CareSource Rx Innovations

PHARMACY POLICY STATEMENT
Indiana Medicaid

DRUG NAME	Ruconest (C1 esterase inhibitor (recombinant))
BENEFIT TYPE	Medical or Pharmacy
STATUS	Prior Authorization Required

Ruconest, approved by the FDA in 2014, is a C1 esterase inhibitor [recombinant] indicated for the treatment of acute attacks in adult and adolescent patients with hereditary angioedema (HAE).
HAE is a rare autosomal dominant disease characterized by episodic unpredictable swelling, which can occur in a variety of anatomic locations. The swelling results from excess production of the vasodilator bradykinin. Attacks may be painful and cause functional impairment but are not associated with pruritis. The most common types of HAE are caused by deficiency (type 1) or dysfunction (type 2) of C1 inhibitor (C1-INH). Type 1 is the most prevalent.

Ruconest (C1 esterase inhibitor (recombinant)) will be considered for coverage when the following criteria are met:

Hereditary Angioedema (HAE)

For **initial** authorization:
1. Member must be 12 years of age or older; AND
2. Medication must be prescribed by or in consultation with an allergist or immunologist; AND
3. Member has a diagnosis of HAE type I or type II confirmed by both of the following:
   a) Low C4 level;
   b) Low (<50% of normal) C1 inhibitor antigenic and/or functional level; AND
4. Medication is being prescribed for the treatment of acute HAE attacks; AND
5. Member has documented trial and failure of or contraindication to both generic Firazyr (if 18 years of age or older) and Berinert; AND
6. Medication is not being used in combination with another acute HAE therapy (e.g., Berinert, Firazyr, Kalbitor); AND
7. Member does not have a history of allergy to rabbits or rabbit-derived products.
8. **Dosage allowed/Quantity limit:** 50 IU per kg IV; not to exceed 4200 IU (2 vials) per dose. May repeat 1 time; no more than 2 doses within 24 hours.
   QL: 8 vials per fill

If all the above requirements are met, the medication will be approved for 6 months.

For **reauthorization**:
1. Chart notes must document improvement such as faster time to symptom relief or resolution of attack.

If all the above requirements are met, the medication will be approved for an additional 12 months.

CareSource considers Ruconest (C1 esterase inhibitor (recombinant)) not medically necessary for the treatment of conditions that are not listed in this document. For any other indication, please refer to the Off-Label policy.

DATE	ACTION/DESCRIPTION
08/28/2017	New policy for Ruconest created. Criteria for each type of HAE specified. Criteria of documentation of attacks, discontinuation of meds that can cause HAE, and restriction on combinations with other meds for acute attacks added.
01/20/2021	Updated references. Clarified the dosing. Removed statement about causative meds. Removed hematology as specialist. Simplified the diagnostic criteria. Removed log book requirement. Reworded the renewal criteria. Added rabbit allergy contraindication. Extended initial auth duration to 6 mo and renewal to 12 mo. Changed “rabbit-derived” to say “recombinant.” Adjusted quantity limit to allow for repeat doses as indicated. Removed exclusion of laryngeal attacks.
07/01/2022	Transferred to new template. Updated references. Added pharmacy as benefit option. Added “generic” in front of Firazyr. Changed age limit from 13 to 12.
06/18/2025	Annual review; no updates.

References:

1. Ruconest [package insert]. Warren NJ: Pharming Healthcare, Inc; 2020.
2. Frank MM, Zuraw B, Banerji A, et al. Management of children with hereditary angioedema due to C1 inhibitor deficiency. Pediatrics. 2016 Nov;138(5). pii: e20160575.
3. Riedl MA, Bernstein JA, Li H, et al. Recombinant human C1-esterase inhibitor relieves symptoms of hereditary angioedema attacks: phase 3, randomized, placebo-controlled trial. *Ann Allergy Asthma Immunol*. 2014;112(2):163-169.e1. doi:10.1016/j.anai.2013.12.004
4. Busse PJ, Christiansen SC, Riedl MA, et al. US HAEA Medical Advisory Board 2020 Guidelines for the Management of Hereditary Angioedema [published online ahead of print, 2020 Sep 6]. *J Allergy Clin Immunol Pract*. 2020;S2213-2198(20)30878-3. doi:10.1016/j.jaip.2020.08.046
5. Betschel S, Badiou J, Binkley K, et al. The International/Canadian Hereditary Angioedema Guideline [published correction appears in Allergy Asthma Clin Immunol. 2020 May 6;16:33]. *Allergy Asthma Clin Immunol*. 2019;15:72. Published 2019 Nov 25. doi:10.1186/s13223-019-0376-8
6. Bork K, Bernstein JA, Machnig T, Craig TJ. Efficacy of Different Medical Therapies for the Treatment of Acute Laryngeal Attacks of Hereditary Angioedema due to C1-esterase Inhibitor Deficiency. *J Emerg Med*. 2016;50(4):567-80.e1. doi:10.1016/j.jemermed.2015.11.008
7. Maurer M, Magerl M, Betschel S, et al. The international WAO/EAACI guideline for the management of hereditary angioedema - The 2021 revision and update. *World Allergy Organ J*. 2022;15(3):100627. Published 2022 Apr 7. doi:10.1016/j.waojou.2022.100627

Effective date: 01/01/2026
Revised date: 06/18/2025