

# Sophie and her swollen hand

A Booklet for Children with Hereditary Angioedema (HAE)



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# Foreword

This book has been written to help introduce younger children to hereditary angioedema (HAE). It is primarily aimed at children who have HAE where the child has been recently diagnosed. It can also be used to help explain the condition to siblings, young relatives and friends of a child with HAE.

The story is centred on Sophie, a school-age child who experiences symptoms of HAE and is diagnosed by a specialist or hospital doctor based on the family history of HAE (via her father). The intention is to create a link between Sophie's HAE and the symptoms that she will be familiar with through her father's HAE. This should make the story easier for the child reader to understand, while also reducing the fear that they may have about their own diagnosis or their friend's condition.

When reading this book with a child, there are parts of the book that can be used to encourage the child to join in – for example, asking them to try to say the words “hereditary angioedema”, and using the pictures to describe the parts of the body that can be affected by an HAE attack. This type of interaction has been shown to improve understanding and learning.

We hope that you find this book useful and informative, and that you enjoy reading it with your child.

For further information please visit the South African HAE support group on: <https://southafrica.haei.org>

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Please note: the information contained in this book should not be used for diagnosis or treatment, and is not intended to be a substitute for consultation with a healthcare provider. Please consult your healthcare provider for further advice.

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# For parents/carers: About HAE

HAE is a rare inherited condition that is caused by a deficiency of a protein called C1-esterase inhibitor (C1 inhibitor). This protein has an important role in the inflammatory process. There is a 50% chance of a child inheriting the condition if one of their parents has HAE, although in around 25% of cases it can occur with no family history. HAE affects about 1 in 10,000 to 1 in 50,000 people worldwide.

During an attack, your child may have localised swellings (oedema). They can affect any part of the body, but most commonly the face, limbs, trunk, genitals and abdomen. Swellings develop over a period of hours and your child may have signs that an attack is about to happen, such as a rash, tiredness or local discomfort. An attack can last for 1–5 days before resolving spontaneously. Abdominal attacks can cause discomfort, pain, nausea (sickness), vomiting or diarrhoea.

If the swelling is on their shoulders or above (i.e. neck, face or throat) it may affect the airway and is potentially life-threatening. This requires prompt treatment. While this is less common than other attacks, it is important to make sure that your child knows to tell an adult immediately if they have any attacks on the face and neck, especially if they have any of the following symptoms.

Signs of an airway attack could include:

- Sore or itchy throat
- Feeling that something is stuck in the throat
- Voice changes (high-pitched, hoarse or rough)
- Barking cough
- Difficulty speaking
- Difficulty breathing

Although there is currently no cure for HAE, treatments are available for children. There are two different ways to manage or treat HAE attacks. The first is to treat the attack when it happens, normally by injection; this will usually be used for more severe or ‘acute’ attacks. The second is to try to prevent attacks by taking regular medication; this may be prescribed if someone is having attacks very frequently. Please speak to your hospital doctor or specialist to find out more about the most appropriate treatments for HAE.

## Meet Sophie.

Sophie is 6 years old, and she loves her dog Patches and her teddy bear Blinky.

She also loves her family – her mummy, her daddy and her younger brother Jack, even though he can be quite annoying sometimes.





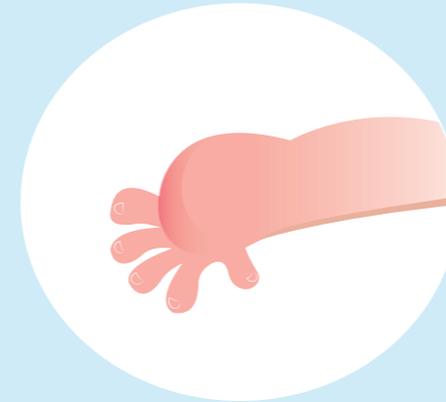
One day Sophie was drawing a picture of mummy and daddy at the beach. After a while she had to stop, because it was hard to hold the pencil. When she looked at her hand she saw that it was all puffy and big.

Sophie was worried so she showed mummy and daddy her hand, and told them what she had been doing.

“Oh dear, your hand is swollen,” said mummy.

“Don’t worry, I think I know what might be wrong,” said daddy.

“We should go and see Dr Brown so that she can make your hand better.”



## At the hospital...

“Hello Sophie,” said Dr Brown.  
“So what seems to be the problem?”

Sophie held up her swollen hand to show Dr Brown.

“Oh dear,” said Dr Brown. “Does it hurt?”

“It feels strange, but it doesn’t hurt too much,”  
said Sophie.

“Hmm. Don’t worry, I think I know what might  
be wrong,” said Dr Brown. “But we will need to do  
some special tests on your blood to make sure.”



“Hello Sophie, my name is Nurse Becky.”

“To help us find out what is wrong with you, I need  
to put a tiny needle in your arm and take out a few  
drops of your blood.”



Sophie looked worried, and held Blinky  
very tight.

“Don’t be scared,” said Nurse Becky.

“I sometimes find that if I close my eyes and  
take a deep breath then I don’t feel the needle.”

Nurse Becky gently rubbed Sophie’s arm  
with a cold wipe, and Sophie closed her eyes.  
Before she knew what was happening,  
Nurse Becky said “All done”.



“That wasn’t so bad,” said Sophie.

“I did feel it, but it didn’t hurt. It just felt a little bit funny.”

“You were a very brave girl Sophie,” said Nurse Becky, giving her a big sticker.

**BRAVE  
GIRL**

**A few days later...**



“How does your hand feel today Sophie?” asked Dr Brown.

“It’s much better thank you, it isn’t puffy anymore and doesn’t feel strange at all,” said Sophie.

“I’m very glad to hear that,” said Dr Brown.

“We have looked at your blood, and we know what the problem is.”

“You have something called hereditary angioedema. See if you can say it after me: her-ed-i-tary angi-o-edema.”

“Because they are tricky words, we normally call it HAE, which means the same thing, but is easier to say.”



“If you have HAE then most of the time you will be able to do all the things you normally do, like playing with your friends and going to school.”

“Every now and then, your hand might get swollen or you could get a swelling somewhere else on your body. This is called an HAE attack. This probably won’t happen very often, and most of the time it shouldn’t be too bad.”

“Here are some of the places where you might have an HAE attack.”



“HAE is something that you are born with. It is not something that you can catch from someone when they are sick, so your friends can’t catch it from you.”

“If a girl or a boy has HAE then their mummy or daddy might also have HAE. In your family Sophie, your daddy has it, but your mummy does not have it.”

“Can you remember a time when your daddy had an attack?”

“Um. I think so,” said Sophie. “There was one time when daddy’s foot got so big that he couldn’t put his shoe on.”



“Even though we can’t stop you having more HAE attacks, we can give you medicine to help make the swellings go away when they happen again.”

“Sometimes you might have an HAE attack that is more serious – it could be on your face or neck, or in your tummy. If that happens, tell a grown up straight away as you might need some medicine to stop the swelling getting worse. Sometimes the medicine will need to be given to you using a needle,” said Dr Brown.”

“Does that sound OK Sophie?” asked Dr Brown.

“Don’t worry, I’m not scared,” said Sophie. “Nurse Becky put a needle in my arm before, and it didn’t hurt really.”

“You really are a very brave girl,” said Nurse Becky.



“Everyone with HAE is different,” explained Dr Brown. “That means that some things might make your daddy have an HAE attack, but might not make you have one.”

“To help us know what things make you have an HAE attack, I would like you to start writing in a special HAE diary,” said Nurse Becky.





“When you feel you might be having an HAE attack, you should write down in your HAE diary what part of your body swelled up, how sore it was, and what you were doing before it started.”

“Your mummy and daddy can help you to write down how long it lasted, what medicine you had and how long it took before you felt better again.”

“And if you ever start to feel like you might be having an HAE attack, make sure you tell a grown up straight away. They can then call your mummy or daddy, who can make sure you feel better as quickly as possible.”



After her hand was back to normal, Sophie could do all the things she normally did, just like Dr Brown said.

She could draw pictures, play football, play with her friends, and play the recorder.

Just like normal.



## One day though...

Sophie fell over in the playground and hurt her foot, which made it swell up. Sophie wasn't scared though, as she knew what to do. Sophie told her teacher, who called Sophie's mummy.

When her mummy asked if it hurt, Sophie said that it hurt a little bit, but not too much. Mummy told Sophie that everyone can get a swelling if they bang their foot, but that sometimes, people with HAE get a bigger swelling. Mummy said that Sophie didn't need to go home or see a doctor, and said that she would see Sophie at home time.

When mummy picked her up, Sophie's foot wasn't hurting anymore. Mummy was very proud of Sophie and gave her a big hug.



## A few months later...

"Hello Sophie," said Dr Brown. "How have you been since we last saw you?"

"I have been very well thank you," said Sophie. "I have been doing all the things I normally do, just like you said."

"I did have one HAE attack. I told a teacher, who called my mummy, and I wasn't scared. And I wrote it all down in my diary just like Nurse Becky said."



"Well done Sophie," said Nurse Becky, "that is excellent. You wrote down when the HAE attack happened, what happened before it, how bad the swelling was, how much it hurt, what medicine you had, and how long it lasted."

"I wish all my patients were as good as you are."

“You were very good to tell your teacher so quickly about your foot,” said Dr Brown.

“Different things can make you have an HAE attack, like something you eat or drink, falling over or doing lots of drawing. Sometimes you might have an attack and we just don’t know why.”



“So, remember to let a grown up know straight away if...



...you have a sore tummy...



...you start to feel puffy in your hands,...

...feet or face...



...your throat feels funny when you swallow, you start to cough a lot or your voice sounds funny or if you find it a little bit hard to breathe.”



“They can then let your mummy or daddy know.”

“We would like to see you again in a few months, to see how you are getting on, and to take another look at your HAE diary.”

Sophie was having a lovely dinner with her family when Jack suddenly said “Are you OK daddy?”

When Sophie looked across the table, she saw that daddy’s face had started to swell up.

“Oh dear,” said daddy, feeling how puffy his face was.

“I think I know what is wrong,” said Sophie. “Don’t worry, daddy. I think you are having an HAE attack. You need to take your medicine and record it in your diary. And let Mummy know if it gets any worse.”





If you would like more information on the references referred to in this booklet, please contact [medinfoEMEA@takeda.com](mailto:medinfoEMEA@takeda.com)

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