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

SPECIALTY GUIDELINE MANAGEMENT

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.

III. 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. Patient is ≥ 6 years of age.
B. Medication is prescribed by, or in consultation with an allergist/immunologist or a physician who specializes in the treatment of HAE or related disorders.
C. Patient has documented diagnosis of HAE type I or type II and meets one of the following (a or b):
    a. Documentation that the patient 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 patient has normal C1 inhibitor as confirmed by laboratory testing and meets one of the following criteria:
        i. Patient has an F12, angiopoietin-1, plasminogen, or kininogen-1 (KNG1) gene, heparan sulfate-glucosamine 3-O-sulfotransferase 6 (HS3ST6), or myoferlin (MYOF) gene mutation as confirmed by genetic testing mutation as confirmed by genetic testing, or
        ii. Patient has a documented family history of angioedema and the angioedema was refractory to a trial of high-dose antihistamine (e.g., cetirizine) 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. Patient is receiving treatment for one of the following:
    a. Short-term HAE prophylaxis prior to a procedure (i.e., dental, or medical procedure)
    b. Patient has a history of one of the following criteria for long term HAE prophylaxis
        i. History of at least one severe HAE attack per month (i.e., airway swelling, debilitating cutaneous or gastrointestinal episodes)

ii. Patient is disabled more than 5 days per month by HAE
iii. History of at least one laryngeal attack caused by HAE; AND
1. Treatment with "on demand" therapy (i.e., Ekterly, Kalbitor, Icatibant, Ruconest or Berinert) did not provide satisfactory control or access to on demand therapy is limited

F. Patient will not use Haegarda concomitantly with Andemry, Cinryze, Orladeyo, or Takhzyro.
G. Dose does not exceed FDA approved labeling.

IV. CONTINUATION OF THERAPY

Authorization of 6 months may be granted for continuation of therapy when all of the following criteria are met:
A. Patient meets all criteria for initial approval; AND
B. Patient has documentation of a favorable clinical response (i.e., decrease in HAE acute attack frequency, decrease in HAE attack severity, or decrease in duration of HAE attacks) since initiating Haegarda prophylactic therapy compared with baseline (i.e., prior to initiating prophylactic therapy).
C. Patient has documentation of reduced the use of medications to treat acute attacks since starting treatment.

VI. 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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4. Bowen T, Cicardi M, Farkas H, et al. 2010 International consensus algorithm for the diagnosis, therapy, and management of hereditary angioedema. *Allergy Asthma Clin Immunol*. 2010;6(1):24.
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