pennsylvania
DEPARTMENT OF HUMAN SERVICES

MEDICAL ASSISTANCE BULLETIN

ISSUE DATE	EFFECTIVE DATE	NUMBER
November 7, 2023	January 8, 2024	*See below

SUBJECT

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

BY

Sally A. Kozak, Deputy Secretary
Office of Medical Assistance Programs

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 and managed care delivery systems. Providers rendering services to MA beneficiaries in the managed care delivery system should address any questions related to the prior authorization of HAE Agents to the appropriate managed care organization.

BACKGROUND:

*01-23-40	09-23-39	27-23-30	33-23-37
02-23-28	11-23-28	30-23-31	
03-23-26	14-23-27	31-23-41	
08-23-43	24-23-36	32-23-26	

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.

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

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

DISCUSSION:

During the September 13, 2023, meeting, the P&T Committee recommended the following revisions to the medical necessity guidelines for HAE Agents:

- Revision of the guidelines for a diagnosis of HAE Type III (with normal C1 inhibitor) that failure to respond to maximum recommended doses of antihistamines (e.g., cetirizine 20 mg twice daily) applies only to beneficiaries with documentation of a family history of HAE, not to beneficiaries with an HAE-causing genetic mutation.
- Revision of the guidelines for non-preferred HAE Agents to consider therapeutically equivalent generics, interchangeable biosimilars, and unbranded biologics.

The revisions to the guidelines to determine medical necessity of prescriptions for HAE Agents submitted for prior authorization, 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 and products 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 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. **Both** of the following:

      a) Has documentation of a family history of HAE
      b) Failed to respond to maximum recommended doses of antihistamines (e.g., cetirizine 20 mg twice daily)

   ii. Has a HAE-causing genetic mutation;

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 of or a contraindication or an 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 (does not apply to non-preferred brands when the therapeutically equivalent generic, interchangeable biosimilar, or unbranded biologic is preferred or to non-preferred generics, interchangeable biosimilars, or unbranded biologics when the therapeutically equivalent brand, interchangeable brand, or brand biologic product is preferred)

    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

1. Berinert Package Insert. Kankakee, Il: CSL Behring LLC; September 2021.
2. Busse PJ, Christiansen SC, Riedl MA, Banerji A, Bernstein JA, Castaldo AJ, Craig T, Davis-Lorton M, Frank MM, Li HH, Lumry WR, Zuraw BL. US HAEA Medical Advisory Board 2020 Guidelines for the Management of Hereditary Angioedema. J Allergy Clin Immunol Pract. 2021 Jan;9(1):132-150.e3. doi: 10.1016/j.jaip.2020.08.046. Epub 2020 Sep 6. PMID: 32898710.
3. Cinryze Package Insert. Lexington, MA:Takeda Pharmaceuticals U.S.A., Inc.; February 2023.
4. Firazyr Package Insert. Lexington, MA: Takeda Pharmaceuticals America, Inc.; October 2021.
5. Frank MM, Zuraw B, Banerji A, et al. Management of Children With Hereditary Angioedema Due to C1 Inhibitor Deficiency. Pediatrics. 2016;138(5):e20160575
6. Haegarda Package Insert. Kankakee, Il: CSL Behring LLC; January 2022.
7. Kalbitor Package Insert. Lexington, MA: Takeda Pharmaceuticals U.S.A., Inc.; November 2021.
8. Maurer, M, Magerl, M, Betschel, S, et al. The international WAO/EAACI guideline for the management of hereditary angioedema—The 2021 revision and update. Allergy. 2022; 77: 1961–1990. doi:10.1111/all.15214
9. Orladeyo Package Insert. Durham, NC: BioCryst Pharmaceuticals, Inc.; March 2022.
10. Ruconest Package Insert. Warren, NJ: Pharming Healthcare Inc.; April 2020.
11. Sajazir Package Insert. Cambridge, United Kingdom: Cycle Pharmaceuticals Ltd; May 2022.
12. Takhzyro Package Insert. Lexington, MA: Takeda Pharmaceuticals U.S.A., February 2023.
13. Zuraw B, Bork K. Hereditary angioedema with normal C1 inhibitor. Saini S, Felweg AM, eds. Waltham, MA: UpToDate Inc. Updated October 11, 2021. Accessed August 08, 2023.
14. Zuraw B, Farkas H. Hereditary angioedema: Acute treatment of angioedema attacks. Saini S, Felweg AM, eds. Waltham, MA: UpToDate Inc. Updated March 23, 2020 May 09. 2023. Accessed July 28, 2020 August 08, 2023.
15. Zuraw B, Farkas H. Hereditary angioedema (due to C1 inhibitor deficiency): Pathogenesis and diagnosis. Saini S, Felweg AM, eds. Waltham, MA: UpToDate Inc. Updated February 08, 2022. Accessed August 08, 2023.
16. Zuraw B, Farkas H. Hereditary angioedema (due to C1 inhibitor deficiency): General care and long-term prophylaxis. Saini S, Felweg AM, eds. Waltham, MA; UpToDate Inc. Updated June 29, 2023. Accessed August 8, 2023.