

## **SPECIALTY GUIDELINE MANAGEMENT**

### **CINRYZE (C1 esterase inhibitor)**

#### **POLICY**

##### **I. 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.

A. FDA-Approved Indications

Routine prophylaxis against angioedema attacks in adolescent and adult patients with hereditary angioedema (HAE)

B. Compendial Uses

Treatment of acute HAE attacks

All other indications are considered experimental/investigational and are not a covered benefit.

##### **II. REQUIRED DOCUMENTATION**

The following information is necessary to initiate the prior authorization review: C4 levels and C1 inhibitor functional and antigenic protein levels.

##### **III. CRITERIA FOR INITIAL APPROVAL**

Authorization for 12 months may be granted for treatment and prevention of HAE attacks when all of the following criteria are met:

1. Diagnostic laboratory testing for HAE has been performed (eg, C4 levels, C1 inhibitor functional and antigenic protein levels).
2. With C1 inhibitor deficiency: C1 inhibitor antigenic protein level and/or C1 inhibitor functional level is below the lower limit of normal as defined by the laboratory performing the test.
3. With normal C1 inhibitor: Other causes of angioedema have been ruled out (eg, drug-induced) and either of the following criteria are met:
  - a. Member has tested positive for the F12 gene mutation or
  - b. The member has a family history of angioedema and the angioedema was refractory to a trial of antihistamine (e.g., cetirizine) for at least one month.

##### **IV. CONTINUATION OF THERAPY**

All members (including new members) requesting authorization for continuation of therapy must meet all initial authorization criteria.

##### **V. DOSAGE AND ADMINISTRATION**

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

**VI. REFERENCES**

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