Haegarda (C1 inhibitor (human)) is a preferred product and will only be considered for coverage under the medical benefit when the following criteria are met:

Members must be clinically diagnosed with one of the following disease states and meet their individual criteria as stated.

**HEREDITARY ANGIOEDEMA (HAE)**

For initial authorization:
1. Member must be 12 years of age or older, and medication is being used for routine prophylaxis to prevent HAE attacks (NOT for treatment of acquired angioedema); AND
2. Medication prescribed by or in consultation with a provider specializing in allergy, immunology, or hematology; AND
3. Member must have a confirmed diagnosis of HAE as one of the following:
   a) Type 1 HAE documented in chart notes with ALL of the following (Note: tests listed below must be repeated for confirmation of diagnosis):
      i) Low levels (below the limits of the laboratory’s normal reference range) of C4, C1-INH antigenic protein and C1-INH functional level; AND
      ii) Positive family history of angioedema OR earlier age of onset (before age 30) with normal C1q antigenic protein level;
   b) Type 2 HAE documented in chart notes with ALL of the following (Note: tests listed below must be repeated for confirmation of diagnosis):
      i) Normal or elevated level of C1-INH antigenic protein (as defined by performing lab); AND
      ii) Low level (below the limits of the laboratory’s normal reference range) C4 and C1-INH functional; AND
4. Documentation in medical chart of at least two attacks per month before treatment initiation; AND
5. Medication is not being used in combination with Cinryze; AND
6. Medications known to cause angioedema (i.e. ACE-Inhibitors, estrogens, angiotensin II receptor blockers) have been evaluated and discontinued when appropriate.
7. Dosage allowed: 60 units/kg of actual body weight twice weekly.

**Note:** Personal documentation (log book, journal, etc.) of medication use will be necessary for reauthorization. Prescribers should be aware and make their patients aware of this requirement for reauthorization.

*If member meets all the requirements listed above, the medication will be approved for 3 months.*
For **reauthorization**:  
1. Member must be in compliance with all other initial criteria; **AND**  
2. Chart notes have been provided that show the member's signs and symptoms of disease have improved and the number of acute attacks per month has decreased; **AND**  
3. Log of medication use supported by medical chart or by claims data has been provided.

*If member meets all the reauthorization requirements above, the medication will be approved for an additional 6 months.*

**CareSource considers Haegarda (C1 inhibitor (human)) not medically necessary for the treatment of the following disease states based on a lack of robust clinical controlled trials showing superior efficacy compared to currently available treatments:**

- Acquired angioedema (AAE)
- Treatment of acute HAE attacks

<table>
<thead>
<tr>
<th>DATE</th>
<th>ACTION/DESCRIPTION</th>
</tr>
</thead>
<tbody>
<tr>
<td>08/25/2017</td>
<td>New policy for Haegarda created.</td>
</tr>
</tbody>
</table>

**References:**

4. Haegarda (C1 Esterase Inhibitor [Human]) [prescribing information]. Kankakee, IL: CSL Behring LLC; June 2017.

Effective date: 09/08/2017  
Revised date: 08/25/2017