PHARMACY POLICY STATEMENT
Indiana Medicaid

DRUG NAME	Cinryze (C1 esterase inhibitor (human))
BILLING CODE	J0598
BENEFIT TYPE	Medical
SITE OF SERVICE ALLOWED	Home/Office
STATUS	Prior Authorization Required

Cinryze is a C1 esterase inhibitor indicated for routine prophylaxis against angioedema attacks in adults, adolescents and pediatric patients (6 years of age and older) with Hereditary Angioedema (HAE).
HAE is a rare autosomal dominant disease characterized by episodic unpredictable swelling, which can occur in a variety of anatomic locations. The swelling results from excess production of the vasodilator bradykinin. Attacks may be painful and cause functional impairment but are not associated with pruritis. The most common types of HAE are caused by deficiency (type 1) or dysfunction (type 2) of C1 inhibitor (C1-INH). Type 1 is the most prevalent.

Cinryze (C1 esterase inhibitor (human)) will be considered for coverage when the following criteria are met:

Hereditary Angioedema (HAE)

For initial authorization:
1. Member must be 6 years of age or older; AND
2. Medication must be prescribed by or in consultation with an allergist or immunologist; AND
3. Member has a diagnosis of HAE type I or type II confirmed by both of the following:
   a) Low Age 12+: 1000 units IV infusion every 3 or 4 days; if response is inadequate, doses up to 2,500 units

(not exceeding 100 units/kg) every 3 or 4 days may be considered.
Age 6-11: 500 units IV infusion every 3 or 4 days; if response is inadequate, doses up to 1000 units every 3 or 4 days may be considered.
QL: 20 vials (500 IU/vial) per 28 days

If all the above requirements are met , 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 all the above requirements are met , the medication will be approved for an additional 12 months.

CareSource considers Cinryze (C1 esterase inhibitor (human)) not medically necessary for the treatment of conditions that are not listed in this document. For any other indication, please refer to the Off- Label policy.

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.
07/05/2022	Transferred to new template. Updated references.

References:

1. Cinryze [package insert]. Exton, PA; ViroPharma Biologics, Inc.; 2020.
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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. 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
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6. Betschel S, Badiou J, Binkley K, et al. The International/Canadian Hereditary Angioedema Guideline [published correction appears in Allergy Asthma Clin Immunol. 2020 May 6;16:33]. *Allergy Asthma Clin Immunol*. 2019;15:72. Published 2019 Nov 25. doi:10.1186/s13223-019-0376-8
7. Maurer M, Magerl M, Betschel S, et al. The international WAO/EAACI guideline for the management of hereditary angioedema - The 2021 revision and update. *World Allergy Organ J*. 2022;15(3):100627. Published 2022 Apr 7. doi:10.1016/j.waojou.2022.100627

Effective date: 01/01/2023
Revised date: 07/05/2022