



PHARMACY POLICY STATEMENT North Carolina Marketplace

DRUG NAME	Haegarda (C1 esterase inhibitor (human))
BILLING CODE	NDC
BENEFIT TYPE	Pharmacy
SITE OF SERVICE ALLOWED	Home
STATUS	Prior Authorization Required

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. HAE is a rare autosomal dominant disease characterized by episodic unpredictable swelling, which can occur in a variety of anatomic locations. The swelling results from excess production of the vasodilator bradykinin. Attacks may be painful and cause functional impairment but are not associated with pruritis. The most common types of HAE are caused by deficiency (type 1) or dysfunction (type 2) of C1 inhibitor (C1-INH). Type 1 is the most prevalent.

Haegarda (C1 esterase inhibitor (human)) will be considered for coverage when the following criteria are met:

Hereditary Angioedema (HAE)

For **initial** authorization:

1. Member must be 6 years of age or older; AND
2. Medication must be prescribed by or in consultation with an allergist or immunologist; AND
3. Member has a diagnosis of HAE type I or type II confirmed by both of the following:
 - a) Low C4 level;
 - b) Low (<50% of normal) C1 inhibitor antigenic and/or functional level; AND
4. Chart notes must document the member's baseline frequency of HAE attacks; AND
5. Member is inadequately controlled with on-demand treatment alone; AND
6. Haegarda is being prescribed for ongoing prophylaxis and will not be used to treat acute attacks.
7. **Dosage allowed/Quantity limit:** 60 units/kg subQ twice weekly (every 3 or 4 days).
QL: 16 vials/28 days

If all the above requirements are met, the medication will be approved for 6 months.

For **reauthorization**:

1. Chart notes must be provided that show a reduced frequency or number of acute attacks since starting treatment.

If all the above requirements are met, the medication will be approved for an additional 12 months.



CareSource considers Haegarda (C1 esterase inhibitor (human)) not medically necessary for the treatment of conditions that are not listed in this document. For any other indication, please refer to the Off-Label policy.

DATE	ACTION/DESCRIPTION
08/25/2017	New policy for Haegarda created.
01/14/2021	Updated and revised all content; consistent with other HAE prophylactics. Added specific J code. Changed age limit to 6 per recent label change. Updated references. Greatly simplified the diagnostic confirmation criteria. Removed minimum required number of attacks, per guidelines; will just ask for baseline measure. Removed the statement about causative medications. Added that they must try on-demand treatment first. Rewrote the renewal criteria and removed log book requirement. Extended initial auth duration to 6 mo and renewal to 12 mo. Inserted the word “esterase” in front of “inhibitor” in the drug name.
07/05/2022	Transferred to new template. Updated references. Switched to pharmacy benefit.

References:

1. Haegarda (C1 Esterase Inhibitor [Human]) [prescribing information]. Kankakee, IL: CSL Behring LLC; 2022.
2. Longhurst H, Cicardi M, Craig T, et al. Prevention of Hereditary Angioedema Attacks with a Subcutaneous C1 Inhibitor. *N Engl J Med.* 2017;376(12):1131-1140.
3. Lumry W. Management and Prevention of Hereditary Angioedema Attacks. *Am J Manag Care.* 2013;19:S111-S118.
4. Lumry WR, Martinez-Saguer I, Yang WH, et al. Fixed-Dose Subcutaneous C1-Inhibitor Liquid for Prophylactic Treatment of C1-INH-HAE: SAHARA Randomized Study. *J Allergy Clin Immunol Pract.* 2019;7(5):1610-1618.e4. doi:10.1016/j.jaip.2019.01.021
5. Busse PJ, Christiansen SC, Riedl MA, et al. US HAEA Medical Advisory Board 2020 Guidelines for the Management of Hereditary Angioedema [published online ahead of print, 2020 Sep 6]. *J Allergy Clin Immunol Pract.* 2020;S2213-2198(20)30878-3. doi:10.1016/j.jaip.2020.08.046
6. Betschel S, Badiou J, Binkley K, et al. The International/Canadian Hereditary Angioedema Guideline [published correction appears in *Allergy Asthma Clin Immunol.* 2020 May 6;16:33]. *Allergy Asthma Clin Immunol.* 2019;15:72. Published 2019 Nov 25. doi:10.1186/s13223-019-0376-8
7. Maurer M, Magerl M, Betschel S, et al. The international WAO/EAACI guideline for the management of hereditary angioedema - The 2021 revision and update. *World Allergy Organ J.* 2022;15(3):100627. Published 2022 Apr 7. doi:10.1016/j.waojou.2022.100627

Effective date: 01/01/2023
Revised date: 07/05/2022