




**Expert Physicians Discuss
GUIDELINE FOCUSED
CARE IN HEREDITARY
ANGIOEDEMA**




US HAEA Medical Advisory Board 2020 Guidelines for the Management of Hereditary Angioedema
Available at: <https://www.sciencedirect.com/science/article/pii/S2213219820308783?via%3Dihub>


Jointly provided by the Postgraduate Institute for Medicine, US Hereditary Angioedema Association (HAEA), and RMEI Medical Education, LLC.

 Postgraduate Institute for Medicine  HAEA US HEREDITARY ANGIOEDEMA ASSOCIATION  RMEI MEDICAL EDUCATION FOR BETTER OUTCOMES

1

Learning Objectives

-  Summarize important considerations in prophylactic treatment for HAE-C1INH, including recent therapeutic advances and patient specific factors
-  Discuss the diagnostic and treatment approaches for patients with HAE-nl-C1INH
-  Develop individualized management plans, including logistics, education, and monitoring for HAE special patient populations

 HAEA US HEREDITARY ANGIOEDEMA ASSOCIATION

2

Long-term Prophylaxis for HAE: When and How to Use in the Management Plan








3

Hereditary Angioedema: Long-term Prophylaxis

? Who should be on long-term prophylaxis (LTP)?

Based on the needs of individual patients':

-  Treatment preferences
-  Attack frequency and severity
-  Comorbid conditions
-  Access to emergent treatment
-  Quality of life



4

Hereditary Angioedema: Long-term Prophylaxis

- LTP decisions cannot be made based on rigid criteria
- Initiation or continuation of LTP periodically reviewed/discussed due to changes in condition, life circumstances, and patient preference



5

Hereditary Angioedema: Long-term Prophylaxis

Treatment options for LTP in HAE-C1INH



First-line Medications

- C1INH Concentrates
 - SQ pdC1INH (Haegarda®)
 - IV pdC1INH (Cinryze®)
- Lanadelumab
 - SQ plasma kallikrein inhibitor (Takhzyro®)
- Berotralstat
 - Oral plasma kallikrein inhibitor (Orladeyo™)



6

Hereditary Angioedema: Long-term Prophylaxis

Treatment options for LTP in HAE-C1INH



Second-line Medications

- Attenuated Androgens
 - Danazol, stanozolol, oxandrolone
- Antifibrinolytics
 - Tranexamic acid, aminocaproic acid



7

Hereditary Angioedema: Long-term Prophylaxis



Treatment considerations when developing LTP management plan

- Efficacy
- Safety including potential long-term effects
- Tolerability, burden of treatment, adherence
- Special considerations (pregnancy, lactation)



Shared Decision Making



8

HAE-nI-C1INH: How to Diagnose and Treat



9

HAE-nI-C1INH: Diagnosis

- Lack of available diagnostic tests
 - Genetic: FXII, PLG, KNG, ANGPT, MYOF
 - Biochemical: Investigational assays
- Challenge of avoiding overdiagnosis or underdiagnosis
 - Similarities to HAE-C1INH, some differences (female predominance, variable penetrance, and estrogen sensitivity)



10



HAE-nl-C1INH: Diagnosis

- Clinical criteria
 - Recurrent angioedema in absence of urticaria
 - Exclusion of causative medication
 - Normal C1INH testing
 - Identified HAE-nl-C1INH mutation OR documented resistance to mast-cell targeted preventative therapy plus family history



11

HAE-nl-C1INH: Diagnosis

- Other supportive data
 -  A history of rapid, durable response to a bradykinin-targeted medication
 -  Predominant documented visible angioedema; or with predominant abdominal symptoms, evidence of bowel wall edema documented by CT or MRI
- Requires close clinical follow-up and assessment with treatment
- Evaluation/diagnosis by an experienced angioedema specialist is recommended

12

HAE-nI-C1INH: Treatment

- Less treatment data compared to HAE-C1INH
- Acute treatment
 - Similar approach to HAE-C1INH; assess for reliable, consistent benefit of medication
- Prophylactic treatment
 - Differences in approach based on available data
 - Tranexamic acid
 - Progestins



13

Enhancing QoL in HAE: Developing a Multidisciplinary Approach

14

Enhancing QoL in HAE



Patient Empowerment

- Shared decision making
- Frequent communication
- What is important to the patient? (treatment goals, work, school, family, leisure activities, and/or travel)



15

Enhancing QoL in HAE



Monitoring Outcomes

- Clinical follow-up with an expert HAE physician
- Patients record HAE symptoms and medication use
- Monitoring efficacy and safety of treatment plan
- Assessing impairment in daily activities from HAE symptoms or treatment burden
- Quality of life assessment (validated tools available)



16

Enhancing QoL in HAE


- Pharmacoeconomic considerations
- Economic costs
 - Direct and indirect
 - Immediate and long-term
- Cost and impact on family
- Self-administration management plans to reduce treatment costs
- Information and valuable resources available at www.HAEA.org
 - A template of a general letter of medical necessity developed at the UCSD US HAEA Angioedema Center is available in the Guidelines located here: <https://www.sciencedirect.com/science/article/pii/S2213219820308783?via%3Dihub>




17

Enhancing QoL in HAE

Recent study data on modern LTP treatments for HAE

 Clinically significant improvements in QoL

 Reduce direct and indirect costs related to on-demand only treatment

Castaldo AJ, et al. *Allergy Asthma Proc.* 2021 Feb 13. Online ahead of print.



18

Thank You for Participating

- In order to receive CME credit for this activity, please complete the evaluation.
- If you complete the evaluation, you will be entered into a drawing to win a \$100 Amazon gift card.*

*The expense for this gift card is solely funded by RMEI Medical Education, LLC. No supporter funding was used for the expense of this gift card.

