UnitedHealthcare Pharmacy
Clinical Pharmacy Programs

Program Number	2026 P 2148-9
Program	Prior Authorization/Medical Necessity
Medication	Haegarda® (C1 esterase inhibitor, human)
P&T Approval Date	7/2018, 7/2019, 6/2020, 3/2021, 3/2022, 3/2023, 3/2024, 3/2025, 3/2026
Effective Date	6/1/2026

1. Background:

Haegarda is a plasma-derived concentrate of C1 Esterase Inhibitor (Human) (C1-INH) indicated for routine prophylaxis to prevent hereditary angioedema (HAE) attacks in patients 6 years of age and older.

2. Coverage Criteria [a]:

A. Initial Authorization

1. **Haegarda** will be approved based on **all** of the following criteria:

   a. Diagnosis of hereditary angioedema (HAE) as confirmed by **one** of the following:

      (1) C1 inhibitor (C1-INH) deficiency or dysfunction (Type I or II HAE) as documented by **one** of the following (per laboratory standard):

         (a) C1-INH antigenic level below the lower limit of normal

         (b) C1-INH functional level below the lower limit of normal

-OR-

      (2) HAE with normal C1 inhibitor levels and **one** of the following:

         (a) Confirmed presence of variant(s) in the gene(s) for factor XII, angiopoietin-1, plasminogen-1, kininogen-1, myoferlin, and heparan sulfate-glucosamine 3-O-sulfotransferase 6

         (b) Recurring angioedema attacks that are refractory to high-dose antihistamines with confirmed family history of angioedema

         (c) Recurring angioedema attacks that are refractory to high-dose antihistamines with unknown background de-novo mutation(s) (i.e., no family history) (HAE-unknown)

-AND-

   b. Prescribed for the prophylaxis of HAE attacks

-AND-

c. Not used in combination with other products indicated for prophylaxis against HAE attacks (e.g., Andembry, Cinryze, Dawnzera, Orladeyo, Takhzyro)

-AND-

d. Prescriber attests that patient has experienced attacks of a severity and/or frequency such that they would clinically benefit from prophylactic therapy with Haegarda

-AND-

e. Prescribed by **one** of the following:

(1) Immunologist

(2) Allergist

Authorization will be issued for 12 months.

B. Reauthorization

1. **Haegarda** will be approved based on **all** of the following criteria:

a. Documentation of positive clinical response to **Haegarda** therapy

-AND-

b. Reduction in the utilization of on-demand therapies used for acute attacks (e.g., Berinert, Ekterly, Firazyr, icatibant, Ruconest) as determined by claims information, while on Haegarda therapy

-AND-

c. **Both** of the following:

(1) Prescribed for the prophylaxis of HAE attacks

-AND-

(2) Not used in combination with other products indicated for prophylaxis against HAE attacks (e.g., Andembry, Cinryze, Dawnzera, Orladeyo, Takhzyro)

-AND-

d. Prescribed by **one** of the following:

(1) Immunologist

(2) Allergist

Authorization will be issued for 12 months.

[a] State mandates may apply. Any federal regulatory requirements and the member specific benefit plan coverage may also impact coverage criteria. Other policies and utilization management programs may apply.

3. Additional Clinical Programs:

- Notwithstanding Coverage Criteria, UnitedHealthcare may approve initial and re-authorization based solely on previous claim/medication history, diagnosis codes (ICD-10) and/or claim logic. Use of automated approval and re-approval processes varies by program and/or therapeutic class.
- Supply limits may be in place.

4. References:

1. Haegarda [package insert]. Kankakee, IL: CSL Behring, LLC; January 2022.
2. Wu, E. Hereditary angioedema with normal C1 inhibitor. In: UpToDate, Saini, S (Ed), UpToDate, Waltham, MA, 2023.
3. Busse, P., Christiansen, S., Riedl, M., et. al. "US HAEA Medical Advisory Board 2020 Guidelines for the Management of Hereditary Angioedema." *The Journal of Allergy and Clinical Immunology.* 2020 September 05.
4. Maurer M, Magerl M, Betschel S, et al. The international WAO/EAACI guideline for the management of hereditary angioedema-The 2021 revision and update. Allergy. 2022;77(7):1961-1990. doi:10.1111/all.15214

Program	Prior Authorization/Medical Necessity - Haegarda (C1 esterase inhibitor, human)
Change Control	
7/2018	New program.
7/2019	Annual review. No changes made to program.
6/2020	Annual review. Aligned criteria with acute and prophylactic therapies.
3/2021	Added diagnosis criteria and aligned combination use language with prophylactic therapies. Updated references.
3/2022	Annual review. Updated background and references.
3/2023	Annual review. Updated combination use language with prophylactic therapies without change to clinical intent. Updated references.
3/2024	Annual review with update to diagnostic criteria for HAE with normal C1 inhibitor levels. Updated language for reauthorization criteria.
3/2025	Annual review. No changes to the clinical criteria.
3/2026	Annual review. Updated list of examples of prophylactic and acute HAE treatments. Updated references.