

PA DEPARTMENT OF HUMAN SERVICES  
MAAC BRIEFING DOCUMENT  
SICKLE CELL ANEMIA AGENTS

**I. Requirements for Prior Authorization of Sickle Cell Anemia Agents**

**Proposed Effective Date:** March 16, 2026

Revisions are noted with a ~~strikethrough~~ for deletions and **bold and underline** for additions.

A. Revisions to Prescriptions That Require Prior Authorization

Prescriptions for Sickle Cell Anemia Agents that meet any of the following conditions must be prior authorized:

1. A non-preferred Sickle Cell Anemia Agent. See the Preferred Drug List (PDL) for the list of preferred Sickle Cell Anemia Agents at: <https://papdl.com/preferred-drug-list>.
2. A Sickle Cell Anemia Agent with a prescribed quantity that exceeds the quantity limit. The list of drugs that are subject to quantity limits, with accompanying quantity limits, is available at: <https://www.pa.gov/agencies/dhs/resources/pharmacy-services/quantity-limits-daily-dose-limits>.
3. **A prescription for Siklos (hydroxyurea) tablet when prescribed for a beneficiary 18 years of age or older.**

B. Revisions to Review of Documentation for Medical Necessity

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

1. Is prescribed the Sickle Cell Anemia 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 Sickle Cell Anemia Agent by or in consultation with a hematologist/oncologist or sickle cell disease specialist; **AND**
5. ~~Had all potential drug interactions addressed by the prescriber (such as discontinuation of the interacting drug, dose reduction of the interacting drug, or counseling of the beneficiary of the risks associated with the use of both medications when they interact);~~ **AND**

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6. **For Adakveo (crizanlizumab-tmca) or L-glutamine powder**, has a history of therapeutic failure of or a contraindication or an intolerance to maximum tolerated doses of hydroxyurea for at least six months; **AND**
7. **For Siklos (hydroxyurea) tablet for a beneficiary 18 years of age or older, is unable to obtain a hydroxyurea dose that is consistent with FDA-approved package labeling, nationally recognized compendia, or peer-reviewed medical literature with the preferred hydroxyurea capsule; AND**
8. **For a non-preferred hydroxyurea Sickle Cell Anemia Agent, one of the following:**
  - a. **Has a history of therapeutic failure of or a contraindication or an intolerance to the preferred hydroxyurea Sickle Cell Anemia Agents that would not be expected to occur with the requested drug**
  - b. **Is unable to obtain a hydroxyurea dose that is consistent with FDA-approved package labeling, nationally recognized compendia, or peer-reviewed medical literature with the preferred hydroxyurea Sickle Cell Anemia Agents;**

**AND**

9. If a prescription for a Sickle Cell Anemia 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.

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 SICKLE CELL ANEMIA AGENTS: The determination of medical necessity of a request for renewal of a prior authorization for a Sickle Cell Anemia Agent that was previously approved will take into account whether the beneficiary:

1. Has documentation of ~~tolerability and~~ a positive clinical response to the drug; **AND**
2. Is prescribed a dose that is consistent with FDA-approved package labeling, nationally recognized compendia, or peer-reviewed medical literature; **AND**
3. Is prescribed the Sickle Cell Anemia Agent by or in consultation with a hematologist/oncologist or sickle cell disease specialist; **AND**
4. ~~Had all potential drug interactions addressed by the prescriber (such as discontinuation of the interacting drug, dose reduction of the interacting drug, or counseling of the beneficiary of the risks associated with the use of both medications when they interact); **AND**~~

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5. **For Siklos (hydroxyurea) tablet for a beneficiary 18 years of age or older, is unable to obtain a hydroxyurea dose that is consistent with FDA-approved package labeling, nationally recognized compendia, or peer-reviewed medical literature with the preferred hydroxyurea capsule; AND**
6. **For a non-preferred hydroxyurea Sickle Cell Anemia Agent, one of the following:**
  - a. **Has a history of therapeutic failure of or a contraindication or an intolerance to the preferred hydroxyurea Sickle Cell Anemia Agents that would not be expected to occur with the requested drug**
  - a. **Is unable to obtain a hydroxyurea dose that is consistent with FDA-approved package labeling, nationally recognized compendia, or peer-reviewed medical literature with the preferred hydroxyurea Sickle Cell Anemia Agents;**

**AND**

7. If a prescription for a Sickle Cell Anemia 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.

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 a Sickle Cell Anemia 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. ~~Adakveo Package Insert. East Hanover, NJ: Novartis Pharmaceuticals Corporation; November 2019.~~
2. ~~Endari Package Insert. Torrance, CA: Emmaus Medical, Inc.; November 2019.~~
3. ~~Oxbryta Package Insert. San Francisco, CA: Global Blood Therapeutics, Inc.; November 2019.~~
4. ~~Siklos Package Insert. Bryn Mawr, PA: Medunik USA, Inc.; May 2018.~~