

Market Applicability														
Market	DC	FL & FHK	FL MMA	FL LTC	GA	KS	KY	MD	NJ	NV	NY	TN	TX	WA
Applicable	X	X	NA	NA	X	NA	X	X	X	X	X	NA	NA	X

\*FHK- Florida Healthy Kids

## Cinryze (C1 esterase inhibitor [Human])

Override(s)	Approval Duration
Prior Authorization Quantity Limit	1 year

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

### APPROVAL CRITERIA

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 and the individual has failed, is intolerant to, or has a contraindication (such as pregnant, or breastfeeding) to 17 alpha-alkylated androgens (for example, danazol) or antifibrinolytic agents (for example, aminocaproic acid); **AND**
- III. Individual is 6 years of age or older; **AND**
- IV. Diagnosis 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, painful facial distortion).

This policy does not apply to health plans or member categories that do not have pharmacy benefits, nor does it apply to Medicare. Note that market specific restrictions or transition-of-care benefit limitations may apply.

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State Specific Mandates		
State name	Date effective	Mandate details (including specific bill if applicable)
N/A	N/A	N/A

**Key References:**

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4. Efficacy and 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 27, 2018.
5. Lexi-Comp ONLINE™ with AHFS™, Hudson, Ohio: Lexi-Comp, Inc.; 2018; Updated periodically.
6. Riedl MA, Bernstein JA, Craig T, et al. An open-label study to evaluate the long-term safety and efficacy of lanadelumab for prevention of attacks in hereditary angioedema: design of the HELP study extension. *Clin Transl Allergy*. 2017;7:36.
7. 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.
8. 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.
9. 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](http://www.jacionline.org/article/S0091-6749(13)00523-X/pdf). Accessed on: July 3, 2018.

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