SENTARA HEALTH PLANS

PHARMACY PRIOR AUTHORIZATION/STEP-EDIT REQUEST*

Directions: **The prescribing physician must sign and clearly print name (preprinted stamps not valid) on this request**. All other information may be filled in by office staff; **fax to 1-800-750-9692**. No additional phone calls will be necessary if all information (including phone and fax #s) on this form is correct. **If the information provided is not complete, correct, or legible, the authorization process can be delayed.**

Drug Requested: Ruconest® IV (C1 Inhibitor Recombinant) **(Pharmacy)**

MEMBER & PRESCRIBER INFORMATION: Authorization may be delayed if incomplete.

Member Name: _____

Member Sentara #: _____ **Date of Birth:** _____

Prescriber Name: _____

Prescriber Signature: _____ **Date:** _____

Office Contact Name: _____

Phone Number: _____ **Fax Number:** _____

DEA OR NPI #: _____

DRUG INFORMATION: Authorization may be delayed if incomplete.

Drug Form/Strength: _____

Dosing Schedule: _____ **Length of Therapy**: _____

Diagnosis: _____ **ICD Code, if applicable:** _____

Weight: _____ **Date:** _____

Dosing Limit (see below):

A. Quantity Limit:

- **Pharmacy Benefit:** Ruconest 2100mg vial: 2 vials per 28 days

B. Dose:

- Body weight < 84kg: 50 international units (IU) per kg body weight by intravenous injection
- Body weight > 84kg: 4200 IU (2 vials) by intravenous injection

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 two doses should be administered within a 24-hour period

- **NDC:** Ruconest 2100 IU single use 25mL vial - 68012-0350-xx
- **Coverage is provided for 12 months and will be eligible for renewal**

☐ Standard Review. In checking this box, the timeframe does not jeopardize the life or health of the member or the member's ability to regain maximum function and would not subject the member to severe pain.

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CLINICAL CRITERIA: Check below all that apply. All criteria must be met for approval. To support each line checked, all documentation, including lab results, diagnostics, and/or chart notes, must be provided or request may be denied.

Initial Approval Criteria – 12 months. The cumulative amount of medication(s) the patient has on-hand, indicated for the acute treatment of HAE, will be taken into account when authorizing. The authorization will provide a sufficient quantity in order for the patient to have a cumulative amount of HAE medication(s) on-hand in order to treat acute attacks for the duration of the authorization (unless otherwise specified)

- ❑ Must be prescribed by, or in consultation with, a specialist in: allergy, immunology, hematology, pulmonology, or medical genetics

AND

Treatment of acute attacks of Hereditary Angioedema (HAE):

- ❑ Member must be at least 6 years of age

AND

- ❑ Member has a history of one of the following:
  - ❑ Severe cutaneous episodes
  - ❑ Abdominal attacks (debilitating gastrointestinal symptoms)
  - ❑ Mild to severe airway swelling attacks of HAE (i.e. laryngeal/ pharyngeal/ tongue swelling)

AND

- ❑ Confirmation the member is avoiding the following possible triggers for HAE attacks **(ALL MUST APPLY)**:
  - ❑ Helicobacter pylori infections **(confirmed by lab test)**
  - ❑ Estrogen-containing oral contraceptive agents, hormone replacement therapy
  - ❑ Antihypertensive agents containing ACE inhibitors

AND

Patient has one of the following clinical presentations below that is consistent with a HAE subtype, which is confirmed by repeat blood testing (please submit chart notes for symptoms and lab value to confirm HAE subtype):

II.A. ❑ HAE I: (all bullet points must apply)

- Low C1 inhibitor (C1-INH) antigenic level (C1-INH antigenic level below the lower limit of normal as defined by the laboratory performing the test); **AND**
- Low C4 level (C4 below the lower limit of normal as defined by the laboratory performing the test); **AND**
- Low C1-INH functional level (C1-INH functional level below the lower limit of normal as defined by the laboratory performing the test) **AND** one of the following:
  - ❑ Member has a family history of HAE; **OR**

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- ❑ Acquired angioedema has been ruled out (i.e., patient onset of symptoms occur prior to 30 years old, normal C1q levels, patient does not have underlying disease such as lymphoma or benign monoclonal gammopathy [MGUS], etc.)

OR

II.B. ❑ HAE II (C1-Inhibitor dysfunction): (all bullet points must apply)

- Normal to elevated C1-INH antigenic level; **AND**
- Low C4 level (C4 below the lower limit of normal as defined by the laboratory performing the test); **AND**
- Low C1-INH functional level (C1-INH functional level below the lower limit of normal as defined by the laboratory performing the test) **AND** one of the following:
  - ❑ Member has a family history of HAE; **OR**
  - ❑ Acquired angioedema has been ruled out (i.e., patient onset of symptoms occur prior to 30 years old, normal C1q levels, patient does not have underlying disease such as lymphoma or benign monoclonal gammopathy [MGUS], etc.)

OR

II.C. ❑ HAE III with normal C1-INH: (all bullet points must apply)

- Normal C1-INH antigenic level; **AND**
- Normal C4 level; **AND**
- Normal C1-INH functional level; **AND**
- Repeat blood testing during an attack has confirmed the patient does not have abnormal lab values indicative of HAE I or HAE II; **AND**
- Patient had an inadequate response or intolerance to an adequate trial of prophylactic therapy with one of following:
  - ❑ antifibrinolytic agent: (tranexamic acid (TXA) **OR** ❑ aminocaproic acid)
  - ❑ 17α- alkylated androgen: danazol
  - ❑ progestins (female patients only)

AND

❑ **One of the following:**

- ❑ Patient has a known HAE-causing mutation (e.g., mutation of coagulation factor XII gene [F12 mutation], mutation in the angiopoietin-1 gene, mutation in the plasminogen gene or kininogen-1, **OR**
- ❑ Patient has a family history of HAE and documented evidence of lack of efficacy of chronic high-dose antihistamine therapy (e.g. cetirizine standard dosing at up to four times daily or an alternative equivalent, given for at least one month or an interval long enough to expect three or more angioedema attacks) AND corticosteroids

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RENEWAL CRITERIA: All criteria must be checked for approval. To support each line checked, all documentation (lab results, diagnostics, and/or chart notes) must be provided or request may be denied.

Reauthorization Approval - 12 months

- ❑ Member must continue to meet the criteria in section I & II (A-C)

AND

- ❑ Significant improvement in severity and duration of attacks have been achieved and sustained

AND

- ❑ Absence of unacceptable toxicity from the drug: Examples of unacceptable toxicity include hypersensitivity reactions.

Medication being provided by (check box below that applies):

❑ **Physician's office** **OR** ❑ **Specialty Pharmacy – PropriumRx**

For urgent reviews: Practitioner should call Sentara Pre-Authorization Department if they believe a standard review would subject the member to adverse health consequences. Sentara's definition of urgent is a lack of treatment that could seriously jeopardize the life or health of the member or the member's ability to regain maximum function.

Not all drugs may be covered under every Plan

If a drug is non-formulary on a Plan, documentation of medical necessity will be required.

****Use of samples to initiate therapy does not meet step edit/ preauthorization criteria.****

Previous therapies will be verified through pharmacy paid claims sor submitted chart notes.*

***Approved by Pharmacy and Therapeutics Committee:** 12/12/2016
REVISED/UPDATED: ~~3/28/2017~~; ~~7/24/2017~~; ~~9/29/2017~~; (Reformatted) ~~3/19/2019~~; ~~7/8/2019~~; 9/24/2019; ~~4/1/202:06/30/20211~~; **10/30/2023**