| Effective date: 7/1/2019 |
| --- |
| Reviewed Date: 6/2019, 9/2020, 2/2021, 2/2022, 3/2023, 12/2023, 3/2024<br>Pharmacy Scope: Medicaid<br>Medical Scope: Medicaid, Commercial, Medicare-Medicaid Plan (MMP) |

**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.

### II. DOCUMENTATION

Submission of the following information is necessary to initiate the prior authorization review:

A. For initial authorization, the following should be documented:
   1. C1 inhibitor functional and antigenic protein levels
   2. F12, angiopoietin-1, plasminogen, kininogen-1 (KNG1), heparan sulfate-glucosamine 3-O-sulfotransferase 6 (HS3ST6), or myoferlin (MYOF) gene mutation testing, if applicable
   3. Chart notes confirming family history of angioedema and the angioedema was refractory to a trial of high-dose antihistamine therapy, if applicable
B. For continuation of therapy, chart notes demonstrating a reduction in frequency of attacks

### 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. 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. 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, 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. Member 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 . Patient will not use Haegarda concomitantly with Cinryze, Orladeyo, or Takhzyro.

E. Dose does not exceed FDA approved labeling.

## III. 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 had 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 reduced the use of medications to treat acute attacks since starting treatment.

## IV. QUANTITY LIMIT

Haegarda 2000 units or 3000 units: 20 vials per 30 days

## V. 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 |

## IV. 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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12. Farkas H, Martinez-Saguer I, Bork K, et al. International consensus on the diagnosis and management of pediatric patients with hereditary angioedema with C1 inhibitor deficiency. *Allergy*. 2017;72(2):300-313.