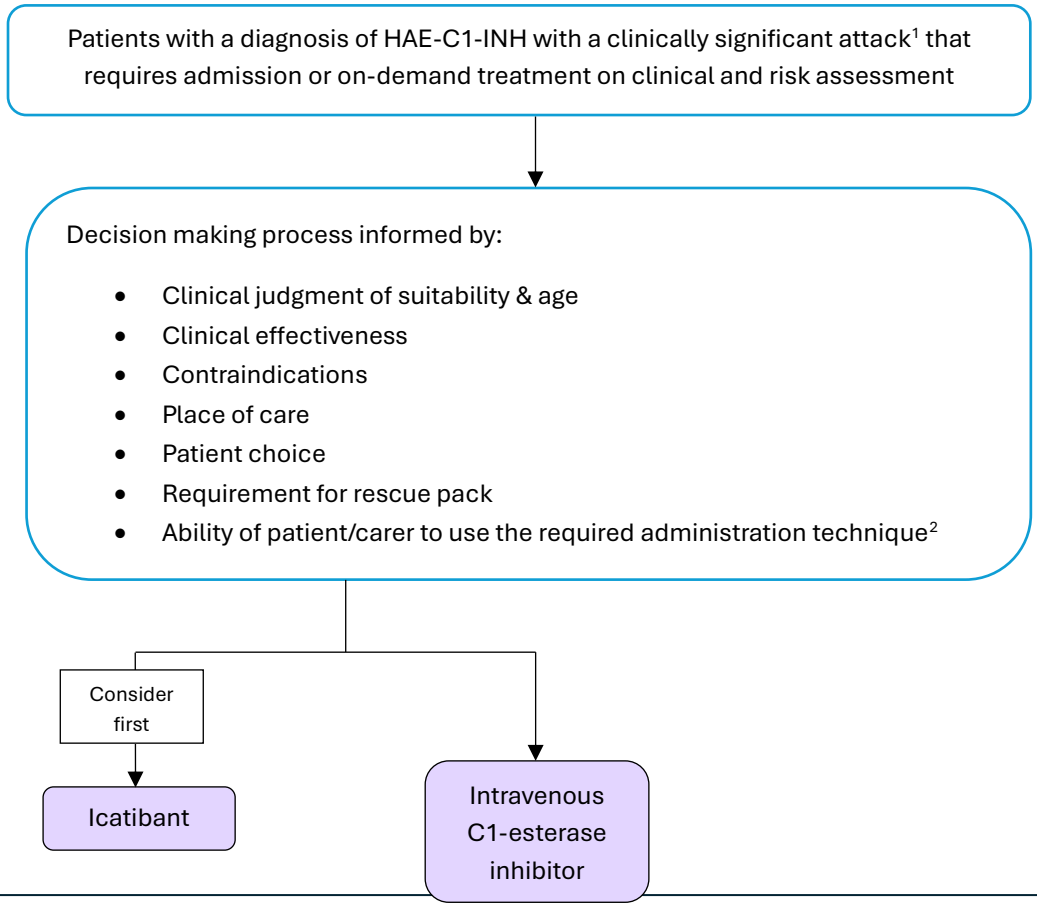
April 2026 update

Commissioned treatment options for patients with hereditary angioedema secondary to C1 esterase inhibitor deficiency (HAE-C1-INH)

- This algorithm provides a framework to aid decision-making for angioedema specialists and patients
- The algorithm is informed by the regulatory status, NICE technology appraisal (TA) guidance and NHS England (NHSE) clinical commissioning policies. Relevant clinical commissioning policies/TAs should be consulted for further details.
- All patients with a diagnosis of HAE-C1-INH should be under the care of specialised immunology centres as outlined in the service specification. HAE-C1-INH is classified as per the IUIS Phenotypical Classification.
- For special circumstances including pregnancy and lactation, please refer to individual product Summary of Product Characteristics (SmPC). Please note that high-dose (2000IU and 3000IU) formulations of subcutaneous C1-esterase inhibitors (C1-INH) may be considered in pregnancy and breastfeeding.
- Where plasma products are used, patients need to be consented to potential risks associated with these products.
- This algorithm is not intended to guide management during critical events including airway threatening or life-threatening emergencies.

On demand treatments

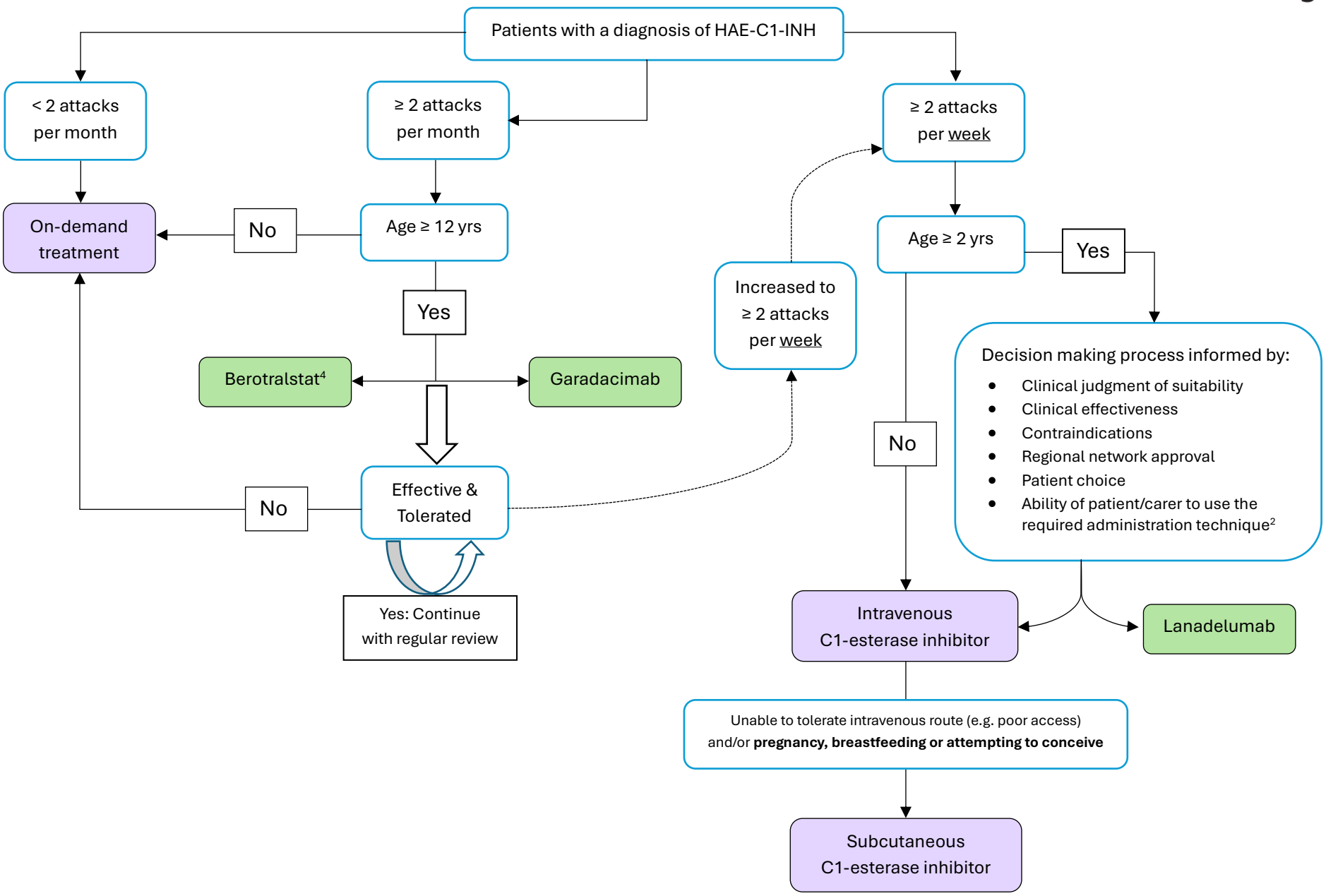


Pre-procedure prophylaxis: Patients with HAE-C1-INH are eligible for prophylaxis with C1-esterase inhibitor before undergoing dental, medical, obstetric or surgical procedures

1. A clinically significant attack is defined as either i) potentially life threatening because it affects the head or neck or ii) causes pain or disability such that the patient cannot continue their normal activities. Frequency should be calculated over a period of at least 56 days.
2. This includes securing venous access for C1-esterase inhibitors, and ability to reconstitute doses from multiple vials.
3. Some adult patients are treated with androgens as oral prophylactic treatment. However, evidence is limited and accessing treatment is difficult so this is not recommended as first-line for patients newly started on prophylaxis. Where existing patients are established on androgen therapy, this may continue if considered clinically appropriate; if established patients do cease treatment with androgen therapy then review the need for any prophylaxis. An individualised assessment to withdraw androgens and commence new prophylaxis should be taken. If a historical attack frequency is documented, it can be used as the basis for selecting other prophylaxis treatment options.
4. Berotralstat should be stopped if, after 3 months of treatment, attack frequency has not reduced by at least 50% compared to baseline.
5. Some patients, including children under 12, are treated with tranexamic acid however evidence is limited.

April 2026 update

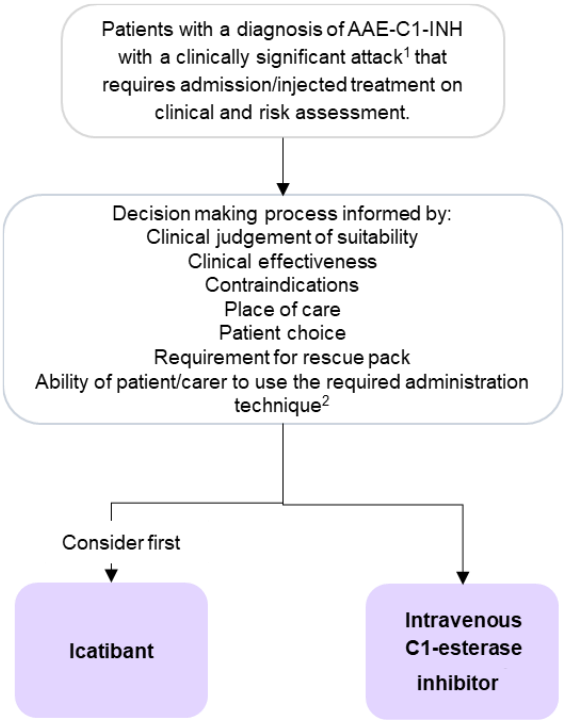
Long-term prophylactic treatment



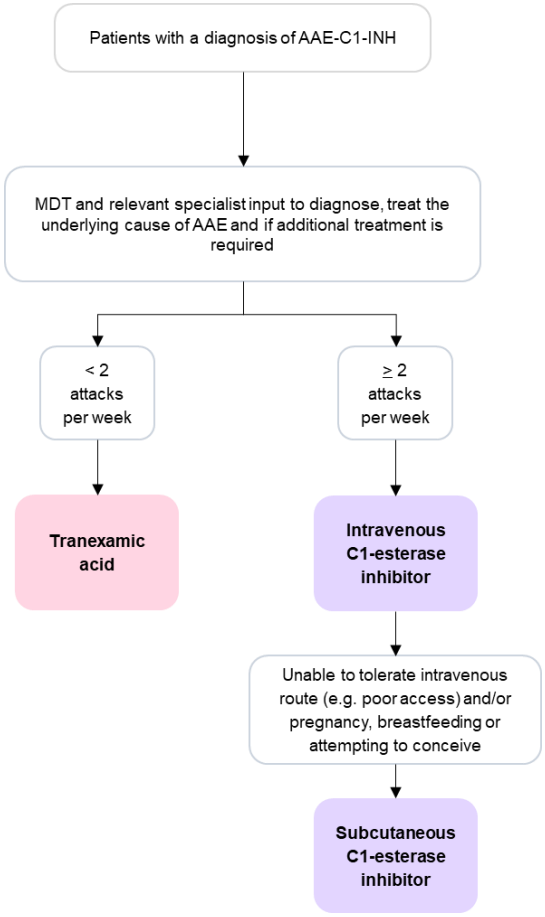
Commissioned treatment options for patients with acquired angioedema secondary to C1 esterase inhibitor deficiency (AAE-C1-INH)

- This algorithm provides a framework to aid decision-making for angioedema specialists and patients
- The algorithm is informed by the regulatory status, NICE technology appraisal (TA) guidance and NHS England (NHSE) clinical commissioning policies. Relevant clinical commissioning policies/TAs should be consulted for further details.
- All patients with a diagnosis of AAE-C1-INH should be under the care of specialised immunology centres as outlined in the service specification. AAE-C1-INH is classified as per the IUIS Phenotypical Classification.
- For special circumstances including pregnancy and lactation, please refer to individual product Summary of Product Characteristics (SmPC). Please note that, high-dose (2000IU and 3000IU) formulations of subcutaneous C1-esterase inhibitors (C1-INH) may be considered in pregnancy and breastfeeding.
- Where plasma products are used, patients need to be consented to potential risks associated with these products.
- This algorithm is not intended to guide management during critical events including airway-threatening or life-threatening emergencies.

On demand treatments



Long-term prophylactic treatment



Pre-procedure prophylaxis: Patients with HAE-C1-INH are eligible for prophylaxis with C1-esterase inhibitor before undergoing dental, medical, obstetric or surgical procedures

1. A clinically significant attack is defined as either i) potentially life threatening because it affects the head or neck or ii) causes pain or disability such that the patient cannot continue their normal activities. Frequency should be calculated over a period of at least 56 days.
2. This includes securing venous access for C1-esterase inhibitors and ability to reconstitute doses from multiple vials
3. Some adult patients are treated with androgens as oral prophylactic treatment. However, evidence is limited and accessing treatment is difficult so this is not recommended as first-line for patients newly started on prophylaxis. Where existing patients are established on androgen therapy, this may continue if considered clinically appropriate; if established patients do cease treatment with androgen therapy then review the need for any prophylaxis. An individualised assessment to withdraw androgens and commence new prophylaxis should be taken.
   If a historical attack frequency is documented, it can be used as the basis for selecting other prophylaxis treatment options.

Key: NHSE Policy | NICE TA | In tariff/ NHSE Policy

Commissioning position at time of publication

The algorithm describes the key criteria for accessing commissioned treatments; the source of commissioning is colour coded in the algorithm (see the key below). Whilst the algorithm will be periodically updated, users are advised to confirm the latest version of any cross-referenced policy or NICE technology appraisal guidance, and to familiarise with each summary of product characteristics for dosing, adverse effects, contra-indications and other useful information on each medicine (www.medicines.org.uk/emc).

Three of the commissioned treatments are recommended in NICE Technology Appraisal Guidance:

- Lanadelumab for preventing recurrent attacks of hereditary angioedema (Technology Appraisal [TA606] 2019)
- Berotralstat for preventing recurrent attacks of hereditary angioedema (Technology Appraisal [TA738] 2021)
- Garadacimab for preventing recurrent attacks of hereditary angioedema in people 12 years and over (Technology Appraisal [TA1101] 2025)

NHS England has published two clinical commissioning policies relating directly to HAE and one general policy of relevance to prescribing for children.

- Clinical commissioning policy: treatment of acute attacks in hereditary angioedema (adult) (April 2013)
- Clinical commissioning policy: intravenous (6 years and above) and subcutaneous (adolescents and adults) C1-esterase inhibitor injections for routine prophylaxis of hereditary angioedema (HAE) type I and II
- Commissioning medicines for children in specialised services (March 2024 update)

Prior approval funding request requirements (known as Blueteq)

Drug	Initiation of treatment	Continuation (review period)	Medicines for Children
Berotralstat	BER2_ver1.0 Age ≥12 yrs	3 months and then every 12 months	No. Standard form for age ≥12 yrs
Garadacimab	GAR1_v1.0 Age ≥12 yrs	3 months and then every 12 months	No. Standard form for age ≥12 yrs
Icatibant	-	-	Age 2 to 17 yrs, HAE and AAE
Lanadelumab	Age ≥12 yrs	3 months and then every 12 months	Age 2 to 11 yrs, initiation and continuation forms
C1-Esterase Inhibitor (Berinert or Cinzryze)	Separate forms for age ≥18 yrs and age 2 to 17 yrs	2 months and then every 12 months	Children 2 to 5 yrs commissioned via Medicines for Children policy, included in ≤17 form

If not listed in this table there is no prior approval requirement at time of publication.