Family’s brave battle against rare disorder

by Max Wieland

A Tameside family struggling from a rare and mostly unheard of illness have spoken of their brave struggle to live ‘as normal a life as possible’. Now their hope is that a worldwide awareness day will help draw attention to the life-threatening condition they have faced for nearly 20 years.

The Harrison family from Droylsden spoke to the Reporter this week about the struggle and suffering of living with hereditary angioedema (HAE). HAE, a rare genetic disorder characterised by severe painful swelling, has now plagued Jen Harrison’s family for just over 18 years.

The condition is often mistaken for an allergic reaction, resulting in improper medications being prescribed, leading to Jen, 36, explaining that greater awareness is essential. She told us: “I had never heard of it until my husband was diagnosed. “Before that he had been misdiagnosed for many years as it is so rare, most doctors haven’t even heard of it. “There are many people who are suffering from this disease and don’t even know that they have it. “To doctors it might seem like an allergic reaction, but if we don’t raise awareness then the mortality rate is only going to rise.”

The illness is a rare and potentially life-threatening inherited disorder that occurs in approximately one in 10,000 to one in 50,000 people worldwide. HAE patients often experience unpredictable and potentially life-threatening attacks of inflammation affecting the throat, abdomen and face, to name but a few, often lasting from one to three days.

Jen, who is a mother to four, three of whom suffer from HAE like their father Phil, 37, explained that the illness is life-consuming. “I encourage my children to live as normal a life as possible,” she said. “As long as they are aware, or who they are with are aware of the symptoms so that they can call for help in an emergency, then their day to day activities wouldn’t have to be so limited. “It’s down to me to keep them calm because anything can cause an attack, lack of sleep, change of weather, vibrations, excitement, writing, stress, knocks – literally anything. “It’s important to be aware of any signs and symptoms and always be prepared for an attack.”

Continued on Page 3

Hope: From left Sienna and Keira with their mum Jen.

TREATMENT: Sienna in hospital.

Honours for Tameside College Adult Learners

Whit a Friday
● pages 28, 29

next week

Scouts tie the knot
● see page 5
Rare disorder can be a living HAE-T

One member of a family being afflicted with hereditary angioedema (HAE) is bad enough but unimaginably, three of Jen Harrison’s children suffer from the illness too.

Jen’s eldest daughter Danielle, 17, is currently sitting her A Levels at Droylsden Academy Sixth Form College and needs to minimise stress levels to reduce triggers of an attack.

The college student suffered her first outbreak at the age of seven when her hands swelled, forcing her to become dependent on others until she was able to attend school a few months later.

The similarity in both Danielle and her father’s condition prompted the family to investigate into a possible shared allergy.

Danielle has since discovered that stress and bodily injuries are the main triggers for attacks.

The 17-year-old has also suffered attacks that have caused temporary loss of vision and abdominal attacks which have resulted in her being admitted to hospital.

She is now also under the care of the Adult Immunology service at Manchester Royal Infirmary like her father and also receives C1 infusions at home when needed.

“The advice we were lucky that they have funding for C1 Inhibitor (which costs £500 per vial) as not all people can have access to treatment,” said Jen.

“They are also lucky that they have a brilliant consultant and specialist nurse at MRI, who definitely shares a great deal of knowledge of HAE.

“He (Dr Mathew Helbert) is very supportive and at Danielle’s last appointment he was particularly concerned about her A-levels.

“He spoke to her about the importance of relieving stress and the different techniques she can use to minimise the chances of an attack.”

Jen’s other two daughters suffering with HAE are Sienna, seven, and Shannon, 15.

Both get swellings that can affect their airways and also suffer from severe abdominal attacks, which led to Sienna admitted to hospital for treatment.

Unfortunately, Jen is not able to administer Shannon’s C1 Inhibitor at home and so the threat of the swellings is always a danger.

“We need authorisation for home therapy for Shannon,” she said.

He alone would improve her quality of life but unless she gets her referral to Dr Helbert then Shannon will continue to suffer.

“Shannon has been diagnosed with depression as a result of HAE and now has regular sessions with a psychologist Dr Joy Harris at Tameside Hospital to help her come to terms with the condition.

“Dr Harris has become Shannon’s strength; her main focus is making sure Shannon is getting enough support at school and at home to keep her stress levels down while she sits her GCSE exams.

“I really can’t thank Dr Joy Harris enough for her support.”

Sienna was diagnosed with HAE when she was five-years-old, often suffering with weekly facial swellings which can last up to five days.

“This concerned me as I knew that swelling of the face can progress to the throat which can cause death by suffocation,” said Jen.

But if that wasn’t tough enough for both Sienna and Shannon, who attend Greenon Primary School and Droylsden Academy respectively, then their wish for a normal life.

“The toughest part for them is coming to terms with HAE and the fact that they could suffer a throat swelling at any time,” said Jen.

“They do in fact live in fear of their lives, they don’t discuss it with people as they don’t want to be viewed differently; they just want to live a normal life.

“And the one thing I always say to them is that the more research that is carried out then the more treatments that will become available.

“They obviously worry about the future but by then they will hopefully know a lot more and up to live around a cure.”

For more information on HAE, you can visit the www.haeuk.org.

INSPIRATION: Caring Jen encourages her daughters to live as normal a life as possible

PAINFUL: Victims of HAE suffer painful swelling of various parts of the body

Now classmate is diagnosed with same condition

With HAE affecting one in 50,000 people worldwide, health officials were stunned to find out that two Droylsden classmates were suffering from the same life-threatening condition!

Sienna Harrison, seven, has been suffering from the illness after inheriting the condition from her father, but it wasn’t until a chance encounter on Facebook that the parents of Ruby Hesford, also seven, discovered she was suffering from HAE as well.

After showing the common swelling symptoms that comes with the condition since October, Ruby’s mother Faye posted a photo of yet another hand swelling on Facebook in March, in the hope of discovering if any of her online friends had come across anything similar.

After one of her friends alerted Sienna’s mum Jen to the picture, who also has two other children and a husband who suffers from HAE – Ruby’s family finally started to have some answers!

Faye, who also has two other children who don’t suffer from the illness, said: “Jen asked me seven questions and said if I could answer yes to three of them then Ruby would definitely be suffering from HAE.

“Scarily, the answer to all seven questions was yes!”

“The hospital said it was too rare for two children in the same class to be suffering from HAE, so it has been a battle to finally have Ruby diagnosed,” she added.

After being diagnosed on April 4, the family finally have an answer to the symptoms that had previously been identified as an allergic reaction.

“It is hard to describe any emotion when we found out the diagnosis,” said Faye.

“We were just relieved to know what Ruby is dealing with.”

Since the diagnosis, Ruby has benefitted from a care package put in place by the Royal Manchester Children’s hospital, leading Faye to explain that increasing awareness of the condition is important.

The difference after diagnosis, when a C1 Inhibitor infusion is available is an insight as the difference within 20 minutes is astonishing.

“For a long while we didn’t have a clue what Ruby was dealing with and I am quite certain there are a lot of kids walking around undiagnosed.

“One thing which is certain is that Ruby would not have been diagnosed if it wasn’t for our chance encounter with Jen.”

With Ruby and Sienna now becoming even closer friends after discovering they share the same illness, Faye would like to encourage anyone with similar symptoms to raise the possibility of HAE with their GP.

“I really don’t think it is as rare as the statistics say,” she added.

Continued from page one

A child has a 50 per cent chance of inheriting HAE if a parent possesses the genetic defect.

However, 25 per cent of HAE cases are due to a spontaneous genetic mutation, meaning the absence of family history does not exclude the diagnosis of HAE.

Phil has suffered from the condition throughout his life, beginning when he was young following a head injury, which led to swelling on his face and head.

He was told that the swellings were a result of allergic reactions, meaning it was a long time before the correct diagnosis was made.

“Phil has suffered from it since he was a child,” said Jen.

“When he was nine he had an accident on his bike and he remembers swelling to a point where he was unrecognisable.

“Then there was a time early on in our relationship where both his hands swelled up after bricklaying one day.

“He explained that it hadn’t happened for a couple of years and until his diagnosis, we had no idea of what it was or how to treat it.”

Jen’s husband has also more recently suffered a throat swelling which needed an emergency treatment of C1 Inhibitor Infusion.

The doctors on duty were not familiar with the treatment and Jen was forced to contact her husband’s consultant who, over the phone, guided a doctor through the process of the C1 inhibitor administration.

A C1 Inhibitor infusion is the only way of halting the swelling as quickly as possible, and in Phil’s case at the time – life-saving.

It was at this time that Immunology Specialist Dr Mathew Helbert encouraged Jen to begin home therapy training, in order for her to be able to control the progression of the attacks.

Upon learning of his correct diagnosis, Jen explained: “It was an extremely traumatic experience, but thankfully I received plenty of support and guidance from the Immunology Specialist Nurse Alex Farragher, she encouraged me to start home therapy training using the C1 inhibitor.

“It was terrifying when I found out because I was told that I had to do as much research into HAE as possible because not that much is known about it.

“Taking all that on board was a lot to deal with but it made me determined to do whatever I could to help.”

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