Ruconest® (recombinant C1 esterase inhibitor)

When requesting Ruconest (recombinant C1 esterase inhibitor), the individual requiring treatment must be diagnosed with the following FDA-approved indication and meet the specific coverage guidelines for the covered indication.

FDA-Approved Indication

Ruconest is indicated for the treatment of acute attacks of Hereditary Angioedema (HAE).

Coverage Guidelines

Hereditary Angioedema (HAE) Due to C1 Inhibitor (C1-INH) Deficiency - Treatment of Acute Attacks

The individual must meet **all** of the following criteria for initial authorization:

- Has HAE type I or type II as confirmed by the following diagnostic criteria:
  - Has low levels of functional C1-INH protein (less than 50% of normal) at baseline, as defined by the laboratory reference values; AND
  - Has lower than normal serum C4 levels at baseline, as defined by the laboratory reference values;
- Medication is prescribed by or in consultation with an allergist/immunologist or a physician that specializes in the treatment of HAE or related disorders.

Approval duration: 12 months

The individual must meet **all** of the following criteria for reauthorization:
Note: An individual who is currently on therapy but has not been previously approved via eviCore coverage determination is reviewed under criteria for initial therapy.

- Has previously received Ruconest for acute HAE type I or type II attacks;
- Has had a favorable clinical response (e.g., decrease in the duration of HAE attacks, quick onset of symptom relief, complete resolution of symptoms, decrease in HAE acute attack frequency or severity) with Ruconest treatment;
- Medication is prescribed by or in consultation with an allergist/immunologist or a physician that specializes in the treatment of HAE or related disorders.

Approval duration: 12 months

Dosing Recommendation

- For individuals < 84 kg, administer 50 U/kg intravenously.
- For individuals ≥ 84 kg, administer 4200 U intravenously.

If symptoms persist, a second dose can be given at the recommended dose up to a maximum of 4200 U per dose. No more than two doses should be given in a 24 hour period.

References

1. Ruconest® [prescribing information]. Warren, NJ: Pharming; April 2020.
2. Zuraw BL. Hereditary angioedema. *N Engl J Med*. 2008;359:1027-1036.
3. Craig T, Shapiro R, Vegh A, et al. Efficacy and safety of an intravenous C1-inhibitor concentrate for long-term prophylaxis in hereditary angioedema. *Allergy Rhinol (Providence)*. 2017 Mar 1;8(1):13-19.
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 Pract*. 2021 Jan;9(1):132-150.e3.
5. Maurer M, Magerl M, Betschel S, et al. The international WAO/EAACI guideline for the management of hereditary angioedema: the 2021 revision and update. Allergy. 2022;77(7):1961-1990.
6. Riedl MA, Grivcheva-Panovska V, Moldovan D, et al. Recombinant human C1 esterase inhibitor for prophylaxis of hereditary angio-oedema: a phase 2, multicentre, randomised, double-blind, placebo-controlled crossover trial. *Lancet.* 2017;390:1595-1602.