


## Prior Authorization Review Panel

### CHC-MCO Policy Submission

A separate copy of this form must accompany each policy submitted for review.  
Policies submitted without this form will not be considered for review.

<b>Plan: PA Health &amp; Wellness</b>	<b>Submission Date: 11/2020</b>
<b>Policy Number: PHW.PDL.501</b>	<b>Effective Date: 01/01/2020</b> <b>Revision Date: 11/2020</b>
<b>Policy Name: Hereditary Angioedema Treatments</b>	
<p><b>Type of Submission – <u>Check all that apply</u>:</b></p> <ul style="list-style-type: none"> <li><input type="checkbox"/> New Policy</li> <li><input checked="" type="checkbox"/> Revised Policy*</li> <li><input type="checkbox"/> Annual Review - No Revisions</li> <li><input checked="" type="checkbox"/> <b>Statewide PDL</b> - <i>Select this box when submitting policies for Statewide PDL implementation and when submitting policies for drug classes included on the Statewide PDL.</i></li> </ul>	
<p><b>*All revisions to the policy <u>must</u> be highlighted using track changes throughout the document.</b></p> <p><b>Please provide any changes or clarifying information for the policy below:</b></p> <p style="margin-top: 20px;">Q1 2021: policy revised according to DHS revisions effective 01/05/2021</p>	
<b>Name of Authorized Individual (Please type or print):</b>  <b>Auren Weinberg, MD</b>	<b>Signature of Authorized Individual:</b>  

## Clinical Policy: Hereditary Angioedema Treatments

Reference Number: PHW.PDL.501

Effective Date: 01/01/2020

Last Review Date: 11/2020

[Revision Log](#)

### Policy/Criteria

*Provider must submit documentation (such as office chart notes, lab results or other clinical information) supporting that member has met all approval criteria.*

It is the policy of health plans affiliated with PA Health and Wellness® that Hereditary Angioedema Treatments are **medically necessary** when the following criteria are met:

### **I. Requirements for Prior Authorization of Hereditary Angioedema (HAE) Agents**

#### **A. Prescriptions That Require Prior Authorization**

All prescriptions for Hereditary Angioedema (HAE) Agents must be prior authorized.

#### **B. Review of Documentation for Medical Necessity**

In evaluating a request for prior authorization of a prescription for an HAE Agent, the determination of whether the requested prescription is medically necessary will take into account whether the beneficiary:

1. Is prescribed the HAE Agent for an indication that is included in the U.S. Food and Drug Administration (FDA)-approved package labeling OR a medically accepted indication; **AND**
2. Is age-appropriate according to FDA-approved package labeling, nationally recognized compendia, or peer-reviewed medical literature; **AND**
3. Is prescribed a dose that is consistent with FDA-approved package labeling, nationally recognized compendia, or peer-reviewed medical literature; **AND**
4. Is prescribed the HAE Agent by or in consultation with an appropriate specialist (i.e., an allergist/immunologist, hematologist, or dermatologist); **AND**
5. Does not have a history of a contraindication to the prescribed medication; **AND**
6. With the exception of requests for short-term prophylaxis (e.g., surgical or dental procedure), will not be using the requested HAE Agent with another HAE Agent for the same indication (i.e., more than one HAE Agent for acute treatment or more than one HAE Agent for long-term prophylaxis); **AND**
7. For a diagnosis of HAE Type I or II (with C1 inhibitor deficiency/dysfunction), has **both** of the following lab values obtained on two separate instances:

- a. Low C4 complement level (mg/dL)
- b. At least **one** of the following:
  - i. Low C1 esterase inhibitor antigenic level (mg/dL)
  - ii. Low C1 esterase inhibitor functional level [ $<65\%$ ] unless already using an androgen or C1 esterase inhibitor];

**AND**

- 8. For a diagnosis of HAE Type III (with normal C1 inhibitor), **all** of the following:
  - a. Has **all** of the following lab values:
    - i. Normal C4 complement level (mg/dL),
    - ii. Normal C1 esterase inhibitor antigenic level (mg/dL),
    - iii. Normal C1 esterase inhibitor functional level,
  - b. Has a history of recurrent angioedema without urticaria,
  - c. **One** of the following:
    - i. Has documentation of a family history of hereditary angioedema
    - ii. Has a hereditary angioedema-causing genetic mutation,
  - d. Failed to respond to maximum recommended doses of antihistamines (e.g., cetirizine 20 mg twice daily);

**AND**

- 9. Is not taking an estrogen-containing medication unless medically necessary or an ACE inhibitor; **AND**
- 10. If prescribed the HAE Agent for long-term prophylaxis, has poorly controlled HAE based on the prescriber's assessment despite use of an HAE Agent for on demand/acute treatment; **AND**
- 11. For a non-preferred HAE Agent, **one** of the following:
  - a. Has a history of therapeutic failure, contraindication, or intolerance to the preferred HAE Agents approved or medically accepted for the beneficiary's indication
  - b. Has a current history (within the past 90 days) of being prescribed the same non-preferred HAE Agent

See the Preferred Drug List (PDL) for the list of preferred HAE Agents at <https://papdl.com/preferred-drug-list>;

**AND**

12. If a prescription for an HAE Agent is for a quantity that exceeds the quantity limit, the determination of whether the prescription is medically necessary will also take into account the guidelines set forth in PA.CP.PMN.59 Quantity Limit Override

NOTE: If the beneficiary does not meet the clinical review guidelines listed above but, in the professional judgment of the physician reviewer, the services are medically necessary to meet the medical needs of the beneficiary, the request for prior authorization will be approved.

**FOR RENEWALS OF PRIOR AUTHORIZATION FOR AN HAE AGENT:** The determination of medical necessity of a request for renewal of a prior authorization for an HAE agent that was previously approved will take into account whether the beneficiary:

1. Is prescribed a dose that is consistent with FDA-approved package labeling, nationally recognized compendia, or peer-reviewed medical literature; **AND**
2. Is prescribed the HAE Agent by or in consultation with an appropriate specialist (i.e., an allergist/immunologist, hematologist, or dermatologist); **AND**
3. With the exception of requests for short-term prophylaxis, will not be using the requested HAE Agent with another HAE Agent for the same indication (i.e., more than one HAE Agent for acute treatment or more than one HAE Agent for long-term prophylaxis); **AND**
4. If prescribed the HAE Agent for acute treatment, has documentation of a positive clinical response to the requested medication; **AND**
5. If prescribed the HAE Agent for long-term prophylaxis, has a documented reduction in the number of HAE attacks; **AND**
6. If a prescription for an HAE Agent is for a quantity that exceeds the quantity limit, the determination of whether the prescription is medically necessary will also take into account the guidelines set forth in PA.CP.PMN.59 Quantity Limit Override

NOTE: If the beneficiary does not meet the clinical review guidelines listed above but, in the professional judgment of the physician reviewer, the services are medically necessary to meet the medical needs of the beneficiary, the request for prior authorization will be approved.

C. Clinical Review Process

Prior authorization personnel will review the request for prior authorization and apply the clinical guidelines in Section B. above, to assess the medical necessity of a prescription for an HAE Agent. If the guidelines in Section B are met, the reviewer will prior authorize the prescription. If the guidelines are not met, the prior authorization request will be referred to a physician reviewer for a medical necessity determination. Such a request for prior authorization will be approved when, in the professional judgment of the physician reviewer, the services are medically necessary to meet the medical needs of the beneficiary.

**D. Approval Duration**

<b>Berinert, Ruconest, Firazyr, Kalbitor</b>	<b>Acute attacks</b>	<b>6 months</b>
<b>Cinryze, Haegarda</b>	<b>Long-term prophylaxis</b>	<b>12 months</b>
	<b>Short-term prophylaxis</b>	<b>2 doses per procedure</b>
<b>Takhzyro</b>	<b>Long-term prophylaxis</b>	<b>New request: 6 months Renewal request: 12 months</b>

**E. References**

1. Berinert Package Insert. Kankakee, IL: CSL Behring LLC; April 2019.
2. Cinryze Package Insert. Lexington, MA: Shire ViroPharma Incorporated; June 2018.
3. Firazyr Package Insert. Lexington, MA: Shire Orphan Therapies, LLC; April 2020.
4. Frank MM, Zuraw B, Banerji A, et al. Management of Children With Hereditary Angioedema Due to C1 Inhibitor Deficiency. *Pediatrics*. 2016;138(5):e20160575
5. Haegarda Package Insert. Kankakee, IL: CSL Behring LLC; September 2019.
6. Kalbitor Package Insert. Burlington, MA: Dyax Corp.; March 2015.
7. Maurer, M., Magerl, M., Ansotegui, I. *et al.* The international WAO/EAACI guideline for the management of hereditary angioedema – the 2017 revision and update. *World Allergy Organ J* 11, 5 (2018). <https://doi.org/10.1186/s40413-017-0180-1>
8. Ruconest Package Insert. Bridgewater, NJ: Pharming Healthcare Inc.; December 2019.
9. Takhzyro Package Insert. Lexington, MA: Dyax Corp.; November 2018.
10. Zuraw B. Hereditary angioedema (due to C1 inhibitor deficiency): General care and long-term prophylaxis. Waltham, MA; UpToDate Inc. Updated May 28, 2020. Accessed July 28, 2020.
11. Zuraw B. Hereditary angioedema: Pathogenesis and diagnosis. Waltham, MA: UpToDate Inc. Updated January 15, 2018. Accessed January 27, 2020.
12. Zuraw B, Farkas H. Hereditary angioedema: Acute treatment of angioedema attacks. Waltham, MA: UpToDate Inc. Updated March 23, 2020. Accessed July 28, 2020.
13. Zuraw BL, Banerji A, Bernstein JA, Busse PJ, Christiansen SC, Davis-Lorton M, 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. <http://dx.doi.org/10.1016/j.jaip.2013.07.002>.

Reviews, Revisions, and Approvals	Date
Policy created	01/01/2020
Q3 2020 annual review: no changes.	07/2020
Q1 2021: policy revised according to DHS revisions effective 01/05/2021	11/2020