BOSTON MEDICAL CENTER HEALTHNet PLAN

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Pharmacy Policy

Hereditary Angioedema

Policy Number: 9.101
Version Number: 2.0
Version Effective Date: 1/1/2022

Product Applicability	☐ **All Plan[+] Products**

Well Sense Health Plan
☐ New Hampshire Medicaid

Boston Medical Center HealthNet Plan
☐ MassHealth - MCO
☐ MassHealth - ACO
☒ Qualified Health Plans/ConnectorCare/Employer Choice Direct
☐ Senior Care Options

Note: Disclaimer and audit information is located at the end of this document.

Prior Authorization Policy

Products Affected:

- **icatibant (Firazyr)**
- **Haegarda (C1 Esterase Inhibitor [Human])**
- **Orladeyo (berotralstat)**
- **Berinert (C1 Esterase Inhibitor [Human])**
- **Cinryze (C1 Esterase Inhibitor [Human])**
- **Kalbitor (ecallantide)**
- **Ruconest (C1 esterase inhibitor [recombinant])**
- **Takhzyro (lanadelumab-flyo)**

The Plan may authorize coverage of the above products for members meeting the following criteria:

Covered Use	All FDA approved indications not otherwise excluded

[+] *Plan* refers to Boston Medical Center Health Plan, Inc. and its affiliates and subsidiaries offering health coverage plans to enrolled members. The Plan operates in Massachusetts under the trade name Boston Medical Center HealthNet Plan and in other states under the trade name Well Sense Health Plan.

Exclusion Criteria	Use of two drugs in this policy for the same indication
Required Medical Information	Diagnosis and Documentation of the following: **<u>Berinert , Icatibant, Kalbitor, Ruconest:</u>** 1. Diagnosis of type I or II Hereditary Angioedema(HAE); ***AND*** 2. Attestation that diagnosis is confirmed by laboratory testing (e.g., low C4 level, reduced C1 esterase inhibitor level or function); **AND** 3. Baseline frequency of HAE attacks is documented; **AND** 4. Member has a history of at least one severe attack within the past 6 months **<u>Cinryze , Haegarda, Orladeyo, Takhyzro :</u>** 1. Diagnosis of Hereditary Angioedema (HAE); ***AND*** 2. Attestation that diagnosis is confirmed by laboratory testing (e.g., low C4 level, reduced C1 esterase inhibitor level or function); **AND** 3. Baseline frequency of HAE attacks must be documented; **AND** 4. Documentation that "on-demand" therapy (i.e. Berinert, icatibant, Kalbitor, or Ruconest) did not provide satisfactory control; **AND** 5. Member has had an inadequate response, intolerance or contraindication to a trial of **<u>both</u>** of the following classes of medication: a. 17α-alkylated androgens (e.g. danazol, stanozolol, oxandrolone, methyltestosterone); **AND** b. Antifibrinolytic agents (e.g., aminocaproic acid, tranexamic acid)
Age Restriction	Berinert: 5 years of age and older Cinryze: 6 years of age and older Haegarda, Kalbitor, Orladeyo, Ruconest, and Takhzyro 12 years of age and older Icabitant: 18 years of age and older
Prescriber Restriction	Prescribed by or in consultation with an allergist, hematologist, or immunologist
Coverage Duration	Initial: 3 months Reauthorization: 12 months
Other criteria	**Reauthorization** **Cinryze, Haegarda, Oraldeyo, and Takhzyro**: 1. Significant improvement in severity and duration of attacks have been achieved and sustained, or the member has had a decrease in attack frequency; **AND** 2. Member has been adherent to therapy **Berinert, Icabitant, Kalbitor, and Ruconest** 1. Significant improvement in severity and duration of attacks have been achieved and sustained, or the member has had a decrease in attack frequency; **AND** 2. If the member is also on prophylactic therapy for HAE, they have been adherent to the

	prophylactic treatment.

Applicable Coding:

Code	Medication
J0597	C1 esterase inhibitor (human), 10 units (Berinert®)- intravenous
J0598	C1 esterase inhibitor (human) 10 units (Cinryze™)- intravenous
J1290	Ecallantide 1mg (Kalbitor®)
J1744	Icantibant 1mg (Firazyr®)- subcutaneous
J0596	C1 esterase inhibitor (recombinant) Ruconest- intravenous
J0599	C1 esterase inhibitor (human) [Haegarda]- subcutaneous

Clinical Background Information and References

Original Approval Date	Original Effective Date	Policy Owner	Approved by
12/1/2020	1/1/2021	Pharmacy Services	Pharmacy & Therapeutics (P&T) Committee

Policy Revisions History			
Review Date	**Summary of Revisions**	**Revision Effective Date**	**Approved by**
12/1/2020	P&T Committee: discontinued policy 9.021 and created a separate policy for QHP; moved Berinert, Cinryze, Kalbitor, Ruconest and Takhzyro to Non preferred; reflected generic availability of Firazyr	1/1/2021	P&T Committee
08/12/2021	P&T Annual Review. Add medical benefit only options back to the policy . Add Orladeyo to coverage. Remove restrictions against ACEIs. Update prescriber restrictions and reauthorization criteria.	1/1/2022	P&T Committee

Next Review Date

8/2022

Other Applicable Policies

1. Atkinson JP, Ciardi M, Zuraw B. Prevention of attacks in hereditary angioedema. UptoDate. Last updated Oct 10, 2014. Accessed February 2015. Available from http://www.uptodate.com. .
2. Atkinson JP, Ciardi M, Zuraw B. Treatment of acute attacks in hereditary angioedema. UptoDate. Last updated Dec 19, 2014. Accessed February 2015. Available from http://www.uptodate.com.
3. Berinert® [package insert]. Kankakee (IL): CSL Behring LLC; September 2016.
4. Bowen T, et al. 2010 International Consensus Algorithm for the Diagnosis, Therapy, and Management of Hereditary Angioedema. *Allergy Asthma & Clinical Immunology.* 2010; 6:24. Available at: http://www.aacijournal.com/content/pdf/1710-1492-6-24.pdf.
5. Cicardi M, Zuraw B. Hereditary angioedema: General care and long-term prophylaxis. UpToDate. Last updated Aug 04,2015. Accessed February 2016. Available from http://www.uptodate.com
6. Cicardi M, Zuraw B. Hereditary angioedema: Treatment of acute attacks. UpToDate. Last updated Aug 04, 2015. Accessed February 2016. Available from http://www.uptodate.com.
7. Cinryze® [package insert]. Exton (PA): ViroPharma Biologics, Inc.; December 2016.
8. Craig, T., Aygören-Pürsün, E., & Maurer, M. (2012). WAO guideline for the management of hereditary angioedema. World Allergy Organ J, 5(12), 182-199.
9. Firazyr® [package insert]. Lexington (MA): Shire; December 2015.
10. Haegarda® [package insert]. Marburg, Germany: CSL Behring GmbH; October 2017.
11. Kalbitor® [package insert]. Burlington (MA): Dyax Corp.; March 2015.
12. Maurer M. et al. The International WAO/EAACI guideline for the management of hereditary angioedema- The 2017 revision and update. *Allergy*. 2018;73:1576-1596
13. Orladeyo [package insert]. Durham (NC): BioCryst Pharmaceuticals, Inc.; December 2020.
14. Ruconest® [package insert]. Raleigh (NC): Santaris; February 2015.
15. Sardana N, Craig TJ. Recent Advances in Management and Treatment of Hereditary Angioedema. Pediatrics. 2011;128:1173-1180.
16. Takhzyro™ [package insert]. Lexington, MA; Dyax Corporation. August 2018
17. Zuraw B. Clinical Practice: Hereditary angioedema. *N Engl J Med*. 2008; 359(10):1027-36.
18. Zuraw BL, Banerji A, Bernstein JA, et al. US Hereditary Angioedema Association Medical Advistory Board 2013 recommendations for the management of hereditary angioedema due to C1 inhibitor deficiency. *J Allergy Clin Immunol Pract.* 2013;1(5):458-467.

Reference to Applicable Laws and Regulations, If Any

Disclaimer Information

Medical Policies are the Plan's guidelines for determining the medical necessity of certain services or supplies for purposes of determining coverage. These Policies may also describe when a service or supply is considered experimental or investigational, or cosmetic. In making coverage decisions, the Plan uses these guidelines and other Plan Policies, as well as the Member's benefit document, and when appropriate, coordinates with the Member's health care Providers to consider the individual Member's health care needs.

Plan Policies are developed in accordance with applicable state and federal laws and regulations, and accrediting organization standards (including NCQA). Medical Policies are also developed, as appropriate, with consideration of the medical necessity definitions in various Plan products, review of current literature, consultation with practicing Providers in the Plan's service area who are medical experts in the particular field, and adherence to FDA and other government agency policies. Applicable state or federal mandates, as well as the Member's benefit document, take precedence over

these guidelines. Policies are reviewed and updated on an annual basis, or more frequently as needed. Treating providers are solely responsible for the medical advice and treatment of Members.

The use of this Policy is neither a guarantee of payment nor a final prediction of how a specific claim(s) will be adjudicated. Reimbursement is based on many factors, including member eligibility and benefits on the date of service; medical necessity; utilization management guidelines (when applicable); coordination of benefits; adherence with applicable Plan policies and procedures; clinical coding criteria; claim editing logic; and the applicable Plan – Provider agreement.