Policy Title: Ruconest (recombinant C1 esterase inhibitor) (Intravenous)

Department: PHA

Effective Date: 01/01/2020

Review Date: 12/20/2019, 1/22/20, 5/06/2021

Revision Date: 12/20/2019, 1/22/20, 5/06/2021

Purpose: To support safe, effective and appropriate use of Ruconest (recombinant C1 esterase inhibitor).

Scope: Medicaid, Commercial, Medicare-Medicaid Plan (MMP)

Policy Statement:
Ruconest (recombinant C1 esterase inhibitor) is covered under the Medical Benefit when used within the following guidelines. Use outside of these guidelines may result in non-payment unless approved under an exception process.

Procedure:
Coverage of Ruconest (recombinant C1 esterase inhibitor) will be reviewed prospectively via the prior authorization process based on criteria below.

Initial Criteria:
- Member is 13 years of age or older; AND
- Ruconest is being used for treatment of acute hereditary angioedema (HAE) attacks
- Patient has documented diagnosis of HAE type I or type II and meets one of the following:
  - Member has C1 inhibitor deficiency or dysfunction as confirmed by laboratory testing; and meets both of the following criteria:
    - Member has a C4 level below the lower limit of normal as defined by the laboratory performing the test, and
    - Member meets one of the following criteria:
      - C1 inhibitor (C1-INH) antigenic level below the lower limit of normal as defined by the laboratory performing the test, or
      - 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
  - Member has normal C1 inhibitor as confirmed by laboratory testing and meets one of the following criteria:
    - Member has an F12, angiopoietin-1, plasminogen, or kininogen-1 (KNG1) gene mutation as confirmed by genetic testing, or
- 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.
- Medication is prescribed by, or in consultation with allergist/immunologist or a physician who specializes in the treatment of HAE or related disorders; AND
- Member has history of moderate to severe cutaneous attacks (without concomitant hives) OR abdominal attacks OR mild to severe airway swelling attacks of HAE (i.e., debilitating cutaneous/gastrointestinal symptoms OR laryngeal/pharyngeal/tongue swelling); AND
- Dose does not exceed FDA approved labeling; AND
- MMP members who have previously received this medication within the past 365 days are not subject to Step Therapy Requirements.

**Continuation of Therapy Criteria:**

- Patient continues to meet initial criteria; AND
- Patient has experienced reduction in severity and duration of attacks since starting treatment; AND
- Documentation supporting a positive clinical response to therapy with Ruconest (e.g., chart notes, medical records)

**Coverage durations:**

- Initial coverage: 6 months
- Continuation of therapy coverage: 6 months

*** Requests will also be reviewed to National Coverage Determination (NCD) and Local Coverage Determinations (LCDs) if applicable.***

**Dosage/Administration:**

<table>
<thead>
<tr>
<th>Indication</th>
<th>Dose</th>
<th>Maximum dose (1 billable unit = 10 units)</th>
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<tbody>
<tr>
<td>HAE</td>
<td>Body weight &lt; 84 kg: 50 international units (IU) per kg body weight by intravenous injection</td>
<td>3360 billable units per 28 days</td>
</tr>
<tr>
<td></td>
<td>Body weight ≥ 84 kg: 4200 IU (2 vials) by intravenous injection</td>
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<td></td>
<td>If the attack symptoms persist, an additional (second) dose can be administered at the recommended dose level. Do not exceed 4200 IU per dose. No more than</td>
<td></td>
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two doses should be administered within a 24-hour period.

**Investigational use:** All therapies are considered investigational when used at a dose or for a condition other than those that are recognized as medically accepted indications as defined in any one of the following standard reference compendia: American Hospital Formulary Service Drug information (AHFS-DI), Thomson Micromedex DrugDex, Clinical Pharmacology, Wolters Kluwer Lexi-Drugs, or Peer-reviewed published medical literature indicating that sufficient evidence exists to support use. Neighborhood does not provide coverage for drugs when used for investigational purposes.

**Applicable Codes:**
Below is a list of billing codes applicable for covered treatment options. The below tables are provided for reference purposes and may not be all-inclusive. Requests received with codes from tables below do not guarantee coverage. Requests must meet all criteria provided in the procedure section.

The following HCPCS/CPT codes are:

<table>
<thead>
<tr>
<th>HCPCS/CPT Code</th>
<th>Description</th>
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<tbody>
<tr>
<td>J0596</td>
<td>Injection, c1 esterase inhibitor (recombinant), Ruconest, 10 units</td>
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**References:**