Highmark Wholecare
Pharmacy Division
Phone 800-392-1147 Fax 888-245-2049

I. Requirements for Prior Authorization of Sickle Cell Anemia Agents

A. 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 prescription for Siklos (hydroxyurea) tablet when prescribed for a beneficiary 18 years of age or older.

B. 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. 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**
6. 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**
7. 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;

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 a positive clinical response to the requested 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. 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**
5. 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;

   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.

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SICKLE CELL ANEMIA AGENTS PRIOR AUTHORIZATION FORM *(form effective 3/30/2026)*

☐New request ☐Renewal request	# of pages: _____	Prescriber name:	
Name of office contact:		Specialty:	
Contact's phone number:		NPI:	State license #:
LTC facility contact/phone:		Street address:	
Beneficiary name:		City/State/Zip:	
Beneficiary ID#:	DOB:	Phone:	Fax:

CLINICAL INFORMATION

Drug requested:	Strength:	Formulation (powder, tablet, etc.):	
Dose/directions:		Quantity:	Refills:
Diagnosis *(submit documentation)*:		Dx code (*required*):	
Is the medication being prescribed by or in consultation with a hematologist/oncologist or sickle cell disease specialist?		☐Yes ☐No	*Submit documentation of consultation, if applicable.*

Complete all sections that apply to the beneficiary and this request.
Check all that apply and submit documentation for each item.

INITIAL requests

1. **For a ADAKVEO (crizanlizumab-tmca) or L-GLUTAMINE powder:**

   ☐Tried and failed or has a contraindication or an intolerance to maximum tolerated doses of hydroxyurea for at least 6 months

2. **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 *(Refer to https://papdl.com/preferred-drug-list for a list of preferred and non-preferred drugs in this class.)*

3. **For a NON-PREFERRED HYDROXYUREA Sickle Cell Anemia Agent** *(Refer to https://papdl.com/preferred-drug-list for a list of preferred and non-preferred drugs in this class.)***:**

   ☐Tried and failed or has a contraindication or an intolerance to the preferred hydroxyurea Sickle Cell Anemia Agents that would not be expected to occur with the requested drug

   ☐ 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

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RENEWAL requests

1. **For ALL renewal requests:**

   ☐ Has documentation of a positive clinical response to the requested drug

2. **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 *(Refer to https://papdl.com/preferred-drug-list for a list of preferred and non-preferred drugs in this class.)*

3. **For a NON-PREFERRED HYDROXYUREA Sickle Cell Anemia Agent** *(Refer to https://papdl.com/preferred-drug-list for a list of preferred and non-preferred drugs in this class.)*:

   ☐ Tried and failed or has a contraindication or an intolerance to the preferred hydroxyurea Sickle Cell Anemia Agents that would not be expected to occur with the requested drug

   ☐ 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

PLEASE FAX COMPLETED FORM WITH REQUIRED CLINICAL DOCUMENTATION

Prescriber Signature:	**Date:**