

Learning to Live with Hereditary Angioedema (HAE)





Introduction

If you are living with hereditary angioedema (HAE), you have probably experienced a range of emotions from frustration and fear to anger and helplessness. It's not easy living with HAE. That's why we created this brochure – to provide you with some suggestions to help you live life to the fullest. HAE may be rare¹⁻⁵, but you are not alone. We are here to help.

This booklet is to be used for general information purposes only. It should not replace the advice and expertise of your healthcare professional. Please consult your healthcare professional should you have any questions about your condition.



Living Well With Hereditary Angioedema (HAE)

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The Importance of Emotional Support

Everyone is different in terms of how they respond to living with HAE. Having a network of support and an individual management plan can help you feel a sense of control over your condition. This can allow you to continue to do the things you want to do while living with HAE.

Some common feelings

While every person with HAE is unique, you may find that you share some common feelings and experiences with other patients. It is perfectly normal to feel frustrated, angry, anxious, and alone from time to time – to feel like no one understands what you are going through. However, having a coping strategy for dealing with these feelings is an important step along the road to living well with HAE. This includes ensuring you have close friends, family, and perhaps others who are living with HAE to lean on when you need them.



Worrying about an unexpected attack

Feeling anxious about the potential for an unexpected attack can sometimes be overwhelming. HAE attacks are unpredictable¹⁻⁵, but having a plan in place for attacks may help ease your mind by making you feel more prepared and in control. Your plan could include things like making sure medication is on hand or ensuring friends, family and colleagues are aware of what to do in the event of an attack.

Feeling that quality of life is suffering

HAE can impact quality of life.^{2,6} An HAE attack or the fear of having one can interfere with some of the things you love to do, or cause you to do those things a bit differently. You may need to adapt your activities to minimize the impact of HAE on your daily life.

For example, if you know an attack will keep you from work for a few days every month you may wish to educate your co-workers on your condition. People are more understanding and helpful when they know what you are going through. In addition, sharing a bit about your condition can help break down fear and stereotypes.

Managing Triggers

Part of looking after your overall well being includes learning to identify and manage triggers of an HAE attack. HAE is unpredictable and attacks can occur for no apparent reason.¹⁻⁵ However, certain triggers have been identified in some people. These include^{4,5}:

- Trauma
- Surgery
- Anesthesia
- Dental procedures
- Emotional stress
- Fatigue
- Infections
- Birth control pills containing estrogen
- Hormone replacement therapy
- Menstruation
- Some medications used to treat high blood pressure
- Alcohol

Knowing what your triggers are and learning how to manage them may help some people avoid attacks.

Dealing with the reactions of others

Sometimes the reactions of others can be difficult to deal with. Yet, helping others understand what it's like to have HAE can help alleviate feelings of awkwardness people often experience when symptoms occur.

Here are a few tips on how to manage your feelings about your condition when around other people:

- Being with friends, family and other people who understand the nature of HAE while symptoms occur, rather than being alone, can often act as a positive distraction from your symptoms.
- If your normal routine is interrupted due to HAE symptoms, use alternative strategies to allow you to get on with what you need to do, such as working or studying from home. At the same time, if you need to rest, do so.
- Talk to your colleagues, teachers, friends and acquaintances if you are comfortable doing so. Let them understand the nature of your symptoms. People are generally very understanding and helpful, particularly if you are able to explain how HAE can impact your life.
- It is perfectly normal to feel self-conscious when symptoms occur in public. Some people find it helpful to prepare a list of standard responses to address the curiosity of strangers rather than allowing HAE to interrupt their routine. Using humor in particular, with a brief explanation that the symptoms are temporary, can help make others – and you – feel more comfortable and in control.



Feeling overwhelmed

While it is normal to feel a range of emotions as a result of having HAE, including those that can be negative from time to time, you don't have to live with these feelings all of the time. Part of looking after your overall well being is being aware of your emotions and knowing when to get help if you feel they are starting to take over. It is important to talk to your doctor if you are starting to feel overwhelmed, constantly anxious, or depressed. The aim of any professional support is to provide people with the skills and resources necessary to cope with their condition and improve the quality of their life.

Family and HAE

An HAE attack can affect different people in different ways. Only you will know how comfortable (or not) you feel being seen by family members during an attack. Feelings of embarrassment are normal and common. Remember, though, that HAE is a medical condition and you do not need to feel ashamed by it.

Relating to other family members

- Talk through your feelings with your family if you are comfortable doing so and, if they're concerned, reassure them that there are no lasting effects on appearance.
- There may be situations in which an attack becomes serious and even a medical emergency. Make sure your family members know what to do in the event that an attack becomes life threatening, so that they don't panic and can offer you the assistance you need.
- Don't feel obliged to share your condition with distant family members, unless an attack should happen in their presence or if it affects a social engagement.

When other family members have HAE

- If you are the first in your family to be diagnosed with HAE, encourage your relatives to be tested for HAE, especially if they have experienced unexplained swelling. A blood test or genetic test can confirm diagnosis.^{4,5}
- If other family members have HAE there will be considerable common knowledge about living with the condition and its effects on family members. Don't be afraid to ask them to share their experiences and get their advice on coping with the condition.
- Everyone's experience of HAE is unique. Just because other family members have HAE, does not necessarily mean you will have the same physical or emotional symptoms. Share your own experiences with family affected by HAE – you may be surprised by the similarities and differences.



Reassuring your children

It's entirely normal for children to be concerned when they first see a parent having an acute HAE attack, particularly if the attack affects the face. Some parents prefer not to let their children see them while they are having a facial attack; however, avoiding your children may not be the best approach in all cases. It might be helpful to be honest and forthright about your condition. Teach your child, in age appropriate terms, what your condition is about, how it might affect you, and reassure your child that the swelling will go down. Also let them know that there are no lasting effects on your appearance. Children can be very sympathetic once they understand more about the condition and their worries have been dealt with. It also sets a good example for children, showing them that we are all different and all deserving of respect and empathy in spite of our differences.

Relationships and HAE

It is important to let your partner know how HAE affects you. This can help you manage expectations right from the outset, helping make it easier to enjoy a happy, fulfilling relationship.

Telling your partner about HAE

- Consider telling your partner about HAE early on in the relationship, especially if you are becoming more physically or sexually intimate. This way your partner will feel more included and be better prepared if an attack happens.
- Explain to your partner what typically happens to you during an attack and that you can self-treat to prevent a mild attack from becoming more serious. In the unlikely event of an emergency, make sure your partner has an emergency contact and that he or she is aware of what to say to medical personnel in such cases.
- Early on in your relationship, you may or may not feel comfortable with your new partner seeing you during an attack. This is perfectly normal. Make sure you explain how you feel to your partner so that he or she knows what to expect should you withdraw or “lay low” for a bit. Reassure your partner that there are no lasting effects with your appearance.
- As your relationship grows, your confidence should grow as well; however, you may still wish to be left alone during an attack. Make sure that your partner knows this – and that you don’t need him or her to be fussing over you during a mild attack.



Sexual activity with your partner

- Care may need to be taken by your partner in sexual activity (and in everyday activities) not to cause any physical trauma, as this may trigger an attack in some people.
- Explain to your partner that an attack involving the sexual organs may cause considerable swelling and painful intercourse.^{2,3} If this is the case reassure your partner that the swelling will go down and that there are no lasting effects on your appearance. Sharing these concerns with your partner will allow you to continue enjoying a close, intimate relationship. It may also bring you both closer as you find ways to manage something so personal. Communicating your needs to your partner is essential.



HAE at school and in the workplace

Because HAE is rare¹⁻⁵ your teacher or employer is unlikely to know much about. Giving him or her advanced warning of how HAE affects you will help you manage his or her expectations better – and allow you to get the most from school and work.

With your permission, your teacher or employer can share more about your condition with your colleagues so that everyone is prepared to handle an attack sensitively should one occur on site, particularly if it involves disfigurement.

Ultimately it's your decision how much you wish to disclose about your condition.

Tips on managing HAE while at school or work

- Planning ahead is important. Make sure your teacher/employer is aware that you may occasionally miss school or work because of an attack. Ensure he or she knows that you are still committed to your work, regardless of how your condition affects you. Arrange to have lecture notes sent to you (if you are in school), or if possible, arrange to be able to work from home on occasion (if you are working) so that you can still get your work done.
- Learn about what accommodations can be made for your condition. Many institutions have webcast capabilities that allow you to link to a lecture remotely should you be unable to attend in person. Likewise, many meetings at work can be held via webcast or teleconference.
- Let your teacher/employer know that occasionally you may require more time to meet a deadline if an HAE attack has prevented you from working on your assignment. This can help him or her manage expectations appropriately.
- Driving to and from school/work may prove to be difficult after an attack. Likewise taking public transportation could make you feel self-conscious. Be sure to plan for alternate transportation ahead of time so that you aren't left scrambling after an attack.

Traveling with HAE

It's normal for you to have concerns about travelling. But remember, going on holiday with family or friends should be an enjoyable time. A little planning can help make your time away less stressful.

Tips on managing HAE while travelling

- Check that your travel medical insurance covers existing conditions.
- Check carry-on luggage regulations and storage conditions with individual airlines before travelling.
- Make sure that you have a signed letter from your doctor stating that, out of medical necessity, you must carry your HAE supplies with you at all times.
- Make a list of all the prescription items you have to take with you.
- Place labels on all of your supplies and check that the name on the prescription medicine matches the name on your airline ticket.



- Take sufficient supplies and medication for extended travel abroad and ensure you check the storage conditions.
- Bring a sharps container (such as a biohazard, hard plastic, or metal container) with you for safe disposal of sharps (syringe/needle).
- Remember to have your physician letter with you at all times, so that you can refer to this in the event of an emergency.
- Many medical personnel in hospital emergency rooms have not heard of HAE and treat it as an extreme allergic reaction.

About HAE: Commonly asked questions

What is HAE? Hereditary Angioedema (HAE) is a rare but serious blood disorder that is passed along in a person's genes.^{1-4,6} HAE is marked by recurring episodes of severe swelling – most commonly of the hands, face, throat and abdomen (belly). While there is no cure for HAE, it can be treated with medication.^{1-4,6}

How common is HAE? HAE affects only about 1 in every 10,000 to 50,000 Canadians.¹ Because it is so rare, it is often misunderstood and misdiagnosed.¹⁻⁵

Will your children get HAE? HAE is a hereditary condition, which means that the likelihood of passing HAE on to your children is greater if one parent has it.⁶ Every case is different, however. It is best to talk to your doctor who can help determine what the risk of passing it along might be.

Is HAE contagious? HAE is not contagious. It cannot be passed on to others like a virus or bacterial infection.

What should I do if I have an attack? It's natural for people to want to help you if you have an attack. However, everyone experiences HAE differently. You be the judge of how you want to be helped by others during an attack. Let people know ahead of time. Carry an emergency card with you that instructs others what to do in the event of an emergency, including who should be contacted.

About HAE: Commonly asked questions

How many types of HAE are there? There are three types of hereditary angioedema:

- Type I - Low C1 inhibitor protein level and function – most common type.
- Type II - Normal C1 inhibitor protein level, but abnormal function.
- HAE with Normal C1 (*formally known as Type III*) - Normal C1 inhibitor protein level and function – estrogen dependent.

Acquired angioedema (AAE) is also documented in the literature, but is not associated with family history. Drug-induced angioedema is another angioedema type found in the literature, associated with such drugs as angiotensin-converting enzyme inhibitors (Bowen et al., 2010). Neither of these are hereditary.

Why was I referred to an immunologist and not a hematologist?

Hereditary angioedema is both a blood disorder and an immunologic disorder in that the deficient protein (C1 esterase inhibitor) is part of the blood and this protein regulates part of the immune system. The disorder involves the complement and contact systems of the immune response and the protein affected in hereditary angioedema is an inhibitor of these systems. C1 esterase inhibitor protein is an enzyme that is used by the body to shut down the immune response, which is responsible for the swelling (Agostoni et al., 2004; Ritchie, 2003). HAE patients can be followed by either immunologists or hematologists, but most tend to be referred to immunologists.

Helpful Links

- HAE Canada: HAECanada.org
- HAE International (HAEi), a global organization dedicated to raising awareness of C1 inhibitor deficiencies around the world: haei.org
- Canadian Hereditary Angioedema Network (CHAEN) Physician Website: chaen-rcah.ca/
- Canadian Allergy, Asthma, and Immunology Foundation (CAAIF): www.allergyfoundation.ca/
- Canadian Society of Allergy and Clinical Immunology (CSACI): csaci.ca/
- Canadian Organization for Rare Disorders (CORD): www.raredisorders.ca/
- World Allergy Organization (WAO), a global alliance of asthma, allergy and immunology societies: www.worldallergy.org

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Where can I go for more support?

While your doctor is an important source for information and advice about your condition, you may also find it helpful to connect with others just like you who are living with HAE.



HAE Canada is a patient group that can be a valuable resource for people living with HAE and membership is free. Get the latest news and information on HAE, and perhaps even share a bit of your time volunteering to help others living with HAE. For more information, please visit HAECanada.org.