Effective date: 7/1/2019
Reviewed Date: 6/2019, 9/2020, 2/2021, 2/2022, 3/2023, 12/2023, 01/2024, 07/2025, 11/2025 Pharmacy Scope: Medicaid Medical Scope: Medicaid, Commercial, Medicare

HAEGARDA (C1 Esterase Inhibitor Subcutaneous [Human])

POLICY

I. INDICATIONS

The indications below including FDA-approved indications and compendial uses are considered a covered benefit provided that all the approval criteria are met and the member has no exclusions to the prescribed therapy.

FDA-Approved Indication
Routine prophylaxis to prevent Hereditary Angioedema (HAE) attacks in patients 6 years of age and older.

All other indications are considered experimental/investigational and are not a covered benefit.

II. CRITERIA FOR APPROVAL

Authorization for 6 months may be granted for prevention of hereditary angioedema attacks when all of the following criteria is met:

A. Member is ≥ 6 years of age.
B. Medication is prescribed by, or in consultation with an allergist/immunologist or a physician who specializes in the management of HAE.
C. Member has documented diagnosis of HAE type I or type II and meets one of the following (a or b):
    a. Documentation that the member has C1 inhibitor deficiency or dysfunction as confirmed by laboratory testing; and meets one of the following criteria:
        i. C1 inhibitor (C1-INH) antigenic level below the lower limit of normal as defined by the laboratory performing the test, or
        ii. Normal C1-INH antigenic level and a low C1-INH functional level (functional C1-INH less than 50% or C1-INH functional level below the lower limit of normal as defined by the laboratory performing the test); OR
    b. Documentation that the member has normal C1 inhibitor as confirmed by laboratory testing and meets one of the following criteria:
        i. Member has an F12, angiopoietin-1, plasminogen, or kininogen-1 (KNG1) gene, heparan sulfate-glucosamine 3-O-sulfotransferase 6 (HS3ST6), or myoferlin (MYOF) pathogenic variant as confirmed by genetic testing, or
        ii. Member has a documented family history of angioedema and the angioedema was refractory to a trial of high-dose antihistamine (e.g., cetirizine 40mg per day or the equivalent) for at least one month.
D. Other causes of angioedema have been ruled out (e.g., angiotensin-converting enzyme inhibitor [ACE-I] induced an angioedema, angioedema related to an estrogen containing drug, allergic angioedema).
E. Member requires long-term prophylactic treatment based on the provider's assessment of the patient's disease activity, quality of life, availability of health care resources, and/or failure to achieve adequate control by appropriate on-demand therapy [i.e., Ekterly (sebetralstat), Kalbitor(ecallantide), Icatibant, Ruconest (C1 esterase inhibitor) or Berinert (C1 esterase inhibitor), etc.]

F. Member will not use Haegarda (C1 esterase inhibitor), concomitantly with Takhzyro (lanadelumab-flyo), Andembry (garadacimab-gxii), Cinryze (C1 esterase inhibitor), Orladeyo (berotralstat) or Dawnzera (donidalorsen)

III. CONTINUATION OF THERAPY

Authorization of 6 months may be granted for continuation of therapy when all of the following criteria are met:

A. Member meets all criteria for initial approval; AND
B. Member has documentation of experiencing a significant reduction in frequency of attacks (e.g., ≥ 50%) since starting prophylactic treatment.; AND
C. Member has documentation of reduced the use of medications to treat acute attacks since starting prophylactic treatment.

V. QUANTITY LIMIT

Haegarda 2000 units or 3000 units: 20 vials per 30 days (daily dose of 0.667)

VII. DOSING AND ADMINISTRATION

Indication	**Dose**	**Maximum dose (1 billable unit = 10 IU)**
Prophylaxis of Hereditary Angioedema (HAE) attacks	60 IU/kg body weight injected subcutaneously twice weekly (every 3 or 4 days)	5,600 billable units per 28 days

The following HCPCS/CPT code is:

HCPCS/CPT Code	**Description**
J0599	Injection, c-1 esterase

VIII. REFERENCES

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3. Cicardi M, Bork K, Caballero T, et al. Hereditary Angioedema International Working Group. Evidence-based recommendations for the therapeutic management of angioedema owing to hereditary C1 inhibitor deficiency: consensus report of an International Working Group. *Allergy*. 2012;67:147-157.
4. Bowen T, Cicardi M, Farkas H, et al. 2010 International consensus algorithm for the diagnosis, therapy,

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