**Berinert (C1 esterase inhibitor [human])**

**Line(s) of Business:**  
HMO; PPO; QUEST Integration  
Akamai Advantage

**Original Effective Date:** 10/01/2015  
**Current Effective Date:** TBD

**POLICY**

**A. INDICATIONS**

The indications below including FDA-approved indications and compendial uses are considered a covered benefit provided that all the approval criteria are met and the member has no exclusions to the prescribed therapy.

**FDA-Approved Indication**

- Treatment of acute abdominal, facial, or laryngeal attacks of hereditary angioedema (HAE) attacks in adult and adolescent patients

**Compendial Use**

- Prophylaxis of HAE attacks

**B. REQUIRED DOCUMENTATION**

The following information is necessary to initiate the prior authorization review:

- Laboratory report with C4 levels, C1 inhibitor functional levels, and C1 inhibitor antigenic protein level and/or C1 inhibitor functional activity

- For the diagnosis of HAE with normal C1 inhibitor, F12 gene mutation testing results (if applicable)

**C. CRITERIA FOR APPROVAL**

**Hereditary Angioedema**

Indefinite Authorization of 12 months may be granted to members who meet ALL of the following criteria:

1. Berinert is being requested for the treatment of acute HAE attacks

2. Member has a diagnosis of HAE. Diagnostic laboratory testing for HAE has been performed including (eg. C4 levels, C1 inhibitor antigenic protein level and/or C1 inhibitor functional activity and antigenic protein levels)
3. HAE has been confirmed by laboratory testing for members with HAE with C1 inhibitor deficiency (HAE type I or type II):
   a. Low C4 level, and
   b. Low C1 inhibitor antigenic protein level and/or low C1 inhibitor functional activity level (is below the lower limit of normal as defined by the laboratory performing the test)

4. For members with HAE with normal C1 inhibitor (HAE type III):
   a. Normal C4 level, normal C1 inhibitor antigenic protein level and normal C1 inhibitor functional activity
   b. Member meets EITHER of the following criteria:
      i. Member tested positive for the F12 gene mutation
      ii. Member has a family history of angioedema
   c. Other causes of angioedema have been ruled out (eg, drug-induced) and the member meets EITHER of the following criteria:
      i. Member tested positive for the F12 gene mutation
      ii. Member has a family history of angioedema

D. CONTINUATION OF THERAPY
1. No previous authorization/precertification:
   All members (including members currently receiving treatment without prior authorization) must meet criteria for initial approval in section C.

2. Reauthorization:
   Authorization of 12 months may be granted to members requesting authorization for continuation of therapy if Berinert was previously authorized by HMSA/CVS and member demonstrated a clinical response to therapy.

D.E. DOSAGE AND ADMINISTRATION
Approvals may be subject to dosing limits in accordance with FDA-approved labeling, accepted compendia, and/or evidence-based practice guidelines.

F. ADMINISTRATIVE GUIDELINES
Precertification is required. Please refer to the HMSA medical policy web site for the fax form.

E.G. IMPORTANT REMINDER
The purpose of this Medical Policy is to provide a guide to coverage. This Medical Policy is not intended to dictate to providers how to practice medicine. Nothing in this Medical Policy is intended to discourage or prohibit providing other medical advice or treatment deemed appropriate by the treating physician.

Benefit determinations are subject to applicable member contract language. To the extent there are any conflicts between these guidelines and the contract language, the contract language will control.

This Medical Policy has been developed through consideration of the medical necessity criteria under Hawaii’s Patients’ Bill of Rights and Responsibilities Act (Hawaii Revised Statutes §432E-1.4), generally accepted standards of medical practice and review of medical literature and government approval status. HMSA has determined that services not covered under this Medical Policy will not be medically necessary under Hawaii law in most cases. If a treating physician disagrees with HMSA’s determination as to medical necessity in a given case, the physician may request that
CVS/caremark reconsider the application of the medical necessity criteria to the case at issue in light of any supporting documentation.

F.H. REFERENCES