

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 and applicable safety criteria for the covered indications.

FDA-approved indication

- Ruconest[®] is indicated for the treatment of acute attacks of Hereditary Angioedema (HAE)

Approved Off-label Compendial use(s)

- None

Coverage Guidelines

Hereditary Angioedema (HAE) Due to C1 Inhibitor (C1-INH) Deficiency [Type I or Type II] – Treatment of Acute Attacks

Initial authorization:

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

- The patient has HAE type I or type II as confirmed by the following diagnostic criteria:
 - Patient has low levels of functional C1-INH protein (Less than 50% of normal) at baseline, as defined by the laboratory reference values; AND
 - Patient has lower than normal serum C4 levels at baseline, as defined by the laboratory reference values; AND
- The 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

Reauthorization:

The patient must meet all of the following criteria for reauthorization:

- The patient has treated previous acute HAE type I or type II attacks with Ruconest; AND
- According to the prescribing physician, the patient 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; AND
- The 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 Guidelines

- o Ruconest: the recommended dose is 50 IU/kg IV (maximum 4,200 IU). A maximum of two doses may be administered in a 24-hour period.

References

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3. Mauer 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(8):1575-1596. Available at: <https://onlinelibrary.wiley.com/doi/epdf/10.1111/all.13384>. Accessed on September 12, 2018.
4. 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:S145-S156.
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6. 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;1:458-467. Available at: <https://haei.org/wp-content/uploads/2015/04/Zuraw-B-L-US-HAEA-MAB-2013-Recommendations.pdf>. Accessed on September 13, 2018.
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8. Zuraw BL. Hereditary angioedema. *N Engl J Med.* 2008;359:1027-1036.
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10. Pharming receives complete response letter from FDA [press release]. Pharming Group NV; September 19, 2018. Available at <https://www.pharming.com/pharming-receives-complete-response-letter-from-fda/>. Accessed on September 21, 2018.