Mass General Brigham
Health Plan

Ruconest (C1 esterase inhibitor [recombinant])
Effective 01/01/2026

Plan	☐ MassHealth UPPL ☒Commercial/Exchange	**Program Type**	☒ Prior Authorization ☐ Quantity Limit ☐ Step Therapy
Benefit	☒ Pharmacy Benefit ☐ Medical Benefit		
Specialty Limitations	This medication has been designated specialty and must be filled at a contracted specialty pharmacy.		
Contact Information	**Medical Benefit** **Pharmacy Benefit**	Phone: 833-895-2611 Phone: 800-711-4555	Fax: 888-656-6671 Fax: 844-403-1029
Exceptions	N/A		

Overview

FDA-Approved Indication

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

Limitation of Use

Effectiveness was not established in HAE patients with laryngeal attacks.

All other indications are considered experimental/investigational and not medically necessary.

Coverage Guidelines

Authorization may be granted for members new to the plan within the past 90 days who are currently receiving treatment with the requested medication, excluding when the product is obtained as samples or via manufacturer's patient assistance programs.

OR

Authorization may be granted for treatment of acute hereditary angioedema attacks when the requested medication will not be used in combination with any other medication used for the treatment of acute HAE attacks and either of the following criteria is met:

A. Member has C1 inhibitor deficiency or dysfunction as confirmed by laboratory testing and meets both of the following criteria.
   1. Member has a C4 level below the lower limit of normal as defined by the laboratory performing the test, and
   2. Member meets one of the following criteria:
      i. C1 inhibitor (C1-INH) antigenic level below the lower limit of normal as defined by the laboratory performing the test; or
      ii. Normal C1-INH antigenic level and a low C1-INH functional level (functional C1-INH less than 50% or C1-INH functional level below the lower limit of normal as defined by the laboratory performing the test)

B. Member has normal C1 inhibitor as confirmed by laboratory testing and meets one of the following criteria:

1. Member has an F12, angiopoietin-1, plasminogen, kininogen-1 (KNG1), heparan sulfate-glucosamine 3-O-sulfotransferase 6 (HS3ST6), or myoferlin (MYOF) gene mutation as confirmed by genetic testing, or
2. Member has a documented family history of angioedema, and the angioedema was refractory to a trial of high-dose antihistamine therapy (i.e., cetirizine at 40 mg per day or the equivalent) for at least one month.

Continuation of Therapy

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

A. Member meets the criteria for initial approval.
B. Member has experienced a reduction in severity and/or duration of attacks when the requested medication is used to treat an acute attack.
C. Prophylaxis should be considered based on the attack frequency, attack severity, comorbid conditions, and member's quality of life.

Limitations

1. All approvals will be granted for 6 months.

References

1. Bernstein J. Severity of hereditary angioedema, prevalence, and diagnostic considerations. *Am J Med.* 2018;24:292-298.
2. Bernstein JA. Update on angioedema: Evaluation, diagnosis, and treatment. *Allergy and Asthma Proceedings.* 2011;32(6):408-412.
3. Bork K, Aygören-Pürsün E, Bas M, et al. Guideline: Hereditary angioedema due to C1 inhibitor deficiency. *Allergo J Int*. 2019;28:16–29.
4. Bowen T, Cicardi M, Farkas H, et al. 2010 International consensus algorithm for the diagnosis, therapy, and management of hereditary angioedema. *Allergy Asthma Clin Immunol*. 2010;6(1):24.
5. 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.
6. Busse PJ, Christiansen, SC, Riedl MA, et al. US HAEA Medical Advisory Board 2020 Guidelines for the Management of Hereditary Angioedema. *J Allergy Clin Immunol: In Practice.* 2021 Jan;9(1):132-150.e3.
7. 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.
8. 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.
9. Craig T, Busse P, Gower RG, et al. Long-term prophylaxis therapy in patients with hereditary angioedema with C1 inhibitor deficiency. *Ann Allergy Asthma Immunol.* 2018;121(6):673-679.
10. 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.
11. Henao MP, Kraschnewski J, Kelbel T, Craig T. Diagnosis and screening of patients with hereditary

angioedema in primary care. *Therapeutics and Clin Risk Management.* 2016;12:701-711.
12. Kanani, A., Schellenberg, R. & Warrington, R. Urticaria and angioedema. *All Asth Clin Immun* 7, S9 (2011), Table 2.
13. Lang DM, Aberer W, Bernstein JA, et al. International consensus on hereditary and acquired angioedema. *Ann Allergy Asthma Immunol*. 2012;109:395-402.
14. Longhurst H, Cicardi M. Hereditary angio-edema. *Lancet.* 2012;379:474-481.
15. 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;73:1575-1596.
16. Ruconest [package insert]. Warren, NJ: Pharming Healthcare Inc.; April 2020.
17. Sharma J, Jindal AK, Banday AZ, et al. Pathophysiology of Hereditary Angioedema (HAE) Beyond the SERPING1 Gene [published online ahead of print, 2021 Jan 14] [published correction appears in Clin Rev Allergy Immunol. 2021 Feb 17]. *Clin Rev Allergy Immunol*. 2021;10.1007/s12016-021-08835-8. Doi:10.1007/s12016-021-08835-8.
18. 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.

Review History

12/13/2023: Reviewed at Dec P&T, switched from SGM to Custom. Effective 1/1/2024
10/08/2025 - Reviewed at October P&T. Updated policy to indicate it no longer applies to the medical benefit. Effective 01/01/2026.