CareSource

PHARMACY POLICY STATEMENT
Arkansas PASSE

DRUG NAME	Cinryze (C1 esterase inhibitor (human))
BILLING CODE	J0598
BENEFIT TYPE	Medical
SITE OF SERVICE ALLOWED	Home/Office
COVERAGE REQUIREMENTS	Prior Authorization Required (Non-Preferred Product) Alternative preferred product: Haegarda QUANTITY LIMIT— 20 vials (500 IU/vial) per 30 days
LIST OF DIAGNOSES CONSIDERED **NOT** MEDICALLY NECESSARY	Click Here

Cinryze (C1 esterase inhibitor (human)) will only be considered for coverage under the **medical** benefit when the following criteria are met:

Members must be clinically diagnosed with one of the following disease states and meet their individual criteria as stated.

HEREDITARY ANGIOEDEMA (HAE)

For **initial** authorization:

1. Member must be 6 years of age or older; AND
2. Member has a diagnosis of HAE type I or type II confirmed by both of the following:
   a) Low C4 level;
   b) Low (<50% of normal) C1 inhibitor antigenic and/or functional level; AND
3. Chart notes must document the member's baseline frequency of HAE attacks; AND
4. Member is inadequately controlled with on-demand treatment alone; AND
5. Cinryze is being prescribed for ongoing prophylaxis and will not be used to treat acute attacks; AND
6. Member has a trial and failure of or contraindication to Haegarda.
7. **Dosage allowed:**
   Age 12+: 1000 units IV infusion every 3 or 4 days; if no adequate response, doses up to 2,500 units (not exceeding 100 units/kg) every 3 or 4 days.
   Age 6-11: 500 units IV infusion every 3 or 4 days ; if no adequate response, doses up to 1000 units IV every 3 or 4 days may be considered.

If member meets all the requirements listed above, the medication will be approved for 6 months.

For **reauthorization**:

1. Chart notes must be provided that show a reduced frequency or number of acute attacks since starting treatment.

If member meets all the reauthorization requirements above, the medication will be approved for an additional 12 months.

CareSource considers Cinryze (C1 esterase inhibitor (human)) not medically necessary for the treatment of the following disease states based on a lack of robust clinical controlled trials showing superior efficacy compared to currently available treatments:

- Acquired angioedema (AAE)
- Treatment of acute HAE attacks

DATE	ACTION/DESCRIPTION
08/25/2017	New policy for Cinryze created. Criteria for each type of HAE specified. Criteria of documentation of attacks, discontinuation of meds that can cause HAE, and restriction on combinations with other meds for acute attacks were added.
07/27/2018	Medication is now approved for 6 years old and older.
01/14/2021	Updated and revised all content; consistent with other HAE prophylactics. Updated references. Greatly simplified the diagnostic confirmation criteria. Removed minimum required number of attacks, per guidelines; will just ask for baseline measure. Removed the statement about causative medications. Added that they must try on-demand treatment first. Rewrote the renewal criteria and removed log book requirement. Extended initial auth duration to 6 mo and renewal to 12 mo. Edited dosing information.
12/21/2021	Removed prescriber specialty requirement.

References:

1. Cinryze [package insert]. Exton, PA; ViroPharma Biologics, Inc.; 2020.
2. Lumry W. Management and Prevention of Hereditary Angioedema Attacks. Am J Manag Care. 2013;19:S111-S118.
3. Busse PJ, Christiansen SC, Riedl MA, et al. US HAEA Medical Advisory Board 2020 Guidelines for the Management of Hereditary Angioedema [published online ahead of print, 2020 Sep 6]. *J Allergy Clin Immunol Pract*. 2020;S2213-2198(20)30878-3. doi:10.1016/j.jaip.2020.08.046
4. 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;73(8):1575-1596. doi:10.1111/all.13384
5. Zuraw BL, Busse PJ, White M, et al. Nanofiltered C1 inhibitor concentrate for treatment of hereditary angioedema. *N Engl J Med*. 2010;363(6):513-522. doi:10.1056/NEJMoa0805538
6. Lumry W, Manning ME, Hurewitz DS, et al. Nanofiltered C1-esterase inhibitor for the acute management and prevention of hereditary angioedema attacks due to C1-inhibitor deficiency in children. *J Pediatr*. 2013;162(5):1017-22.e222. doi:10.1016/j.jpeds.2012.11.030

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