

OPTIMA HEALTH PLAN

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 1-844-202-5034. No additional phone calls will be necessary if all information (including phone and fax #s) on this form is correct. Incomplete form will delay authorization process.*

Drug Requested (select applicable drug below): *Medical*

Cinryze® (C1 Esterase Inhibitor Human) (J0598) **Haegarda®** (C1 Esterase Inhibitor Human) (J3590)

DRUG INFORMATION: Complete information below. If incomplete, authorization process will be delayed.

Drug Name: _____

Drug Form/Strength/Quantity: _____

Dosing Schedule: _____ Length of Therapy: _____

Diagnosis: _____ ICD Code: _____

Dosing Limit: (see below)

- A. **Quantity Limit (max daily dose):** Pharmacy Benefit: None
- B. **Max Units (per dose and over time):** Medical Benefit:
- 1,000units every 3-4 days=7,000 units every 30 days (14 vials)
- Cinryze* 7,000 IU vial: 700 billable units per 30 days
- 10 units=1billable
- **J0598** 500 unit: 10 unit=1billable **AND** NDC 42227-0081-xx 500unit

- Coverage is provided for **12 months** and will be eligible for renewal

CLINICAL CRITERIA: Check boxes below to qualify. If **not** checked, authorization process will be delayed.

Initial Approval Criteria:

I. Treatment of acute attacks of Hereditary Angioedema (HAE):

- Patient must be at least 9 years of age; **AND**
- Patient has a history of one of the following criteria for long-term HAE prophylaxis:
 - History of **four (4) or more** severe HAE attacks per month (i.e., airway swelling, debilitating cutaneous or gastrointestinal episodes); **OR**
 - Patient is disabled more than 5 days per month by HAE; **OR**
 - History of recurrent laryngeal attacks caused by HAE; **AND**
- Treatment of patient with “on-demand” therapy (i.e., Kalbitor, Firazyr, Ruconest, or Berinert) did not provide satisfactory control or access to “on-demand therapy” is limited (*defined as more than 5 attacks/month for 4 months consecutively within the same year*); **AND**
- Patient has tried and failed, is intolerant, or has a contraindication to attenuated (17 alpha-alkylated) androgens (i.e., Danazol) for HAE prophylaxis; **AND**
- Confirmation the patient is avoiding the following possible triggers for HAE attacks:
 - Helicobacter pylori infections (confirmed by lab test)
 - Estrogen-containing oral contraceptive agents OR hormone replacement therapy
 - Antihypertensive agents containing ACE inhibitors; **AND**

II.A. Patient has the following clinical presentation consistent with HAE I:

- Treatment of patient with “on-demand” therapy (i.e., Kalbitor, Firazyr, Ruconest, or Berinert) did not provide satisfactory control or access to “on-demand therapy” is limited (*defined as more than 5 attacks/month for 4 months consecutively within the same year*); **AND**

(continued on next page)

- 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**
- Patient has a family history of HAE; **OR**
- Normal C1q level; **OR**

II.B. Patient has the following clinical presentation consistent with HAE II:

- Treatment of patient with “on-demand” therapy (i.e., Kalbitor, Firazyr, Ruconest, or Berinert) did not provide satisfactory control or access to “on-demand therapy” is limited (*defined as more than 5 attacks/month for 4 months consecutively within the same year*); **AND**
- 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); **OR**

II.C. Patient has the following clinical presentation consistent with HAE III:

- Treatment of patient with “on-demand” therapy (i.e., Kalbitor, Firazyr, Ruconest, or Berinert) did not provide satisfactory control or access to “on-demand therapy” is limited (*defined as more than 5 attacks/month for 4 months consecutively within the same year*); **AND**
- Normal C1-INH antigenic level); **AND**
- Normal C4 level; **AND**
- Normal C1-INH functional level; **AND**
- Patient has a known HAE causing C1-INH mutation (i.e., mutation of coagulation factor XII gene); **OR**
- Patient has a family history of HAE; **AND**

Renewal Criteria

- Patient 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 applicable box below):

- Physician's office **OR** Specialty Pharmacy: BriovaRx

****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.

Patient Name: _____

Member Optima #: _____ Date of Birth: _____

Prescriber Name: _____

Prescriber Signature: _____ Date: _____

Office Contact Name: _____

Phone Number: _____ Fax Number: _____

DEA OR NPI #: _____

*Approved by Pharmacy and Therapeutics Committee: 9/19/2013

REVISED/UPDATED: 11/20/2013; 10/31/2014; 4/3/2015; 5/22/2015; 12/30/2015; 1/29/2016; 8/17/2016; 9/22/2016; 12/11/2016; 7/30/2017; 9/13/2017; 11/16/2017; 3/28/2018