C1 esterase inhibitor [human] (Haegarda®)	**Place of Service** **Self-Administration** **HCPCS**: **J0599** per 10 units

Condition(s) listed in policy *(see criteria for details)*:
• Hereditary Angioedema (HAE), prophylaxis

AHFS therapeutic class: Blood product derivative
Mechanism of action: C1 esterase inhibitor (human)

(1) Special Instructions and Pertinent Information

Haegarda is managed under the outpatient Pharmacy Benefit for self-administration. Please contact the member's Pharmacy Benefit for information on how to obtain this drug.

To submit a request for Haegarda under the medical benefit, please submit clinical information for prior authorization review and include medical rationale why the patient cannot self-administer this drug in the home.

(2) Prior Authorization/Medical Review is required for the following condition(s)

All requests for Haegarda® (C1 esterase inhibitor [human]) must be sent for clinical review and receive authorization prior to drug administration or claim payment.

Hereditary angioedema (HAE), prophylaxis

1. Chart documentation for the clinical diagnosis of Type I or Type II Hereditary Angioedema (HAE), including serum C4 and C1-INH (antigenic or functional level) that are below the limits of the laboratory's normal reference range, **AND**
2. Patient has a history of frequent or severe attacks (i.e., an HAE attack at least once per month, a history of serious attacks with laryngeal/ upper airway involvement or attacks resulting in impaired daily living), **AND**
3. Not used in the combination with other HAE therapies for the prophylaxis of HAE attacks (e.g., Berinert, Cinryze, Orladey, Takhzyro)

Covered Doses
Up to 60 IU/kg SC twice weekly (every 3 or 4 days)

Coverage Period
Indefinite

ICD-10:
D84.1

(3) The following condition(s) DO NOT require Prior Authorization/Preservice

All requests for Haegarda® must be sent for clinical review and receive authorization prior to drug administration or claim payment.

(4) This Medication is NOT medically necessary for the following condition(s):

Coverage for a Non-FDA approved indication, requires that criteria outlined in Health and Safety Code § 1367.21, including objective evidence of efficacy and safety are met for the proposed indication.

Please refer to the Provider Manual and User Guide for more information.

(5) Additional Information

How supplied:

- 2000 IU lyophilized powder single-use vial
- 3000 IU lyophilized powder single-use vial

HAE Diagnosis[1,2]:

- HAE is an inherited autosomal disorder.
- ≥ 75% of patients with HAE report a family history of attacks.
  - A diagnosis of HAE (C1 INH deficiency) is suggested by a history of recurrent attacks of angioedema and abdominal pain. Swelling may affect the extremities, face, upper respiratory tract and gastrointestinal tract.
  - Typically swelling develops gradually over hours, increasing slowly for 12 – 36 hours and subsiding after 2 – 5 days. Urticaria is not a feature of HAE.
  - The onset of HAE attacks is in childhood or young adulthood and worsens around the time of puberty.
  - Attacks do not respond to antihistamines or corticosteroids.
  - Laboratory complement tests are used to confirm the diagnosis of HAE.
  - Virtually all patients with hereditary angioedema have a persistently low antigenic C4 level with normal antigenic C1 and C3 levels.
  - Measurement of C4 levels can rule out hereditary angioedema, although in rare cases, the C4 level is normal between attacks. Subsequent measurement of antigenic and functional C1-inhibitor levels confirms the diagnosis of hereditary angioedema and distinguishes between type I (low antigenic and functional C1-inhibitor levels) and type II (normal antigenic C1-inhibitor level but low functional C1-inhibitor activity).
  - In rare cases, patients with inherited angioedema have normal functional C1-inhibitor levels; some but not all of these patients are found to have a factor XII mutation.

Table 2. Diagnostic criteria for HAE [1,2]

Type of Angioedema	Laboratory Findings		
	C4 Level	C1 INH Level	Functional C1 INH Level
HAE – Type I	↓	↓	↓
HAE – Type II	↓	↔ or ↑	↓

Key: ↓ - decreased, ↑ - increased, ↔ - normal

(6) References

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1. Gompels MM, Lock RJ, Abinum M, et al. C1 inhibitor deficiency: consensus document. Clinical and Experimental Immunology. 2005; 139: 379 – 394.
2. Zuraw BL. Hereditary Angioedema. The New England Journal of Medicine. 2008; 359: 1027 –36.

(7) Policy Update

Date of last review: 1Q2022
Date of next review: 1Q2023
Changes from previous policy version:
No clinical change to policy following routine annual review.

BSC Drug Coverage Criteria to Determine Medical Necessity
Reviewed by P&T Committee