



Your HAE patient support association

HAEUK

# Newsletter

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

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## Welcome to our winter newsletter

I started with HAE UK on 1st July 2015 so have just completed my fifth month with HAE UK..... the words 'feet' 'ground' 'touch' and 'not' can be composed into a simple sentence that describes those five months!

Ann and John Price were incredibly generous with their time and expertise during my first three months, introducing me to key people, teaching me all about the running of the charity and generally setting me up with all the basic knowledge I needed. They then (wisely) went on holiday for a month and left me to it! I don't think too many disasters happened.....

*Laura*

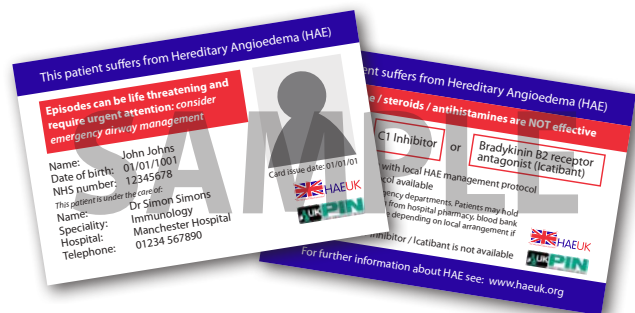
## THE QUARTERLY QUESTION:

How did my clinician decide what product to prescribe for me and my HAE?

## The HAE A&E Card

### Reasons to have a HAE A&E Card

- The photographic identification card contains your details and helps you to explain the need for urgent treatment in A&E in the event of an airway swell or a severe abdominal swell
- The card tells first responders that you have HAE and how to treat you in an emergency, if you were hurt or too unwell to speak
- The card contains the contact details of your HAE consultant should the A&E clinician treating you need advice
- Along with the card you will also receive a letter you can take to A&E which contains more detail about HAE, the severity and required treatment
- Your local A&E department will also receive a copy of the letter so they are aware of you as a HAE patient



### How to get your HAE A&E Card

Obtaining your card is easy. You will need to request an application pack, complete and return it to HAE UK, along with a clear photograph. Once your application has been approved by your HAE consultant, your card will be produced and posted to you. Please e-mail: [rachel.annals@haeuk.org](mailto:rachel.annals@haeuk.org) to request your application pack.

# HAE UK Patient Day 2015

Our 2015 Patient Day was held at Maple House in the centre of Birmingham. It was easy to access from New Street Station and the new shopping complex on top of the station gave quite a few attendees to get in some early Christmas shopping (you know who you are!). The event proved very popular with over a hundred attendees and this year we seated everyone 'cabaret style' around tables, which worked quite well although some tables suffered slightly obscured views because of pillars in the way, something to look out for next year.



Our brilliant speakers were mostly from the Birmingham Heartlands Immunology Centre which has a large cohort of HAE patients and this year's theme was '**HAE through the changing stages of life**'. First to present was Dr Scott Hackett, paediatric immunologist, who gave a very comprehensive presentation on diagnosing HAE in infants and treatment of children from babyhood to adolescence. The next speaker, (possibly slightly in the wrong order – think chicken and egg), was Dr Sarah Goddard speaking on conception and pregnancy, with a following patient story from Izzy Richards who was at the meeting with 2 month old baby Benjamin as evidence of a successful pregnancy!

Dr Hilary Longhurst then gave a detailed and interesting presentation on managing the transition from paediatric services to adult, which can be a difficult time for teenagers because in the UK this move happens at age 16, which is just when they are in the middle of examinations as well. Dr Richard Baretto led us through the management of HAE in older patients and showed some very good slides on quality of life. The morning was finished off by Dr Aarn Huissoon who changed the theme with a presentation on drug trials; what possibilities there are in the various company portfolios and the importance of taking part in clinical trials, with Nicola Bowen speaking excellently on the patient experience of being involved in trials.....and then it was time for lunch!



After lunch Laura gave a short presentation for HAE UK, outlining some events and plans for next year. We then held break-out groups with topics similar to the morning and extra groups on HAE and Employment Issues, Women's Issues and Preparing your Child for School. Everyone had the opportunity to attend two of the break-out groups. After a short coffee break, a final question and answer session concluded the day.



We have had a great deal of good feedback from the day and many suggestions to make next year even better! And we have already started planning it.....



Our thanks to all the speakers, and to our Medical Advisory Panel: Dr Longhurst, Dr Gompels, Dr Crouch, John Dempster, Christine Symons and Fran Ashworth, also our Trustees John Price, Ann Price, Ed Price and Ann Harding. Thank you also to Furkhanda who greeted everyone on arrival, handed out the delegate packs, ran the raffle and was generally indispensable. She was aided and abetted by Rose and Leyon, huge thanks to them, also to Ray Frappell who brought posters of the HAE lorry, to Jimmy and Marion and to everyone else who helped make the day a success. And special thanks to our wonderful Rachel Annals without whom I would have been completely at sea!

We look forward to seeing many of you at the 2016 patient day.

## **Fundraising** - A huge **THANK YOU** to everyone fundraising for HAE UK

A huge thank you to Dana Shapiro who climbed Mount Kilimanjaro in August to raise money for HAE UK. Dana said it was an amazing experience and we look forward to featuring some photo's of this event in our next newsletter

One of our members, Rick Talbot, is going to don a lion 'onesy' in order to take part in the annual Boxing Day Sea Dip at Llandudno in aid of the Lions Club and HAE UK. For those of us who cannot get there to support him, he has agreed to provide photographic evidence, so look out for this in the March newsletter!

Rick has already raised a tremendous amount of money and is willing to accept more, so anyone wishing to donate can do so through his JustGiving page: [www.justgiving.com/Rick-Talbot](http://www.justgiving.com/Rick-Talbot).

Another mad but wonderful person is Danny Owen whose wife suffers with HAE. Danny is taking part in the London Revolution, cycling over 185 miles in two days around London in May. You can support Danny in his challenge by visiting his JustGiving page: [www.justgiving.com/Danny-Owens](http://www.justgiving.com/Danny-Owens)

For anyone else wanting to raise funds for HAE UK, Rachel can provide a 'fundraising pack' with sponsor forms, ideas for fundraising events and so on. We are also able to print t-shirts, sweatshirts and tabards with our logo for you to wear for your event! Please get in touch by e-mailing: [rachel.annals@haeuk.org](mailto:rachel.annals@haeuk.org)

We have also received individual one off donations through JustGiving and via our Text Donate number 70070, monthly donations from members and friends, as well as kind donations in memory of loved ones.

## **Shopping online this Christmas?**

...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 Amazon, John Lewis, Argos, eBay, Next, ToysRUs, Tesco and many many more.

HAE UK currently has 14 supporters who have raised £25.30 without paying a penny themselves. Thank you for your support. To register see: [www.easyfundraising.org.uk](http://www.easyfundraising.org.uk)

## **The HAE Lorry**

Another exciting thing to happen this Autumn is that Ford have kindly allowed one of their lorries to have panels advertising HAE UK. This was organized by Ray Frappell who works for Ford at Dagenham and it will not only be driving around the UK but it will be going over to Europe too! So keep a look out for it and let us know if you spot it!



## **HAE DAY** **16th May 2015**

At the patient day we asked for ideas for a theme for HAE day – along the same lines as the brain cancer 'Wear a Hat', the Anaphylaxis campaign with their orange wigs.....we need something to make us stand out!

Are we going to all wear onesies for the day, a la Rick Talbot?

All ideas appreciated! Please e-mail us with any suggestions you may have: [support@haeuk.org](mailto:support@haeuk.org)

## UK PIN

Laura attended the UK PIN meeting in Belfast in November. This is a meeting open to all the clinical immunologists, immunology nurses and lab staff in the UK. Dr David Edgar, currently Chair of UKPIN is based in Belfast and made us very welcome.

There were many interesting presentations and posters and quite a few based around HAE. The accreditation of Immunology Centres has been taken over by the Royal College of Physicians and Laura had an interesting conversation with them about how the process of continually improving services is quantified. There is quite a large sector of this process dedicated to the HAE service provision, which is great news.

One of the highlights of the meeting was Faye Davenport who gave a fantastic presentation for the Shire symposium on HAE. Faye spoke on how the diagnosis of HAE, first of Ruby and then of herself, has changed their lives and gave real insight into the day to day reality of HAE. Her presentation was sometimes almost tragic, often humorous but never boring. Our heartfelt thanks to Faye for being so brave – she spoke to a packed meeting – and the very many comments received afterwards from clinicians showed that she had really struck home.

Which leads us onto another idea!

At the Patient day, one of the questions was 'What is HAE UK doing to raise awareness of the condition, particularly with GPs?'. Looking at ways of promoting the condition with GPs is one of Laura's priorities, as often the saddest stories heard from HAE people are all based around how long diagnosis took, and how they were shunted from pillar to post before anything constructive happened. Now, the big problem with rare diseases is that GPs rarely see them; so they are often at a loss to diagnose the condition.

We are sure that the best educators are you yourselves!

Almost every practice will have regular meetings of the doctors, nurses, health visitors etc. Why not ask if you can go along to one of these and make a short presentation about HAE? Our patient booklet and our website provide a huge amount of information and Laura is looking into making a short presentation that can be used. If you don't want to speak yourself, Laura, Rachel or one of our other contacts would always be prepared to.

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

This exciting conference 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, to clinicians, nurses and research scientists – and this year there will be a separate track for young patients. The organisers expect to have about 600 attendees.

Registration will be open in the coming weeks and we will notify all of our members as soon as we hear ourselves.

## Studies

There are several important studies we would love members of HAE UK to be involved in, some are treatment based, others non interventional. Details are posted on the website's News & Events page. More will be added as they come in, so please all get involved and help improve the knowledge and treatment of HAE.

## HAE UK YOUTH WING

At the patient day Laura announced that she wants to set up a group for and run by our younger members. Obviously we will have to keep a little bit of an eye on you for all the boring stuff like Health and Safety and Child Protection, but she already has a couple of adult volunteers with the requisite clearances.

What we need now are some interested people between the ages of 9 and 16 who would like to be involved in the first project which will be to design a patient information booklet especially for this age group.

Laura will be hoping to have our first meeting in about March and the location will be determined by where the majority of attendees are based.

Please get in touch with us if you are interested in helping out with this project.

# THE QUARTERLY QUESTION

## How did my clinician decide what product to prescribe for me and my HAE?

*Dr Hilary Longhurst answers;*

In the UK we are fortunate to have a choice of 3 approved and funded treatments for HAE attacks. NHS guidance suggests that anyone with hereditary or acquired angioedema due to C1 inhibitor deficiency should receive C1 inhibitor (Berinert, Cinryze or Ruconest) or icatibant for any attack which is likely to cause severe pain or which could spread to affect the throat.

All treatments probably act to stop the build-up of oedema very quickly. However, it takes some time for the body to reabsorb the oedema which is already there, so after treatment it takes between half an hour and several hours before there is noticeable improvement and several hours to a day or more for the swelling to completely disappear.

Icatibant is supplied in a prefilled syringe and is injected into subcutaneous (skin) of the tummy. It is very quick and easy to administer at home, at work or when travelling. After a brief training, almost everyone can master self-administration. Some people with very rapid onset attacks, or who have had laryngeal attacks in the night, like to keep a syringe of icatibant by their bed, in case they wake in the night with a severe attack needing rapid treatment. Icatibant is undergoing trials for use in children and is only used for under 18's in exceptional circumstances. It has not been tested in pregnancy, so should be avoided

C1 inhibitor is given intravenously and usually requires several sessions of training before self-administration is mastered. Berinert and Cinryze have the advantage that, assuming an adequate dose is given, there is a period of protection from recurrent attacks lasting 24-48 hours. For this reason, Berinert or Cinryze are usually used in A&E and are preferred by people who have very frequent attacks (more than once per week).

The dosing regimen is different: Berinert is usually given in a slightly higher dose, which is adjusted for weight, compared with Cinryze which is a fixed dose. Berinert is also available in a high concentration, low volume form. This may be an advantage for bigger people, although the dose of Cinryze can also be adjusted if necessary.

Cinryze has also been shown to be effective for long term prophylaxis and if this is required, your specialist will usually prescribe this, although Berinert is sometimes also used for prophylaxis.

Ruconest is a C1 inhibitor purified from the milk of genetically modified rabbits. It comes in a much higher dose and some people find that it may work more quickly. The dose and the lower number of vials make this a popular option for larger people. Since, unlike Cinryze and Berinert, it is not purified from human blood plasma, it is the C1 inhibitor of choice for those who wish to avoid blood products. Ruconest has a shorter duration of action than other C1 inhibitors so is not usually used for prophylaxis and is less popular with people with extremely frequent attacks.

In summary, each treatment has its benefits. The choice of treatment is a very personal decision which should be based on discussions between the patient and specialist, taking into account the frequency and severity of the HAE, and social, work and family factors that may influence matters. Many people are trained to use both icatibant and C1 inhibitor and will choose the most appropriate treatment for each attack. However, most people end up preferring one or the other and use either icatibant or a C1 inhibitor for the majority of their attacks.

Do you have a question you'd like answered for The Quarterly Question? If so, please submit it via e-mail to: [support@haeuk.org](mailto:support@haeuk.org)

*We wish you all a very  
Merry Christmas and health  
and happiness for 2015.*

*With our best wishes from  
Laura & Rachel  
and Ann, John, Ann & Ed (Trustees)*