CareSource

PHARMACY POLICY STATEMENT
Georgia Medicaid

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 includes 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)) is a **non-preferred** product and 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 12 years of age or older, and medication is being used **for routine prophylaxis to prevent HAE attacks** (NOT for treatment of acquired angioedema); AND
2. Medication prescribed by or in consultation with a provider specializing in allergy, immunology, or hematology; AND
3. Member has documented trial and failure of or contraindication to Heagarda (Chart notes required); AND
4. Member must have a confirmed diagnosis of HAE as **one** of the following:
   a) Type 1 HAE documented in chart notes with ALL of the following (Note: tests listed below must be repeated for confirmation of diagnosis):
      i) Low levels (below the limits of the laboratory's normal reference range) of C4, C1-INH antigenic protein and C1-INH functional level; AND
      ii) Positive family history of angioedema OR earlier age of onset (before age 30) with normal C1q antigenic protein level;
   b) Type 2 HAE documented in chart notes with ALL of the following (Note: tests listed below must be repeated for confirmation of diagnosis):
      i) Normal or elevated level of C1-INH antigenic protein (as defined by performing lab); AND
      ii) Low level (below the limits of the laboratory's normal reference range) C4 and C1-INH functional; AND
5. Documentation in medical chart of at least **two** attacks per month before treatment initiation; AND
6. Medication is **not** being used in combination with Haegarda; AND
7. Medications known to cause angioedema (i.e. ACE-Inhibitors, estrogens, angiotensin II receptor blockers) have been evaluated and discontinued when appropriate.
8. **Dosage allowed:** 1,000 units intravenously every 3-4 days.

Note: Personal documentation (log book, journal, etc.) of medication use will be necessary for reauthorization. Prescribers should be aware and make their patients aware of this requirement for reauthorization.

If member meets all the requirements listed above, the medication will be approved for 3 months.

For **reauthorization**:

1. Member must be in compliance with all other initial criteria; AND
2. Chart notes have been provided that show the member's signs and symptoms of disease have improved and the number of acute attacks per month has decreased; AND
3. Log of medication use supported by medical chart or by claims data has been provided.

If member meets all the reauthorization requirements above, the medication will be approved for an additional 6 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.

References:

1. Cinryze [package insert]. Exton, PA; ViroPharma Biologics, Inc.; February 2014.
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3. ClinicalTrials.gov web site. Bethesda, MD. U.S. National Institutes of Health. Identifier NCT02584959, Study to Evaluate the Clinical Efficacy and Safety of Subcutaneously Administered C1 Esterase Inhibitor for the Prevention of Angioedema Attacks in Adolescents and Adults With Hereditary Angioedema; October 20, 2015. Available at: https://clinicaltrials.gov/ct2/show/NCT02584959.
4. Craig T, Pursun EA, Bork K, Bowen T, et al. World Allergy Organization Guideline for the Management of Hereditary Angioedema. WAO J. 2012; 5:182-199.
5. Lang DM, Aberer W, Bernstein JA, et al. International consensus on hereditary and acquired angioedema. Ann Allergy Asthma Immunol. 2012;109:395-402.
6. Lexicomp Online® , Lexi-Drugs® , Hudson, Ohio: Lexi-Comp, Inc.; July 17, 2017.
7. Longhurst H, Cicardi M, Craig T, et al. Prevention of Hereditary Angioedema Attacks with a Subcutaneous C1 Inhibitor. N Engl J Med. 2017;376(12):1131-1140.
8. Lumry W. Management and Prevention of Hereditary Angioedema Attacks. Am J Manag Care. 2013;19:S111-S118.
9. Cinryze. Micromedex Solutions. Truven Health Analytics, Inc. Ann Arbor, MI. Available at: http://www.micromedexsolutions.com. Accessed August 8, 2017.
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(5): 458-467.

Effective date: 09/08/2017
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