

STANDARD MEDICARE PART B MANAGEMENT

RUCONEST (C1 esterase inhibitor [recombinant])

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 Indication

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

All other indications will be assessed on an individual basis. Submissions for indications other than those enumerated in this policy should be accompanied by supporting evidence from Medicare approved compendia.

II. CRITERIA FOR INITIAL APPROVAL

Hereditary angioedema (HAE)

Authorization of 12 months may be granted for treatment of acute HAE attacks when either of the following criteria are 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 kininogen-1 (KNG1) gene mutation as confirmed by genetic testing.
 - 2. 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.

III. CONTINUATION OF THERAPY

All members (including new members) requesting authorization for continuation of therapy must be currently receiving therapy with the requested agent.

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

- A. The member is currently receiving therapy with Ruconest.
- B. Ruconest is being used to treat an indication enumerated in Section II.
- C. The member is receiving benefit from therapy.

IV. REFERENCES

1. Ruconest [package insert]. Bridgewater, NJ: Pharming Healthcare Inc.; December 2019.

Reference number(s)
2194-A

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3. Cicardi M, Bork K, Caballero T, et al. Evidence-based recommendations for the therapeutic management of angioedema owing to hereditary C1 inhibitor deficiency: consensus report of an International Working Group. *Allergy*. 2012;67:147-157.
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.
5. Zuraw BL, Bork K, Binkley KE, et al. Hereditary angioedema with normal C1 inhibitor function: consensus of an international expert panel. *Allergy Asthma Proc*. 2012;33(6):S145-S156.
6. Maurer M, Magerl M, Ansotegui I, et al. The international WAO/EAACI guideline for the management of hereditary angioedema – the 2017 revision and update. *Allergy*. 2018;00:1-22.
7. Lang DM, Aberer W, Bernstein JA, et al. International consensus on hereditary and acquired angioedema. *Ann Allergy Asthma Immunol*. 2012;109:395-202.
8. Cicardi M, Aberer W, Banerji A, et al. Classification, diagnosis, and approach to treatment for angioedema: consensus report from the Hereditary Angioedema International Working Group. *Allergy*. 2014;69:602-616.
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.
10. Bernstein JA. Update on angioedema: Evaluation, diagnosis, and treatment. *Allergy and Asthma Proceedings*. 2011;32(6):408-412.
11. Longhurst H, Cicardi M. Hereditary angio-edema. *Lancet*. 2012;379:474-481.