

Establishing an Emergency Care Plan for HAE patients at a local hospital



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Step One: Making the Connection

How to put an Emergency Care
Plan in motion

Connect with your HAE specialist



Make an appointment with your HAE specialist to get a current physician's letter.



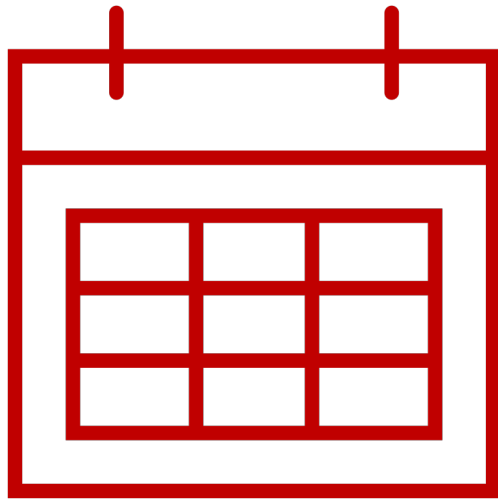
The letter should contain:

- your diagnosis
- individualized care plan
- treatment details



You can find a sample physician letter on our website:

<https://haecanada.org/travel/>



Helpful Tip

It is a good idea to be re-assessed annually by your HAE specialist to discuss issues and to explore possible new treatment options.

If you do not see your treating HAE specialist (at least) annually, you will require a new referral from your family doctor before setting up your next appointment

Visit your local hospital



Set up an appointment with your hospital's Director of Emergency Services (or equivalent) and / or the hospital educator.

They are responsible for setting up a care plan and will ensure proper information and training reaches hospital staff.



The appointment may need to be at least 30 minutes, and a follow up appointment may be required.



Helpful Tips

You should set up your care plan appointment through your hospital's main switchboard, *not through the Emergency Department*. Emergency Department staff do not have time to set up care plans for individual patients.

Always meet with hospital staff swell-free. Attacks are stressful and you cannot advocate effectively during an attack. Staff will understand if you need to reschedule for a later date.

Step Two: The Meeting

How to put a plan in place



Bring appropriate HAE resources to familiarize hospital staff with HAE and how it can be managed. Most hospital staff have little or no knowledge of HAE, how it presents, treatments, or its severity.

You will need to educate them about how HAE affects you personally. You might find it helpful to prepare notes so you don't forget anything.

What to Bring



Letter from your HAE specialist




Your specialist's contact information



HAE Resources such as:


1. HAE Fact Sheet
2. Tear sheet
3. Infographic
4. Wallet Card

HAE Resources



Hereditary Angioedema (HAE)

What? Who? When? Where? Why? How?



What is HAE?

- HAE is a rare genetic disorder characterized by recurring painful attacks of swelling in different areas of the body. The severity of the attacks can vary between patients and throughout an individual patient's lifetime. Attacks can also be disfiguring and when they occur in the throat, can be life-threatening due to the risk of suffocation.¹

Who gets HAE?

- HAE Affects between 1 in 10,000 to 1 in 50,000 people.^{2,3}
- Most people with HAE have inherited it from a parent. About 20-25% of cases of HAE occur spontaneously, without previous family history.⁴
- Children of someone with HAE have a 50% chance of inheriting HAE.⁵
- HAE affects men and women⁶, with higher rates in women, and people of all ethnic groups⁷ at approximately the same rates.

When does HAE occur?

- Inherited HAE symptoms can start at different ages, even in children just a few years old. Half are symptomatic by age 7. For some, symptoms only appear in their 30's or 40's.⁸
- Majority of HAE swelling attacks are unpredictable.⁹ Some can have attacks as frequently as every three days while others have attacks less often.¹⁰
- Swelling attacks generally occur without warning, though in some cases an attack is preceded by symptoms such as fatigue, irritability, or a pink rash.¹¹
- Attacks usually worsen steadily over the first 24 hours then gradually subside over the next two to three days.¹²
- Some attacks can be as short as four hours and others can last as long as five days.¹³
- In general, people with HAE are incapacitated by attacks from 20 to 100 days per year.¹⁴
- Certain triggers or events can bring on the attacks such as an injury, surgery, dental procedures, anesthesia, stress, infection, fluctuating hormone levels, menstruation or use of oral contraceptives.¹⁵

Where in the body does HAE affect?

- Almost any part of the body can be affected by a swelling attack, but attacks most commonly occur in the face, extremities (arms, hands, legs, feet), genitals, abdomen, or throat.¹⁶
- Attacks can be localized in one area of the body or can spread to another location before resolving.¹⁷
- Attacks in the abdomen and intestines are usually the most painful and can cause severe vomiting, diarrhea and cramping that can be mistaken for other conditions, such as appendicitis, that require surgical intervention.¹⁸
- Attacks leading to swelling in the throat are life threatening medical emergencies that require prompt attention due to the risk of suffocation.¹⁹

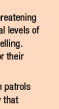
HAE Fact Sheet

Hereditary Angioedema (HAE)

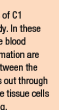
What is hereditary angioedema?

Hereditary angioedema (HAE) is a rare, potentially life-threatening genetic disorder. It is caused by deficient or dysfunctional levels of C1 inhibitor, a protein in the blood that helps prevent swelling. People with HAE either don't have enough C1 inhibitor, or their C1 inhibitor does not work properly.

- In people with normal levels of C1 inhibitor, the protein patrols the blood vessels, regulating the systems in your body that cause inflammation.
- People with HAE have deficient or dysfunctional levels of C1 inhibitor, which triggers a cascade of events in the body. In these people, C1 inhibitor can no longer effectively patrol the blood vessels, which means the system that control inflammation are no longer regulated. Eventually, this opens up gaps between the cells that make up the blood vessel walls. Fluid moves out through the leaky blood vessel walls and builds up between the tissue cells underneath the skin, which results in localized swelling.




Normal levels of C1 inhibitor



Levels of C1 inhibitor in a person with HAE

Signs and symptoms of HAE



<https://www.haenrgs.com>


The nature of HAE varies greatly and symptoms of HAE may change over time. Attacks typically last between 2 and 5 days, and patients can experience 1 to 3 attacks per month, resulting in 20 to 100 days of decreased functioning a year.

Typical symptoms of HAE include:

- Intense, sometimes painful attacks of swelling in any part of your body, especially the extremities (hands, feet, arms, and legs), abdomen, face, genital area, and throat
- Attacks of abdominal pain, nausea, and vomiting that are caused by swelling in the intestinal wall
- Swelling of the throat, which can block the breathing passage and result in a life-threatening emergency

Talking with your healthcare provider

It is important that you share any family history of angioedema with your healthcare provider. Be prepared to discuss your symptoms, including the severity and duration of your attacks.




Tear sheet


HEREDITARY ANGIOEDEMA (HAE)

What?

...is HAE?




HAE is a **RARE AND SERIOUS GENETIC DISORDER** characterized by recurring painful attacks or **SWELLING** in different areas of the body. The severity of the attacks can vary between patients and throughout an individual patient's lifetime. Attacks can be painful and disfiguring and, when they occur in the **THROAT**, can be **LIFE-THREATENING** due to the risk of suffocation.



Where?


...in the body DOES HAE affect?




Almost any part of the body can be affected by a swelling attack, but attacks most commonly occur in the face, extremities (arms, hands, legs, feet), abdomen or throat.

When?

...does HAE occur?



Although **HAE SWELLING ATTACKS ARE UNPREDICTABLE**, some can have attacks as frequently as every three days, while others have attacks once a month or even less often.




Who?

...is HAE?

HAE affects **1 in 10,000** to **1 in 50,000**

- About **20-25%** of cases of HAE occur **SPONTANEOUSLY**, without previous family history.
- HAE affects men and women and people of all ethnic groups at approximately the same rates.



How?

...is HAE diagnosed and treated?

TREATMENTS

6 Health Canada approved treatments

There are three blood tests to confirm HAE:

1. C4.
2. C1-inhibitor quantitative (antigenic)
3. C1-inhibitor functional

• Treatments exist to help reduce symptoms of an attack but

PREVENTION

• Treatments can also be administered on a regular basis to help prevent attacks.

This infographic was made possible through an investigator grant from the Canadian Institutes of Health Research.

Revised March 2019

Infographic

Infographic

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Wallet Card

Step Three: Living with HAE

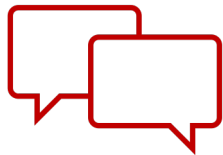
How to continue to manage
your HAE using your care plan
and other resources

Understand your care plan and its limits



Get to know your Emergency Department staff.

It will take time for staff to recognize you, how you present with HAE, and how to put your Care Plan into effect



Be patient & provide the hospital with feedback.

If things are not working, let the person responsible for the Care Plan know so they can work with you to improve it. It may take time & experience for the care plan to work as planned.

Helpful Tip

If you get to know your hospital, blood bank and/or pharmacy staff, that may translate into you receiving better treatment.

Ask if they would like information on HAE; their familiarity with your HAE could help you get more effective treatment.





This is the standard Emergency Department Protocol that we at HAE Canada believe is attainable and are striving for throughout Canada