LRI Emergency Department

Clinical guideline for:

Hereditary angioedema (HAE): ED management of acute attacks in adults

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Background

Hereditary angioedema (HAE) is characterized by recurrent episodes of (often painful) swelling under an area of skin or mucous membrane, without urticaria. It is due to low circulating levels, absence or malfunction of the protein C1-esterase inhibitor (C1-INH). Patients most commonly experience asymmetric swelling of hands, feet, eyelids, lips, and/or genitals but the mucosa of the upper respiratory and gastrointestinal tract can also be affected.

Laryngeal attacks are rare but a medical emergency and may cause fatal airway obstruction.

A small group of patients (around 50) with known HAE live within the catchment area of our Trust. Most are adults and have been provided with two pre-filled syringes of icatibant (a bradykinin B2 receptor antagonist) at home to treat their acute attacks. This has markedly reduced ED attendances.

Our HAE patients have been educated to attend the emergency department if they experience laryngeal or facial angioedema, or angioedema not responding to icatibant. They will usually carry a letter from the UHL immunology department with details of their disease and the required treatment.

Emergency management in the ED

Clinical presentation of HAE shares features with that of anaphylaxis but features usually develop much more slowly.

Emergency replacement of C1-INH is usually effective. One of the three C1-INH products licensed in the UK will be stocked in the ER treatment room at any one time, depending on availability.
NB: Dosing is specific for each product – further details below.

In patients with signs of airway compromise (such as stridor, dyspnea, hoarseness or reduced SpO_2), request anaesthetic and ENT support immediately. Unless there is rapid improvement following C1-INH administration, early fiberoptic intubation should be considered.

NB: The procedure should be undertaken jointly by an experienced anaesthetist and ENT surgeon. Failed intubation attempts or other instrumentation may worsen airway compromise; the team therefore has to be ready to proceed to a surgical airway (‘front of neck access’).

Please note that peripheral and gastrointestinal attacks can be very painful: Give effective analgesia.

Berinert ®

Recommended dose is 20units/kg (rounded to the nearest 500 units), given by slow IV injection:

Weight	Dose	Rate of injection
< 38kg	500units (1 vial)	2.5min
38kg - 62kg	1000units (2 vials)	5min
63kg - 87kg	1500units (3 vials)	7.5min
> 87kg	2000units (4 vials)	10min

Cinryze ®

Give by IV injection over 10 minutes (rate = 1mL/min):

Weight	Dose
All patients	1000units (2 vials)

A second dose may be given if inadequate response after 60mins.

Further details on the administration of Berinert and Cinryze are available on Medusa (type 'berinert' or 'cinryze' into the search box and click 'show monograph').

Ruconest ®

Contraindicated in those with a rabbit allergy.

After reconstitution, concentration is 150units/mL. Give by IV injection over 5 minutes:

Weight	Dose	Volume (mL)
< 84kg	50units/kg	kg body weight divided by 3
84kg and above	4200units	28 (= 2 full vials)

A second dose may be given if inadequate response after 120min.

No information on Medusa – see Summary of Product Characteristics (SPC) on EMC for more details.

Notes on icatibant

Our ED does not stock icatibant, but patients arriving with their own supply can also be managed with icatibant instead of C1-INH. It is administered by slow subcutaneous injection over several minutes. A second injection can be given after six hours if clinical response to the first dose is inadequate, followed if required by a third after a further six hours. **DO NOT** give more than 3 doses within 24h.

Disposition following ED emergency management

Rarely, patients whose symptoms are not resolving as expected despite appropriate emergency management will require admission to AMU/ACB or ICU. Otherwise, disposition should be as follows:

Peripheral swellings	Discharge once symptoms resolving
Abdominal symptoms	Discharge once pain completely resolved & other symptoms resolving (consider admission to EDU using the consultant-only or EPIC pathway as applicable if more time is needed for symptoms to settle)
Head and neck including laryngeal swelling	Admit to EDU using consultant-only or EPIC pathway as applicable. Observe for at least 6h; discharge only once symptoms fully resolved.