

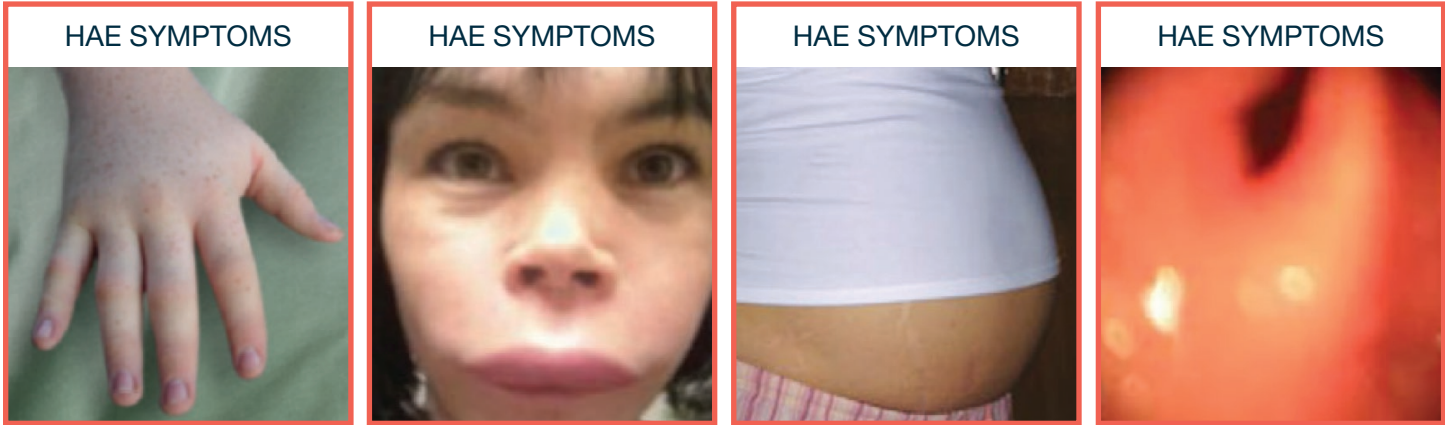
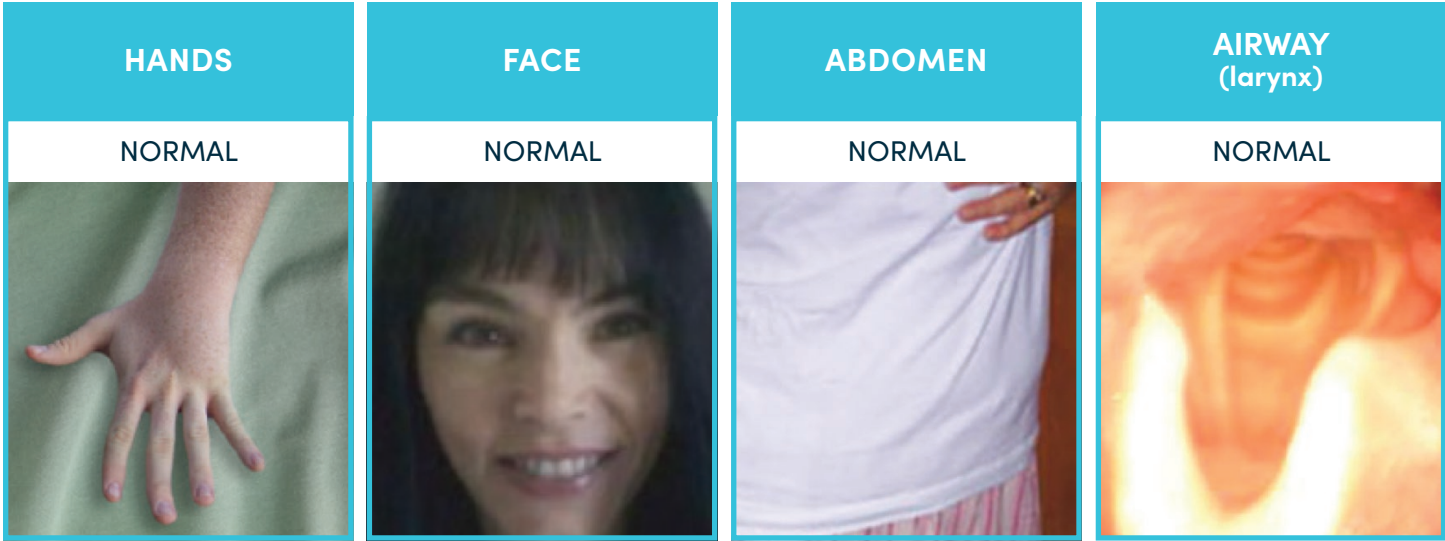
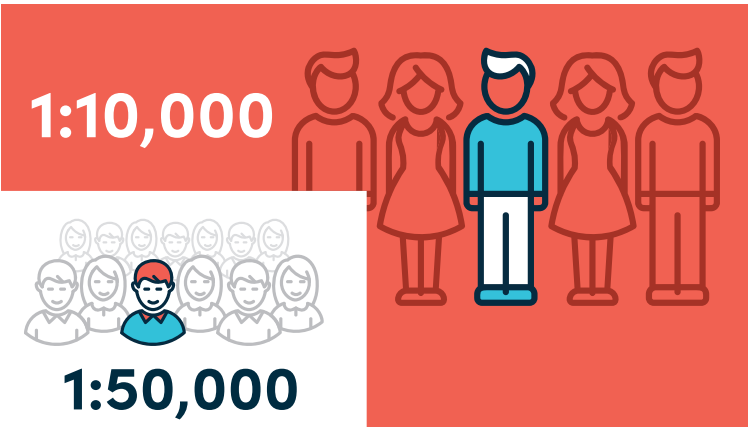


UNDERSTANDING HEREDITARY ANGIOEDEMA (HAE)

WHAT IS HAE?

HAE is a very rare genetic condition that occurs in about 1 in 10,000 to 1 in 50,000 people.

HAE symptoms include episodes of edema (swelling) in various body parts including the hands, feet, face, abdomen, and airway. Excruciating pain, nausea and vomiting can accompany abdominal attacks.



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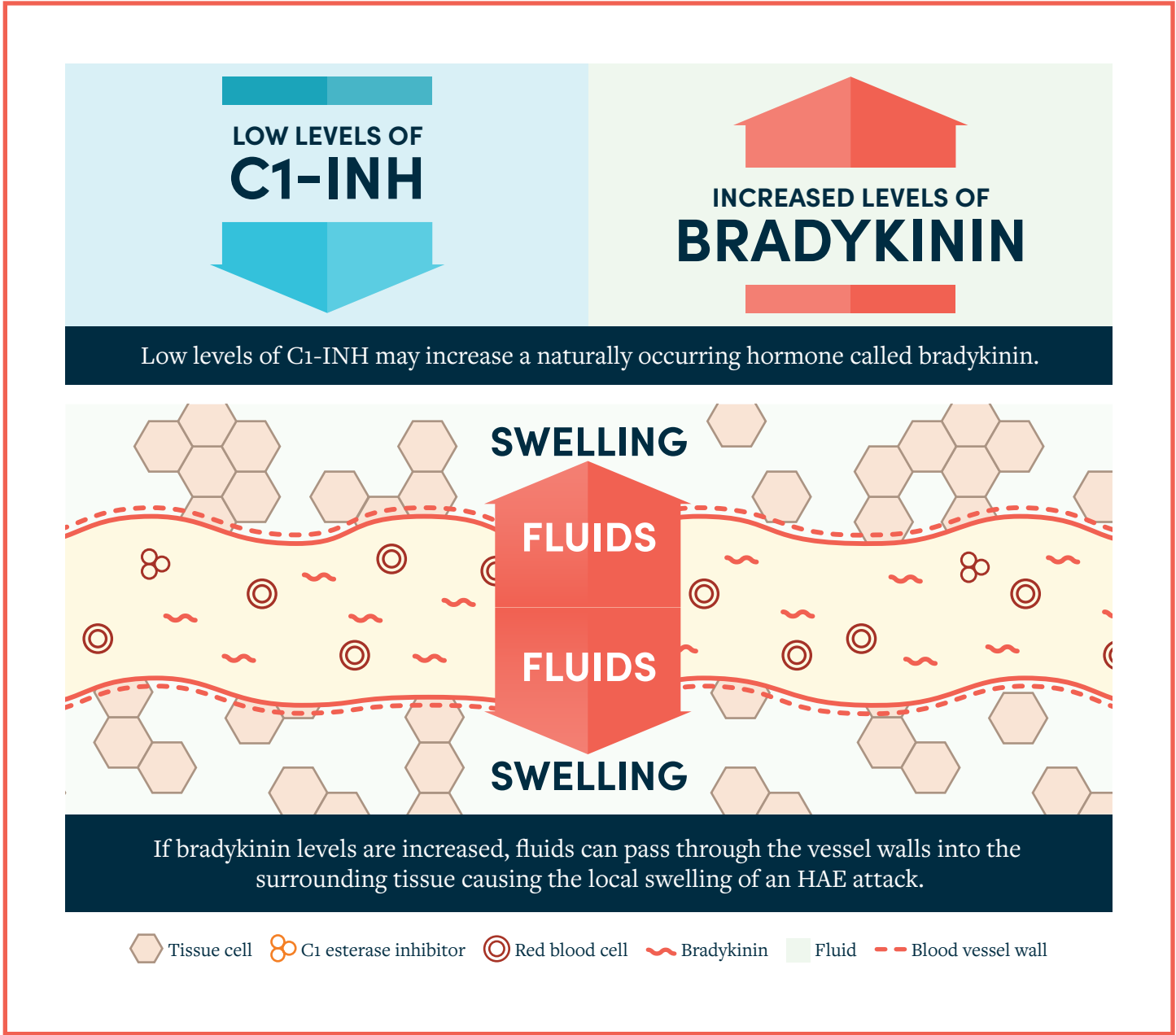
Courtesy of Bas M, et al. *Allergy* 2006;61(12):1490-1492.

WHAT CAUSES HAE?

Type I HAE and Type II HAE are caused by a defect (mutation) in the gene responsible for producing the protein C1 esterase inhibitor (C1-INH).

Unlike other hereditary diseases, the healthy gene cannot compensate for the defect in the other gene in patients with HAE.

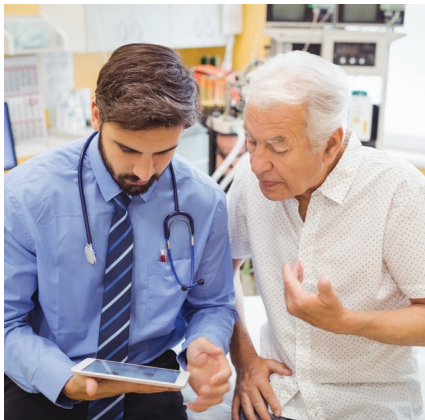
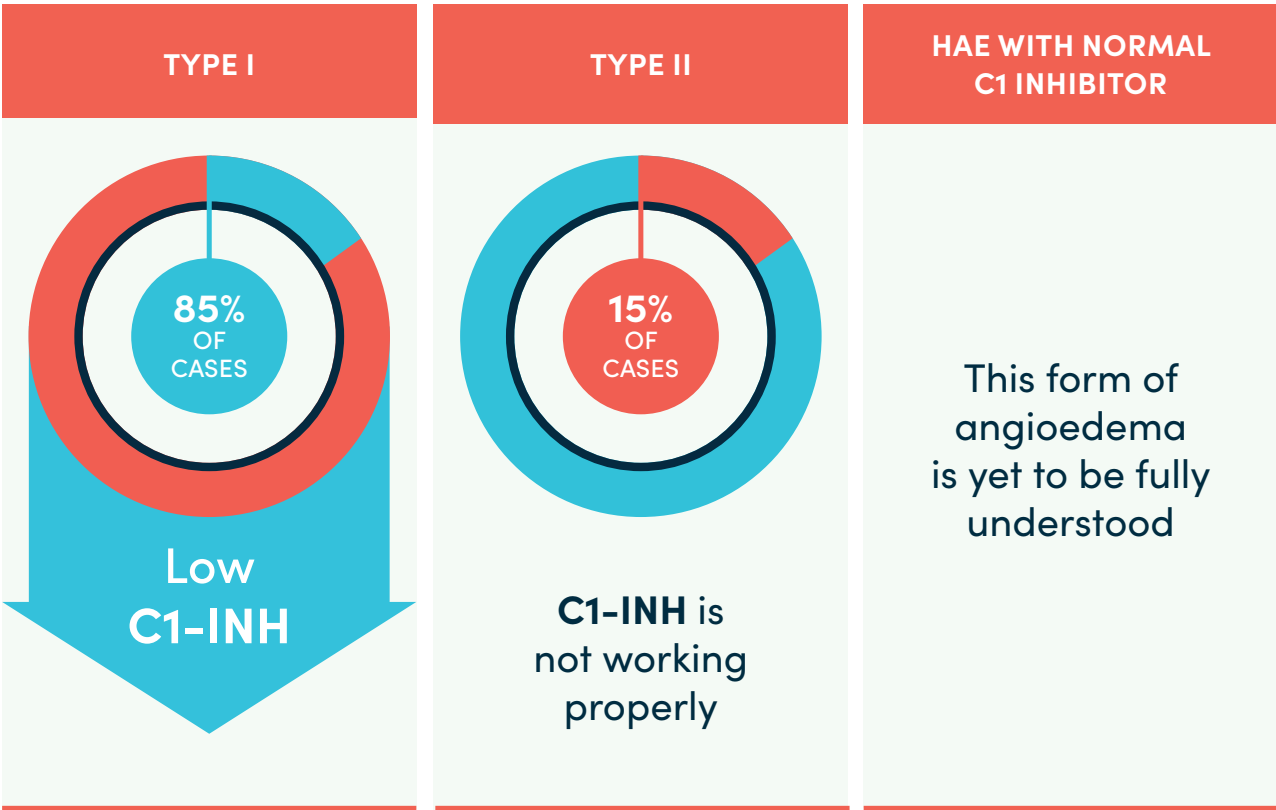
- Low levels of C1-INH may increase a naturally occurring hormone called bradykinin.
- If bradykinin levels are increased, fluids can pass through the vessel walls into the surrounding tissue causing the local swelling of an HAE attack.



HOW IS HAE DIAGNOSED?

Laboratory analysis of blood samples or genetic testing is required to establish the HAE diagnosis.

There are three types of hereditary angioedema:



The diagnosis of HAE requires a detailed personal and family history, and laboratory measure of C1-INH, C1-INH activity, and serum C4 levels.

C4 is reduced in **98%** of cases for both HAE Type I and Type II, and nearly **100%** of the time during an attack.

HOW IS HAE TREATED?

Routine prevention of HAE attacks

Long-term prophylaxis (LTP) refers to ongoing treatment of HAE.

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| The aim of LTP is to reduce the frequency and/or severity of HAE attacks. | Some patients may be candidates for LTP and the benefits and risks associated with such treatments should be explored. | LTP does not eliminate the risk of attacks completely and those attacks can still be fatal. | All patients must have a plan to treat attacks on demand despite being on LTP therapy. |
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On-demand treatment of HAE symptoms

On-demand treatment refers to the treatment of acute attacks.

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| Attacks should be treated as early as possible. | All attacks of angioedema involving the upper airway are medical emergencies and must be treated immediately. In addition, we recommend emergency department assessment. | All patients should have on-demand treatment for two attacks. | All patients should carry their on-demand treatment at all times. |
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Treat HAE attacks early

All attacks – anywhere in the body – should be treated as soon as you recognize the symptoms

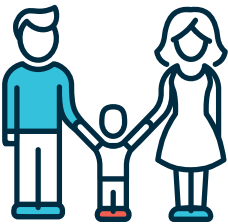
Use the form provided in the back pocket to track your HAE attacks and share the results with your healthcare provider.

- Record when the attack started, when on-demand treatment was administered, when you experienced initial symptom relief, and the time to the end of your attack.

An individualized treatment plan should be carefully developed in partnership between the physician and the patient. It should address preventive measures, home care, self-administration, and should include an effective on-demand treatment plan, with clear instructions on how to best use medications to treat HAE attacks. The local hospital should have therapies available for emergency treatment.

WHY IS FAMILY TESTING IMPORTANT?

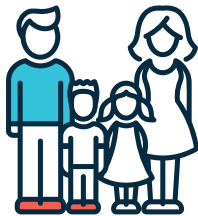
HAE due to C1-INH deficiency is a genetic disorder, and the child of an affected parent has a 50% chance of inheriting the disease.



- HAE symptoms often begin in early childhood, however some patients experience their first attack later in life and some do not experience any symptoms.
- There is great variability in the frequency, location, and severity of attacks; this can also be true between members of the same family.
- Members of your family may have the disease even if they do not present with any symptoms, highlighting the importance of family testing.
- It is important to establish the diagnosis as early as possible, ideally before the onset of clinical manifestations.

Family members of patients with HAE should be screened to prepare for the future.

It is recommended that the following members of your family should be tested for HAE:



- Grandparents
- Parents
- Children



- Grandchildren
- Siblings

Your HAE family health tree

Keeping a record of your family history of HAE may help you determine which family members you should contact about the possibility of getting tested:

- Who has already been screened?
- Who has already been diagnosed?

This information may also be useful in the future to help you talk to relatives and clinicians about your condition.

How to approach your family members about testing for HAE

Look at your family history to help you determine which family members you should contact about testing for HAE. Use the letter provided to help you inform your relatives about the importance of family testing. Each letter contains a brief introduction to the disease and encourages your relative to talk to their doctor about contacting your HAE clinician about testing.

Read the letter carefully. Complete the letter with the name of your relative and the contact details of your HAE clinician, then sign your name at the end. It is then your relative’s decision as to whether they want to talk to their doctor about contacting your HAE clinician.

Reassure your relatives that in the event of a positive diagnosis, your HAE clinician can help them manage their symptoms.

Treatment tips to remember:

- If on LTP, follow your doctor's treatment instructions to prevent attacks.
- LTP does not eliminate attack risk and you may still experience HAE attacks.
- Treat all breakthrough attacks early with your on-demand treatment.
- Ensure you carry your on-demand treatment at all times.

WHERE CAN I GO FOR MORE SUPPORT?


HAE Canada Patient Organization

HAE Canada is a patient group formed in 2010 to work with physicians, nurses, and other healthcare related professionals to create a better life for HAE patients living in Canada. All HAE patients in Canada are encouraged to join this group. There is no membership fee and you will find many helpful resources. For more information, visit www.haecanada.org.



If you are looking for a Canadian Hereditary Angioedema Network (CHAEN) physician and/or clinic, please visit: <http://chaen-rcah.ca/patient-resources/>.



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