Cinryze (C1 esterase inhibitor [Human])

Override(s)	Approval Duration
Prior Authorization Quantity Limit	Initial authorization: 6 months Continuation authorization: 1 year

Medications	Quantity Limit
Cinryze (C1 esterase inhibitor [human]) intravenous solution	May be subject to quantity limit

APPROVAL CRITERIA

Initial requests for Cinryze (C1 Esterase Inhibitor [Human]) may be approved if the following criteria are met:

I. Individual has a diagnosis of hereditary angioedema; **AND**
II. Individual is using for prophylaxis against acute attacks of hereditary angioedema for either of the following:
    A. Short term prophylaxis prior to surgery, dental procedures or intubation; **OR**
    B. Long-term prophylaxis to minimize the frequency and/or severity of recurrent attacks; **AND**
III. Individual is 6 years of age or older; **AND**
IV. Documentation is provided that, dDiagnosis of Hereditary Angioedema (HAE) confirmed by a C4 level below the lower limit of normal as defined by the laboratory performing the test **AND** any 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; **OR**
    C. Presence of a known HAE-causing C1-INH mutation; **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 Cinryze may be approved for continuation of use in prophylactic care if the following criteria are met:

I. Confirmation of a positive clinical response defined as a clinically significant reduction in the number and/or frequency of HAE attacks occurred.

Requests for Cinryze may not be approved for all other indications not included above.

Key References:

1. Bark, Konrad. Diagnosis and treatment of hereditary angioedema with normal C1 inhibitor. Allergy, Asthma & Clinical Immunology. 2010; 6:15. Available from: https://aacijournal.biomedcentral.com/track/pdf/10.1186/1710-1492-6-15 . Accessed July 15, 2020.
2. Clinical Pharmacology [database online]. Tampa, FL: Gold Standard, Inc.: 2020. URL: http://www.clinicalpharmacology.com. Updated periodically.
3. DailyMed. Package inserts. U.S. National Library of Medicine, National Institutes of Health website. http://dailymed.nlm.nih.gov/dailymed/about.cfm. Accessed: July 8, 2020.
4. DrugPoints® System [electronic version]. Truven Health Analytics, Greenwood Village, CO. Updated periodically.
5. 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. Accessed on July 8, 2019
6. Lexi-Comp ONLINE™ with AHFS™, Hudson, Ohio: Lexi-Comp, Inc.; 2019; Updated periodically.
7. 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.
8. 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**.
9. 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.
10. 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. Accessed on: July 8, 2020.

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.