

## ***Rose's story***

### **Living with HAE**

---

I was five years old when I first started to experience stomach upsets, they would come on very suddenly and without any warning. They would start with excruciating pain in my stomach and then progress to vomiting. Once I had vomited the pain would ease, but not go away. This cycle lasted for 24 hours, nothing would stay in my stomach, not even sips of water and I would be bed-bound during this period. After 24 hours the vomiting and pain would subside leaving me completely weak. I would gradually build up my strength again over the next couple of days. These attacks would happen about every two weeks, meaning that I lost a lot of time at School.



My parents took me to visit the doctor on numerous occasions and many explanations were given, possibly worms, a cold in my stomach, I even had a barium meal examination at the hospital, but nothing conclusive was found.

Miraculously and without any explanation when I was about 10 years old, these stomach upsets suddenly stopped! Everybody thought that it must have just been a childhood illness that I had now grown out of, how wonderful was that.

Unfortunately at the age of 13 the stomach attacks returned with a vengeance, but this time they were accompanied by swellings. I would go to bed perfectly fine and awake in the morning with a completely swollen hand, so swollen that it was rigid and painful and I was unable to move it. On occasions this would be both, but it could also be my ankles, feet and private parts. I would feel very lethargic and within a couple of hours a stomach attack would occur. The swellings would take around a week for my limbs to return to their normal state. Once again this resulted in lots of time off school. It was around this time that my Mother remembered that she'd had a swelling in her arm and hand whilst being pregnant with my eldest brother Michael, but she'd had nothing since.

During this time I continued to visit my doctor and when I was 18 and still suffering every other week I was referred to a dermatology specialist at my local hospital West Middlesex in Isleworth, my consultant was Dr Cream a lovely old gentleman. I was still a complete mystery to all of the doctors. One day Dr Cream had a junior doctor present in his clinic and he had an idea what could be wrong with me. He arranged for me to be tested for Hereditary Angioedema (C1 inhibitor deficiency). The results showed that his theory was correct and I was immediately put onto Danazol 200mg per day. My family were also tested; my mother had the illness, but neither of my siblings.

My life was completely transformed, from that day forward I never had any more upset stomachs or swellings. I continued to visit Dr Cream at the dermatology department who did regular blood tests and Liver scans.

Although Danazol was a miracle drug it came with its own side effects and I suffered horrendous acne

for which Dr Cream gave me all kinds of lotions and pills. My breasts seemed to shrink in size, my periods completely stopped, I started to get stubbly facial hair, redness to my face and my voice became husky! I also suffered from random headaches. That being said my life was 100% better and I was able to live a much more normal life and hold down a regular job with minimal sickness.

I continued with Danazol, until my late 20's when I wanted to try for a baby. I was advised that I needed to stop taking Danazol. And was given C1 inhibitor to keep at home in my fridge. The local A&E department were advised that I would be coming in for them to administer it to me. The attacks soon restarted and I was at A&E a couple of times a month. Sadly my attempts to get pregnant didn't work out and I returned to my full dosage of Danazol this is where I remained for the next 10 years with no attacks.

About 15 years ago I started to see another dermatology consultant at Guys Hospital London, who suggested that I tried to cut down my Danazol as far as possible. I managed to reduce the dosage by half to 100mg per day, but going lower than that meant, the attacks started to break through, so I stuck with the 100mg per day and was totally fine.

It was during this time that I attended Hammersmith Hospital London on a regular basis for junior doctor patient sessions. A team of junior doctors would quiz me about the symptom of my illness and then try to work out what was wrong with me. Amazingly that in most groups one of the doctors would be able to work out the problem.

In 2012 I attended my first HAE patient day in Manchester and was astonished to discover how far treatment for HAE patients had moved on. I found it to be quite an emotional experience, as before this meeting the only person that had any idea of how I had felt living with HAE was my late Mother, who had passed away back in 2001, since then I had felt completely alone with this illness and now I was in a room full of like-minded people who understood completely.

After this meeting I asked my current consultant to refer me to Dr Longhurst at the Royal London Hospital. Dr Longhurst offered me home therapies to self-inject with Berinert into my veins or Icatibant into my stomach as an alternative. Within a couple of months I had been trained, along with my husband Leyon, to do this and, with her support and a stress-free lifestyle, have managed to reduce the Danazol to 100mg twice a week, with very minimal breakthrough attacks. We did at one point reduce to 100mg per week, but found that the attacks were too frequent. On my lower dosage of Danazol the side effects that I currently suffer are minimal. My voice, periods and breast size have not returned to normal, I think that is due to the length of time that I have been taking the drug and it is too late, but the headaches, redness of the face and acne are greatly improved.

Last year I embarked on training and completed a 62 mile bike ride to raise money for HAEUK to give something back to this worthy charity with a group of friends and work colleagues.

So, here I am, aged nearly 52 and with the tremendous support of HAEUK, ingenious thinking of the junior doctor (sadly his name escapes me) and currently Dr Longhurst, and the dual care of my local consultant Dr Keohane, I feel that I am completely in control of my illness and fully enjoying all that life has to offer with my wonderful husband, family and friends.