Adakveo (crizanlizumab-tmca)
PAM – 025

Iowa Medicaid Program	Prior Authorization	**Effective Date**	01/01/2021
Revision Number	6	**Last Reviewed**	07/17/2026
Reviewed By	Medicaid Medical Director	**Next Review**	07/16/2027
Approved By	Medicaid Clinical Advisory Committee	**Approved Date**	12/23/2020

Overview

Medication: [1]	crizanlizumab-tmca
Brand Name:	Adakveo®
Pharmacologic Category:	Sickle Cell Disease Agents; P-selectin blocker
FDA-Approved Indication(s):	Reduce the frequency of vaso-occlusive crises (VOCs) in adults and pediatric patients aged 16 years and older with sickle cell disease
How Supplied:	100 mg/10 mL (10 mg/mL) single-dose vial
Dosage and Administration:	• Intravenous (IV) infusion: 5 mg/kg at weeks 0 and 2, then 5 mg/kg every 4 weeks thereafter • Provider should reevaluate the treatment at least yearly to assess response to treatment and consider discontinuation if no perceived benefit is achieved.
Benefit Category:	Medical

Descriptive Narrative

Sickle cell disease (SCD) affects about 100,000 persons in the United States and primarily affects African Americans, Latinos, and other minorities. SCD results in a host of acute and chronic complications including vaso-occlusive pain, organ damage, hemolytic anemia, countless disease complications, and shortened survival. Persons with this genetic disorder have an average life expectancy of 40 to 60 years.

Sickle hemoglobin (Hb S, alpha2:beta^{S}2) is a result of a specific point mutation in the gene *HBB*, which encodes hemoglobin beta chains. It substitutes valine for the normal glutamic acid at the seventh amino acid (*HBB* p.glu7val). SCD is an autosomal recessive trait. For the disease to manifest, there must be homozygosity for Hb S or compound heterozygosity for Hb S and beta thalassemia or a different *HBB* variant that interacts with Hb S.[2]

Episodes of acute pain (previously called “sickle cell crises”) are one of the most common types of vaso-occlusive events in SCD and the most common reason for seeking medical attention. Vaso-occlusive pain in SCD is intense, although there can be significant variability in the severity and frequency of acute painful episodes. Pain can co-occur with (and mask) other potentially life-threatening complications of SCD, and so treatment should be accompanied by additional evaluations as appropriate for these complications.[3]

After an acute vaso-occlusive pain episode resolves, it is prudent to ensure that appropriate measures have been taken to decrease the likelihood of future vaso-occlusive pain events and to reduce the intensity of chronic pain. For children and adults with Hb SS or Hb S-beta^0-thalassemia, hydroxyurea is the main prevention therapy for decreasing the incidence rate of acute vaso-occlusive pain. Hydroxyurea has the greatest long-term evidence for benefit in reducing acute painful episodes as well as reducing other vaso-occlusive complications of SCD such as acute chest syndrome and in some cases stroke, decreasing the requirement for blood transfusions, and improving survival. If hydroxyurea is not tolerated or is ineffective, treatment with Adakveo® is an option for patients who are 16 years of age or older.[4]

Guidelines

Guidelines for the management of sickle cell disease are published by the American Society of Hematology (ASH) as five separate guidelines:

1. Guidelines for sickle cell disease: Management of acute and chronic pain (2020).[5]
2. Guidelines for sickle cell disease: Cerebrovascular disease (2020).[6]
3. Guidelines for sickle cell disease: Transfusion support (2020).[7]
4. Guidelines for sickle cell disease: Cardiopulmonary and kidney disease (2019).[8]
5. Guidelines for sickle cell disease: Stem cell transplantation (2021).[9]

In 2024, in response to long-standing member interest, ASH approved a plan to develop a 6th set of guidelines: Dosing and Monitoring of Hydroxyurea for Sickle Cell Disease. Like other ASH guidelines, these new guidelines will be developed using methods that meet the highest standards for rigor and trust. The development process will include formation of an expert panel, systematic reviews of available evidence, structured recommendations, and public review. The project plan has been posted for review.[10]

Criteria

Prior authorization is required.

Adakveo® is considered medically necessary when **ALL** of the following are met:

1. Diagnosis of sickle cell disease (any genotype); **AND**
2. Member is 16 years of age or older; **AND**
3. The member has experienced two or more vaso-occlusive crises (VOCs) within the previous 12 months prior to initiation of therapy; **AND**
4. One of the two following conditions are met (a or b):
    a. The member is currently receiving and will continue to receive hydroxyurea in conjunction with Adakveo®; **OR**
    b. The member has a documented history of treatment failure, intolerance, or contraindication to hydroxyurea; **AND**
5. Adakveo® will not be used in conjunction with Oxbryta® (voxelotor); **AND**
6. Prescribed by, or in consultation with, a hematologist; **AND**
7. Request meets one of the following (a or b):
    a. Regimen prescribed does not exceed 5 mg/kg at weeks 0 and 2, then 5 mg/kg every 4 weeks thereafter; or
    b. Regimen is supported by clinical practice guidelines. Supporting clinical documentation must be provided with any request for which regimen prescribed does not align with FDA-approved labeling.

Adakveo® is considered medically necessary for continuation of therapy when **ALL** of the following are met:

1. Member is currently receiving medication through the Iowa Medicaid benefit or has previously met initial approval criteria; **AND**
2. Documentation of positive clinical benefit to treatment with Adakveo® (as evidenced by a reduction in the frequency of vaso-occlusive crises [VOC] from baseline); **AND**
3. Prescribed by, or in consultation with, a hematologist; **AND**
4. Request meets one of the following (a or b):
    a. Regimen prescribed does not exceed 5 mg/kg every 4 weeks; or
    b. Regimen is supported by clinical practice guidelines. Supporting clinical documentation must be provided with any request for which regimen prescribed does not align with FDA-approved labeling.

Approval Duration and Quantity Limits

	Initial Authorization	Subsequent Authorization(s)
Approval Duration	12 months	12 months
Quantity Limits	5 mg/kg at weeks 0 and 2, then 5 mg/kg every 4 weeks thereafter	5 mg/kg every 4 weeks

Coding and Product Information

The following list(s) of codes and product information are provided for reference purposes only and may not be all inclusive. The inclusion of a code does not imply any right to reimbursement or guarantee claim payment, nor does the exclusion of a code imply that its association to the HCPCS code is inappropriate.

HCPCS	Description
J0791	Injection, crizanlizumab-tmca, 5 mg

ICD-10	Description
D57.00 – D57.819	Sickle cell disease

NDC (Strength)	Labeler	Dosage	Pkg Size	Pkg Qty	Units/ Pkg
00078-0883-61 (100 mg/10 mL single-dose vial)	Novartis Pharmaceuticals Corporation (00078)	5 mg	1	EA	20

Compliance

1. Should conflict exist between the policy and applicable statute, the applicable statute shall supersede.
2. Federal and State law, as well as contract language, including definitions and specific contract provisions or exclusions, take precedence over medical policy and must be considered first in determining eligibility for coverage.
3. Medical technology is constantly evolving, and Iowa Medicaid reserves the right to review and update medical policy on an annual or as-needed basis.

Medical necessity guidelines have been developed for determining coverage for member benefits and are published to provide a better understanding of the basis upon which coverage decisions are made. Medical necessity guidelines are developed for selected physician-administered medications found to be safe and proven to be effective in a limited, defined population or clinical circumstances. They include concise clinical coverage criteria based on current literature review, consultation with practicing physicians in the service area who are medical experts in the particular field, FDA and other government agency policies, and standards adopted by national accreditation organizations. Criteria are revised and updated annually, or more frequently if new evidence becomes available that suggests needed revisions.

References

[1] Adakveo® prescribing information (06/2024). Novartis Pharmaceuticals Corporation: East Hanover, NJ. Available online: www.us.adakveo.com. Accessed May 5, 2026.

[2] Steinberg MH. Pathophysiology of sickle cell disease. Tirnauer JS, ed. UpToDate. Waltham, MA: UpToDate Inc. www.uptodate.com. Accessed May 5, 2026.

[3] Vichinsky EP, Field JJ. Overview of the clinical manifestations of sickle cell disease. Tirnauer JS, ed. UpToDate. Waltham, MA: UpToDate Inc. www.uptodate.com. Accessed May 5, 2026.

[4] Vichinsky EP. Disease-modifying therapies to prevent pain and other complications of sickle cell disease. Tirnauer JS, ed. UpToDate. Waltham, MA: UpToDate Inc. www.uptodate.com. Accessed May 5, 2026.

[5] Brandow AM, Carroll CP, et al. American Society of Hematology 2020 guidelines for sickle cell disease: management of acute and chronic pain. *Blood Adv* 2020; 4 (12): 2656–2701. PMID 32559294.

[6] DeBaun MR, Jordan LC, et al. American Society of Hematology 2020 guidelines for sickle cell disease: prevention, diagnosis, and treatment of cerebrovascular disease in children and adults. *Blood Adv* 2020; 4 (8): 1554-1588. PMID 32298430.

[7] Chou ST, Alsawas M, et al. American Society of Hematology 2020 guidelines for sickle cell disease: transfusion support. *Blood Adv* 2020; 4(2): 327-255. PMID 31985807.

[8] Liem RI, Lanzkron S, et al. American Society of Hematology 2019 guidelines for sickle cell disease: cardiopulmonary and kidney disease. *Blood Adv* 2019; 3(23): 3867-3897. PMID 31794601.

[9] Kanter J, Liem RI, et al. American Society of Hematology 2021 guidelines for sickle cell disease: stem cell transplantation. Blood Adv. 2021 Sep 28;5(18):3668-3689. PMID 34581773.

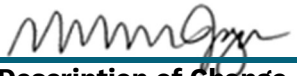
[10] ASH Clinical Practice Guidelines on Sickle Cell Disease. Dosing and Monitoring of Hydroxyurea for Sickle Cell Disease: Plan approved and available for review online at www.hematology.org/education/clinicians/ guidelines-and-quality-care/clinical-practice-guidelines/sickle-cell-disease-guidelines/ash-clinical-practice-guidelines-on-dosing-and-monitoring-of-hydroxyurea-for-sickle-cell-disease. Accessed June 9, 2025.

Development of utilization management criteria may also involve research into other state Medicaid programs, other payer policies, consultation with experts and review by the Medicaid Clinical Advisory Committee (CAC). These sources may not be referenced individually unless they are specifically published and are otherwise applicable to the criteria at issue.

Criteria Change History

Change Date	Changed By	Description of Change	Version
[mm/dd/yyyy]	CAC		
Signature			

Change Date	Changed By	Description of Change	Version
[mm/dd/yyyy]	CAC		
Signature			

Change Date	Changed By	Description of Change	Version
07/17/2026	CAC	Annual review. Updated references.	6
Signature William (Bill) Jagiello, DO			

Change Date	Changed By	Description of Change	Version
07/18/2025	CAC	Annual review. Updated Guidelines section: project plan for the 6th ASH guideline, "Dosing and Monitoring of Hydroxyurea for Sickle Cell Disease, is posted online for review and comments. Updated references.	5
Signature William (Bill) Jagiello, DO			

Change Date	Changed By	Description of Change	Version
07/19/2024	CAC	Annual review. Overview table: added "Provider should reevaluate the treatment at least yearly to assess response to treatment and consider discontinuation if no perceived benefit is achieved." (language approved by FDA June 26, 2024). Descriptive Narrative: added information on vaso-occlusive pain in sickle cell disease and options for treatment. Guidelines: added guidelines on stem cell transplantation, as well as information on a sixth guideline planned for hydroxyurea dosing and monitoring in SCD. Added dosing information into criteria. Updated references.	4
Signature William (Bill) Jagiello, DO			

Change Date	Changed By	Description of Change	Version
07/21/2023	CAC	Annual review. Updated references.	3
Signature William (Bill) Jagiello, DO			

Change Date	Changed By	Description of Change	Version
07/15/2022	CAC	Added overview of sickle cell genetics to Descriptive Narrative. Added criteria language "The regimen/dosing prescribed is within the FDA-approved labeling." Added dosing information to Approval Duration/Quantity Limits section. Initial authorization duration changed from every 6 months to every 12 months. Format updated.	2
Signature William (Bill) Jagiello, DO			

Change Date	Changed By	Description of Change	Version
07/16/2021	CAC	Criteria implementation.	1
Signature William (Bill) Jagiello, DO			

CAC = Medicaid Clinical Advisory Committee