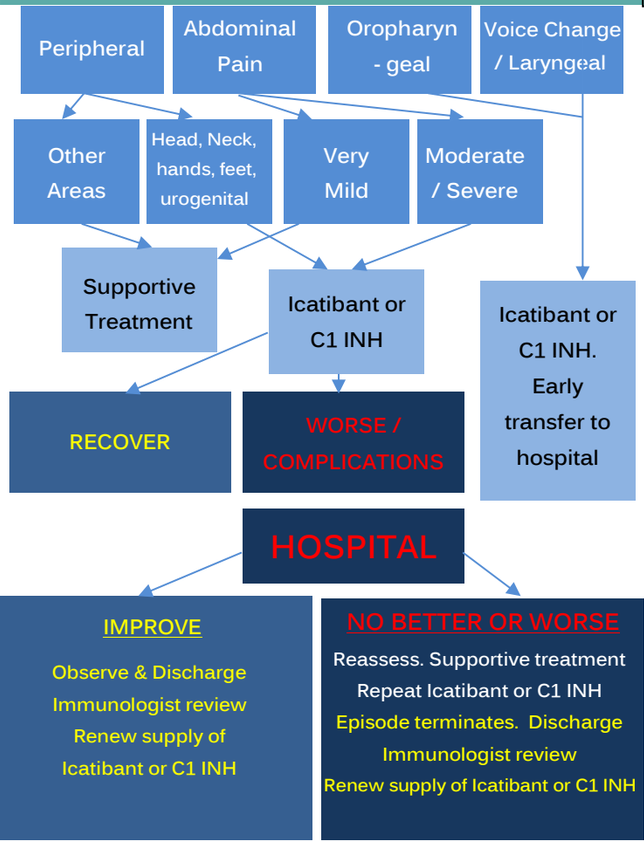
HAE Attack Algorithm



ASCIA Algorithm for Management of HAE in Australia and New Zealand

Clinical Presentations

HAE DOES NOT RESPOND TO TREATMENT WITH: ADRENALINE, ANTIHISTAMINE, CORTICOSTEROIDS

- Areas affected by oedema may be one or more of the following:
  - upper airway
  - face, mouth, tongue
  - genitalia
  - gastrointestinal tract
  - hands, arms
  - feet, legs
  - trunk
- Oedema without urticaria that is not itchy or pitting
- Symptoms of upper airway swelling: difficulty breathing, difficulty swallowing, changes to voice.
- Symptoms of gastrointestinal tract oedema: abdominal pain, abdominal distention, nausea, vomiting, dehydration, diarrhoea or constipation.
- May be prodromal indicators (tingling, non-itchy rash)
- Takes 24 hrs to peak & resolving over: 48 – 72hrs
- Unpredictable in onset

Triggers

Often there are no apparent triggers. However, the following may trigger an oedema attack:

- Stress
- Infection
- Injury
- Dental & other Surgeries
- Oestrogens
- ACE Inhibitors

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HAE Australasia
hope • advocacy • education
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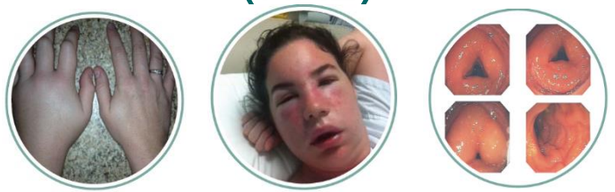
Reference for this Brochure:

Katelaris C, Smith W, Wong M, Jordan A. Position Paper on Hereditary Angioedema (HAE). *Australasian Society of Clinical Immunology and Allergy (ASCIA).* Revised 2017

Hereditary Angioedema (HAE)



Information for:

Patients, Carers, Families
Physicians treating HAE Patients

Hereditary Angioedema

Hereditary Angioedema (HAE) is a rare, autosomal dominant condition. HAE has been described in 3 types (see below). Without sufficient levels of C1 Inhibitor (C1 INH), excessive production of bradykinin occurs, which results in subcutaneous and submucosal oedema. Patients with HAE display symptoms of recurrent oedema of the limbs, trunk, face, genitals, gastrointestinal tract or upper airway (which, if left untreated, may cause death by asphyxiation). The oedema is without uticaria, does not itch and is non-pitting.

HAE Type 1	Approx. 85% of HAE patients	Low antigenic & functional C1 INH levels.
HAE Type 2	Approx. 15% of HAE patients	Normal antigenic C1 INH. Low functional C1 INH
HAE Type 3 *	Very rare cases	Normal C1 INH antigenic & functional levels. Symptoms are similar to HAE Type 1 & 2

* HAE Type 3 is outside the scope of this brochure. Please refer to the ASCIA HAE Position Paper for information

Rate of Population

Estimates of the occurrence of HAE range from 1 in 10,000 to 1 in 150,000 people with approximately the same frequency in men and women. Based on the current quoted number of cases in other populations, there could be up to 480 cases in Australia and 90 in New Zealand.

More than 50% of patients experience first symptoms of HAE before age of 10. 35% of patients experience first episodes in their teenage years and 7% occur in the first year of life. Historically, average time to diagnosis has been 13 to 21 years but with increasing awareness there has been improvement in this delay in diagnosis.

Testing & Diagnosis

To determine if a patient has HAE, C4 level is the initial screening test performed followed by C1 INH level and function which may be done at the same time if there is a high index of suspicion. If results indicate HAE, a repeat test needs to be done for confirmation. If a diagnosis is made, family members of that HAE patient should also be tested. See ASCIA HAE Position Paper for discussion on genetic testing.

Special Circumstances

Surgical & Dental Procedures

There is an increased risk of potentially life-threatening airway oedema with procedures that involve the mouth or laryngopharyngeal area (including intubation). Planning and consultation between immunologist, surgical teams and anaesthetis should occur prior to surgical and dental procedures where possible. Patients undergoing dental procedures need to ensure that they have prompt access to emergency treatments in case of failure of prophylaxis.

Travel

It is advisable that travelling HAE patients carry:
- Medical bracelet, ASCIA Action Plan and Physicians Letter
- Prescriptions for supplies of prophylactic medications
- Supply of C1 INH or Icatibant (with letter for airport controls)

Ensure legality of medications in destination country is checked.

Pregnancy

Published cases of pregnant HAE patients have demonstrated patients may experience increased HAE symptoms, decreased HAE symptoms, or no change. Some studies show decreased incidence of attacks in the second and third trimesters.

Childbirth: Even though trauma is considered a trigger of HAE attacks, attacks at the time of delivery are rare.

Post-Partum: Australian studies have found approx. 57% of HAE patients saw an increase in frequency and severity of attacks post-partum compared to pre-pregnancy.

Treatments

Patients should discuss treatments with their physician. Refer to product information for further information and side effects

	Purified C1 INH	Icatibant	Danazol	Tranexamic Acid
Brand Name	Berinert ® Cinryze ®	Firazyr ®	Azol ®	Cyklokapron ®
Adminis-tration	Intravenous	Pre-filled syringe for sub-cutaneous injection	Tablet form	Tablet form or injectable
At Home Use	In appropriate cases after adequate training	In appropriate cases after adequate training		
Special Notes	Preferable for use during pregnancy, breastfeeding & for children	Used for emergency treatment of acute episodes in HAE patients over 18	Contra-indicated in pregnancy/ breastfeeding	Lack of efficacy when compared to C1 Inh & Icatibant
For Prophylaxis	Short Term: Prior to Surgical or dental procedures Long Term (Berinert) Available in Aust.	-	Long or short term prophylaxis (adj dose for short term)	Long or short term prophylaxis, but not preferred
Other Info	Safe & well tolerated	Safe & well tolerated	Use limited by side effects	Use limited by side effects
Access	Aust & NZ: Berinert is subsidised for specific uses in Type 1 & Type 2	Subsidised in Aust & NZ	Subsidised in Aust & NZ	Subsidised in Aust & NZ

Note: Cinryze ®is registered but not reimbursed in Australia