judge who they consider will be the best choice for the Ambassador.

Please accompany your essay with your full name and address, age, name of parent/guardian and preferred airport in the UK to fly from. It will also be necessary for the lucky applicant to have permission from their school for leave to attend the conference (Thursday 19th and Friday 20th May).

Please forward your applications to Laura as soon as possible: laura.szutowicz@haeuk.org

Registering with HAE UK

We are currently reviewing the information we hold about our members and updating our records accordingly.

To ensure the information we hold about you is current, we are e-mailing you all on a person-by-person basis, asking you to re-register using the new form on our website. We would also like you to encourage all of your family members with HAE to also register, this will enable us to build an accurate

picture of patients across the UK and we can better support you all.

Please keep an eye out for this e-mail in the coming weeks. Once re-registered, all new members will receive one of our new Welcome Packs through the post.

You can register here by following this link: www.haeuk.org/advice-support/register

THE QUARTERLY QUESTION

Where does my C1 come from?

Here in the UK, most of the C1 INH used is manufactured from human plasma, although there is a small amount of 'recombinant' product also used.

The plasma used to produce C1 INH is sourced from plasma donations from countries with very low risk of vCJD, UK plasma is not permitted to be used because of this. All donor centres, donors and donations are carefully selected, screened and tested for viruses in compliance with International regulations, and all donations are subject to 'Inventory Hold' for 60 days which further reduces the risk of viral contamination entering the manufacturing cycle.

The plasma is then 'pooled' ie plasma from 1500-2000 individual donors is mixed together and the process begins with 'fractionation' which separates all the active components of the plasma out in stages.

The manufacturing process includes multiple steps that reduce the risk of virus transmission. The virus inactivation/reduction capacity consists of minimum 3 steps such as;

- Pasteurisation in aqueous solution at 60°C for 10 hours
- Hydrophobic interaction chromatography
- Virus filtration (also called nanofiltration) by 2 filters (20 nm and 15 nm filtration)

The final product is then packed, labelled and packaged for you to use. Even the most modern procedures result in a yield of less than 150 milligrammes per litre of plasma, which is why it cannot be manufactured from single donations. Every time you use your C1 INH, it can be traced back to the every single one of the original plasma donors. Quite a mind-boggling thought!

NOT TO MISS:

Medical Mysteries Thursday 31st March 8:00pm, Channel 5

Medical Mysteries is a medical documentary meeting patients in search of answers to their mysterious conditions. Next Thursday, 31st March, is episode two which stars Izzy Richards and her HAE story.

The programme will also be shown on Channel 5+1 at 9:00pm and on Channel 5+24 on Friday 1st April at 8:00pm.

Upcoming dates for your diary:

Saturday 26th March
Sam's Cardiff Half Marathon

Thursday 31st March
Medical Mysteries, 8:00pm, Channel 4

Saturday 14th and Sunday 15th May
Danny's London Revolution Cycle Ride

Sunday 15th - Tuesday 17th May
El Camino Walk, Spain

Thursday 19th - Saturday 21st May
HAEi Global Conference, Madrid, Spain

We wish you all a very Happy Easter and look forward to seeing some of you in Madrid in May.

*With our best wishes from Laura & Rachel
and the Trustees*

www.haeuk.org
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