# When is a swelling not an allergy?

HAE think

## Hereditary angioedema (HAE) is a rare disease that can easily be missed in an emergency setting.

If someone presents with a swelling, it is typically thought to be a histamine-mediated allergic response and, as a result, patients may inadvertently be treated and managed incorrectly.[1-3] In the worst cases, where laryngeal swelling is concerned, there is a risk of potentially fatal outcomes.[1,4] Due to its rarity, one of the barriers to progress in the management and treatment of HAE is lack of experience and knowledge.[1,5] The information below will help you to better identify the signs and symptoms of HAE in your patients.

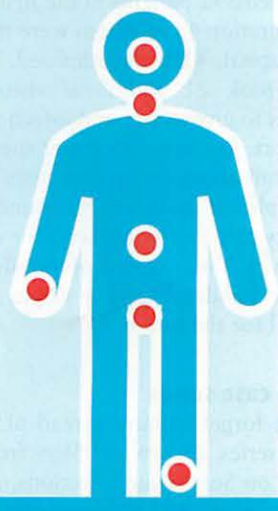
## What is HAE?

HAE is a rare autosomal dominant disorder resulting from C1-esterase inhibitor (C1-INH) deficiency[1] with an estimated prevalence of between 1:10,000 to 1:50,000.[1,3,6]

Characterised by recurrent attacks of localised, non-pruritic subcutaneous or mucosal oedema,[7] HAE occurs most commonly on the face, abdomen, urogenital area and extremities with individual attacks varying in location, frequency, duration and severity.[7-9] Abdominal attacks may be accompanied by severe pain, nausea, vomiting, diarrhoea and constipation.[7,10] Around 50% of patients will have at least one laryngeal attack risking life-threatening airway obstruction.[7,11]

**Unlike histamine-mediated angioedema, HAE is not associated with:**[2,12,13]

- **Food allergens**
- **Insect bites or stings**
- **Latex exposure**
- **Side effect of medication**



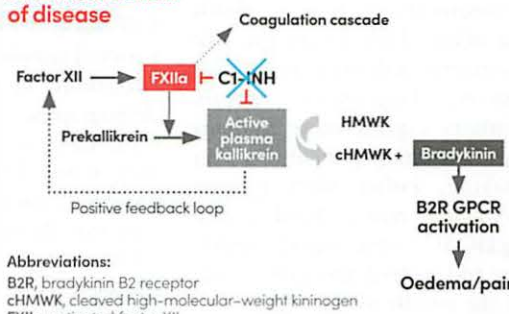
Patients may experience prodromal symptoms before attack onset, with symptoms intensifying over a 12- to 24-hour period, sometimes spreading to other areas of the body, and then taking around 5 days to resolve.[7,10]

- **Mouth/Face**
- **Throat**
- **Abdomen**
- **Genitals**
- **Hands/Feet**

## HAE is mediated by bradykinin[2,14-16]

In the kallikrein-kinin pathway, deficient or dysfunctional C1-INH results in uncontrolled kallikrein activity, leading to increased generation of the vasoactive peptide bradykinin.[2,14-16]

### HAE mechanism of disease




**Abbreviations:**
B2R, bradykinin B2 receptor
cHMWK, cleaved high-molecular-weight kininogen
FXIIa, activated factor XII
GPCR, G protein-coupled receptor
HMWK, high-molecular-weight kininogen

Adapted from: Bernstein JA, et al. Int J Emerg Med 2017;10:15.
Kaplan A, Joseph K. Ann Allergy Asthma Immunol 2010;104:193–204.

> *"Many ED staff may be unaware of how to recognise or manage HAE and hospitals may not have specific medications or protocols in place in the department.[2] A greater awareness of the different pathophysiologic pathways that lead to angioedema will improve diagnosis and patient care."*[2,17]

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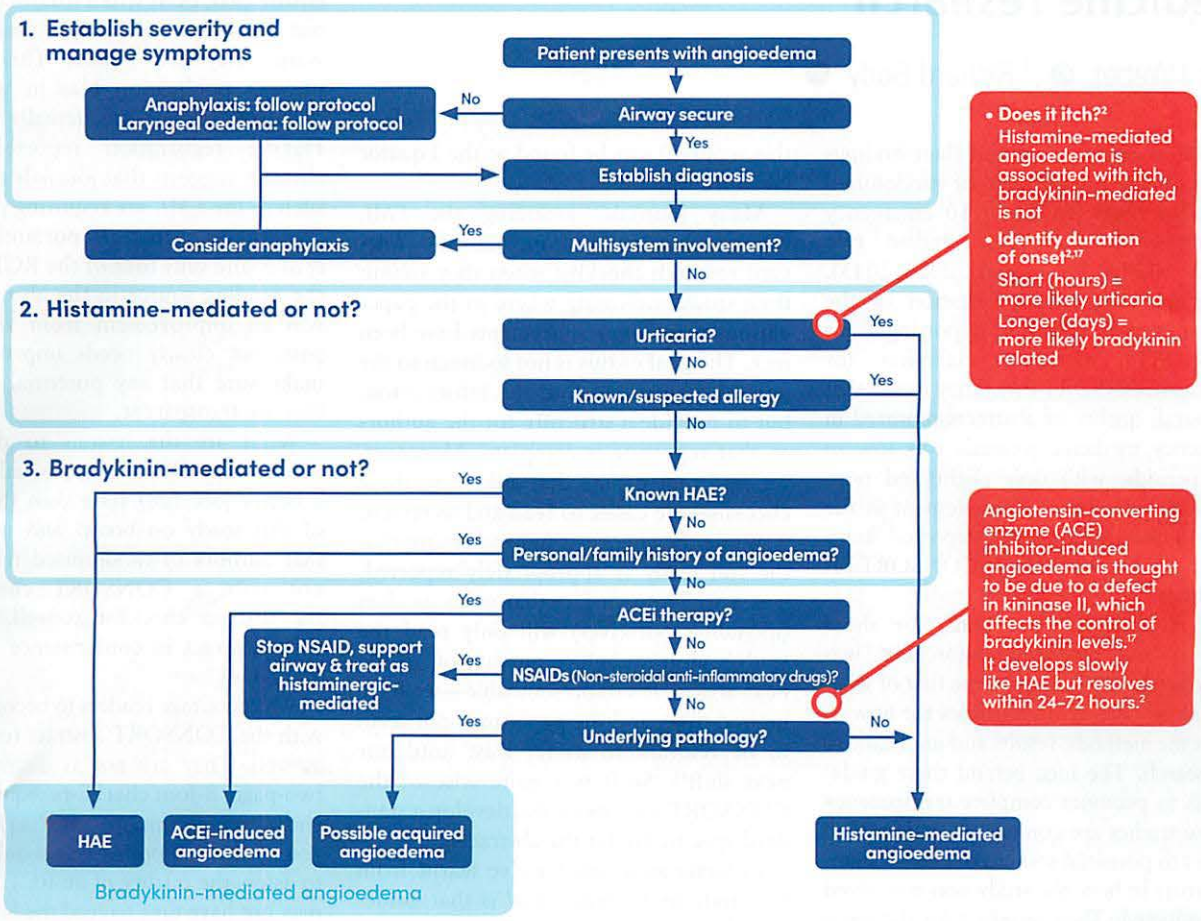
# Assessment of acute angioedema in an emergency setting

(adapted from Bernstein et al. 2017)[2]




## Meeting the need for information about HAE

**www.knowHAE.co.uk** has been launched to provide accurate information about the symptoms, treatment, and ongoing management of HAE. Alongside a range of resources for patients and caregivers, the website includes a separate section developed specifically for healthcare professionals working outside the field of immunology.

**Visit www.knowHAE.co.uk to find out more.**

knowHAE
awareness, answers, action


**References**

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