pennsylvania
DEPARTMENT OF HUMAN SERVICES

MEDICAL ASSISTANCE BULLETIN

ISSUE DATE	EFFECTIVE DATE	NUMBER
November 9, 2020	January 5, 2021	*See below

SUBJECT

Prior Authorization of Hereditary Angioedema (HAE) Agents – Pharmacy Services

BY

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IMPORTANT REMINDER: All providers must revalidate the Medical Assistance (MA) enrollment of each service location every 5 years. Providers should log into PROMISe to check the revalidation dates of each service location and submit revalidation applications at least 60 days prior to the revalidation dates. Enrollment (revalidation) applications may be found at: https://www.dhs.pa.gov/providers/Providers/Pages/PROMISe-Enrollment.aspx.

PURPOSE:

The purpose of this bulletin is to issue updated handbook pages that include the requirements for prior authorization and the type of information needed to evaluate the medical necessity of prescriptions for Hereditary Angioedema (HAE) Agents submitted for prior authorization.

SCOPE:

This bulletin applies to all licensed pharmacies and prescribers enrolled in the Medical Assistance (MA) Program. The guidelines to determine the medical necessity of HAE Agents will be utilized in the fee-for-service delivery system and by the MA managed care organizations (MCOs) in Physical Health HealthChoices and Community HealthChoices. Providers rendering services in the MA managed care delivery system should address any questions related to the prior authorization of HAE Agents to the appropriate managed care organization.

*01-20-25	09-20-24	27-20-20	33-20-21
02-20-18	11-20-18	30-20-17	
03-20-18	14-20-19	31-20-25	
08-20-28	24-20-40	32-20-17	

COMMENTS AND QUESTIONS REGARDING THIS BULLETIN SHOULD BE DIRECTED TO:

The appropriate toll-free number for your provider type.

Visit the Office of Medical Assistance Programs website at https://www.dhs.pa.gov/providers/Providers/Pages/Health%20Care%20for%20Providers/Contact-Information-for-Providers.aspx.

BACKGROUND:

The Department of Human Services' (Department) Pharmacy and Therapeutics (P&T) Committee reviews published peer-reviewed clinical literature and recommends the following:

- Preferred or non-preferred status for new drugs in therapeutic classes already included in the Preferred Drug List (PDL);
- Changes in the status of drugs on the PDL from preferred to non-preferred and non-preferred to preferred;
- New quantity limits;
- Classes of drugs to be added to or deleted from the PDL; and
- New guidelines or revisions to existing guidelines to evaluate the medical necessity of prescriptions submitted for prior authorization.

DISCUSSION:

During the August 11, 2020, meeting, the P&T Committee recommended the following revisions to the guidelines to determine medical necessity of HAE Agents:

- Addition of guidelines to determine that the beneficiary's indication, age, and prescribed dose are consistent with U.S. Food and Drug Administration (FDA)-approved labeling or medical literature;
- Revision of a guideline to determine that the beneficiary is being prescribed the HAE Agent by or in consultation with an appropriate specialist;
- Addition of a guideline that the beneficiary does not have a history of a contraindication to the prescribed HAE agent;
- Addition of a guideline that the beneficiary will not be using the requested HAE Agent with another HAE Agent for the same indication;
- Revision of the guideline that a beneficiary with a diagnosis of HAE Type I or II (with C1 inhibitor deficiency/dysfunction) has lab values obtained on two separate instances confirming a diagnosis of HAE;
- Addition of guidelines for a diagnosis of HAE Type III (with normal C1 inhibitor);
- Removal of the guidelines that if prescribed a human C1 esterase inhibitor, the beneficiary was tested for hepatis B, hepatitis C, and HIV and received vaccination for hepatitis B;
- Addition of a guideline that specifies that HAE Agents are subject to the guidelines in the Quantity Limits Chapter;
- Revision to the guideline for a beneficiary being prescribed an HAE Agent for long-term prophylaxis;
- Addition of a renewal guideline that if prescribed the HAE Agent for acute treatment, the beneficiary has documentation of a positive clinical response to the requested medication; and

- Revision to the renewal guideline that if prescribed the HAE Agent for long-term prophylaxis, the beneficiary has a documented reduction in the number of HAE attacks.

The revisions to the guidelines to determine medical necessity of HAE Agents, as recommended by the P&T Committee, were subject to public review and comment and subsequently approved for implementation by the Department.

PROCEDURE:

The procedures for prescribers to request prior authorization of HAE Agents are located in SECTION I of the Prior Authorization of Pharmaceutical Services Handbook. The Department will take into account the elements specified in the clinical review guidelines (which are included in the provider handbook pages in the SECTION II chapter related to HAE Agents) when reviewing the prior authorization request to determine medical necessity.

As set forth in 55 Pa. Code § 1101.67(a), the procedures described in the handbook pages must be followed to ensure appropriate and timely processing of prior authorization requests for drugs that require prior authorization.

ATTACHMENTS:

Prior Authorization of Pharmaceutical Services Handbook - Updated pages

RESOURCES:

Prior Authorization of Pharmaceutical Services Handbook – SECTION I
Pharmacy Prior Authorization General Requirements
https://www.dhs.pa.gov/providers/Pharmacy-Services/Pages/Pharmacy-Prior-Authorization-General-Requirements.aspx

Prior Authorization of Pharmaceutical Services Handbook – SECTION II
Pharmacy Prior Authorization Guidelines
https://www.dhs.pa.gov/providers/Pharmacy-Services/Pages/Clinical-Guidelines.aspx

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 the Quantity Limits Chapter. The list of drugs that are subject to quantity limits, with accompanying quantity limits, is available at: https://www.dhs.pa.gov/providers/Pharmacy-Services/Pages/Quantity-Limits-and-Daily-Dose-Limits.aspx.

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 the Quantity Limits Chapter. See Quantity Limits List: https://www.dhs.pa.gov/providers/Pharmacy-Services/Pages/Quantity-Limits-and-Daily-Dose-Limits.aspx.

   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. References

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6. Kalbitor Package Insert. Burlington, MA: Dyax Corp.; March 2015.
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11. Zuraw B. Hereditary angioedema: Pathogenesis and diagnosis. Waltham, MA: UpToDate Inc. Updated January 15, 2018. Accessed January 27, 2020.
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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.