Hereditary Angioedema (HAE)
A patient’s guide

This patient guide is only to be shared with patients diagnosed with HAE

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What is Hereditary Angioedema (HAE)?

Hereditary Angioedema (HAE) is a rare genetic disorder which is estimated to affect between 1 in 10,000 and 1 in 50,000 people worldwide. HAE is marked by recurrent episodes of swelling (oedema) of the skin in different parts of the body, as well as in the airways and internal organs.

- **Hereditary** means that it can be passed on from one generation to the next in your genes
- **Angio** means it is related to blood vessels
- **Oedema** means excess watery fluid collects in the body’s tissues (causing the swelling)

HAE runs in the family

- A family history is found in 75% of cases
- According to international guidelines, family members of people with HAE should be tested, including grandparents, parents, siblings, children, and grandchildren
  - Encourage them to be tested, especially if they have experienced unexplained swelling or abdominal pain – a simple blood test is all that is required
- Speak to your doctor/nurse specialist to find out more

Images courtesy of Professor M Bas, Klinikum Rechts der Isar der Technischen Universität München, Germany.
Common symptoms

Some common symptoms are listed here. But HAE can present differently between patients and each attack can vary even within the same person.

IMPORTANT

Treat an HAE attack as soon as you can with your emergency medicine.
What can trigger an HAE attack?

- Physical and psychological stress
  - Exams
  - Weddings
  - Other emotional events (positive and negative)

- Dental procedures

- Illness and physical injury
  - Injury
  - Medical and surgical procedures
  - Illness
  - Physical injury

- Hormonal changes
  - Puberty
  - Contraceptives containing oestrogens
  - Hormone replacement therapy
  - Menstrual cycle
  - Pregnancy
  - Changes in estrogen levels

- ACE inhibitors (a type of medicine used to treat blood pressure or heart conditions)
Emotional triggers

- Emotional stress, both positive and negative in nature, can be a key trigger for HAE attacks

- During stressful times in your life you may require a change to your treatment. You should discuss this with your doctor/nurse specialist
Are there any signs an attack may occur?

- Most patients are able to predict that an attack will occur based on symptoms that come before the swelling (these are known as prodromal symptoms)
- These early symptoms can last 0–48 hours, and include:
  - Excessive tiredness
  - Feeling sick
  - Aching
  - Rash
  - Tingling
  - Feeling anxious
  - Mood changes
Swelling of the airway

- Swelling of the upper airways can be fatal, as the swelling may obstruct the airways and lead to asphyxiation (choking)
- Patients experiencing this type of oedema (swelling) must seek immediate emergency medical assistance
- You must take your emergency treatment immediately

Signs that you might be about to have an HAE attack of the throat, nose or tongue include:
- Difficulties swallowing
- Tightness in the throat
- Hoarseness
- In extreme cases, asphyxiation (choking)
- Changes in the voice
What to do during an attack

- Ensure you carry your emergency medication with you
- Carry a medical ID/A&E card explaining what HAE is
- Have a letter from your doctor/nurse specialist for emergencies and keep it with your medication
- Ensure family, friends and colleagues know what to do

Only you will know how comfortable you feel being seen by family members during an attack:
- Because attacks can affect your appearance, it is normal to feel self-conscious sometimes
- If you feel embarrassed, remember that HAE is a medical condition, and that you are not a burden to those around you. It’s something you cannot control, and you should not feel ashamed by it

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What to say in A&E

- I have hereditary angioedema (HAE)
- I may be having an HAE attack
- I need urgent attention
- I am being treated with

• My doctor/nurse specialist is

IMPORTANT
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Planning in advance with pre-procedural treatment to prevent HAE attacks

Contact your doctor/nurse specialist if you are planning:

• Dental treatment
• Surgery
• Any invasive treatment

Travelling with HAE

To help your journey go as smoothly as possible, you may want to consider the following **before** you travel:

• Check that your travel medical insurance covers existing conditions:
  - Explain to your travel insurance company that HAE attacks can be unpredictable and life-threatening, especially in the case of attacks involving swelling of the throat
• Ensure that you can carry your medication with you when you travel
  - Check carry-on luggage regulations and medication storage conditions with individual airlines before travelling
• Confirm you will have proper medication storage while travelling and where you are staying (you may need a container for disposal of sharps)
• Contact your doctor/nurse specialist ahead of time and ask that they write you a letter addressed (in the same name as your passport) to the airline and customs and immigration personnel stating that:
  - Out of medical necessity, you must carry your HAE supplies (including the syringe and needle) with you at all times
• Make a list of all the prescription items you need to take with you; all of which you should obtain from your pharmacist before leaving
Let’s talk about HAE

Communication is key

• Because HAE is a rare condition, it’s likely that your friends, family or partner won’t know much about it. Make those in your social circle aware of how it affects you to help manage their expectations and keep channels of discussion open.

• There’s more to you than HAE – but when everyone is informed, the unpredictability of HAE will feel more manageable.

• In the event of an emergency, make sure your partner, family and key friends have your doctor/nurse specialist’s information and that they’re aware of what to say to medical personnel.

With family

• It is a good idea to reassure children that you have medication that will help reduce swelling, and that it isn’t permanent. Children tend to worry much less when they know what to expect.

• Don’t be afraid to ask any family members with HAE to share their experiences and get their advice on coping with the condition.

With your partner

Making your partner aware of how HAE affects you, so that they know what to expect if an attack does occur, may help you both enjoy a happier, more stable relationship.

• Consider telling your partner about HAE early on in the relationship, especially if becoming more physically or sexually intimate. That way your partner will be better prepared if an attack happens.

• Explain to your partner what to expect during an attack, and that you can self-treat it to prevent a mild attack becoming more serious.

• You may not feel comfortable with your partner seeing you during an HAE attack, especially if it is early in your relationship. Reassure him/her that the attack should pass in time.
Feelings of embarrassment are normal and common for a person with HAE. Talk through your feelings with your partner.

If you prefer to be left alone during an HAE attack, let your partner know.

As with any physical interaction, urogenital attacks can result from sexual intercourse. You may find it useful to explain to your partner that an HAE attack may increase the size of your sexual organs and intercourse can be painful for both men and women during an HAE attack. If this happens, reassure your partner that the swelling will reduce following treatment.

With friends and acquaintances

Because you can't control when attacks happen, you are likely to have at least one in front of a friend or acquaintance at some point. Don't be embarrassed if an attack disrupts plans or affects how you look — more likely than not your friends will be more concerned for your well-being. Let them know how they can help!

You are not obliged to share anything with anyone if you don't feel comfortable.
Adolescent patients

Tell your school about your HAE – by letting them know how it might affect you, they will be more prepared to offer you the flexibility you may need.

• Ensure you have treatment available and adults who know what to do
• Stress can make HAE worse
• Talk to your nurse and plan ahead for stressful times
• Your HAE may affect you differently to other family members with HAE
• It is useful to keep a diary to understand what is normal for you

Studying with HAE

• An attack might mean taking time off school or university or even studying from home
• If you occasionally miss a class or lecture due to an attack, ask your teacher or lecturer if you can have copies of that day’s notes to help you stay on track with your studies
  - Alternatively, ask a classmate to share their notes, or take notes for you
  - Ask for more time if assignments are delayed due to an attack
• Discuss options with your teacher or lecturer
  - Some institutions have facilities to record classes and lectures or dial in to a live stream video
• The possibility of attacks may mean being selective about playing contact sport – you may wish to discuss available options with a teacher or lecturer
• If driving or taking public transport is difficult after an attack, ask a family member or a classmate to help get you where you need to go

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Managing HAE at work

While you don’t have to mention it in a job interview, letting your employer and co-workers know what HAE is and how it can affect you on a daily basis will help you to manage their expectations better.

• You may find it beneficial to remind your employer that having a chronic medical condition does not affect your enthusiasm or commitment to the job. What’s more, you will continue to complete your responsibilities to the best of your ability

• Make it clear to him or her that valuable support, sensitivity and understanding from colleagues about your condition will give you the confidence you need to perform your job well

• Work with your employer to come up with a contingency plan should an attack happen at work
  - This might mean choosing to work flexible hours or arranging temporary cover if the situation arises
  - You can agree on the tasks you are best suited to, or you can adapt during an attack
  - When you have an attack, you may need to go home and rest, and even take some time off work
Where to find more information

Living in the UK:
• HAE UK is a Patient Association focusing on the needs of people with HAE and acquired angioedema (AAE) and their families
  - HAE UK is affiliated to the International HAE Patient Association
• If you or someone in your family has hereditary angioedema, HAE UK can help with:
  - Information about diagnosis and symptoms
  - Signposting to specialist HAE centres
  - Information about medications and individual HAE management strategies
  - One-to-one support and advocacy

Contact details for HAE UK:
E-mail: support@haeuk.org
Telephone: 07975 611787
Website: www.haeuk.org

Living in Ireland:
The Health Service Executive website is an independent source of unbiased and relevant advice for people living with HAE.

Contact details:
www.hse.ie/eng/health/az/a/angioedema
With HAE you can still lead a normal life

With a proper understanding of the condition and its treatment options, people with HAE and their families may be able to manage their symptoms and lead normal and active lives.

Sharing details about your HAE is extremely personal, and as you learn to manage your condition, it may be something that brings you and those important to you even closer. Communicating your needs to those you love — and who love you — is essential.

In addition to support from family and friends, connecting with other HAE patients through a patient support group can be extremely valuable. Sharing experiences and feelings with people who know what it’s like to live with HAE can be both comforting and empowering.

REMEMBER
Your Healthcare Team are there to provide information, advice and support to help minimise the risks and burdens of HAE.

YOU ARE NOT ALONE!