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## Frequently Asked Questions

For parents/carers of children diagnosed with hereditary angioedema (HAE)

This booklet is for the parents and carers of children with HAE, and supports the '**Being Me with HAE**' materials your child has been given.

If your child has recently been diagnosed with HAE then you may have lots of questions. We have brought together answers to some of the common questions about caring for a young person with HAE and the role of the parent/carer.

Some of the questions refer to a child's 'parent with HAE'. Even if you are familiar with HAE, it is worth knowing that your child's experience of HAE might be different to other family members with the condition, and so we hope that this information will be useful to all those caring for a young person with HAE, regardless of their own experience.

If you have any specific questions or concerns about your child's HAE, you should always ask your immunology doctor or nurse. They will be happy to help you understand anything you are unsure about.



# Introduction to HAE

## What is HAE?

Hereditary angioedema (HAE) is a rare, inherited condition. Inherited means it can be passed from parent to child.

This may mean that you have the condition yourself, in which case you will know that HAE causes episodes of considerable swelling in various parts of the body. You will also know that it is a life-long condition, but treatment is available to help manage the symptoms.

## What symptoms will my child experience?

HAE can cause swelling in various parts of the body. These swellings, sometimes called 'acute attacks', or 'attacks', can be painful.

In HAE, swelling can occur almost anywhere, but attacks often affect the hands, legs, arms, feet or genitals, usually in the tissue just below the skin ('subcutaneous' tissue). Attacks in these areas of the body are called 'peripheral attacks' and they make up more than half of all attacks of HAE.

Other attacks can occur in the abdomen and cause pain in the stomach.

Attacks can also take place in the throat or face. If such an attack occurs it is very important that your child receives immediate medical attention.

See the section "**Dealing with attacks**" for further information.

*If your child starts to develop symptoms anywhere from their neck upwards – including a tingling feeling on their skin, a hoarse voice, a rash, trouble breathing, or swelling anywhere on their face or neck – you should call 999 or take them to the nearest hospital emergency department immediately.*

## How often will attacks happen?

Depending upon the individual and the severity of the disease, some people may have many swelling attacks each month, while others may go months or even years without one.

## What causes HAE?

HAE is a hereditary condition, which means that most (but not all) people with HAE inherit it from one of their parents. Although it is possible to be the first person in your family to have HAE, in most cases other family members have also had similar swelling attacks.

Swellings of HAE are caused by the poor functioning or lack of a protein called C1-esterase inhibitor (C1-INH). If there is not enough C1-INH in the body, or it is not working as it should, blood vessels can become leaky, allowing fluid to build up in the tissue and causing the swelling that is characteristic of HAE.

More information can be found at [www.AllAboutHAE.co.uk](http://www.AllAboutHAE.co.uk)

## Is HAE dangerous for my child?

The risk is determined by the frequency, location, and severity of swelling. The majority of attacks are not life-threatening. However, swelling in the throat may block the airway and these attacks **can** be life-threatening. It is important that you understand the treatment plan your doctor or nurse has provided for your child, and what you should do in an emergency. You may have heard this referred to as an emergency management plan.

Swelling in the intestines may cause severe, abdominal (stomach) pain and discomfort. These kind of attacks may also require medical attention if the pain is severe so that other reasons for this pain can be ruled out.

## Are there any warning signs that may help me predict when my child is going to have an attack?

Attacks are often unpredictable, but there are sometimes signs that can alert you to their onset. Over time, you and your child may learn to recognise the warning symptoms specific to their attacks.

Common symptoms are:

- sudden mood changes
- rash
- irritability
- aggressiveness
- anxiety
- extreme fatigue
- a tingling sensation of the skin where the swelling will begin

These symptoms are called prodrome or prodromal symptoms.

A hoarse voice or laryngitis, difficulty in swallowing, a feeling of tightness in the throat, and voice changes may be the first signs of a life-threatening laryngeal attack. It is important your child learns to watch out for these symptoms, and knows to alert you or another responsible adult if they feel them developing. If your child does complain of these symptoms, you should seek emergency help for them as soon as possible.

## Will my child's experience of HAE be the same as their parent with HAE?

It is important to know that each person will experience HAE differently. So the symptoms your child has, the treatment they are prescribed, or the impact HAE has on their life could all be very different from the experience of their parent with HAE. Your child's doctor and nurse will develop a care plan that is specific to your child, and this may also be very different from the care plan used by their parent with HAE to manage their condition.

# Managing HAE

## What do I need to know about my child's treatment for HAE?

Your immunologist is the first person you should ask about any aspect of your child's care. It is worth noting that the treatment your child is prescribed (if any) may be different from the treatment taken by their parent with HAE, so it is important to be aware of their specific needs. Your child should not be treated with the same treatment as their parent with HAE unless specifically instructed by their immunologist. If your child is prescribed a home treatment for their HAE, their immunologist will make sure you know how to help them take it properly if needed.

## What are the most important things to communicate to my child about their HAE?

Safety always comes first. The most important thing for your child to be aware of is that they must let you or another adult know if they develop symptoms anywhere from their neck upwards.

Any strange feelings in their head, neck or throat could be a sign of a laryngeal attack, which requires immediate medical attention.

It will therefore be important to help your child to:

- **Recognise the signs** of an attack developing
  - You can help them to become familiar with and begin to name their own prodromal symptoms (explained in the section 'Introduction to HAE')
- **Know their triggers** and how to avoid them where possible
  - This may include foods, stressful situations, or any medications found to trigger their attacks
- **Be prepared** in the event of an attack
  - This includes keeping details of their emergency management plan accessible at all times and knowing who they must tell if they get symptoms

The 'Being Me with HAE' booklets for children may help you explain HAE to your child, and the things they can do to help manage their condition.

*Know your child's emergency management plan and make sure you keep details of this with you at all times. This information should also be given to any adult supervising your child in your absence.*

## Will my child need prophylactic (preventative) treatment?

Every person with HAE has a different experience of the condition. Your child's immunologist will decide whether or not preventative treatment is necessary and what type of treatment this may be.

## Will I need to help my child take injections (in an emergency or otherwise)?

If your child's immunologist feels that home treatment is appropriate, this will be discussed with you at clinic appointments. If your child requires home treatment, your immunologist or specialist nurse will arrange the appropriate training.

It is important to remember that if your child has swelling anywhere from their neck upwards, you should dial 999 or take them to the nearest hospital emergency department as a matter of urgency, even if you have been able to treat them at home.

## Who needs to know that my child has HAE?

The golden rule is to make sure your child could always get emergency help if they were to have a swelling anywhere from their neck upwards. It is therefore recommended that you provide a letter from your child's doctor to anyone supervising your child, to help them understand their condition. This may include nursery staff, teachers, activity leaders and babysitters.

If your child is referred for any type of surgery, including any dental work (and especially extractions), it is important to tell both the surgeon/dentist and your immunologist. Any type of procedure may raise the risk of an attack of HAE, and your child's immunologist may suggest a preventative treatment.

Ask your child's immunologist about letters they can provide for the purpose of informing other healthcare professionals.

*If your child is anxious during visits to the dentist or doctor, this in itself may bring on an attack, so be mindful to look out for symptoms.*

## Is there anything I should do to help my child avoid attacks?

There is no reason your child's HAE should prevent them from doing anything their friends do. It may, however, help to be mindful that certain things can bring on an attack. These can be different for every person, but can include stress, hormones and bumps and bruises. It can be a good idea to look out for signs of an attack if any of these arise.

For some people, certain foods or medications may be triggers. Do not be afraid to let your child try new things, but try to look out for symptoms after giving them new foods or medicines so that you can act quickly if they begin to show signs of an attack.

## What advice should I give my child for when they are at school / out with friends / on school trips, etc.?

Your child should be aware of the importance of getting help right away if they feel the signs of an attack developing, especially anywhere from their neck upwards.

They should also know who they can speak to (ideally an adult who knows about their HAE and what to do in an emergency) if they begin to feel the symptoms of an attack. This is so that they can get help as soon as possible without having to explain their condition.

Make sure they have details of their emergency management plan with them, or ensure an adult in charge is provided with this information.

# My child's future with HAE

## What do I need to be aware of when taking my child on holiday?

You should always have details of your child's emergency management plan with you, but this can be especially important when travelling. If you are travelling to a country where you do not speak the language, it may be a good idea to find out the translation for hereditary angioedema, and write it down to keep with you. This will allow you to communicate your child's condition to medical staff in the case of an emergency. It is also a good idea to be aware of the location of the nearest hospital emergency department.

If your child has been prescribed a non-emergency treatment for their HAE, make sure you have enough treatment to last the duration of the trip.

If your child has a history of having serious attacks, speak to your immunologist about special situations such as long flights where it may be more difficult to get help in an emergency. They can advise you on the things you can do to minimise risk.

*If your child gets nervous in new situations, you may need to keep an even closer eye out for symptoms while travelling, as stress may trigger an attack.*

## How do I explain HAE to my child's brothers/sisters without HAE? / What should I explain to them about their sibling's condition?

It is up to you to decide how much you feel your child(ren) without HAE will be comfortable knowing about and understanding HAE. However, it is worth keeping in mind that children may be more frightened by what they do not understand.

They might benefit from being aware of what needs to happen in an emergency. This will help prepare them, and may reduce feelings of fear and anxiety. Brothers and sisters may even be able to help spot the early signs of an attack.

It is also important that your child with HAE has as little reason as possible to feel they are different to siblings and peers. It may be a good idea to talk to your children together about the condition. You might find that the 'Being Me with HAE' booklets can act as useful prompts for these types of conversations, to help you talk to your children about the different aspects of living with HAE.

## Where can I find out about support groups for families affected by HAE?

HAE UK is an association of HAE patients and families, working together to improve the situation for all HAE patients in the UK. Further information can be found at [www.HAEUK.org](http://www.HAEUK.org). You can also visit [www.facesofhae.co.uk](http://www.facesofhae.co.uk) to read the stories of real people living with HAE around the UK.

## Will my child's HAE get better or worse as they get older?

Each individual with HAE experiences the condition differently, so it is impossible to know how your child's HAE will progress. You should always be alert to any changes in their condition and keep your immunology doctor and nurse informed. With the right support, children can learn to manage the condition effectively. Whatever course your child's HAE takes, it will be important for you to encourage and enable them to learn to manage their own condition as they get older. This will help them develop a sense of confidence and independence in living the life they choose.

## Will my child have the same prodromal symptoms and triggers as their parent with HAE?

Your child's symptoms, attacks and treatment may not be the same as those of their parent with HAE. It will be important for you to help them look out for their own prodromal symptoms and triggers, which may be very different to the ones with which you are familiar.

*HAE is different for everyone. Symptoms and triggers may be completely different among members of the same family with HAE.*

## Will my child's HAE take the same course as their parent with HAE's condition as they get older?

It is impossible to predict the course that an individual's HAE will take. Each person experiences HAE differently. Be sure to keep an eye out for changes in their condition which their parent with HAE may not have experienced.

## Will my child be suitable for the same treatment as their parent with HAE?

Not everyone with HAE needs to take a treatment regularly, or even at all. A treatment taken by your child's parent with HAE is not necessarily suitable for your child.

Any decisions about treatment will be made by you and your child's immunology team. Treatments will be tailored specifically to meet your child's individual needs.

# Dealing with attacks

## How do I tell which attacks need treating and which do not?

Always follow the advice provided by your immunologist in the first instance. In general:

**For attacks from the neck upwards you should immediately take your child to the nearest hospital emergency department or call an ambulance (whichever will be quicker).**

If your child has been started on a home treatment, you can administer this in the meantime if the person who is trained is available to give them the treatment.

If your child has extreme pain in their abdomen (stomach area) you should also take them to hospital so that they can be checked to make sure there is no other cause of the pain.

### For all other attacks from the shoulders down:

- If your child has been started on a home treatment for their attacks, your immunologist will likely have recommended that **all** attacks are treated. They may also have asked you to notify them each time your child has an attack.
- If your child has not been prescribed a treatment for their attacks, you should tell your immunology team that your child has had an attack. Do not wait for their next appointment.

*A young child may have difficulty conveying feelings of discomfort, pain and other medical symptoms. If you have **any reason** to suspect that your child is having symptoms of an attack anywhere from their neck upwards, you should seek emergency medical advice immediately.*

## What do I need to tell emergency medical staff if my child has a serious attack?

### When you go to the hospital for treatment of a serious attack (swelling anywhere from the neck upwards):

- Make sure the emergency medical staff understand that your child has HAE.
- Make it clear to the medical staff on call that it is an emergency and provide them with details of your child's emergency management plan. This will provide them with all the information needed to treat your child for a serious attack.

*If you are not satisfied that the treatment your child is given by emergency medical staff follows your child's emergency management plan, be firm and insist that they speak to an immunologist on call.*

### For visits to the hospital for severe abdominal pain:

- In this scenario, you will also need to make it clear to the medical team that your child has HAE. This will help them to understand that this is a potential cause of their pain which needs to be investigated.

## What are the signs of a serious attack/at what point should I call 999?

If you have any reason to believe your child is developing a serious attack (an attack anywhere from their neck upwards) you should take them to the nearest hospital emergency department or call 999 right away.

For example, if your child complains of tingling or any unusual sensation anywhere from their neck upwards, or develops a rash anywhere on their neck or face, this could be a sign of a serious attack.

The symptoms listed are not the only ones that could signal a serious attack. If there are other symptoms your child usually gets before a serious attack, or that you suspect may be a sign of a serious attack developing for any other reason, then you should also treat these as an emergency.

*If your child is experiencing symptoms that you suspect signal a serious attack, you should treat this as an emergency and seek treatment.*

## What should I do to take care of my child following an attack?

Depending on your child's experience of HAE, it may take a while for the swelling of an attack to go down. While they are waiting for their symptoms to subside, your child should be encouraged to drink plenty of water. They should try to rest and avoid any activity that may aggravate the swelling.

If they are in pain, you can provide them with painkillers, if needed, at the dose recommended on the packaging, or as otherwise discussed with your immunologist.

After the swelling has gone down, your child should continue to rest and drink plenty of fluids. They can also be given further painkillers, in line with the recommended dose, as required.

*While your child is resting, it may be a good opportunity to record the details of their attack with them. Ask your immunologist if they can provide a symptoms tracker for this.*

# Visits to your child's immunology clinic

## How often will I need to make appointments for my child to see their immunologist?

How often your child will need to see their immunology team will depend on the frequency and severity of their attacks. Your immunologist will tell you how often you should see them for routine appointments.

If your child has an attack between appointments, no matter how mild, it is important to let your immunology team know right away. They may ask to see your child before their next scheduled appointment.

*Remember to let your child's immunologist know about each attack right after it happens, unless you have been advised otherwise.*

## What can I expect from my child's immunology team?

There are many aspects of your child's care that their immunology team will consider. If there is anything about your child's HAE you are unsure of, they will be able to provide you with more information that is specific to your child.

Some of the things your child's immunology team will generally take responsibility for are listed below:

- Maintaining a detailed history of your child's HAE attacks, including:
  - Frequency and timing of attacks, and for each individual attack:
    - Trigger
    - Location on body
    - Duration
    - Treatment administered (if applicable)
- Staying informed and mindful of any other health conditions that affect your child
- Advising on whether treatment is needed for your child's attacks of HAE, and prescribing this treatment for them
- Advising on pain medication that you can give your child, including dose, if needed, during or following attacks
- Providing an emergency management plan, details of which should be shown to the hospital emergency department (ED) in the case of a serious attack. This may be provided directly to the ED of the hospital, and/or you may be given details to hand over, in case you visit a different ED, for example.

**If you have any further questions that are not answered in this booklet, or if anything is still unclear, your immunology team will be happy to help you.**

You can also find more information about HAE in general at:

[www.AllAboutHAE.co.uk](http://www.AllAboutHAE.co.uk)

## What can I do to help my child's immunology team and other medical professionals involved in their care?

Having comprehensive information about your child will help the medical team caring for your child to provide the most appropriate care. You can help by taking responsibility for the following things:

- Keeping details of your child's emergency management plan with you at all times, and providing a copy of this information to your child/an adult in charge if you will not be with them
- Notifying your GP, dentist, your child's school and any other places they regularly attend about their condition (your immunologist may be able to provide you with documents for this purpose, e.g. letters)
- Making your immunologist aware of any attacks of HAE that your child experiences as soon as they happen, unless you have been advised otherwise
- Informing your immunologist about any changes in medication or any planned surgeries or dental procedures that your child will be undergoing, as they may need to adjust or provide medication as a precaution
- Keeping a detailed record of your child's HAE attacks to be passed on to your immunologist:
  - Frequency and timing of attacks, and for each individual attack:
    - Trigger
    - Location on body
    - Duration
    - Treatment administered (if applicable)
- Knowing your child's prodromal symptoms (early HAE attack warning signs), and seeking emergency medical attention for serious attacks
  - For younger children, this includes being aware of symptoms they may not be able to describe

*Ask your immunologist about a symptoms tracker to help you keep note of the details of your child's attacks.*

## Reporting of side effects

If you get any side effects, talk to your doctor, pharmacist or nurse. This includes any possible side effects not listed in the package leaflet. You can also report side effects directly via the Yellow Card Scheme at:

[www.mhra.gov.uk/yellowcard](http://www.mhra.gov.uk/yellowcard)

By reporting side effects, you can help provide more information on the safety of this medicine.

Provided as a service to medicine by CSL Behring UK Ltd.

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