# 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: Ekterly®** (sebetralstat)

**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:** _____

**NPI #:** _____

**DRUG INFORMATION:** Authorization may be delayed if incomplete.

**Drug Name/Form/Strength:** _____

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

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

**Weight (if applicable):** _____ **Date weight obtained:** _____

### A. Quantity Limit (max daily dose):

- Ekterly® 300 mg tablets: 4 tablets (1200 mg) per 24-hour period
- **NDC: 82928-0300-xx**

### B. Recommended Dosage:

- One dose of 600 mg (2 tablets) taken orally at the earliest recognition of an HAE attack. A second dose of 600 mg (2 tablets) may be taken 3 hours after the first dose if response is inadequate, or if symptoms worsen or recur. Maximum Recommended Dosage: 1,200 mg in any 24-hour period.

**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 Authorization: 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).

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## Treatment of acute attacks of Hereditary Angioedema (HAE):

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

**AND**

- ❑ Member must be at least 12 years of age

**AND**

- ❑ Member has a history of one of the following (choose one):
  - ❑ Moderate to severe cutaneous attacks (without concomitant hives)
  - ❑ Abdominal attacks (pain and swelling)
  - ❑ Mild to severe airway swelling attacks of HAE (i.e. debilitating cutaneous/gastrointestinal symptoms **OR** 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**

- ❑ Member must have trial and failure of **BOTH** of the following **(submit documentation)**:
  - ❑ icatibant ***requires prior authorization***
  - ❑ Berinert ***requires prior authorization***

**AND**

## Member 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 the HAE subtype:

- ❑ **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**
    - ❑ 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.)

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**OR**

- ❑ **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**

- ❑ **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**
  - ❑ Member 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:**
  - ❑ Member 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**

  - ❑ Member 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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**Reauthorization: 12 months.** 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.

- ❑ Member must continue to meet all initial authorization criteria

**AND**

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

**AND**

- ❑ Member has experienced an absence of unacceptable toxicity from the drug. Examples of unacceptable toxicity include hypersensitivity reactions.

**Medication being provided by Specialty Pharmacy – Proprium Rx**

***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 or submitted chart notes.****



***Approved by Pharmacy and Therapeutics Committee**: 9/25/2025
**REVISED/UPDATED/REFORMATTED: 10/22/2025**