Ruconest (C1 esterase inhibitor [recombinant])

Override(s)	Approval Duration
Prior Authorization	1 year

Medications	Quantity Limit
Ruconest (C1 esterase inhibitor [recombinant])	Up to two 50 units/kg doses [max of 4200 units (2 vials) per dose] per attack (Max: 16 vials/30 days)

APPROVAL CRITERIA

Requests for Ruconest (C1 esterase inhibitor [recombinant]) may be approved if the following criteria are met:

I. Individual has a diagnosis of hereditary angioedema; **AND**
II. Individual is using for the treatment of acute attacks (not prophylaxis); **AND**
III. Individual is 13 years of age or older; **AND**
IV. Documentation is provided that diagnosis is confirmed by a C4 level below the lower limit of normal as defined by the laboratory performing the test AND one of the following:
    A. C1 inhibitor (C1-INH) antigenic level below the lower limit of normal as defined by the laboratory performing the test with documentation provided; **OR**
    B. C1-INH functional level below the lower limit of normal as defined by the laboratory performing the test with documentation provided;

AND

V. Individual has a history of moderate or severe attacks such as airway swelling, severe abdominal pain, facial swelling, nausea and vomiting, or painful facial distortion**.**

Requests for Ruconest may not be approved for the following:

I. All other indications not included above; **OR**
II. Individuals using to treat laryngeal attacks; **OR**
III. In combination with other Hereditary Angioedema (HAE) agents for acute attacks (including but not limited to Berinert, Icatibant (Firazyr, Sajazir), or Kalbitor; **OR**
IV. Individual has a known or suspected allergy to rabbits or rabbit -derived products.

Key References:

1. Bork, K., Anderson, J.T., Caballero, T. *et al.* Assessment and management of disease burden and quality of life in patients with hereditary angioedema: a consensus report. *Allergy Asthma Clin Immunol* 17, 40 (2021). https://doi.org/10.1186/s13223-021-00537-2. Accessed on July 9, 2022.

2. 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;9:132-50.
3. Clinical Pharmacology [database online]. Tampa, FL: Gold Standard, Inc.: 2022. URL: http://www.clinicalpharmacology.com. Updated periodically.
4. DailyMed. Package inserts. U.S. National Library of Medicine, National Institutes of Health website. http://dailymed.nlm.nih.gov/dailymed/about.cfm. Accessed: July 9, 2022.
5. DrugPoints® System [electronic version]. Truven Health Analytics, Greenwood Village, CO. Updated periodically.
6. Efficacy ans Safety Study of DX-2930 to Prevent Acute Angioedema Attacks in Patients with Type I and Type II HAE. NCT02586805 (HELP Study). Available at https://www.clinicaltrials.gov/ct2/show/study/NCT02586805.
7. Lexi-Comp ONLINE™ with AHFS™, Hudson, Ohio: Lexi-Comp, Inc.; 2022; Updated periodically.
8. 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 Jan 10.
9. Riedl MA. Creating a Comprehensive Treatment Plan for Hereditary Angioedema. *Immunol Allergy Clin N Am*. 2013; 33 (4): 471-485. doi:10.1016/j.iac.2013.07.003.
10. 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; 1:458-67. doi:10.1016/j.jaip.2013.07.002.
11. Zuraw BL, Bernstein JA, Lang DM, et al. A focused parameter update: Hereditary angioedema, acquired C1 inhibitor deficiency, and angiotensin-converting enzyme inhibitor–associated angioedema. *J Allergy Clin Immunol*. 2013; 131(6):1491-1493.e1-e25. Available from: http://www.jacionline.org/article/S0091-6749(13)00523-X/pdf.

Federal and state laws or requirements, contract language, and Plan utilization management programs or polices may take precedence over the application of this clinical criteria.