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

HAE AOH CANADA

Created: May 2022

Step One:

How to put an Emergency Care Plan in motion

Connect with your HAE specialist

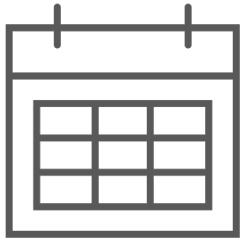
Make an appointment with your HAE specialist to get a current physician's letter.

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

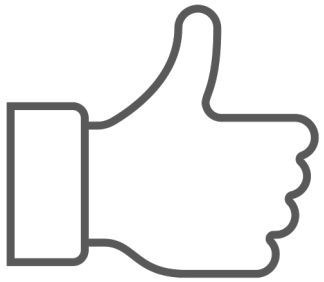
Visit your local hospital

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:

How to put a plan in place

Be Prepared

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

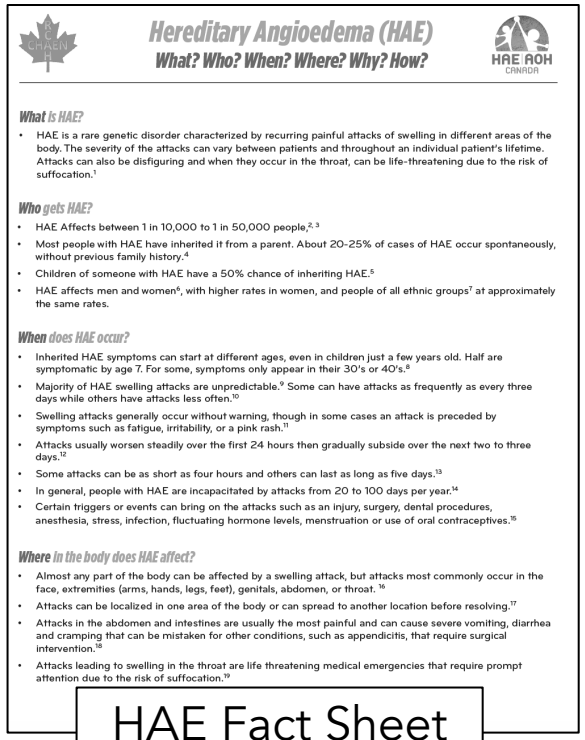
Letter from your HAE specialist

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?

HAE AOH CANADA

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 women[6], with higher rates in women, and people of all ethnic groups[7] at approximately the same rates.

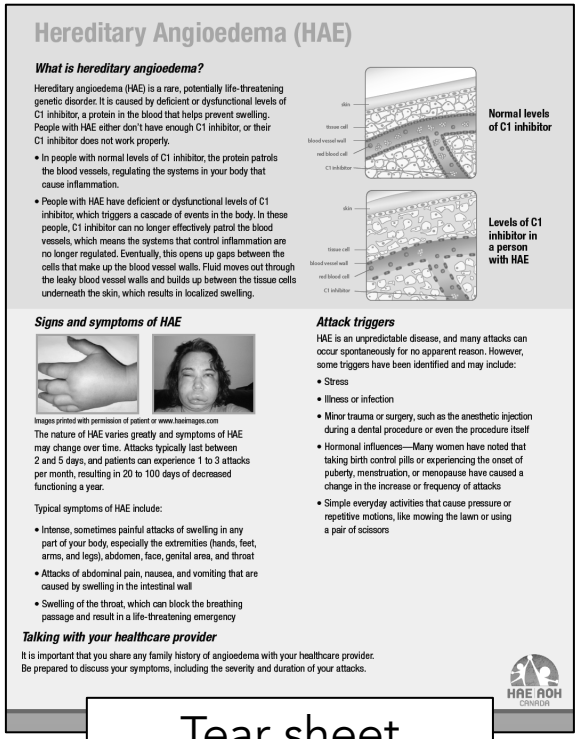
When does HAE occur?

- 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 systems 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

Images printed with permission of patient or www.haeimages.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

Attack triggers

HAE is an unpredictable disease, and many attacks can occur spontaneously for no apparent reason. However, some triggers have been identified and may include:

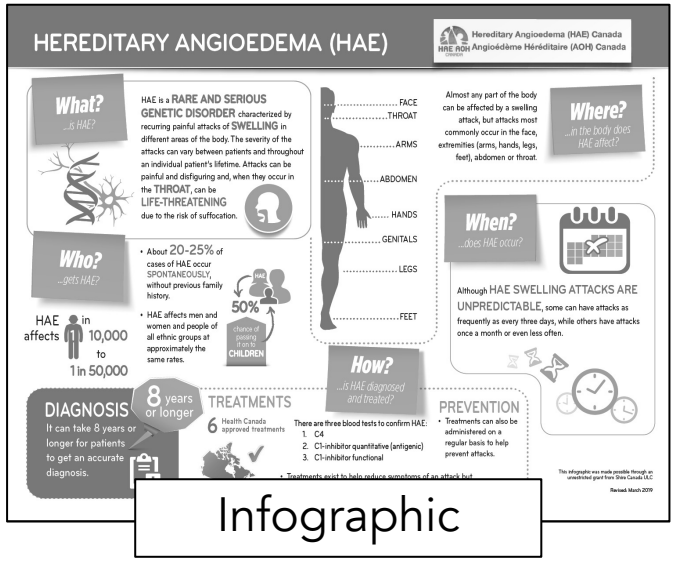
- Stress
- Illness or infection
- Minor trauma or surgery, such as the anesthetic injection during a dental procedure or even the procedure itself
- Hormonal influences—Many women have noted that taking birth control pills or experiencing the onset of puberty, menstruation, or menopause have caused a change in the increase or frequency of attacks
- Simple everyday activities that cause pressure or repetitive motions, like mowing the lawn or using a pair of scissors

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.

HAE AOH CANADA

Tear sheet



HEREDITARY ANGIOEDEMA (HAE)

Hereditary Angioedema (HAE) Canada
Angioédème Héréditaire (AOH) Canada

What? ...is HAE?

HAE is a **RARE AND SERIOUS 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 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.

FACE
THROAT
ARMS
ABDOMEN
HANDS
GENITALS
LEGS
FEET

Who? ...gets HAE?

- 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.

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

50% chance of passing it on to CHILDREN

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.

How? ...is HAE diagnosed and treated?

DIAGNOSIS

It can take 8 years or longer for patients to get an accurate diagnosis.

8 years or longer

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

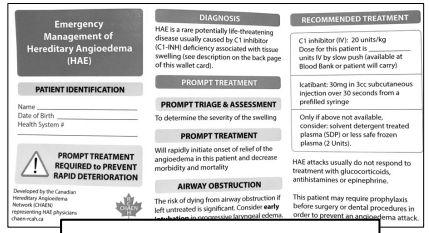
PREVENTION

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

This infographic was made possible through an unrestricted grant from Shire Canada ULC

Revised: March 2019

Infographic



Emergency Management of Hereditary Angioedema (HAE)

PATIENT IDENTIFICATION

Name _____
Date of Birth _____
Health System # _____

PROMPT TREATMENT REQUIRED to PREVENT RAPID DETERIORATION

Developed by the Canadian Hereditary Angioedema Network (CHAEN) representing HAE physicians chaen-rcah.ca

DIAGNOSIS

HAE is a rare potentially life-threatening disease usually caused by C1 inhibitor (C1-INH) deficiency associated with tissue swelling (see description on the back page of this wallet card).

PROMPT TREATMENT

PROMPT TRIAGE & ASSESSMENT

To determine the severity of the swelling

PROMPT TREATMENT

Will rapidly initiate onset of relief of the angioedema in this patient and decrease morbidity and mortality

AIRWAY OBSTRUCTION

The risk of dying from airway obstruction if left untreated is significant. Consider **early** [illegible] in progressive laryngeal edema.

RECOMMENDED TREATMENT

C1 inhibitor (IV): 20 units/kg
Dose for this patient is _____ units IV by slow push (available at Blood Bank or patient will carry)

Icatibant: 30mg in 3cc subcutaneous injection over 30 seconds from a prefilled syringe

Only if above not available, consider: solvent detergent treated plasma (SDP) or less safe frozen plasma (2 Units).

HAE attacks usually do not respond to treatment with glucocorticoids, antihistamines or epinephrine.

This patient may require prophylaxis before surgery or dental procedures in order to prevent an angioedema attack.

Wallet Card

You can download resources from the HAE Canada website: https://haecanada.org/resources/

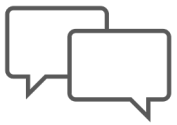
Step Three: Living with HAE

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