Cinryze is a C1 esterase inhibitor indicated for routine prophylaxis against angioedema attacks in adults, adolescents and pediatric patients (6 years of age and older) with Hereditary Angioedema (HAE). HAE is a rare autosomal dominant disease characterized by episodic unpredictable swelling, which can occur in a variety of anatomic locations. The swelling results from excess production of the vasodilator bradykinin. Attacks may be painful and cause functional impairment but are not associated with pruritis. The most common types of HAE are caused by deficiency (type 1) or dysfunction (type 2) of C1 inhibitor (C1-INH). Type 1 is the most prevalent.

Cinryze (C1 esterase inhibitor (human)) will be considered for coverage when the following criteria are met:

**Hereditary Angioedema (HAE)**

For **initial** authorization:
1. Member must be 6 years of age or older; AND
2. Medication must be prescribed by or in consultation with an allergist or immunologist; AND
3. Member has a diagnosis of HAE type I or type II confirmed by both of the following:
   a) Low C4 level;
   b) Low (<50% of normal) C1 inhibitor antigenic and/or functional level; AND
4. Chart notes must document the member’s baseline frequency of HAE attacks; AND
5. Member is inadequately controlled with on-demand treatment alone; AND
6. Cinryze is being prescribed for ongoing prophylaxis and will not be used to treat acute attacks; AND
7. Member has a trial and failure of or contraindication to Haegarda.
8. **Dosage allowed/Quantity limit:**
   - Age 12+: 1000 units IV infusion every 3 or 4 days; if response is inadequate, doses up to 2,500 units (not exceeding 100 units/kg) every 3 or 4 days may be considered.
   - Age 6-11: 500 units IV infusion every 3 or 4 days; if response is inadequate, doses up to 1000 units every 3 or 4 days may be considered.
   - QL: 20 vials (500 IU/vial) per 28 days

*If all the above requirements are met, the medication will be approved for 6 months.*

For **reauthorization**:
1. Chart notes must be provided that show a reduced frequency or number of acute attacks since starting treatment.

*If all the above requirements are met, the medication will be approved for an additional 12 months.*
CareSource considers Cinryze (C1 esterase inhibitor (human)) not medically necessary for the treatment of conditions that are not listed in this document. For any other indication, please refer to the Off-Label policy.

<table>
<thead>
<tr>
<th>DATE</th>
<th>ACTION/DESCRIPTION</th>
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</thead>
<tbody>
<tr>
<td>08/25/2017</td>
<td>New policy for Cinryze created. Criteria for each type of HAE specified. Criteria of documentation of attacks, discontinuation of meds that can cause HAE, and restriction on combinations with other meds for acute attacks were added.</td>
</tr>
<tr>
<td>07/27/2018</td>
<td>Medication is now approved for 6 years old and older.</td>
</tr>
<tr>
<td>01/14/2021</td>
<td>Updated and revised all content; consistent with other HAE prophylactics. Updated references. Greatly simplified the diagnostic confirmation criteria. Removed minimum required number of attacks, per guidelines; will just ask for baseline measure. Removed the statement about causative medications. Added that they must try on-demand treatment first. Rewrote the renewal criteria and removed log book requirement. Extended initial auth duration to 6 mo and renewal to 12 mo. Edited dosing information.</td>
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<tr>
<td>07/05/2022</td>
<td>Transferred to new template. Updated references.</td>
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</table>

References:


Effective date: 01/01/2023
Revised date: 07/05/2022