

ENHANCED SPECIALTY GUIDELINE MANAGEMENT

RUCONEST (recombinant 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.

FDA-Approved Indications

Treatment of acute attacks in adults and adolescent patients with hereditary angioedema (HAE)

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 of 12 months may be granted for treatment of acute hereditary angioedema attacks when either of the following criteria is met:

- A. Member has C1 inhibitor deficiency or dysfunction as confirmed by laboratory testing.
- B. Member has normal C1 inhibitor as confirmed by laboratory testing and meets one of the following criteria:
 1. Member has an F12, angiotensin-converting enzyme, or plasminogen gene mutation as confirmed by genetic testing, or
 2. Member has a family history of angioedema and the angioedema was refractory to a trial of high-dose antihistamine (e.g., cetirizine) for at least one month.

IV. CONTINUATION OF THERAPY

Authorization of 12 months may be granted for continuation of therapy when all of the following criteria are met:

- A. Member meets all initial authorization criteria.
- B. Member has experienced reduction in severity and duration of attacks since starting treatment.

V. REFERENCES

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4. Zuraw BL, Banerji A, Bernstein JA, et al. US Hereditary Angioedema Association Medical Advisory Board 2013 recommendations for the management of hereditary angioedema due to C1 inhibitor deficiency. *J Allergy Clin Immunol: In Practice*. 2013; 1(5): 458-467.

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9. Bowen T. Hereditary angioedema: beyond international consensus – circa December 2010 – The Canadian Society of Allergy and Clinical Immunology Dr. David McCourtie Lecture. *Allergy Asthma Clin Immunol.* 2011;7(1):1.
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12. Farkas H, Martinez-Saguer I, Bork K, et al. International consensus on the diagnosis and management of pediatric patients with hereditary angioedema with C1 inhibitor deficiency. *Allergy.* 2017;72(2):300-313.