



Medical Policy Bulletin

Title:

Crizanlizumab-tmca (Adakveo®)

Policy #:

MA08.109b

This Policy Bulletin document describes the status of CMS coverage, medical terminology, and/or benefit plan documents and contracts at the time the document was developed. This Policy Bulletin will be reviewed regularly and be updated as Medicare changes their regulations and guidance, scientific and medical literature becomes available, and/or the benefit plan documents and/or contracts are changed.

Policy

Coverage is subject to the terms, conditions, and limitations of the member's Evidence of Coverage.

Company reserves the right to reimburse only those services that are furnished in the most appropriate and cost-effective setting that is appropriate to the member's medical needs and condition.

MEDICALLY NECESSARY

INITIAL THERAPY

Crizanlizumab-tmca (Adakveo®) is considered medically necessary and, therefore, covered to reduce the frequency of vaso-occlusive crises (VOCs) in individuals aged 16 years or older with sickle cell disease, when used as monotherapy or concomitantly with hydroxyurea, when all of the following criteria, including Dosing and Frequency, are met:

- Individual has a documented diagnosis of sickle cell disease confirmed by one of the following tests:
 - Molecular genetic testing that reveals pathogenic variation(s) in the *HBB* gene causing sickle cell disease
 - Hemoglobin electrophoresis
- There is documentation of two or more vaso-occlusive crises (VOCs) in the past 12 months that required a visit to a medical facility and/or healthcare professional and receipt of treatments for condition such as acute pain episodes, acute chest syndrome, hepatic or splenic sequestration, priapism
- Individual is not concomitantly receiving voxelotor (Oxbryta®)
- There are no long-term transfusion therapies planned
- Dosing and Frequency: 5 mg/kg as an intravenous (IV) infusion at Week 0, 2, and every four weeks thereafter

CONTINUATION THERAPY

Continuation of crizanlizumab-tmca (Adakveo®) is considered medically necessary and, therefore, covered for individuals who have demonstrated a documented reduction in the annual rate of sickle cell-related VOCs.

NOT MEDICALLY NECESSARY

When molecular genetic testing reveals established benign variation(s) or wild-type genotype in the *HBB* gene, crizanlizumab-tmca (Adakveo®) is considered not medically necessary and, therefore, not covered because the available published peer-reviewed literature does not support its use in the treatment of this disease.

EXPERIMENTAL/INVESTIGATIONAL

When molecular genetic testing reveals likely pathogenic or variations of unknown significance (VUS) in the *HBB* gene, the use of crizanlizumab-tmca (Adakveo®) is considered experimental/investigational and, therefore, not covered because the safety and/or effectiveness of this service cannot be established by review of the available published peer-reviewed literature.



All other uses for crizanlizumab-tmca (Adakveo®) are considered experimental/investigational and, therefore, not covered unless the indication is supported as an accepted off-label use, as defined in the Company medical policy on off-label coverage for prescription drugs and biologics.

DOSING AND FREQUENCY REQUIREMENTS

The Company reserves the right to modify the Dosing and Frequency Requirements listed in this policy to ensure consistency with the most recently published recommendations for the use of crizanlizumab-tmca (Adakveo®). Changes to these guidelines are based on a consensus of information obtained from resources such as, but not limited to: the US Food and Drug Administration (FDA); Company-recognized authoritative pharmacology compendia; and published peer-reviewed clinical research. The professional provider must supply supporting documentation (i.e., published peer-reviewed literature) in order to request coverage for an amount of crizanlizumab-tmca (Adakveo®) outside of the Dosing and Frequency Requirements listed in this policy. For a list of Company-recognized pharmacology compendia, view our policy on off-label coverage for prescription drugs and biologics.

Accurate member information is necessary for the Company to approve the requested dose and frequency of this drug. If the member's dose, frequency, or regimen changes (based on factors such as changes in member weight or incomplete therapeutic response), the provider must submit those changes to the Company for a new approval based on those changes as part of the utilization management activities. The Company reserves the right to conduct post-payment review and audit procedures for any claims submitted for crizanlizumab-tmca (Adakveo®).

REQUIRED DOCUMENTATION

The individual's medical record must reflect the medical necessity for the care provided. These medical records may include but are not limited to: records from the professional provider's office, hospital, nursing home, home health agencies, therapies, and test reports.

The Company may conduct reviews and audits of services to our members, regardless of the participation status of the provider. All documentation is to be available to the Company upon request. Failure to produce the requested information may result in a denial for the service.

When coverage of crizanlizumab-tmca (Adakveo®) is requested outside of the Dosing and Frequency Requirements listed in this policy, the prescribing professional provider must supply documentation (i.e., published peer-reviewed literature) to the Company that supports this request.

Guidelines

There is no Medicare coverage determination addressing crizanlizumab-tmca (Adakveo®); therefore, the Company policy is applicable.

DRUG INFORMATION

Crizanlizumab-tmca (Adakveo®) is administered as an intravenous (IV) infusion at Week 0, 2, and every four weeks thereafter. Crizanlizumab-tmca (Adakveo®) may be used with or without hydroxyurea.

BENEFIT APPLICATION

Subject to the applicable Evidence of Coverage, crizanlizumab-tmca (Adakveo®) is covered under the medical benefits of the Company's Medicare Advantage products when the medical necessity criteria and Dosing and Frequency Requirements listed in this medical policy are met.

For Medicare Advantage members, certain drugs are available through either the member's medical benefit (Part B benefit) or pharmacy benefit (Part D benefit), depending on how the drug is prescribed, dispensed, or administered. This medical policy only addresses instances when crizanlizumab-tmca (Adakveo®) is covered under a member's medical benefit (Part B benefit). It does not address instances when crizanlizumab-tmca (Adakveo®) is covered under a member's pharmacy benefit (Part D benefit).

US FOOD AND DRUG ADMINISTRATION (FDA) STATUS

Crizanlizumab-tmca (Adakveo®) was approved by the FDA on November 15, 2019, indicated to reduce the frequency of vasoocclusive crises (VOCs) in adults and pediatric individuals aged 16 years and older with sickle cell disease.

The safety and effectiveness of crizanlizumab-tmca (Adakveo®) for sickle cell disease have been established in pediatric individuals aged 16 years and older, supported by evidence from adequate and well-controlled studies in adults and pediatric individuals (SUSTAIN Trial), which enrolled one pediatric individual treated with ADAKVEO 5 mg/kg aged 16 years old. The safety and efficacy of crizanlizumab-tmca (Adakveo®) in pediatric individuals below the age of 16 years have not been established.

Description

Sickle cell disease (SCD) is the most common inherited blood disorder in the United States, affecting 70,000 to 80,000 Americans. The disease is estimated to occur in 1 in 500 African Americans and 1 in 1,000 to 1,400 Hispanic Americans. In sickle cell disease, hemoglobin, a molecule in red blood cells (RBC) that carries oxygen to cells throughout the body, distorts RBCs from a round shape into a sickle, or crescent shape, which becomes hard and sticky. The sickle cells die early, which causes a constant shortage of red blood cells. Symptoms of SCD can vary among individuals in type and severity, and consist of anemia, episodic pain requiring hospitalization (due to blood vessel occlusion), repeat infections, and more serious chronic complications, such as stroke. The episodic pain occurs when the distorted RBCs occlude small blood vessels and deprive the tissues and organs of oxygen-rich blood and can lead to multiorgan dysfunction and early death. Sickle cell-related pain crises are the primary cause of health care encounters in individuals with sickle cell disease. These crises result in a decrease in quality of life and an increase in the risk of death.

SCD is caused by mutations in the *HBB* gene, diagnosed during prenatal screening for hemoglobinopathies. Hemoglobin consists of four protein subunits, typically two subunits called alpha-globin and two subunits called beta-globin. The *HBB* gene provides instructions for making beta-globin. Individuals with SCD have at least one of the beta-globin subunits in hemoglobin replaced with hemoglobin S. Common genotypes of sickle cell disease (SCD) include:

- HbSS: Most severe form of SCD, commonly known as sickle cell anemia. Individuals inherit two sickle cell genes "S", one from each parent
- HbSβ⁰-thalassemia, or HbSβ⁺-thalassemia): Individuals inherit one sickle cell gene "S" from one parent, and one gene for beta thalassemia, another type of anemia, from the other parent. There are two types of beta thalassemia: "0" and "+". Those with HbS beta⁰ -thalassemia usually have a severe form of SCD; those with HbS beta⁺ -thalassemia tend to have a milder form of SCD
- HbSC: Moderate form of SCD. Individuals inherit one sickle cell gene "S" from one parent, and a gene for an abnormal hemoglobin called "C" from the other parent.

P-selectin is a molecule on the surface of endothelial cells and platelets in the blood vessels that promotes the inflammation and adhesion involved in vaso-occlusive crises (VOC). Crizanlizumab-tmca (Adakveo®) is a US Food and Drug Administration (FDA)-approved humanized monoclonal antibody that binds to and blocks the effects P-selectin, therefore preventing RBC occlusion in small blood vessels and maintaining blood flow, and thereby reducing the occurrence and severity of pain crises.

The treatment of sickle cell disease VOCs includes chronic transfusions or oral agents such as hydroxyurea or *L-Glutamine* (Endari™). Both oral agents have different mechanisms of action to treat SCD and are sometimes used in combination. Hydroxyurea has been the standard of care for SCD for many years, with efficacy outcomes that include the ability to decrease the annual rate of SCD pain crises and acute chest syndrome, increase hemoglobin levels, decrease transfusion rates, and prevent primary and secondary stroke in pediatric individuals at risk. There are limitations of these agents, including non-compliance, variable efficacy, as well as monitoring for hematologic toxicity in those receiving hydroxyurea. The only cure for sickle cell disease is a stem cell transplant; however, transplant-related morbidity and mortality remains high.

PEER-REVIEWED LITERATURE

Summary



The safety and efficacy of crizanlizumab-tmca (Adakveo®) was studied in a Phase 2, placebo-controlled, double-blind trial (SUSTAIN) of 198 adolescents and adults 16-65 years of age who were diagnosed with sickle cell disease of any genotype (HbSS, HbSC, HbS β^0 -thalassemia, HbS β^+ -thalassemia, and others) and had, out of two to ten sickle cell--related pain crises in the past 12 months appropriate symptoms to require a visit to a medical facility and/or healthcare professional, plus receipt of pain medication for the crises. Approximately 70% all participants had HbSS, the most severe genotype. Approximately 62% of all participants were receiving hydroxyurea prior to the trial and were able to continue its use, as long as they remained on a stable dose. Key exclusion criteria included planned transfusions, hemoglobin less than 4 g/dL, or planned initiation, termination, or dose alteration of hydroxyurea. Participants were randomized 1:1:1 to receive one of three protocols as an IV infusion every four weeks through Week 52 (after two loading doses two weeks apart): crizanlizumab-tmca (Adakveo®) 5 mg/kg or 2.5 mg/kg, or placebo. The primary outcome of this study was the annual rate of sickle cell--related pain crises leading to a healthcare visit, with high-dose crizanlizumab-tmca (Adakveo®) versus placebo. This trial demonstrated a reduced median annual rate of sickle cell pain crises (SCPCs) by 45.3% compared to placebo (1.63 vs 2.98, $p=0.01$), regardless of concomitant hydroxyurea use or sickle cell disease genotype.

OFF-LABEL INDICATION

There may be additional indications contained in the Policy section of this document due to evaluation of criteria highlighted in the Company's off-label policy, and/or review of clinical guidelines issued by leading professional organizations and government entities.

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Coding

Inclusion of a code in this table does not imply reimbursement. Eligibility, benefits, limitations, exclusions, precertification/referral requirements, provider contracts, and Company policies apply.

The codes listed below are updated on a regular basis, in accordance with nationally accepted coding guidelines. Therefore, this policy applies to any and all future applicable coding changes, revisions, or updates.

In order to ensure optimal reimbursement, all health care services, devices, and pharmaceuticals should be reported using the billing codes and modifiers that most accurately represent the services rendered, unless otherwise directed by the Company.

The Coding Table lists any CPT, ICD-10, and HCPCS billing codes related only to the specific policy in which they appear.

CPT Procedure Code Number(s)

N/A

ICD - 10 Procedure Code Number(s)

N/A

ICD - 10 Diagnosis Code Number(s)

D57.00 Hb-SS disease with crisis, unspecified
D57.01 Hb-SS disease with acute chest syndrome
D57.02 Hb-SS disease with splenic sequestration
D57.03 Hb-SS disease with cerebral vascular involvement
D57.04 Hb-SS disease with dactylitis
D57.09 Hb-SS disease with crisis with other specified complication
D57.1 Sickle-cell disease without crisis
D57.20 Sickle-cell/Hb-C disease without crisis
D57.211 Sickle-cell/Hb-C disease with acute chest syndrome
D57.212 Sickle-cell/Hb-C disease with splenic sequestration
D57.213 Sickle-cell/Hb-C disease with cerebral vascular involvement
D57.214 Sickle-cell/Hb-C disease with dactylitis
D57.218 Sickle-cell/Hb-C disease with crisis with other specified complication
D57.219 Sickle-cell/Hb-C disease with crisis, unspecified
D57.40 Sickle-cell thalassemia without crisis
D57.411 Sickle-cell thalassemia, unspecified, with acute chest syndrome
D57.412 Sickle-cell thalassemia, unspecified, with splenic sequestration
D57.413 Sickle-cell thalassemia, unspecified, with cerebral vascular involvement
D57.414 Sickle-cell thalassemia, unspecified, with dactylitis
D57.418 Sickle-cell thalassemia, unspecified, with crisis with other specified complication
D57.419 Sickle-cell thalassemia, unspecified, with crisis
D57.431 Sickle-cell thalassemia beta zero with acute chest syndrome
D57.432 Sickle-cell thalassemia beta zero with splenic sequestration
D57.433 Sickle-cell thalassemia beta zero with cerebral vascular involvement



D57.434 Sickle-cell thalassemia beta zero with dactylitis
 D57.438 Sickle-cell thalassemia beta zero with crisis with other specified complication
 D57.439 Sickle-cell thalassemia beta zero with crisis, unspecified
 D57.451 Sickle-cell thalassemia beta plus with acute chest syndrome
 D57.452 Sickle-cell thalassemia beta plus with splenic sequestration
 D57.453 Sickle-cell thalassemia beta plus with cerebral vascular involvement
 D57.454 Sickle-cell thalassemia beta plus with dactylitis
 D57.458 Sickle-cell thalassemia beta plus with crisis with other specified complication
 D57.459 Sickle-cell thalassemia beta plus with crisis, unspecified
 D57.80 Other sickle-cell disorders without crisis
 D57.811 Other sickle-cell disorders with acute chest syndrome
 D57.812 Other sickle-cell disorders with splenic sequestration
 D57.813 Other sickle-cell disorders with cerebral vascular involvement
 D57.814 Other sickle-cell disorders with dactylitis
 D57.818 Other sickle-cell disorders with crisis with other specified complication
 D57.819 Other sickle-cell disorders with crisis, unspecified

HCPCS Level II Code Number(s)

J0791 Injection, crizanlizumab-tmca, 5 mg

Revenue Code Number(s)

N/A

Policy History

Revisions From MA08.109b:

10/01/2023	<p>This version of the policy will become effective 10/01/2023.</p> <p>The following ICD-10 CM codes have been added to this policy:</p> <p>D57.04 Hb-SS disease with dactylitis D57.214 Sickle-cell/Hb-C disease with dactylitis D57.414 Sickle-cell thalassemia, unspecified, with dactylitis D57.434 Sickle-cell thalassemia beta zero with dactylitis D57.454 Sickle-cell thalassemia beta plus with dactylitis D57.814 Other sickle-cell disorders with dactylitis</p>
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Revisions From MA08.109a:

2/8/2023	This policy has been reissued in accordance with the Company's annual review process.
03/09/2022	This policy has been reissued in accordance with the Company's annual review process.
05/05/2021	This policy has been reissued in accordance with the Company's annual review process.
10/01/2020	<p>This policy has been identified for the ICD-10 CM code update, effective 10/01/2020.</p> <p>The following ICD-10 CM codes have been added to this policy:</p> <p>D57.03 Hb-SS disease with cerebral vascular involvement D57.09 Hb-SS disease with crisis with other specified complication D57.213 Sickle-cell/Hb-C disease with cerebral vascular involvement D57.218 Sickle-cell/Hb-C disease with crisis with other specified complication D57.413 Sickle-cell thalassemia, unspecified, with cerebral vascular involvement D57.418 Sickle-cell thalassemia, unspecified, with crisis with other specified complication D57.431 Sickle-cell thalassemia beta zero with acute chest syndrome D57.432 Sickle-cell thalassemia beta zero with splenic sequestration D57.433 Sickle-cell thalassemia beta zero with cerebral vascular involvement D57.438 Sickle-cell thalassemia beta zero with crisis with other specified complication D57.439 Sickle-cell thalassemia beta zero with crisis, unspecified D57.451 Sickle-cell thalassemia beta plus with acute chest syndrome</p>



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	<p>D57.452 Sickle-cell thalassemia beta plus with splenic sequestration D57.453 Sickle-cell thalassemia beta plus with cerebral vascular involvement D57.458 Sickle-cell thalassemia beta plus with crisis with other specified complication D57.459 Sickle-cell thalassemia beta plus with crisis, unspecified D57.813 Other sickle-cell disorders with cerebral vascular involvement D57.818 Other sickle-cell disorders with crisis with other specified complication</p> <p>The following ICD-10 CM narratives have been revised in this policy: FROM: D57.411 Sickle-cell thalassemia with acute chest syndrome TO: D57.411 Sickle-cell thalassemia, unspecified, with acute chest syndrome</p> <p>FROM: D57.412 Sickle-cell thalassemia with splenic sequestration TO: D57.412 Sickle-cell thalassemia, unspecified, with splenic sequestration</p> <p>FROM: D57.419 Sickle-cell thalassemia with crisis, unspecified TO: D57.419 Sickle-cell thalassemia, unspecified, with crisis</p>
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Revisions From MA08.109:

09/14/2020	The following new policy has been developed to communicate the Company's coverage criteria for crizanlizumab-tmca (Adakveo®).
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Version Effective Date:

10/01/2020

Version Issued Date:

10/02/2020

Version Reissued Date:

03/09/2022