HAEA
US HEREDITARY ANGIOEDEMA ASSOCIATION

Women

WITH HEREDITARY ANGIOEDEMA

The information provided in this Guide is not meant to be a substitute for medical advice, diagnosis, or treatment recommendations. The US HAEA is product and company neutral and does not endorse or recommend HAE medicines.

A GUIDE FOR WOMEN WITH HEREDITARY ANGIOEDEMA

Pediatrics Through Puberty

Family Planning and Pregnancy

Menopause and Aging

WOMEN WITH HAE

The purpose of this Guide for Women with Hereditary Angioedema (HAE) is to provide information regarding the unique challenges faced by women with HAE. The Hereditary Angioedema Association (HAEA) collaborated with HAE expert physicians to develop an authoritative resource that includes frequently asked questions, stories from patients, and relevant source material.

Studies reveal that HAE symptoms are more severe in women than men. The HAEA created this resource to help women with HAE navigate three specific life stages:

1) pediatrics and puberty;

2) family planning and pregnancy; and,

3) menopause and aging.


"This is your moment.
Own it."
— Oprah Winfrey

UNDERSTANDING HAE AND ITS EFFECT ON THE FEMALE BODY

From the onset of puberty to the late stages of menopause, women experience considerable hormonal fluctuations, particularly in estrogen. Changes in estrogen levels can affect the frequency and severity of HAE attacks. As women move through the various life stages, it is important to be mindful of how hormonal variations may impact HAE symptoms and one's approach to treating attacks.

Included are blank pages for notes and journaling, as well as positive affirmations to support you on your journey.

TESTING FOR HEREDITARY ANGIOEDEMA

Early testing is important to confirm an HAE diagnosis. Blood tests are required to diagnose Type I and Type II HAE.

These blood tests can be ordered at most physician offices.

THE BLOOD TESTS REQUIRED FOR AN HAE DIAGNOSIS ARE:

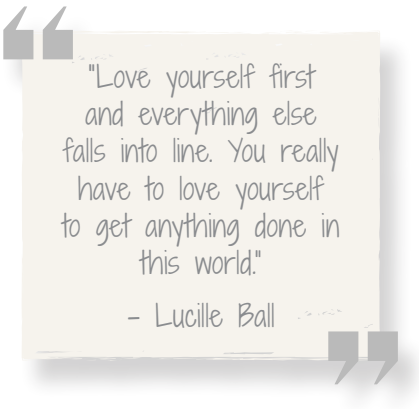
C4 Level

C1 Esterase Inhibitor Level

C1-INH Function

HAE WITH NORMAL C1-INHIBITOR

There are a number of people who have an HAE diagnosis but have blood tests that return with normal levels of C1-Inhibitor. It is important to discuss this diagnosis with your physician.



HAE Testing Notes:

My Notes:

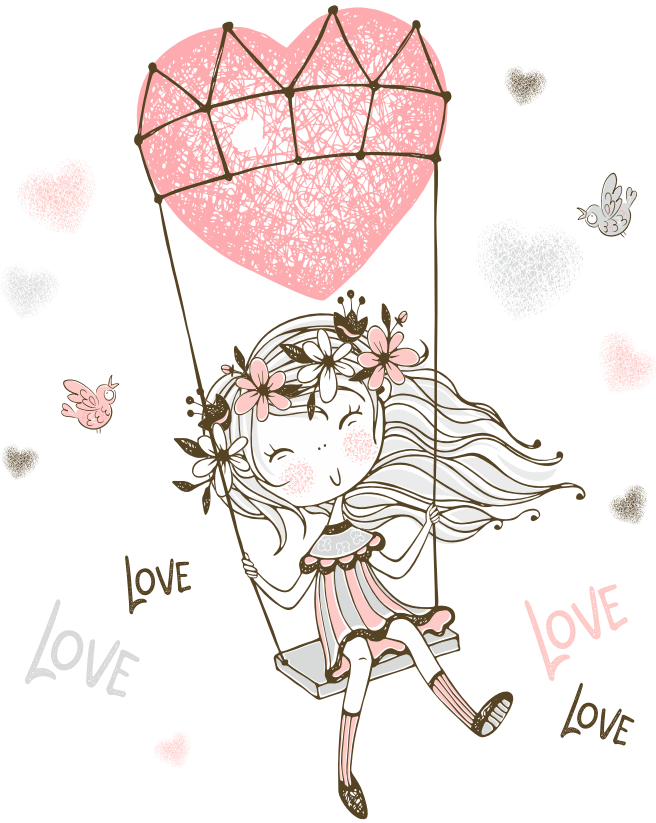
PEDIATRICS THROUGH PUBERTY

TIME TO CHASE YOUR DREAMS

PEDIATRICS THOUGH PUBERTY: THE FEMALE CHILD AND HAE

If you are the parent and/or caregiver of a child with HAE, it is important to be prepared in the event that your child begins to display symptoms. Obtaining an early diagnosis is critical to ensuring that you, your child's pediatrician, and HAE treating physician have a comprehensive plan in place to treat HAE in the event of an attack.

This section includes personal stories and recommendations from parents, frequently asked questions answered by physicians, information about school accommodations for a child with HAE (504 plan), a sample letter for your child's school nurse, and more.



DEVELOPING A TREATMENT PLAN

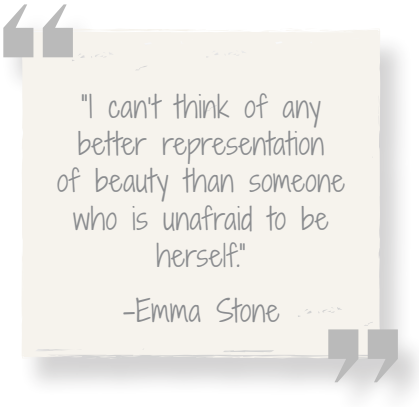
While many children don't begin to exhibit HAE symptoms until early puberty, it is important to work with your physician to develop a treatment plan so your family feels confident and prepared if HAE-related swelling does occur.

The treatment plan should include the following:

- A prescription and HAE medication that is available when needed,
- A diagnosis and treatment recommendation letter from your child's HAE treating physician to share with Emergency Department staff, and
- A coordinated plan to deal with an attack if the child is at school.

Young children and those who have not had many swelling attacks are often not able to identify and recognize HAE swelling. It is important to talk with your child about what to expect if HAE-related swelling occurs.

The age of HAE onset varies considerably from person to person, however, studies show that half of the patients reported onset of their symptoms by the age of seven, and over two-thirds became symptomatic by the age of thirteen. There also seems to be an increased frequency of attacks during puberty or adolescence and the arrival of menstruation.



ADVOCATE

As a parent and/or caregiver, you will be your child's most staunch advocate. Therefore, it is important that you (1) understand how HAE can affect your child's life, and (2) can speak knowledgeably on your child's behalf. For more information on the various types of angioedema, symptoms, triggers, and current treatments, please visit the HAEA website at www.haea.org.

CREATE A TREATMENT PLAN

Parents and/or caregivers should work with their child's physician to create a plan that allows for swift and effective treatment of an HAE swelling attack.

SET UP AN EMERGENCY PLAN

It is imperative that everyone involved in your child's care knows your plan for an HAE emergency. Prepare and provide concise information with everything needed to take care of your child in an HAE emergency, including information on your child's current therapy, your wishes for how and where your child should receive treatment, and your best contact information.

If you're traveling with your child, make sure you know where the nearest emergency medical facility is located.

My Notes:

Amelia's Story

ONE FAMILY'S EXPERIENCE WITH THEIR DAUGHTER'S FIRST HAE ATTACK

"Something is off with Amelia today. She's not walking on her right foot and asking me to take off her shoe! She seems to be in a lot of pain." The chilling concern evident in Amelia's kindergarten teacher's voice had me quickly packing up my work and rushing out to the car, but I already knew what the rest of the day would entail for my four-year-old girl. I also knew this day would eventually come, but why now? Why so early in her bright life as a young, happy four-year-old who ran around without a care in the world, loving school and her friends with no limitations or worry? I pulled up and Amelia was crying near the fence, her hands wrapped around her stomach in pain while her teacher iced her swollen foot. They had no idea how to help her, no protocol or instruction, no education on how they could keep Amelia more comfortable or warning signs to look for. My denial and false hope were to blame.

Amelia is afflicted with a rare disease called Hereditary Angioedema (HAE), which she inherited from me, and this was her first attack. I was not prepared. I hurried to the school's entrance and scooped Amelia up, wiping her tears and thanking the teachers for their care and love. I could feel my lips shaking but managed to hold back all the tears I wanted to cry with her. "It came on so fast, she was happy and playing, then all of a sudden." Her teachers huddled around, and I promised to keep them posted. They had no idea.

"Mommy, is this the HAE? Will I need a shot like the one you take? I don't want a shot Mommy!" Her little voice shaking with fear while I frantically dialed her allergist and pediatrician. "Dr. B wants you to go straight to the ER. She's calling in to make sure Amelia has a bed right away." The receptionist was firm and direct. "You'll be feeling better soon baby, just hang in there," I was able to blurt out between phone calls to my husband, and family members. Amelia's discomfort was growing as was my urgency to get this fixed.

Things moved quickly at the hospital. Amelia was triaged immediately while residents and interns took turns evaluating her foot and asking questions. "You are the first person I've ever met with HAE," was the comment of the day from medical staff at Yale New Haven Hospital. Too many

questions, too many doctors coming in asking me to hold out for treatment of HAE medicines and consider an x-ray instead, just to make sure Amelia didn't break her foot. "I KNOW WHAT THIS IS! The longer we wait, the more pain she will be in; I'm not going to let that happen." I demanded to see the attending allergist/immunologist who agreed to start Amelia on an acute HAE drug. The nurses soothed Amelia through the IV stick.

My little girl was so brave. Within a few hours Amelia's color was coming back, she could wiggle her toes more and more and her stomach pain was subsiding, evidenced by the copious amounts of popsicles the nurses were fetching and she was happily accepting. By the end of the night, we left the hospital with a more knowledgeable medical staff, a happy girl who viewed the whole ordeal as triumphant, and a mom who experienced a huge wake-up call.

For me, the next steps were critical in ensuring Amelia's teachers were prepared for a future attack, especially one that could manifest into an emergency situation. I met with the school nurse and reviewed educational literature developed by the HAEA to provide information on the signs and symptoms, and to create an individualized treatment plan. In the case of an emergency throat swelling, all her teachers were thoroughly trained on how to administer an acute HAE medication since the school nurse was only available in a part-time capacity.

Knowing I had a 50/50 chance of passing this dreaded disease on to my children, my husband and I decided to still start a family. The landscape for people with HAE is different now. There are effective treatments available and even more on the horizon. Before getting pregnant with my first child, I myself had been living a normal life for almost a decade, a life I thought was comfortable and manageable to pass on to my children. I still hung on to that 50%, always wishing the coin dropped more in my favor. Two out of three of my children have been diagnosed with HAE through blood testing. As of today, my daughter, Amelia is the only one to suffer from HAE attacks, for which we have now developed a robust treatment plan and she can be treated acutely from the comfort of our home. While scary at times, she has used this condition as a source of empowerment and an opportunity to show her courage. We've also established our preparedness plan to manage her HAE so she can go back to living a normal, healthy and happy life!

Hannah's Story

"My diagnosis has allowed me to become more in tune with my body, and my triggers."

-Hannah

A YOUNG GIRL'S EXPERIENCE LEARNING TO MANAGE HER HAE WHILE GOING THROUGH PUBERTY

I was 13 years old when I was officially diagnosed with HAE. However, my first attack was around the age of 9 years old. I began to see a large increase in the number of attacks around the ages of 11-12, just around puberty. Before I reached puberty, I had little to no attacks. When my attacks became more frequent, they were mainly abdominal (about 99%) and were happening about once a week. When I got my first period, I would experience crippling abdominal swells that would last for about 3 days or longer and would result in extreme bloating, vomiting, fatigue, and pain.

Each month, as menstruation approached, I would experience abdominal swell symptoms (my most frequent attacks are still abdominal). I would become more bloated than usual and feel slight aching within my stomach that did not feel like cramps. I can almost always tell when my menstruation is about to begin because I can feel attack symptoms coming on.

During this time, the increase in attacks caused me to frequently miss school and other social activities (like hanging out with friends, going to sporting events, etc). I would sometimes have to leave school early when I felt abdominal attacks coming on and miss crucial tests or quizzes. My teachers and coaches were aware of my medical condition and were very flexible in allowing me to make up tests/quizzes and sit out on some practices.

Before being diagnosed and having medication available, I had little to no coping mechanisms. I was completely unaware of my condition, and what was happening to my body. Once my diagnosis was official, I was able to cope a little easier and accept my condition. I turned to friends and family and educated them about HAE, as well as how they could help me, whether it be with an infusion, or just simply offering a hand to hold! I kept encouraging and pushing myself to live a normal life, not letting HAE stop me from doing the things I love like playing varsity lacrosse or snowboarding in the winter. My diagnosis has allowed me to become more in tune with my body, and my triggers. Stress, physical exertion, and menstruation are all big triggers for me, and this knowledge allows me to do what I can to prevent attacks.

Having access to HAE medication drastically decreased the frequency of my attacks. I was finally able to infuse within minutes of feeling symptoms of an attack and recover quickly. I went from having attacks once a week, to once a month, and now once every 6 months. Treating preventively, as well as immediately after symptoms arise, is very important and can greatly decrease the number and severity of attacks and improve the quality of your life.

FAQ

FREQUENTLY ASKED QUESTIONS

Answered by Dr. Raffi Tachdjian

QUESTIONS FROM A CHILD OR TEEN'S PERSPECTIVE

What adults in my life might I need to talk to about my HAE?

Talking to adults about your HAE is important. Adults in your life are in a position to support you if you have an attack and need help. These adults might be people at your school, including teachers, the school nurse, coaches, and counselors. Other adults to speak to might be a girl scout troop leader, your friends' parents, adults who run after-school programs, and anyone with whom you have regular contact. Having a conversation with them about your HAE helps them to be prepared to support you if you have an HAE attack.

What are some tips to help monitor my HAE so I can track attacks and triggers?

Keeping track of when attacks occur, where on the body you swell, and possible triggers can help you to better understand your HAE attacks. You can keep a journal or use the Advance HAE mobile app to monitor and document your attacks. You can also chart your menstrual cycles and see if they correlate with attacks and not feeling your best. Bring that information to your next HAE check-up visit, as it may be very important.

How might puberty and menstruation affect HAE symptoms and what should I expect during this time?

Hormones can play a role in the symptoms and severity of HAE. Puberty often causes increased (or first) symptoms of swelling to occur. Puberty comes with many changes. It is important to embrace the changes and

challenges that happen during the transition to adulthood. Menstruation may lead to more frequent and severe attacks in women. If this becomes a burden impacting quality of life, a conversation with the treating physician would be advised.

Is genital swelling common?

The location of swells on the body can be different for everyone. Some people never experience genital swelling, while others experience swelling in this area more frequently. Genital swelling can be the first sign of HAE for some people. This can be scrotal swelling for boys and labial swelling for girls. It is often due to trauma to the area (i.e., riding a bicycle) but can occur without trauma.

How should I address having HAE with my friends?

Some of my patients say they have special blood. Educating your friends will make everyone understand and help you on your journey.

How should I manage HAE and extracurricular/physical activities?

Be active and live life to the fullest. Talk to your physician about developing a plan to maximize your activities in a safe and progressive manner. Remember, an active and healthy lifestyle approach for your mind, body, and soul generally yields excellent outcomes. You should be smiling on the inside and on the outside.

QUESTIONS FROM A PARENT OR ADULT FEMALE PERSPECTIVE

What should an emergency plan look like for my child? What should I prepare in advance with my child's physician?

Make sure you have the medical records that include diagnosis, labs, and contact information for your child's HAE specialist printed and/or stored on a flash drive (even on a necklace, if available). Have a good access line (phone number) to your specialist so that they can speak to emergency and other hospital personnel should your child be transported urgently to a health care facility that is not familiar with your child or HAE. Ensure that you and your child are frequently following up with the HAE specialist to be sure the most appropriate and up-to-date therapy is in place.

HAE and Birth Control - What do I need to know? What kind of birth control options should I discuss with my doctor?

Speak to your physician about the type of birth control method you plan on using. For many patients, estrogen-based hormones can increase the rate and severity of HAE swelling bouts.

Is there a connection between Urinary Tract Infections and HAE?

As with any infection or inflammation, these can trigger HAE attacks.

How should I approach my desire to try different treatment options with my physician?

Always discuss upcoming therapies and see if your current regimen is working for you. This is why regular visits and follow-ups with your physician can be helpful, if not life-changing. If we stop to think and reflect, it hasn't been long since the dawn of modern HAE therapy. The treatments are always evolving and advancing, and so is our knowledge of our patients and this condition. Open and frequent communication between you and your physician is as important as ever. Remember that your HAE attacks and triggers/stressors in life may change from month to month or year to year. If nothing else, a visit sooner rather than later can update you on the latest treatment options that may be best suited for you.

Do you have any stress management tips?

Introduce nature and natural means of stress management. This can include anything from a hike to a walk by the water (river/ocean), or low-impact mindful activities such as meditation, music, and other methods that promote deep relaxation.

Resources

FOR PARENTS OF CHILDREN WITH HAE

YOU, YOUR CHILD, AND 504 PLANS

What is a 504 plan?

The "504" in "504 plan" refers to Section 504 of the Rehabilitation Act and the Americans with Disabilities Act, which specifies that no one with a disability can be excluded from participating in federally funded programs or activities, including elementary, secondary, or postsecondary education. "Disability" in this context refers to a "physical or mental impairment which substantially limits one or more major life activities." This can include physical impairments; illnesses or injuries; communicable diseases; chronic conditions like HAE, asthma, allergies and diabetes; and learning disabilities.

A 504 plan spells out the modifications and accommodations that might be needed for a child with HAE if the condition is preventing him/her from the opportunity to perform at the same level as their peers. Accommodations might include such things as an extra set of textbooks, home instruction, or technology to support learning.

It is important to note that policies and procedures for 504 plans may vary by state, and even among individual school districts. Parents and/or caregivers of children who need a 504 plan should contact the child's school district to find out who they should work with to get their 504 plan initiated. Determining eligibility for services under section 504 is a team decision. Team members often include teachers, school administrators, school nurses, school psychologists, counselors, therapists, the parent and/or caregiver, and the child, if appropriate. You should insist that the child's physician has significant input into the team's decision-making process.

How does a 504 plan differ from an IEP?

It is important to know the difference between a 504 plan and an Individualized Education Plan (IEP). An IEP is a comprehensive document that (1) serves as a blueprint or roadmap for a child with special education needs, and (2) specifies what a child will be learning in school.

A 504 plan deals specifically with how a child will be learning within the school. A 504 plan is issued to students who are able to participate in a general education classroom but need unique accommodations.

Are 504 plans really necessary?

The kind of accommodations and modifications offered by a 504 plan is often similar to what you may have worked out privately with your child's school or teacher in the past. Drafting an outline in a legal document may seem like too much trouble, but individual schools and their policies vary by district and state, so it is always best to put your plan in writing. A new teacher, new principal, new superintendent, or a move to a new school or town can render all your handshake agreements invalid. Having a legally binding plan lets everyone know what's to be done and how to go about it while promoting consistency and accountability.

SAMPLE SCHOOL NURSE LETTER

School name:

Date:

Dear School Nurse,

My child, _____, has been diagnosed with a rare and potentially life-threatening genetic disease called Hereditary Angioedema (HAE).

HAE symptoms include episodes of swelling (edema) in body parts, including the hands, feet, face, and throat. In addition, abdominal swelling can be accompanied by nausea and vomiting, and excruciating pain. Airway swelling is particularly dangerous and can lead to death by suffocation. HAE related swelling is not caused by an allergic reaction and, therefore, does not respond to antihistamines or corticosteroids. HAE attacks often occur spontaneously and can be triggered by many things – including stress, minor trauma, a cold, or flu.

It is important to call 911 immediately if my child reports any signs of swelling in the face, mouth, or throat. This requires emergency medical treatment to ensure that my child's airway is not compromised. In this situation, please also contact me as soon as possible.

Detailed information about HAE can be found at www.haea.org.

My child, _____, may recognize signs of HAE swelling and they may need to be excused from school. _____ is currently under the care of Dr. _____ who can be reached during office hours at _____. Attached is the current treatment plan for _____ that their doctor has developed. My child is treated with _____ for their HAE attacks. Please call me at _____ if you have any questions.

Signed _____ Relationship to Student _____

Attachments:
Prescribing physicians treatment letter
Medication informational brochure
HAE informational flyer

My Notes:

My Notes:


She BELIEVED,
she COULD,
so she DID!

HAEA CHILDREN'S BOOK SERIES

The HAEA partnered with children's author, Caryn Sonberg Seiler, to develop a three-book series that helps kids learn about HAE in a relatable and interesting way. These books are designed for kids with HAE and their siblings who are just learning about the condition. Parents and kids alike will enjoy reading these books together. Sharing these books with local communities is also a wonderful way to help spread awareness.

To learn more about the HAEA Children's Book Series and how to order books for your family, please visit: www.haea.org.

What parents are saying about the HAEA Children's Book Series:

"As a four-year-old with HAE, my daughter doesn't know another child that is going through what she goes through. Nico felt relatable to her and gave her the feeling that she wasn't alone in her experiences!"

"I think the books are a great tool to help open the conversation with kids and the people in their lives about HAE. It has helped spark questions in my child and even his cousins that I never even thought to address before."

HAEA CARES PROGRAM

The HAEA is always thinking of our youngest community members. We know that learning to cope with HAE isn't easy, and presents unique challenges. To help kids manage their condition we developed personalized support kits that we mail directly to them.

HAEA Cares Kits are available for children, teens, and young adults who reside in the United States and have a confirmed HAE diagnosis. One kit per person and the recipient must be a member of the HAEA to qualify.

Contact the HAEA at www.haea.org for more information.

THE BRADY CLUB

The Brady Club is a series of interactive programs and content developed exclusively for children diagnosed with HAE, their siblings, and children who have diagnosed parents. The Brady Club helps young people with HAE to better understand, manage, and cope with their condition while offering fun ways for them to feel inspired, empowered, and connected to other kids in the community. Joining is FREE, and once registered, kids benefit from exciting membership perks including a quarterly activity book, access to themed webinars and meet-ups, and much more!

Register for the Brady Club by visiting: https://brady.haea.org/.

My Notes:

BELIEVE YOU CAN AND YOU'RE HALFWAY THERE

FAMILY PLANNING & PREGNANCY

PREGNANCY AND FAMILY PLANNING WITH HAE

Planning to have a family and going through pregnancy is an exciting time in life. For women with HAE, it is important to understand how the condition could affect your pregnancy. Developing a plan, as well as working with your OBGYN and HAE treating physician, will establish the basis for a healthier and happier pregnancy.

Just like the symptoms of HAE, each pregnancy experience can be different. The good news is that there are a variety of effective treatments that can reduce or eliminate any pregnancy-related HAE symptoms. We hope the information in this section is useful as you plan for a new addition to your family.

COMMUNICATION IS KEY

Talk to your HAE treating physician, as well as your OBGYN, about establishing an HAE treatment plan. Ensure ongoing and open communication between your HAE treating physician and OBGYN.

GENETIC IMPLICATIONS

HAE is an inherited condition and each baby born to a parent with HAE has a 50 percent chance of inheriting the condition. You may wish to speak to your HAE treating physician if you have specific questions about the genetics of HAE.

FERTILITY THERAPIES

Couples struggling to conceive might be considering fertility treatment options. Treatment-related fluctuations in the female hormone estrogen can have an influence on HAE symptoms. It is important that women choosing to pursue fertility treatments discuss whether changes may be needed to their current HAE treatment plan.

DURING PREGNANCY

Like HAE, no two pregnancy experiences are the same. Although HAE attacks can fluctuate in frequency or severity during pregnancy, thankfully, there are FDA-approved therapies available that can help you throughout pregnancy, delivery, and postpartum.

BE PREPARED FOR HAE SYMPTOMS

While some women do not experience any HAE attacks while pregnant, others report an increase in their frequency and/or severity during this time of hormonal change. It is important to pay attention to the early signs of HAE symptoms so you can quickly administer treatment and limit the severity of an attack as soon as swelling is recognized. Keep in mind that during pregnancy, treatment with anabolic steroids (also known as androgens) such as Danazol, Oxandrolone, and Stanozolol is not recommended.

SECURE YOUR MEDICATIONS

Ensure you have an acute HAE treatment available at the hospital where you plan to deliver. You should also secure any additional treatments you may need (acute and prophylactic) for post-delivery, and after you are discharged.

POSTPARTUM CONSIDERATIONS

Keep Medications Handy

HAE attacks are rare at the time of delivery, but there is some indication that an increased frequency and severity of attacks is possible postpartum. Ensure you have enough medication on hand in the weeks and months following delivery while your body is undergoing hormonal changes.

Breastfeeding

Some HAE medications are safe and effective treatments to take while breastfeeding. If you are considering breastfeeding your child, speak with your physician about which treatment options may be right for you.

Testing Your Child for HAE

While you may be eager to learn whether your child also has HAE, it is generally recommended that you wait until your baby is at least one year old to test for HAE to ensure a more accurate test result.

My Notes:

Lisa's Story

"I was determined to be proactive
in my care and treatment to ensure that
I was in control during this time,
and that my HAE was well managed."
-Lisa

A FIRST TIME MOM'S EXPERIENCE MANAGING HER HAE WHILE PREGNANT

Like many young girls growing up with HAE, I often worried about how the condition might impact my ability to have children. I was concerned about what this would mean for me and for my baby.

Like many women, my pregnancy came as a surprise, so I didn't have much time to prepare in advance with my HAE treating physician. Once my pregnancy was confirmed, I knew I needed to set up a meeting with my HAE physician to talk about how hormonal changes during pregnancy might affect HAE symptoms, and to establish a treatment plan. It was going to be important to find an OBGYN willing to learn about the condition and establish an open line of communication with my HAE treating physician.

I met with my HAE treating physician to discuss how pregnancy might affect the frequency of my attacks. He informed me that HAE attacks in pregnant women vary. The frequency decreases or remains the same in some while others experience an increase. He emphasized that if I noticed changes in attack frequency, I should notify him immediately and he would adjust the medication to accommodate my individual experience.

As I entered my second trimester, I started to notice my attacks were becoming more frequent so I contacted my physician. He adjusted my medication to ensure that I had enough to prevent and treat attacks. I found that treating immediately when I felt an attack coming on was key to managing my HAE during this time. Although I was having more attacks, I felt I was able to successfully control HAE symptoms during the entire course of the pregnancy. I accepted the fact that I was having more attacks and made the decision that I was going to do whatever it took, even if that meant infusing more frequently, to prevent attacks from becoming severe.

Finding the right OBGYN was another very important step. After choosing an OBGYN, during the first visit I informed her that I had HAE, and connected her with my HAE treating physician. Even though I was her first HAE patient, I knew she was the right OBGYN because as soon as I told her about my HAE, she stepped out of the room, researched the condition, then came back to ask me questions. Following that first visit, we

worked to build a successful partnership between the OBGYN, HAE treating physician, and me. We prepared for the birth of my child by taking the necessary steps to document my condition at the hospital where I would deliver, developed a plan in the event I needed a cesarean section, and coordinated access to my medication during the hospital stay.

All of this planning proved necessary when I arrived at the hospital to deliver my son. I was in labor for 36 hours and eventually decided to move forward with a cesarean section delivery. Fortunately, I did not experience HAE-related swelling during labor. Once I decided to move forward with delivery via cesarean section, the anesthesiologist wanted to ensure my HAE was not going to cause complications during surgery. At this point, the letter from my HAE treating physician became important. It stated that I was well-managed and had medication available to treat an attack, if necessary. The letter also provided a direct phone number to my HAE treating physician who was standing by to answer any questions. Ultimately, the surgery went well. I did not experience any HAE-related swelling and gave birth to a beautiful 10-pound baby boy. Upon returning home, I continued with my treatment plan and scheduled a follow-up meeting with my HAE treating physician to discuss my treatment and dosing.

I could not have asked for a better experience. Despite my HAE attacks increasing in frequency during pregnancy, I was determined to be proactive in my care and treatment to ensure that my HAE was well managed. All the time and effort that went into planning ahead and preparing a plan paid off, and I now have a beautiful and healthy baby boy.

FAQ

FREQUENTLY ASKED QUESTIONS

Answered by Dr. Sandra Christiansen

Should my HAE diagnosis influence my decision to have children?

Having children is a very personal decision, however, there have been significant advances in HAE treatment options over the last decade, with several promising new therapies on the horizon. What I can say is that at this point, symptoms are well controlled for most people in the HAE community.

What medications would I want to use to treat my HAE while pregnant?

While there have been no clinical trials to test the safety of HAE medications on pregnant women and their babies, there is a long history of pregnant women both in the US and Europe who have used plasma-derived C1-Inhibitor replacement therapies during pregnancy. You should speak with your HAE physician to determine which C1-Inhibitor therapy would be best for you.

Medications to specifically avoid during pregnancy include all forms of androgens such as Danazol, Stanozolol, and Oxandrolone, which are relatives of the male hormone testosterone. These medications could affect fetal development and should be avoided if you are planning to have a baby, and stopped immediately if you discover that you are pregnant.

Does having HAE classify my pregnancy as “high risk”?

It is usually the OBGYN and their team who make the determination as to whether or not a pregnancy is considered high risk. OBGYN physicians, however, will often classify patients with HAE as ‘high risk’ due to

the extra considerations that need to be addressed during pregnancy. This is nothing to feel alarmed about. It just means that health care professionals are paying special attention to you and your baby to ensure a complication-free delivery.

What if I need to have an unexpected cesarean section?

It is recommended that you have a C1-Inhibitor intravenous injection prior to the surgery to ensure that your C1-Inhibitor levels are high enough to protect you from HAE swelling. It is also important to have several doses of medication on hand in the event swelling does occur after the surgery. This question also underscores the importance of discussing plans in advance with your OBGYN and HAE specialists so you are confident that everyone will be prepared.

Will being pregnant affect my HAE symptoms?

Research has shown that there is no way to predict how your HAE symptoms might change during pregnancy. It is possible that your symptoms could get better, they could get worse, or they could stay the same. Adjustments to your medications can be made if your pregnancy causes changes in your HAE symptoms.

Does the gender of the baby influence the severity of HAE while pregnant?

There is no evidence to suggest that the baby's gender will have any impact on the frequency and severity of HAE symptoms.

I've already had one child, and I am pregnant with my second. Can I expect my HAE symptoms to be similar to my first pregnancy?

It has been widely reported that each pregnancy experience is different. While someone may have little to no change in their attack frequency during their first pregnancy, there can be variation in frequency and severity of symptoms during subsequent pregnancies.

Are there resources or tools available to help me track my attacks and HAE symptoms during pregnancy?

The HAEA has a mobile app "Advance HAE" that is available on Android and IOS compatible devices which provides tools for tracking attack frequency and symptoms. Pregnant women are encouraged to join the Advance HAE Scientific Registry.

You can sign up for the Scientific Registry by visiting:

https://www.haea.org/form/registry. By joining the HAEA's Scientific Registry, you can provide researchers with information needed to unlock the remaining mysteries of HAE, and find innovative therapies and potential cures.

What should I expect and/or prepare for during the delivery of my child?

The majority of women report that everything goes quite well during delivery. As noted earlier, it is important to have several doses of medication on hand in the event that HAE-related swelling does occur. It is also important to maintain vigilance after the delivery or postpartum period when women with HAE could have increased HAE attacks.

What is the chance of passing HAE on to my child?

If either parent has HAE Type I or Type II (C1-Inhibitor deficiency) there is a 50/50 chance of the condition being passed to their child.

Scientists suspect that the inheritance pattern for HAE with Normal C1-Inhibitor is similar to what is seen in HAE Types I and Type II, but research is ongoing.

At what age is it recommended to have my child tested for HAE?

It is recommended that all children in your family be tested for HAE as early as the first year of life. It is important to know whether your child has HAE so you can be prepared in the event of an attack. For HAE-C1-INH (type I & type II), this involves simple blood tests and can usually be ordered at the pediatrician's office, measuring levels of C4, C1-Inhibitor (quantitative), and C1-Inhibitor (functional). There is currently little information regarding testing children for HAE with Normal C1-INH.

How do I talk about HAE with my OBGYN?

It is important that you establish a communications pathway between your HAE treating physician and OBGYN. Doing so will ensure that questions are answered and that the physicians work together to create a detailed treatment plan in place.

Your treatment plan should:

- Ensure everyone is aware of medication administration procedures and that the medication is available at all times,

- Address how the person with HAE will access medication while in the care of the hospital (will it be brought with them or will they need to use the pharmacy at the hospital?), and

- Confirm there is a plan in case a cesarean section is performed (address any questions/concerns that the anesthesiologist might have).

What should I expect postpartum?

Recovery from either a vaginal or cesarean section delivery could trigger HAE symptoms, so it is important to treat at the onset of any swelling that is suspected to be HAE-related. As we discussed, while the actual birth usually goes well, it is during postpartum that many women might have increased issues with HAE symptoms.

Should I expect any changes in my HAE while breastfeeding?

This is another area where we need more information. While not common, some women have indicated they have experienced more HAE symptoms during breastfeeding. It is recommended that you continue with C1-INH derived therapies to treat or prevent HAE attacks while breastfeeding -- please discuss optimizing your treatment with your HAE specialist.

My Notes:

Resources

FOR FAMILY PLANNING AND PREGNANCY

COORDINATING CARE FOR PREGNANT WOMEN WITH HAE

On the next page, you will find tips to create an individualized HAE treatment plan for your pregnancy. There are studies published on FDA-approved HAE therapies and pregnancy that you may wish to read and discuss with your physician. Talk with your OBGYN about having a treatment available where you are to deliver.

MY PREGNANCY JOURNAL

Track your pregnancy experience. Note your water intake, attack frequency, and any changes that you notice throughout your pregnancy and postpartum experience.

> Taking care of yourself makes you stronger for everyone in your life... including you."
>
> – Kelly Rudolph

MY PREGNANCY CHECKLIST

- ☐ Inform your OBGYN that you have HAE
- ☐ Establish communications between your health care team (HAE treating physician and OBGYN)
- ☐ Work with your health care team to create a treatment plan that covers all aspects of your pregnancy including HAE treatments for emergencies and any symptoms occurring while you're at the hospital
- ☐ Use this journal to keep track of your HAE symptoms during Pregnancy

One month - 1-4 weeks

My Notes:

Two months - 5-8 weeks

My Notes:

Three months - 9-13 weeks

SECOND TRIMESTER (PREPARING FOR DELIVERY):

- ☐ Obtain a letter from the HAE treating physician or OBGYN noting treatment procedures in the event of a cesarean or other necessary procedures

Four months - 14-17 weeks

My Notes:

Five months - 18-21 weeks

My Notes:

Six months - 22-26 weeks

THIRD TRIMESTER:

- ☐ Have a printed letter outlining the treatment plan with the contact information of both the OBGYN and the HAE treating physician

Seven months - 27-30 weeks

My Notes:

Eight months - 31-35 weeks

My Notes:

Nine months - 36-40 weeks

My Post-Partum Notes:

My Notes:

THE BIG SECRET
IN LIFE IS...

THAT THERE IS NO SECRET

MENOPAUSE & AGING

MENOPAUSE AND AGING

Menopause typically results in a decrease in estrogen production that can significantly affect a woman's HAE symptoms. Physicians often prescribe estrogen replacement therapy for menopausal women. It is important to discuss the risks and benefits of hormone replacement with your HAE treating physician.

My Notes:

Troyce's Story

"Developing a plan for the symptoms you might experience, as well as possible side effects can not only help you deal with these symptoms, but also look at all the options available for you."

-Troyce

A WOMAN'S EXPERIENCE NAVIGATING MENOPAUSE WHILE MANAGING HER HAE

As I began transitioning into menopause, I had discussions with my HAE treating physician and OBGYN regarding my concerns about intimacy and symptoms commonly experienced by other women such as hot flashes, mood swings, and vaginal dryness. When menopause began, we decided against hormone replacement therapy because it contains estrogen, which was an attack trigger when I was younger. Fortunately, I was glad to discover that the frequency of HAE attacks did not increase with menopause. Even though I am on a highly effective preventive medicine that controls HAE, my doctor and I have decided not to take the risk associated with hormone replacement therapy. Instead, I have developed other ways to deal with my menopause symptoms.

I encourage women approaching menopause to discuss this change in life with your OBGYN and HAE treating physician. They can help you with a plan for dealing with menopause symptoms while controlling your HAE. It is important that you have a conversation with your spouse/partner to help them understand menopause and enlist their support as you enter into this phase of your life. I found that open communication helps with the stress that can be associated with this new stage in life.

FAQ

FREQUENTLY ASKED QUESTIONS

Answered by Dr. Marc Riedl

I'm approaching the age of menopause. How might this affect my HAE?

Studies investigating the effect of menopause on HAE symptoms have shown mixed results. The largest published study to date showed that following menopause, HAE symptoms improved in 13% of women, remained the same in 55%, and worsened in 32%. Another large HAE center has reported experience that after menopause 50% of women improved but 15% worsened, in some cases with severe symptoms. HAE is highly variable and unpredictable during menopause. Therefore, it's important to follow up with your HAE treating physician to adjust and, when necessary, fine tune your HAE management plan.

Can I use hormone replacement during menopause?

Systemic (oral or injected) medications containing estrogen are known to increase angioedema symptoms in most women (up to 80%) with HAE. The effect of topical estrogen-containing medications (patches or lotions) on HAE has not been studied in detail, however, extreme caution should be taken with any estrogen-containing medication. Avoidance of estrogen treatments is generally recommended during menopause and the use of other non-estrogen treatments or strategies is encouraged to treat symptoms of menopause.

Can I take hormone replacement therapy if I'm on an HAE preventive treatment?

The answer to this question is unknown because this topic has not been adequately studied. Estrogen-containing medications are known to increase the frequency of HAE attacks, therefore, extreme caution should be taken if estrogen treatment is introduced.

What can I do to help with menopause symptoms if I cannot take estrogen replacement?

Treatment options for symptoms associated with menopause should be carefully discussed with your gynecologist or primary care physician and HAE treating physician. As noted earlier, estrogen medications are generally best avoided. The data on transdermal or topical estrogens is less clear, with a few reports of these formulations being tolerated in some women with HAE, but worsening symptoms in others. Thus, while certain women may tolerate transdermal or topical estrogen treatment, such measures should be undertaken with extreme caution. Progestin-only medications (without estrogen) are beneficial for some menopausal symptoms and can sometimes have a preventative effect for HAE symptoms. Non-hormonal medications have also shown benefit in managing symptoms of menopause. These include selective serotonin reuptake inhibitors (SSRIs) such as paroxetine or citalopram, serotonin-norepinephrine reuptake inhibitors (SNRIs) such as venlafaxine, desvenlafaxine, gabapentin, clonidine, and oxybutynin. Given the numerous options, it is important to discuss the treatment of menopausal symptoms in detail with your health care team.

What can I expect if I have to get a hysterectomy?

There are two primary HAE-specific issues to consider with hysterectomy. The first is the risk of the surgical procedure triggering an HAE attack as this can occur with surgical trauma or manipulation of the airway if general anesthesia is used. The surgical and anesthesia team should be aware of the HAE diagnosis and have a management plan for HAE in collaboration with your HAE treating physician. The second issue relates to any long-term hormonal effects of the surgery. This is dependent on whether the ovaries are removed (oophorectomy) at the same time as the uterus (hysterectomy). The decision about removal of the ovaries is

an important issue to discuss with your gynecologist/surgeon, as there are potential long-term health risks and benefits. Removal of the ovaries will essentially cause menopause by reducing estrogen production. While this may influence HAE symptoms, the clinical effect is unpredictable and unreliable. Hysterectomy/oophorectomy is not currently recommended as a treatment approach for HAE, and these procedures should only be done for other medical reasons.

Should I change/decrease my C1-INH replacement if the symptoms decrease during menopause?

Adjustment of any HAE medications, including C1-INH replacement, should be discussed and made in collaboration with your HAE treating physician. Such changes are generally based on the clinical course of HAE symptoms, as well as the quality of life factors and potential adverse effects of the medication. Often, adjustments to treatment are important during different life phases or events, particularly with long-term prophylactic regimens. Menopause is a time when HAE symptoms may improve or worsen due to the influence of hormonal changes so it may be very reasonable to consider adjustments to the treatment plan. Make sure to discuss this with your HAE treating physician to ensure this is done as safely as possible.

Things to consider when switching to Medicare

Women with HAE nearing 65 years old and older, should review available benefits and coverage for your HAE therapies under Medicare/Medicare Advantage plans. For those already on Medicare, open enrollment provides an opportunity to look at alternatives if you are unhappy with your current plan.

Resources

FOR MENOPAUSE AND AGING

Research your options ahead of time to ensure that you meet the open enrollment deadline and can select the best coverage. It is important to note that if you miss the deadline, you risk having no insurance for the entire year, affecting your access to healthcare and HAE therapies.

For more information visit the following websites:

Healthcare.gov

Medicare.gov

The compassionate and caring HAE Advocates at the HAEA are available to assist you. Contact us for a consultation or with any questions regarding insurance coverage of HAE therapies, and other important aspects of your healthcare plan by visiting www.haea.org.

My Notes:

My Notes:

US HEREDITARY ANGIOEDEMA ASSOCIATION

The HAEA is an advocacy and research organization committed to actively engaging our community in a wide variety of grassroots activities that promote HAE education and awareness. We provide personalized services to address the unique needs of people with HAE and their families, which includes helping them secure access to and reimbursement for modern HAE medicines. Our great success in supporting clinical research has resulted in a variety of FDA-approved therapeutic options. We work closely with expert physicians to continuously upgrade our community's quality of life through improving diagnosis and knowledge of the condition, and encourage an individualized approach to selecting an optimal treatment. The HAEA is product and company neutral, and continues to enthusiastically support drug discovery research aimed at the next generation of HAE therapies.

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