

**Ruconest (C1 esterase inhibitor [recombinant])**  
**Effective 01/01/2024**

<b>Plan</b>	<input type="checkbox"/> MassHealth UPPL <input checked="" type="checkbox"/> Commercial/Exchange	<b>Program Type</b>	<input checked="" type="checkbox"/> Prior Authorization <input type="checkbox"/> Quantity Limit <input type="checkbox"/> Step Therapy
<b>Benefit</b>	<input checked="" type="checkbox"/> Pharmacy Benefit <input checked="" type="checkbox"/> Medical Benefit		
<b>Specialty Limitations</b>	This medication has been designated specialty and must be filled at a contracted specialty pharmacy.		
<b>Contact Information</b>	<b>Medical and Specialty Medications</b>		
	All Plans	Phone: 877-519-1908	Fax: 855-540-3693
	<b>Non-Specialty Medications</b>		
	All Plans	Phone: 800-711-4555	Fax: 844-403-1029
<b>Exceptions</b>	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 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, angiotensin-converting enzyme 1, plasminogen, kininogen-1 (KNG1), heparan sulfate-glucosaminase 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**

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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. 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.
- 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, Anotegui I, et al. The international WAO/EAACI guideline for the management of hereditary angioedema – the 2017 revision and update. *Allergy*. 2018;73:1575-1596.
- 7. Lang DM, Aberer W, Bernstein JA, et al. International consensus on hereditary and acquired angioedema. *Ann Allergy Asthma Immunol*. 2012;109:395-402.
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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.



10. Bernstein JA. Update on angioedema: Evaluation, diagnosis, and treatment. *Allergy and Asthma Proceedings*. 2011;32(6):408-412.
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13. Henaó 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.
14. Bernstein J. Severity of hereditary angioedema, prevalence, and diagnostic considerations. *Am J Med*. 2018;24:292-298.
15. Bork K, Aygören-Pürsün E, Bas M, et al. Guideline: Hereditary angioedema due to C1 inhibitor deficiency. *Allergo J Int*. 2019;28:16–29.
16. 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.
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. Kanani, A., Schellenberg, R. & Warrington, R. Urticaria and angioedema. *All Asth Clin Immun* 7, S9 (2011), Table 2.

### Review History

12/13/2023: Reviewed at Dec P&T, switched from SGM to Custom. Effective 1/1/2024

